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RELATO DE CASO

TL 1104864

A POTENTIALLY TREATABLE HEREDITARY SPASTIC PARAPLEGIA MIMICKER: SPINAL CEREBROTENDINOUS XANTHOMATOSIS

Heloisa Lopes Cohim Moreira, Vinícius Lopes Braga, Wladimir Bocca Vieira de Rezende Pinto, Igor Braga Farias, Paulo Victor Sgobbi de Souza, Acary Souza Bulle Oliveira

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Case Presentation: A 23-year-old Brazilian man presented with a slowly progressive history of unsteady gait, lower limb stiffness and numbness involving both feet since 15 years old. Parents were both first degree cousins (consanguineous parents). Examination disclosed global brisk tendon reflexes, extensor plantar responses, moderate spastic paraparesis, and mild reduction in vibratory sense in the lower limbs. Mignarri score: 100. General lab and biochemical screening were unremarkable. Nerve conduction studies showed demyelinating and axonal sensorimotor polyneuropathy. Needle electromyography studies remained unremarkable. Neuroimaging studies disclosed mild cervical spine atrophy, marked signal changes involving the dorsal column and lateral corticospinal tracts of the cervical and thoracic spinal cord. Gene panel testing including Hereditary Spastic Paraplegia (HSP) and Charcot-Marie-Tooth disease genes disclosed the pathogenic variant c.1183C>T (p.Arg395Cys) in homozygosity in CYP27A1 gene, defining the genetic diagnosis of Cerebrotendinous Xanthomatosis (CTX). Plasma cholesterol levels were assessed and showed increased levels. Serum cholesterol and triglyceride levels were normal. Ophthalmologic evaluation was normal. Patient was started on statins (simvastatin). Chenodeoxycholic acid was not available. **Discussion:** CTX is a rare autosomal recessive inherited metabolic disorder due to pathogenic variants in the CYP27A1 gene. Typical systemic and neuroradiological findings represent important clinical clues for high suspicion in different neurological contexts. CTX presents with different neurological phenotypes, including spastic paraparesis (spinal CTX), lower motor neuron disease, cerebellar ataxia, dystonia, parkinsonism, epilepsy, and polyneuropathy. Spinal CTX has been reported in association with other common systemic signs, including chronic diarrhea, juvenile cataracts, and osteopenia. However, pure neurological presentations without early systemic signs may also occur, representing a complex diagnostic challenge, mainly in contexts with broad differential diagnosis. Spinal CTX is commonly a late diagnosis. Mignarri index for CTX suspicion must not limit diagnostic evaluation in patients with pure neurological presentation, as early CTX treatment changes disease natural history. **Final Comments:** Spinal CTX must be included in the differential diagnosis of HSP phenotypes, even in cases without marked multisystem involvement.

Ataxias

TL 1105157

PROGRESSIVE ATAXIA AND PALATAL TREMOR: CASE PRESENTATION OF A RARE DISEASE

Pedro Machry Pozzobon, Ana Beatriz Marangoni Baston, Patrick Emanuel Mesquita Sousa Santos, Tarcísio Nunes Alvarenga, João Vitor Mortari Lisboa, Igor Oliveira Fonseca, Ana Flavia Andrade Lemos, José Eduardo Trentim Longo, Helio Aquaroni Farao Gomes, Laura Cardia Gomes Lopes

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Case Presentation: A 54-years-old female patient, with hypertension and obesity, presented with a 3-years history of progressive speech disorder, evolving with hoarse voice and slurred speech. She also had gait instability and dysphagia. Such changes were better perceived by family members than by the patient herself. The physical examination showed slurred speech, palatal tremor, gait with a slightly widened base and presence of dysmetria and dysidiadochokinesia in left upper limb. A MRI brain showed only moderate cerebellar atrophy and bilateral and signal abnormality in the inferior olivary nuclei bilaterally. Her clinical presentation and neuroimaging finding support the diagnosis of progressive ataxia and palatal tremor (PAPT). **Discussion:** PAPT is a rare and progressive neurodegenerative disease associated with brainstem and cerebellum lesions, especially at Guillain-Mollaret Triangle or dentato-rubro-olivary pathway (DROP). PAPT may be divided into sporadic and familial forms. Familial causes include Alexander's disease, polymerase gamma mutations, spinocerebellar ataxia type 20, and GM2 gangliosidosis. Sporadic cases are still poorly understood, with adulthood onset, and an uncertain cause. In some previous reports, histopathological analysis point to TAU 3R and 4R pathology and hypertrophic olivary degeneration (HOD) as the cause of sporadic form. PAPT clinical features are palatal tremor (usually incidental), ataxia of limbs and gait, dysarthria and dysphagia, in addition to other signs and symptoms of brainstem or autonomic disturbances. Typical MRI abnormalities are high signal in the ION and cerebellar atrophy. At the present moment, there is no treatment to prevent the disease progression. **Final Comments:** Progressive Ataxia and Palatal Tremor (PAPT) is a rare neurodegenerative disease and there are relatively few studies describing this syndrome. Careful examination is important to identification of this presentation.

Ataxias

TL 1105253

FRAGILE X-ASSOCIATED TREMOR/ATAXIA SYNDROME: CASE PRESENTATION

Fabiana Carla dos Santos Correia, André Iglesias Brandão, Sérgio Roberto Pereira da Silva Júnior, Marcus Vinícius de Souza, Rafael Henrique Neves Gomes, Daniel Sabino de Oliveira, Thiago Cardoso Vale

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Case Presentation: A 66-year-old man presented with imbalance and falls, upper-limb tremor and erectile dysfunction for the last two years. Symptoms progressively worsened during the period and patient became more irritable, impulsive and developed impaired memory. Neurological examination revealed ataxic gait, dysmetria, dysidiadochokinesia, bilateral postural and action tremor, and global areflexia with distal lower-limb hypopallessthesia. MOCA test was 19/30 with 11 years of schooling. Brain MRI showed cerebral and cerebellar atrophy, and symmetrical hyperintensities in the middle cerebellar peduncles, periventricular regions and corpus callosum, which was also atrophic (Figure). Electrophysiological study revealed a sensory-motor polyneuropathy. Genetic testing for the fragile X mental retardation 1 (FMR1) gene showed 111 CGG trinucleotide repeats, confirming the diagnosis of fragile X-associated tremor-ataxia syndrome (FXTAS). **Discussion:** FXTAS is caused by the expansion of the CGG trinucleotide in the FMR1 gene (CGG between 55-200 repeats. Clinical picture comprises the presence of one or more characteristics, in different combinations: cerebellar ataxia, parkinsonism, appendicular tremor, neuropathy, dysautonomia, behavioral and cognitive symptoms. Diagnosis is made by major and / or minor clinical and radiological signs. The most important finding is the middle cerebellar peduncles hyperintensities seen on T2-weighted brain MRI sequences. Minor diagnostic criteria include other radiological findings such as white matter hyperintensities and diffuse cerebral atrophy. Corpus callosum atrophy with hyperintensity of the splenium, dentate nucleus and pons may also occur. Definitive diagnosis requires the presence of the premutation associated with the presence of one major radiological sign plus one major clinical symptom or the presence of FXTAS inclusions in neuropathology. **Final Comments:** It is to be emphasized the importance of FXTAS investigation in patients with late onset ataxia especially when associated to parkinsonism, tremor and peripheral polyneuropathy. However, there is, at present, no specific targeted treatment for FXTAS and management remains only symptomatic.

Ataxias

TL 1105253

FRAGILE X-ASSOCIATED TREMOR/ATAXIA SYNDROME: CASE PRESENTATION

Fabiana Carla dos Santos Correia, André Iglesias Brandão, Sérgio Roberto Pereira da Silva Júnior, Marcus Vinicius de Souza, Rafael Henrique Neves Gomes, Daniel Sabino de Oliveira, Thiago Cardoso Vale

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Case Presentation: A 66-year-old man presented with imbalance and falls, upper-limb tremor and erectile dysfunction for the last two years. Symptoms progressively worsened during the period and patient became more irritable, impulsive and developed impaired memory. Neurological examination revealed ataxic gait, dysmetria, dysdiadochokinesia, bilateral postural and action tremor, and global areflexia with distal lower-limb hypopalles-thesia. MOCA test was 19/30 with 11 years of schooling. Brain MRI showed cerebral and cerebellar atrophy, and symmetrical hyperintensities in the middle cerebellar peduncles, periventricular regions and corpus callosum, which was also atrophic (Figure). Electrophysiological study revealed a sensory-motor polyneuropathy. Genetic testing for the fragile X mental retardation 1 (FMR1) gene showed 111 CGG trinucleotide repeats, confirming the diagnosis of fragile X-associated tremor-ataxia syndrome (FXTAS). **Discussion:** FXTAS is caused by the expansion of the CGG trinucleotide in the FMR1 gene (CGG between 55-200 repeats. Clinical picture comprises the presence of one or more characteristics, in different combinations: cerebellar ataxia, parkinsonism, appendicular tremor, neuropathy, dysautonomia, behavioral and cognitive symptoms. Diagnosis is made by major and / or minor clinical and radiological signs: The most important finding is the middle cerebellar peduncles hyperintensities seen on T2-weighted brain MRI sequences. Minor diagnostic criteria include other radiological findings such as white matter hyperintensities and diffuse cerebral atrophy. Corpus callosum atrophy with hyperintensity of the splenium, dentate nucleus and pons may also occur. Definitive diagnosis requires the presence of the premutation associated with the presence of one major radiological sign plus one major clinical symptom or the presence of FXTAS inclusions in neuropathology. **Final Comments:** It is to be emphasized the importance of FXTAS investigation in patients with late onset ataxia especially when associated to parkinsonism, tremor and peripheral polyneuropathy. However, there is, at present, no specific targeted treatment for FXTAS and management remains only symptomatic.

Ataxias

TL 1105438

ATYPICAL MANIFESTATIONS OF LATE-ONSET FRIEDREICH'S ATAXIA – CASE PRESENTATION

André Bertola Vanzan Filho, Mariana Ribeiro Pererira, Vivian Pinto de Almeida, Patricia Gomes Pinheiro, Rayanne da Silva Souza, Deborah Santos Sales, Clarissa de Araújo Davico, Daniel Lucas de Lima Silva Santos, Francisco Ramon Canale Ferreira, Karina Lebeis Pires

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Case Presentation: A 57-year-old female who four to five years before, started with a progressive ataxia in four limbs and gait. She also presents progressive dysarthria and dysphagia, and severe weight loss. At examination, she presented exacerbated deep tendon reflexes with Babinski sign, vibratory sensory loss in distal inferior limbs, head tremor, cognitive impairment in executive functions and verbal fluency. She had no vestibular symptoms or parkinsonism findings upon examination. No family history of neurological diseases was reported. Laboratory investigation of neoplastic, infectious, immune-mediated, endocrine and nutritional deficiencies showed no abnormalities. Brain magnetic resonance imaging (MRI) showed extensive microangiopathy findings. Genetic testing presented 100 repetitions in both alleles of Frataxin (FXN) gene for Friedreich ataxia. Friedreich ataxia is an autosomal recessive disease, the main cause of hereditary ataxia. Patients usually present symptoms before 25 years old. Our patient started symptoms at 52 years old, an atypical presentation. Patients usually will have a positive family history for the disease. In most cases, patients present aguanine-adenine-adenine (GAA) trinucleotiderepeat in intron 1 of both alleles of the FXN gene. Positive cases vary from 66-1700 repeats. Normal alleles will have between 7-34 repeats. Our patient's number of repeats is located on the low end of positive cases, probably the cause for her atypical presentation. Ataxia in four limbs and gait is a cardinal sign, present in most patients. Deep tendon reflexes are usually lost. Exacerbated reflexes are atypical. Motor weakness involving inferior limbs is a common symptom. Also, sensory loss in the distal limbs affects could be present, predominately involving proprioception and vibration sense. Dysarthria and dysphagia are common features. The main cause of death in these patients is cardiac dysfunction, due to hypertrophic cardiomyopathy frequently present in the disease, absent in our patient, probably related to the late onset of the disease. Even though, case presentation of late onset Friedreich ataxia are frequently published, we present a report with mainly atypical manifestations and in which the patient has no family history of the disease. Therefore, it is a diagnosis solely relying in genetic testing. We should consider in clinical practice hereditary diseases, even when symptoms are atypical, and patients have no family history.

Ataxias

TL 1105507

A MAN WITH SPORADIC PERRAULT SYNDROME – A CASE PRESENTATION

Thábata Emanuelle Martins Nunes, Patricia Aurea Andreucci Martins Bonilha, Beatriz Cassarotti, Luiz Eduardo Novis, Léo Coutinho, Francisco Manoel Branco Germiniani, Helio Afonso Ghizoni Teive

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Case Presentation: Male patient, 35 years old, 12 years ago started with gait ataxia and vertigo. Previous history of bilateral hearing loss at the age 8 years, had normal neurodevelopment and no other systemic diseases. He is an only child, has no family history of neurological disease and denies consanguinity between his parents and other family members. Physical examination showed ataxic gait, monocular diplopia, dysmetria, dysdiadochokinesia and hyperreflexia with an exhaustible clonus in the lower limbs. He performed an extensive complementary investigation, audiometric exam with bilateral sensorineural hearing loss, brain MRI with cerebellar atrophy, EEG and blood laboratory tests were unremarkable. Afterwards, an exome sequence was performed, and mutation of the HSD17B4 gene was identified, a missense compound heterozygosity. Thus, the diagnosis of Perrault Syndrome was made. **Discussion:** Perrault syndrome is a rare disease, with autosomal recessive inheritance. It manifests with bilateral sensorineural hearing loss in both men and women and varies in severity from moderate to severe, with onset in early childhood. Ovarian dysfunction in women with a 46, XX karyotype is expected, ranging from primary ovarian failure to ovarian dysgenesis. Fertility in affected men is generally reported as normal, although the number of men diagnosed is limited. Neurological manifestations are not common but may include learning disabilities and developmental delay, cerebellar ataxia and peripheral sensory and motor neuropathy. The diagnosis is confirmed molecularly by the presence of biallelic pathogenic variants in one of six genes: CLPP, ERAL1, HARS2, HSD17B4, LARS2 and TWNK. Markedly, our patient manifested this disease with a wide range of neurologic symptoms, besides the expected hearing loss. Therefore, this was a challenging diagnosis, since he had no sister, making it difficult to conclude without genetic testing. **Final Comments:** Perrault syndrome is a rare disease and there are no formal criteria for its diagnosis. The degree of diagnostic difficulty increases when related to male patients who do not have an affected sister. Genetic tests play an important role in the early diagnosis of the disease.

Ataxias

TL 1105666

HYPEREMESIS GRAVIDARUM AND WERNICKE'S ENCEPHALOPATHY: A CASE PRESENTATION

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Case Presentation: Two cases of pregnant patients are described, who presented with hyperemesis gravidarum (HG), evolving with Wernicke's Encephalopathy (WE). In the 1st case, a 33-year-old patient, with gestational diabetes, started presenting HG during the 2nd trimester of pregnancy. Denied alcoholism and drug use. During the 15th gestational week, she underwent a laparoscopic cholecystectomy procedure due to acute cholecystitis, without complications. She had a miscarriage at 20 weeks and 6 days of pregnancy. After the abortion, she evolved with temporo-spatial disorientation, bradypsychism and bradykinesia, and with bilateral decomposition of eye movement. In the 2nd case, a 24-year-old patient with hyperthyroidism developed HG during the 3rd trimester of pregnancy, with gait ataxia, nystagmus with horizontal and vertical downward gazes, and loss of recent memory, with improvement in symptoms after 4 days of Thiamine replacement. **Discussion:** Nausea and vomiting are typical symptoms of the gestational period, affecting up to 70% of patients. A small portion of this population – about 0.3-10.8% – has a worsening of this condition, called HG. In this context, due to the restrictive condition, some patients may develop episodes of hypovitaminosis, such as Thiamine Deficiency (vitamin B1), which is associated with WE. WE is an acute neurological syndrome, with a reversible characteristic, which leads to changes in mental status, ocular abnormalities and cerebellar dysfunction. The association of HG with WE is rare, however, it is capable of causing serious complications if not diagnosed early, with fetal death in up to 50% of cases and maternal death in 10-20%. **Final Comments:** Among the differential diagnoses regarding neurological alterations during pregnancy, the hypothesis of WE should be considered. This study is relevant because it exposes cases of the rare correlation between HG and WE, whose delay in diagnosis can lead to morbid complications, and whose treatment is often simple, capable of reversing neurological symptoms in their entirety and minimizing damage to pregnancy.

Ataxias

TL 1105732

KINETIC TREMOR IN PATIENT WITH TYPE 2 SPINOCEREBELLAR ATAXIA (SCA2) AND FAMILY HISTORY OF ESSENTIAL TREMOR: ATYPICAL PRESENTATION OR OVERLAP OF MOVEMENT DISORDERS?

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Case Presentation: A 59 years-old male patient presented at our outpatient clinic with a medical history of imbalance and dysarthria initiated at the age of 39 and progressed slowly. By the age of 57, the condition has worsened dramatically, with recurrent falls and wheelchair dependence. At the same period, he presented with cephalic and upper limbs tremor, hypersomnia, depressive symptoms, and anxiety. A cranial CT scan showed cerebral atrophy and genetic test for ataxia panel confirmed the diagnosis of type 2 Spinocerebellar Ataxy (SCA2) with 22 and 36 CAG repetitions. Genealogy noticed many cases of SCA2 (father, paternal grandmother, maternal grand-grandfather, two sisters, nephew, and daughter). His mother had cephalic essential tremor. On physical examination, he showed severe gait ataxia, impaired walk, postural tremor on upper limbs and head, dysidiadochokinesia and ocular hypsometrical movements. He was taking Levodopa, Amantadine e Paroxetine with no good response. **Discussion:** Type 2 Spinocerebellar ataxia (SCA2) is a rare neurodegenerative disease, dominant autosomal, with a prevalence of 1-2 cases in 100000. The clinical findings are ataxic gait, dysarthria, dysmetria, dysidiadochokinesia and ocular hypsometrical movements. About 10% of cases of SCA2 present kinetic tremor. Essential tremor is the most common cause of kinetic tremor, with a global prevalence of 1%, occurring specially on males. It presents a dominant autosomal heritage with reduced penetration. Additionally ET (essential tremor) possesses a genetical and phenotypical heterogeneity and represents a series of syndromic symptoms, instead of just one disease. **Final Comments:** In the face of these statements, we can understand this particular case as a rare disease, SCA2, associated with the cephalic tremor, opening the possibility of a diagnosis which considers the overlapping, of pathologies, ataxia and essential tremor or an atypical presentation of SCA2.

Ataxias

TL 1105738

LONG-TERM CEREBELLAR ATAXIA ASSOCIATED WITH CONFOUNDING FACTORS: A COMPLEX CASE FOR ASSESSMENT AND MANAGEMENT

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Case Presentation: Patient 77 years old, 5 months of evolution of rotational vertigo, tremors of upper limbs, loss of balance with a tendency to fall and tonic-clonic seizures. Performing treatment for gastritis (proton pump inhibitor – PPI) for a long time and heavy drinker. The patient had multidirectional nystagmus with limited gaze fixation, bilateral upper limb myoclonus. Global muscular hypotrophy with preserved strength, reflexes and sensitivity. She associated movement decomposition to the coordination exam, trunk instability, did not remain standing without bilateral support and the gait was ataxic. She was investigated for the etiology of cerebellar ataxia and tests were performed to exclude autoimmune, vascular, metabolic and paraneoplastic causes, with no changes tests performed. During the investigation, a hypothesis of autoimmune cerebellar ataxia was raised as a result of high anti GAD (406Ui/ml RV < 10) with IV immunoglobulin, but with no change in the clinical picture. In addition, she maintained a persistent condition of hypomagnesemia (serum mg 0.3), initially attributed to the refeeding syndrome, was later noticed an association between normalization of serum levels and consistent clinical improvement. Renal losses and intestinal disorders were ruled out to justify hypomagnesemia, and therefore, given the prolonged use of PPIs. **Discussion:** Cerebellar ataxia can be associated with numerous causes including metabolic, paraneoplastic, vascular and autoimmune disorders. There are descriptions in the literature not only of magnesium-related ataxia but also other neurological manifestations such as seizures and involuntary movements. Hypomagnesemia caused by chronic PPI use is also described and should be investigated when other causes for the disorder have been excluded. Anti GAD may be positive in approximately 0.4 to 1.7% of the general asymptomatic population, highlighting the need to properly interpret the results obtained. **Final Comments:** Proton pump inhibitors are widely used but they can lead to absorptive changes including for magnesium and, as a consequence, lead to significant deficit and be one of the metabolic causes of neurological symptoms, including cerebellar ataxia. Patients who present compatible conditions should be investigated for the different etiologies that may trigger the symptoms, taking into account the patient's clinical improvement to correctly determine the diagnosis.

Ataxias

TL 1105790

PENGUIN GAIT, LYNX EARS AND BULLDOG SCALP, A DIAGNOSTIC DILEMMA

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Case Presentation: A 29-year-old male reported a 6-year-history of gait disturbance and cognitive impairment. His parents were first cousins, and a brother started with the same symptoms at 18-year-old. Clinical examination demonstrated dysarthria and spastic paraplegia, with a grade IV muscular strength, spasticity, and hyperreflexia in the lower limbs; his patellar and Achilles reflexes were brisk, and he presented a spastic gait. In addition, head and skull examination demonstrated a curious feature: The presence of cutis verticis gyrata. Brain Magnetic Resonance Imaging (MRI) revealed bilateral hyperintensities at the periventricular white matter. Whole-exome sequencing revealed a mutation in the SPG11 gene (SPG11:NM_001160227.2: exon35: c.6493_6494del: pS2165Lfs). **Discussion:** Hereditary spastic paraplegia (HSP) is a heterogeneous group of neurodegenerative disorders characterized by progressive lower limb weakness and spasticity, caused by mutations in the spastic paraplegia gene (SPG). SPG11 gene (located on chromosome 15q13-15), has 100.982 coding nucleotides distributed in 40 exons and translates into spatacsin. Descriptions include more than 160 pathogenic variants, mostly missense mutations. SPG 11 can present as a pure HSP with autosomal recessive inheritance, but patients frequently develop a complicated HSP phenotype with cognitive impairment, peripheral neuropathy, cerebellar, lower motor neuron, and extrapyramidal features. A recognized hallmark of SPG11 is the presence of a thin corpus callosum (TCC) in the brain MRI. HSP-TCC also occurs in peripheral neuropathy with agenesis of the corpus callosum and other HSPs. **Final Comments:** Clinical findings such as the age of onset, gait disturbance, and presentation of motor and cognitive impairment were usual for SPG11, but this was not enough to settle the diagnosis. This case presented no additional clues to help in the diagnosis, such as a thin corpus callosum or "ears of the Lynx". The dermatologic sign was a surprise and can open our eyes to possible future genotype-phenotype correlations toward SPG11. Although there are no reported cases correlating cutis verticis gyrata and SPG11, we suggest that clinicians be aware of this possible association. It was an unusual case, where clinical findings alone led to doubt. Genetic testing was necessary, leading to a surprising outcome.

Ataxias

TL 1105804

SEVERE TOXIC AND IMMUNE SENSORY-CEREBELLAR ATAXIA IN AN ONCOLOGIC PATIENT

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Case Presentation: A 73-year-old female patient presents with progressive worsening of gait in the last 6 months, ranging from walking without devices to using a wheelchair in 3 months. Besides chronic type 2 diabetes, she also had a history of breast and ovarian cancer diagnosed in the last 5 years, both with surgical and chemotherapy treatment. On admission, the patient had preserved strength, pseudoathetotic movements in the fingers of the hands, severe sensory impairment composed by hypopallesthesia in all four limbs, worse in the legs, and proprioceptive dysfunction of the halluxes. She had scanning speech and bidirectional horizontal nystagmus accompanied by hypometric saccades with corrective movements. Axial and appendicular ataxia shown as the inability to remain seated without aid and dysmetria in all four limbs, markedly worse when eyes are closed. In orthostasis, she showed abasia accompanied by extremely cautious gait and Romberg's sign. Cranial magnetic resonance imaging revealed marked cerebellar atrophy and electroneuromyography showed severe sensory and motor neuropathy in the 4 limbs. Cerebrospinal fluid (CSF) with 1 leukocyte and 65mg/dL proteins. Vitamin dosages and inflammatory markers without alterations. The autoimmunity test was positive for anti-Yo in CSF and blood samples, with titers of 1:256 and 1:15360, respectively. After receiving 1 g of Methylprednisolone per day for 5 days, the patient had a slight improvement in ataxia and was discharged for outpatient follow-up. Neurotoxicity is a common side effect in patients undergoing chemotherapy, with peripheral nerves being more affected than the central nervous system. Toxicity is usually worse for sensory fibers, generating paresthesias or proprioceptive deficits when thick fibers are affected. In this same context, neuronal death can also be induced by paraneoplastic syndromes, which are mediated by antibodies produced by immune activation in response to cancer. Among them, Paraneoplastic Cerebellar Degeneration mediated by anti-Yo (also known as anti-Purkinje cell cytoplasmic antibody type-1) stands out, an antibody specifically directed to the cerebellum responsible for generating an ataxic syndrome. Ataxia and sensory changes are common findings in cancer patients and may have a multifactorial origin. The final outcome can be influenced by previous pathologies, chemotherapeutic-induced toxicity and even autoimmunity in a context of paraneoplastic syndrome.

Ataxias

TL 1105808

A RARE CAUSE OF SENSORY-MOTOR AXONAL NEUROPATHY WITH HEARING IMPAIRMENT AND CEREBELLAR ATAXIA

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Case Presentation: A 32-year-old adopted man with a history of frequent falls since childhood presented to our neurogenetic outclinic. He had normal motor milestones achievement but presented with progressive unsteady gait and tremors from the age of five. He has also complained of sensory disturbance, especially at distal legs and feet, dysarthria and hearing loss since 8 years old. He denied any cognitive impairment, seizures, cardiologic or pulmonary involvement. In the examination he had multidirectional nystagmus and mild dysarthria. His strength and DTR were normal throughout. Pinprick and vibration sense were impaired distally. He had bilateral severe dysmetria and dysidiadochokinesia, and an ataxic gait with a broad base, besides a positive Romberg test, suggesting a mixed sensory-cerebellar ataxia. Nerve conduction studies revealed a non length dependent sensory-motor nerve axonal impairment. Needle examination revealed diffuse chronic denervation affecting cranial, cervical, thoracic and lumbo-sacral segments. Brain MRI showed only diffuse mild cortical atrophy. Severe neurosensory hearing loss in the right ear and mild loss in the left ear were evinced in audiometry. His muscle biopsy was unremarkable. Initial molecular analysis ruled out Friedreich ataxia, SCA2, SCA3, CMT1A, CMT1B and CMTX1. WES revealed a hemizygous missense pathogenic variant (c.512T>C; p.Met171Thr) in the AIFM1 gene. The pathogenicity of the variant is supported by this location in a hotspot, deleterious computation prediction (13 out of 16), high conservation score (phyloP100: 8.692) and absence on Genome Aggregation Database. **Discussion:** AIFM1 gene is a nuclear gene related to the transcription of a mitochondrial homodimeric protein that is required to maintain the mitochondrial respiratory complex I. AIFM1 related disorders include X-linked deafness, combined oxidative phosphorylation deficiency 6, spondyloepimetaphyseal dysplasia with hypomyelinating leukodystrophy and Cowchock syndrome, CMTX4. Our case highlights the clinical heterogeneity of this rare variant, pointing out that in a patient with sensory-motor axonal neuropathy with hearing impairment and ataxia, features mostly seen in mitochondrial disorders, AIFM1 gene must be considered. **Final Comments:** AIFM1 gene variants are extremely rare. Here we describe a case of progressive ataxia, hearing impairment and sensory-motor neuropathy in a male patient without known family history, in that a hemizygous AIFM1 variant has been found.

Ataxias

TL 1106188

ACUTE CEREBELLAR ATAXIA ASSOCIATED WITH DENGUE FEVER: CASE PRESENTATION AND LITERATURE REVIEW

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Case Presentation: A 70-year-old female was admitted due to vertigo and imbalance associated high fever, headache, photophobia, myalgia, nausea, and emesis for 5 days. She denied other symptoms, as well as exposure to predisposing agents. At physical examination, there was a global asymmetric cerebellar syndrome, without other abnormalities. Laboratory work-up showed lymphocytosis, leucopenia, thrombocytopenia, and increased liver enzymes. Serologic exams were negative for syphilis, human immunodeficiency virus, hepatitis B and C viruses, cytomegalovirus (CMV), rubella, toxoplasmosis, and trypanosomiasis in peripheral blood, and for CMV, herpes simplex and varicella-zoster viruses, and toxoplasmosis in cerebrospinal fluid. Brain MRI with angiography was unremarkable (Figure 1). During Hospital stay, there was complete recovery of the systemic symptoms and gradual improvement of the cerebellar syndrome. She was discharged for outpatient follow-up, during which a positive IgM (MAC-ELISA) for Dengue virus was identified. Fifty-four days after symptom onset, she had a normal neurological exam, remaining with a mild subjective imbalance complaint. **Discussion:** Despite uncommon, neurological involvement in Dengue fever is well recognized, both due to direct viral lesion and to immunomediated mechanism. Encephalitis and encephalopathy are the most frequent manifestations; however, meningitis, stroke, myelitis, Guillain-Barré syndrome, and neuritis have also been reported. Association between cerebellar syndrome and Dengue, however, is extremely rare, with only seven other cases published (Table 1). Acute cerebellar ataxia, part of a spectrum of cerebellar dysfunction related to inflammatory conditions, is characterized by normal neuroimaging and complete recovery in two months to three years. As etiologies are numerous, a throughout investigation must be performed before establishing this causal relationship. In our patient, it was possible to confirm recent infection by Dengue and to exclude other agents associated with ataxia. **Final Comments:** The present report corroborates with the recognition of Dengue virus as a cause of acute cerebellar ataxia. Considering the infection's incidence worldwide, it is probable that this association is underdiagnosed, especially considering the number of cases of acute ataxia without an identified etiology. Active investigation of Dengue infection should be a part of the work-up in such cases.

Ataxias

TL 1106199

A PATIENT WITH CEREBELLAR ATAXIA AND A ITPR1 GENE MUTATION

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Case Presentation: A previously healthy 40-year-old female presented a 4-year history of gait imbalance and dysarthria. In the past 2 years, she developed head and upper limb tremor. Her parents were both alive at the age of 66 (father) and 60 (mother), both reported by the patient as asymptomatic. She was of Portuguese ancestry on the maternal side and Italian and German ancestry on her paternal side. She had a 16-year-old daughter, also reported asymptomatic. On the neurological examination, she presented tandem gait ataxia, dysidiadochokinesia, dysmetria, nystagmus, hypermetric saccades, and mild lower limb spasticity. Her ocular movements, cognition, and peripheral system were unaffected. Unfortunately, her family members could not be examined, as they currently live in another city. Ancillary workup for acquired ataxias, including vitamin E, alpha-fetoprotein, serology testing, and B12 vitamin were all normal. Brain magnetic resonance imaging (MRI) showed only mild cerebellar atrophy. Cervical MRI and nerve conduction studies were unremarkable. The patient was tested for spinocerebellar ataxias (SCA) type 3 and type 10, both negative. Whole-exome sequencing revealed a heterozygous Arg275Trp variant in the PRKN gene and a heterozygous Asp1882Asn variant in the ITPR1 gene. With the identification of the ITPR1 mutation, the diagnosis of SCA 15 was established. **Discussion:** The ITPR1 gene is in the 3p26 chromosome, and encodes the inositol 1,4,5-triphosphate receptor type 1. Mutation of the gene is transmitted in an autosomal dominant manner, and this phenotype was correlated to several neurological syndromes, such as SCA 15, SCA 29, Under-Tan syndrome, and Gillespie syndrome. The mild symptomatology showed by the patient suggests the diagnosis of SCA 15. **Final Comments:** Slowly progressive cases of cerebellar ataxia can often give the impression of a sporadic or acquired etiology due to an apparent lack of family history. After reasonable exclusion of such causes, the possibility of a genetic etiology should be raised and properly evaluated.

Ataxias

TL 1106279

PNKP MUTATION IN BROTHERS: ONE SYNDROME, TWO PHENOTYPES

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Case Presentation: Two brothers attend at our clinical practice with dysarthria, gait ataxia, polyneuropathy and oculomotor apraxia. The older one, now with 40 years old, began at the age of 1 year with gait ataxia, seizures and neuropsychomotor developmental delay, at 18 years old was restricted to wheelchair and, actually has severe dysarthria, fragmented saccades, ocular apraxia and severe muscular atrophy and anesthesia and loss of vibratory sensation in legs and arms, totalizing 40 at SARA score. The younger one is 21 years old and had history of falls during childhood, but only with 16 years old the gait ataxia was evident and, currently, walks with strong support due to intense postural instability, associated with ocular apraxia, mild dysarthria, mild symmetric atrophy of hands muscles and also with compromise of tactile and vibratory sensory in legs and arms. Both had cerebellar atrophy seen in neuroimaging, nerve conduction study with severe sensorimotor axonal and demyelinating polyneuropathy, elevated alfa-fetoprotein and dyslipidemia. The patients underwent exome evaluation and missense mutation of PNKP gene in homozygosis was identified and both were diagnosed with Ataxia-oculomotor apraxia type 4. **Discussion:** First described in 2015, ataxia-oculomotor apraxia due to PNKP mutation was identified in 11 patients of 8 unrelated Portuguese families. Classified as hereditary autosomal-recessive cerebellar ataxia, this is a rare neurodegenerative disorder and, at the same time, is the second most frequent cause for recessive ataxia in Portugal. In literature review of ataxia-oculomotor apraxia 4 cases, we can find that almost all patients have cerebellar atrophy in brain MRI. The PNKP gene has a important role in DNA-oxidative damage repair and its mutation is associated with Ataxia-oculomotor apraxia type 4, Charcot-Marie-Tooth disease type 2B2 and Microcephaly, seizures, and developmental delay. **Final Comments:** PNKP gene-related ataxia-oculomotor apraxia has a wide spectrum of the age of onset of symptoms, and its diagnosis can only be made with a genetic test, which implies a barrier due to the cost of the test. Although rare, it should be among the possible etiologies in ataxia-oculomotor syndromes.

Ataxias

TL 1106317

CEREBELLAR ATAXIA IN PRIMARY SJÖGREN'S SYNDROME WITHOUT CEREBELLAR ATROPHY

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Case Presentation: An 86-years-old woman presented with imbalance, hand tremors, and frequent falls for 3 years. She reported systemic hypertension, coronary artery disease, previous hepatitis B and C, and a history of alcoholism interrupted for more than 10 years. The neurological evaluation showed hypometric saccades in the pursuit of the gaze, altered vestibulo-ocular reflex, axial and appendicular signs of cerebellar ataxia more evident on the left side. Sensory examination was normal. She had signs of sicca syndrome, with tongue fissures and thick saliva, as well as ocular biomicroscopy with altered But test. Brain MRI showed no cerebellar atrophy, and there was elevated erythrocyte sedimentation rate and positive anti-RO. Dosages of vitamin E, thiamine, anti-GAD, protein electrophoresis, cryoglobulins, and CSF analysis were normal. Nerve conduction studies were unavailable. In order to exclude occult neoplasm, mammography, thyroid and endovaginal ultrasound, and CT scans of the chest, abdomen and pelvis were unremarkable. Antineuronal antibodies were not available. **Discussion:** This patient met the criteria for Primary Sjögren's syndrome (PSS) according to the EULAR-2016. PSS is an autoimmune inflammatory disease characterized by lymphocytic infiltration of the exocrine glands. Up to 70% of patients may have neurological manifestations, predominating the involvement of the peripheral nervous system. Cerebellar ataxia is rarely described, especially in the absence of cerebellar atrophy. In a series of cases that included 14 patients and sought to characterize ataxia in patients with PSS, only one 71-year-old woman with dysmetria and associated sensory ataxia did not have cerebellar atrophy. Other study reported a 56-year-old woman with PSS, cerebellar ataxia and limbic encephalitis without MRI alterations. Immunohistochemistry for antineuronal antibodies revealed cerebellar Purkinje cell staining. **Final Comments:** This is the third case of PSS with cerebellar ataxia without cerebellar atrophy described in the literature. Antibodies against Purkinje cells may be involved in the pathogenesis of the condition. It is worth mentioning that due to the latency between the appearance of neurological symptoms and the detection of cancer, a paraneoplastic syndrome in similar cases remains possible, although extensive screening has been carried out.

Ataxias

TL 1106382

ACUTE POSTINFECTIOUS CEREBELLAR ATAXIA: CASE PRESENTATION

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Case Presentation: A previously healthy 16-year-old boy presented onset of his clinical condition pain in his right leg and diarrhea, subsequent odynophagia, fever, and headache. Diagnosed with a viral infection, prescribed azithromycin, which he used without symptom improvement. After a three days his symptoms evolved into mental confusion, dysphonia, photophobia, fever, emesis, asthenia. While in Hospital, he evolved with nystagmus, ataxia, slow speech and vertigo. Brain MRI was performed, showing hypersignal T2/FLAIR on cerebellar topography suggestive of viral cerebellitis. Lumbar puncture with CSF analysis was performed, without changes. The patient was initially treated with acyclovir, with significant improvement of symptoms and ataxic syndrome. Later on, pulse therapy with methylprednisolone 1g/day for 5 days was performed, also with improvement of symptoms, leading to him being discharged from the Hospital. Patient returned for follow-up of postinfectious cerebellitis after three months. Neurological examination evidenced that the patient was oriented, without speech alterations, grade 5 strength in both MS and MI bilaterally, gait with no alterations, with unaltered cerebellar tests and complete improvement of vertiginous symptoms. **Discussion:** The acute cerebellitis is a rare condition with an inflammatory disorder that can cause a different range of symptoms. The most common symptoms are ataxia, vomiting and headache. The most affected population is the pediatric one. It is important to know that acute post-infectious cerebellar ataxia is a subgroup of acute cerebellitis and they both have symptoms of cerebellar ataxia. By definition an acute cerebellitis is a dysfunction attribute to a recent infectious illness, vaccination or ingestion of medication and with a magnetic resonance imaging (MRI) showing isolated cerebellar inflammation. There are three outcomes for cerebellar ataxia: full recovery, mild neurologic sequelae not affecting play and school activities or mobility, or moderate/severe sequelae requiring special treatment or education. The patient in question showed total improvement in his condition. **Final Comments:** Considering that there is a lack of knowledge about the incidence of this neurological condition, it is necessary to discuss and share data related cases of post-infectious cerebellitis, in order to possible future prospective studies about the incidence and risk factors.

Ataxias

TL 1106533

SPINOCEREBELLAR ATAXIA 17 OF ACCELERATED EVOLUTION

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Case Presentation: Patient, male, 35 years old, previously healthy, parents healthy and not consanguineous, started at age 32 with a complaint of imbalance, characterized by an insidious and progressive cerebellar ataxic syndrome, and in less than 6 months of progression, he already depended on walking support. Associated with this, he presented with amnesic and dysexecutive mild cognitive impairment, in addition to depressive-type mood disorder. In the same period, he started with epileptic seizures characterized by behavioral arrest and sphincter release, without motor tremors. Due to the accelerated progression of neurological symptoms, despite the innocent cerebrospinal fluid, intravenous immunoglobulin was performed for 5 days, thinking about autoimmune encephalitis, but without improvement. Advanced brain neuroimaging showed pancerebellar atrophy without contrast enhancement. Genetic testing confirmed it to be Spinocerebellar Ataxia type 17 (SCA 17). **Discussion:** SCA 17 is a rare, autosomal dominant genetic disease identified by the abnormal expansion of the CAG/CAA repeat in the TATA-box-binding protein (TBP) gene, with affected individuals having more than 41 repeats. The clinical manifestation begins between 3 and 75 years (mean: 34.6 years), being defined by cerebellar ataxia (95%), cognitive symptoms (90%), psychiatric symptoms, epilepsy, and involuntary movements (chorea, dystonia). By 2005, fewer than 100 families had been reported to have the disease worldwide. Treatment is currently limited to controlling symptoms, improving quality of life, but there is still no cure. **Final Comments:** Genetic tests are increasingly gaining space and usefulness in clinical practice, elucidating cases and directing conducts, which, although often not curative, are substantial for the individual and family. In this specific case of SCA 17, by configuring an autosomal dominant inheritance, it made it possible to structure a family genetic counseling that would guide the family, since the patient had siblings and children.

Ataxias

TL 1105101

ATYPICAL MIGRAINE WITH TRANSIENT BINOCULAR BLINDNESS: CASE PRESENTATION

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Case Presentation: A 58 year-old male with systemic arterial hypertension, rheumatoid arthritis and HIV started, 3 years ago, episodes of acute bilateral blindness lasting about 20 minutes, 5 episodes a year, without other symptoms. He also had a previous history of intense pulsating headache, which started 20 years ago, with nausea, photophobia and phonophobia, about 3 times a week, independent of the blindness episodes. His neurological examination showed no alterations. Brain MRI, intracranial and cervical CT angiography, 24h holter and transthoracic echocardiogram were within the normal limits. He was started on treatment with valproic acid 250 mg bid, which was suspended due to trunk urticaria. Amitriptyline 50 mg daily was started, with a reduction in intensity and frequency of both headache and blindness episodes. The diagnosis was of migraine with binocular blindness aura. **Discussion:** Visual aura is the most frequent type of migraine associated aura, which mechanism is cortical spreading depression in the occipital cortex. However, complete vision loss in both eyes associated with migraine is extremely rare. Recent publications described binocular blindness as migraine aura and its main hypothesis was retinal spreading depression. Studies with chick retinas showed that retinal spreading depression occurs similarly to cortex. Furthermore, migraine with binocular blindness does not appear to be a subform of basilar-type migraine as the blindness occurs in isolation without other symptoms required for ICHD-3 basilar type migraine diagnosis, and retinal migraine is characterized by monocular blindness. A recent case presentation described two unrelated families with repetitive transient daily blindness in five members that had clinical characteristics of a retinal electrical signal abnormal propagation that may be a retinal spreading depression. One study with many migraine patients showed low incidence of binocular blindness. They were all female, had some abnormality of clotting factors and no history of previous aura. Their episodes were infrequent and may even occur only once in life. The authors suggest classifying episodes of transient blindness as an aura-like event of acute onset. Our patient had no other cause to explain his symptoms. Both episodes of bilateral blindness and headache improved with amitriptyline, contributing to the diagnosis.

Cefaleia

TL 1105197

ENCEPHALITIS ASSOCIATED WITH COVID-19 AND THUNDERCLAP HEADACHE: AN UNUSUAL ASSOCIATION

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Case Presentation: 51 years-old woman, previous history of depression and systemic arterial hypertension reporting sudden holocranial headache that has progressed to severe intensity within minutes at onset (thunderclap headache) started seven days before, which progressed with mental confusion, inattention inability to meet family members and difficulty of communication at the day of admission. General physical examination, including respiratory auscultation and oxygen saturation, was normal except for the presence of borderline pyrexia (37.7°C). Neurological examination showed only mental and cognitive alterations described as inattention, somnolence, disorientation in time and space, inability to say her own name or recognize her husband and language disturbance, characterized by relative fluent speech, incapacity to name objects, repeat words or phrases and to obey commands. Computed angio-tomography (CTA) of the brain excluded bleeding but showed one aneurysm in the supraclinoid internal carotid artery. Cerebrospinal fluid had no xanthochromia and evidenced a lymphomonocytic pleocytosis with discrete hyperproteinorraquia. Electroencephalogram showed symmetrical and disorganized brain electrical, theta rhythm diffuse and presence of triphasic waves. Due to Hospital protocol during the Covid-19 pandemic, the patient underwent computed tomography of thorax and we found ground-glass opacities suggesting viral infection. Then, RT-PCR for Sars-CoV-2 with nasopharyngeal swab and in CSF was performed with both positives. Therefore, the diagnosis was encephalitis associated with Covid-19. We opted for supportive care. Patient evolved with many complications, need of ventilatory support and renal replacement therapy with discharged of the Hospital after 35 days, with no symptoms. At follow-up, two months later, the only finding was mild inattention. **Discussion:** Covid-19 is characterized mainly by respiratory symptoms, however this virus frequently damages the nervous system, although the mechanism involved is still unclear. Headache is the most common neurological symptom and has a great heterogeneity, including thunderclap headache, which is a red flag and investigation is mandatory. **Final Comments:** Headache is common in patients with Covid-19 and can be the first signal of this infection or of a complication of that, like encephalitis. In addition, the spectrum of the pain is range and includes thunderclap headache mimicking an SAH, as we approach here.

Cefaleia

TL 1105686

CHALLENGES OF INTRACRANIAL HYPERTENSION

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Case Presentation: A 13 years old female, in December 2021, she started to have daily, moderate-intensity, tightening holocranial headache, lasting for hours and improved after the use of simple analgesics. And it was a headache without warning signs. With persistence of the headache, but with worsening intensity, the patient was admitted to the Hospital on 02/19/2022, complaining of binocular diplopia, with worsening at side views. She presented new episodes of nausea and vomiting 7 days after admission. We found on the neurological examination: she had esotropia and limitation for abduction of the left eye; Fundoscopy: bilateral papilledema. Cerebrospinal Fluid (CSF): Opening pressure (OP) 58 cmH₂O, erythrocytes 15 / cells 23 (Poly 10%; Mono 90%) / Glucose 48 / Proteins 25 / VDRL non-reactive / Gram absent / China ink negative. Culture for bacteria and fungi: No growth. PCR Herpes virus: negative. Capillary glucose: 110 mg/dL. Patient received Ceftriaxone 4g/day for 7 days, Topiramate 100 mg/day and Acetazolamide 500 mg/day for 12 days. Venous Angioresonance was performed: transversus sinus hypoplasia. After 12 days of Hospitalization, patient was without diplopia with improvement of esotropia. A new CSF collect was performed: OP: 38 cmH₂O, cells: 0, erythrocytes: 30, glucose 52. Capillary glucose: 115 mg/dL. **Discussion:** Our patient had a syndromic diagnosis of Intracranial Hypertension (IH): she had a prolonged clinical picture with non-spontaneous clinical resolution (that needed topiramate and acetazolamide to resolve), in the context of an altered CSF. In fact, this was the main reason why our patient did not meet the criteria for Idiopathic IH. The patient presented criteria for aseptic meningitis (AM), with pleocytosis greater than four cells in CSF associated with a process free of contamination by fungi, bacteria or parasites. AM has as main etiologies: systemic diseases with meningeal involvement, induced by drugs, because of neoplastic and the most common, viral. **Final Comments:** Therefore, our patient presented IH associated with CSF with characteristics of AM with a prolonged clinical picture. Due to the various etiologies of AM, diagnosis and treatment becomes a challenge, especially when associated with IH in a young patient with a severe impairment who arrives in the emergency room with CSF so altered. Please note the context of a public Hospital with difficulties in identifying viral causes. for AM.

Cefaleia

TL 1105693

RAEDER SYNDROME: CASE PRESENTATION

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Case Presentation: A 33-year-old male patient, without any previous disease, was admitted due to a headache in the left frontal region of the head, pulsating, with strong intensity that started 03 days ago. He denied and previous similar episodes. On admission examination, he had left palpebral semiptosis associated with ipsilateral myosis, with no other clinical signs. During clinical investigation, a brain magnetic resonance was performed, which showed no alterations. However, the brain angiotomography elucidated an important tapering of the left internal carotid artery. Later, an angiography was performed that confirmed carotid artery dissection on the left of the cervical portion with severe stenosis in the segment, which led the patient to undergo to angioplasty. Patient evolved with improvement of pain and normalization of the neurological examination. **Discussion:** Paratrigeminal oculosympathetic syndrome or Raeder's Syndrome is a rare condition, with an incidence of 1 in 60,000, and was first described in 1924, consisting of a constant unilateral headache, accompanied by Horner's syndrome. Our patient fulfilled all the criteria according to the ICHD-3 classification. This syndrome can have several causes, such as tumor, infection, and trauma, but carotid dissection is the most common cause. It is divided into two groups, group I, which includes patients with additional involvement of some parasellar nerve (III, IV or VI) and group II, where the parasellar nerves are not affected. This second group usually has a good and self-limiting evolution. **Final Comments:** We present a rare cause of secondary headache that, after its correct etiological investigation, had a favorable clinical outcome for the patient.

Cefaleia

TL 1105734

TRIGEMINAL AUTONOMIC CEPHALALGIA ATTRIBUTED TO ISCHAEMIC STROKE: A CASE PRESENTATION

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Case Presentation: A 42-year-old man, with hypertension presented with complete right hemi-hypoesthesia, hemiparesis, and sudden aphasia 2 years ago. He had no previous history of headaches. Just a few minutes before these neurological symptoms, he had a severe left temporal headache. Since then, he continued presenting episodes of headache with the same characteristics, two to three times a week, associated with psychomotor restlessness, ipsilateral conjunctival injection, and lacrimation, which lasts about 90 minutes but occasionally exceeds 10 hours. The admission CT of the brain in the emergency room was normal, but the MRI three months later evidenced a hypointense lesion in the left pons region. Indomethacin 75mg three times a day, carbamazepine 200mg twice a day, and propranolol 40mg twice a day were started, but without a good response. Due to the suspicion of secondary cluster headache, a neural block with lidocaine and dexamethasone of the greater occipital nerve on the left was performed and the use of sodium divalproate 500mg daily was started. A 2-month follow-up showed a 90% decrease in headache attacks. **Discussion:** Primary trigeminal autonomic cephalalgias (TACs) are a rare group of headache disorders. (1) A high number of secondary cases of TACs were described with clinical features equal to primary TACs, and become a diagnostic challenge, being fundamental for the diagnosis of a complete and careful anamnesis. (3) Qi Lei et al. described a case of SUNC secondary to brainstem infarction, demonstrating the association of infarction of this topography and trigeminal autonomic cephalgia. (4) **Final Comments:** We reported a case of trigeminal autonomic cephalgia secondary to ischemic vascular injury. Despite being a rare condition, it can lead to high morbidity. Be aware of this diagnostic possibility promotes a better functional outcome for the patient.

Cefaleia

TL 1105777

HEADACHE ATTRIBUTED TO SLEEP APNEA: CASE PRESENTATION

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Case Presentation: A 61-year-old female presented to our service with chronic headache for 5 years. Initially, she had two episodes per week; however, frequency increased up to daily attacks throughout the years. Headache was bilateral, pulsating, moderate to intense, and was eventually associated with nausea. It lasted on average two hours and always occurred during the morning, usually right after awakening. There was no worsening with physical activity, nor history of photophobia, phonophobia, aura or autonomic signs. She had multiple comorbidities (i.e., type 2 diabetes, chronic renal failure, obesity, hypertension, and glaucoma), as well as nonrestorative sleep, fatigue, snoring, and nocturnal awakenings owing to pain. Neurological examination and magnetic resonance imaging were unremarkable. Amitriptyline was started and increased up to 75 mg daily. After 8 months, the patient had no improvement of the symptoms. A polysomnography (PSG) was posteriorly performed, yielding an apnea/hypopnea index of 62 events per hour. Diagnosis of obstructive sleep apnea (OSA) was made. After three months of continuous positive airway pressure (CPAP) treatment, frequency decreased to one attack per week. **Discussion:** OSA is associated with chronic headache. According to the International Classification of Headache Disorders – 3rd edition (ICHD-3), sleep apnea headache is a specific entity, characterized by morning headache presenting upon awakening and lasting for less than 4 hours, in more than 15 days per month, associated with confirmed OSA by PSG and pain improvement with apnea treatment. Our patient had a pattern of tension-type headache without any response to tricyclic antidepressant. Besides, she had apnea risk factors (e.g., age, body mass index, hypertension, and post-menopausal status) and fulfilled all the criteria above-mentioned. The expressive improvement in frequency and intensity after the gold standard therapy with CPAP confirms the diagnosis. **Final Comments:** The relationship of sleep disturbances and headache is well known. Pain modulation and circadian rhythm share neuroanatomical structures, such as brainstem and hypothalamus. Insomnia, sleep-related breathing disorders and circadian rhythm disorders are associated with increased headache intensity, attack frequency and risk of chronification. Therefore, it should always be remembered in the evaluation of patients with risk factors and non-responsiveness to first-line treatment of primary headache.

Cefaleia

TL 1105821

SHOCKING, SHARP AND UNILATERAL PAIN OF ONSET IN THE EIGHTH DECADE OF LIFE: A CASE-REPORT OF PAROXYSMAL HEMICRANIA-TIC SYNDROME

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Case Presentation: A 72-year-old woman with a five-month history of headaches was admitted at our headache outpatient clinic. The pain was localized in the periorbital and left temporal regions. It was daily, sharp, intense, sometimes throbbing, and with diffuse ipsilateral irradiation. Episodes usually lasted about 30 minutes and occurred five times a day. The patient reported left hemiface heating during the attacks, however without conjunctival hyperemia, lacrimation, nasal congestion, or rhinorrhea. She sometimes referred shocking, short-lasting pain triggered by biting food and touching certain points of the face. Previous treatments with duloxetine, amitriptyline, lamotrigine, and carbamazepine, initiated by a neurologist in another clinic, provided no relief. Her previous medical history was remarkable for treatment of hypothyroidism and presence of temporomandibular joint dysfunction (TMJD). Physical examinations during pain-free periods were normal except for TMJD signs. Blood counts, liver, renal and thyroid function were normal, as well as Brain magnetic resonance imaging (MRI). Still using CBZ, the patient had pain in only the left side of the face. Therapy was changed to 75 mg a day mg of indomethacin and was increased to 100 mg a day, with headache frequency improvement and up to five pain-free consecutive days. After two months she was admitted due to recurrence of pain, with the same pattern, however associated with conjunctival hyperemia and eyelid edema. Reintroduction of CBZ 400 mg a day in combination with indomethacin resulted in complete improvement. **Discussion:** Paroxysmal Hemicrania-Tic (PH-TIC) syndrome is a rare association between chronic paroxysmal hemicrania (CPH) and trigeminal neuralgia (TN). The age of onset is usually between the fifth and seventh decades of life. Features such as pain of different durations, unilateral involvement of the face and partial response to indomethacin should alert to the overlap between PH and TN. Frequently, no abnormalities are found in laboratory tests or in brain MRI. CPH was improved with initiation of indomethacin and trigeminal neuralgia with reintroduction of CBZ. **Final Comments:** PH-TIC is uncommon, however it should be hypothesized in patients with short-lasting headaches associated with facial pain that partially improve with carbamazepine or indomethacin. by case series, over single pharmacotherapy.

Cefaleia

TL 1106039

HALOPERIDOL IN THE TREATMENT OF MIGRATION STATUS : CASE SERIES

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Case Presentation: Headache is one of the most common complaints in the medical activity, especially, in emergency care. The migraine is, among the primary headaches, the most common. However, this is a pain that is often crippling and is characterized by being strong, pulsatile, worsening to physical activities, being still duration of 4 to 72 hours, with a unilateral character in the most cases in addition to being accompanied by nausea, photophobia and phonophobia. In this context, there is an unusual event, however of great relevance: the "migraine status". This is a framework for pain that lasts beyond the 72 hours verified in migraine crises. In this way, it is common for abuse of analgesics in the context of "migraine status" the use of several anti-inflammatory drugs, ergotamine, tryptans in addition to the non-recommended opium derivatives. Thus there is in the literature the indication of haloperidol in the treatment of migraine, especially in "migraine status". Therefore, a series of seven cases of treatment of migraine seizures are reported, taking use of haloperidol as pain control medication. **Discussion:** It is verified in the literature, for the treatment of migraine, many analgesic options, specially in recent years, with the increasingly frequent use of tryptans with gradual reduction in the use of ergotamines due their adverse effects. Many patients seeking emergencies have already taken many groups of analgesic medications even before get to the Hospital. The use of haloperidol in a series of seven cases, in an intravenous environment (5 mg) in five of these cases and intramuscular application (5mg) in two patients between December 2019 and June 2021, is discussed. Three patients are male and the other female. The ages ranging from 28 years to 65 years. In all cases, there was improvement after the use of the substance haloperidol, although several analgesics had been used previously showing the effectiveness of haloperidol compared to other treatments. **Final Comments:** This study reinforces the findings in the literature that support the use of haloperidol in a dose of 5mg in the treatment of migraine crises, especially in "migraine status", making use of the safety and good results that the substance offers.

Cefaleia

TL 1106126

UNILATERAL OCULOMOTOR PALSY SECONDARY IDIOPATHIC INTRACRANIAL HYPERTENSION: A RARE CASE PRESENTATION

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Case Presentation: A 38 years-old-woman, caucasian, civil engineer, residing in Belo Horizonte, Minas Gerais State, Brazil. She has a depressive disorder and has been orally using Sertraline daily. She denied using hormonal contraceptives. She reported chronic headaches for 10 years characterized by monthly episodes of moderate holocranial pressure starting two days before menstruation and lasting up to three days after, and episodes of bilateral frontal pressure headache of severe intensity three to four times per year. Arose in December 2020 with incipient right eyelid ptosis. In August 2021, she had a headache episode that lasted about 30 days. Vitamin D on June 2021 was 31.8 ng/dL. On January, 10th 2022 presented a new episode with exacerbation. Presented to the neurological examination with a medial deviation of the right eye when looking up vertically and looking centered, right pupil mydriatic and photoreactive, and right eyelid ptosis without worsening of the deficit to repetition stimulation. Magnetic Resonance Imaging of the brain on January 17th without relevant abnormalities. On March 04th during clinical reassessment, she reported spontaneous improvement of the headache. She had symmetrical pupils but maintained right eyelid ptosis. Intracranial Pressure non-invasive monitoring by Brain4 Care on March 30th showed normal oscillation of pressure levels, however, the P2/P1 ratio altered the change to decubitus, reaching a value of 1.8, compatible with intracranial hypertension. Magnetic Resonance angiography of the intracranial venous system on April 27th revealed a slight volumetric reduction of the lateral ventricles associated with elongation and tortuosity of optic nerves, notably on the left. A lumbar puncture with opening pressure of 31cm H2O was performed. Ophthalmoscopy of May 19th without abnormalities. Optical coherence tomography this day showed a borderline reduction of the inferior temporal thickness of both eyes and a reduction of the superior temporal area of the left eye. On May, 25th started oral polyvitamin supplement weekly, cholecalciferol weekly, and acetazolamide twice a day. **Discussion:** The sixth cranial nerve is most commonly affected. Third cranial nerve involvement is distinctly unusual. **Final Comments:** Systematic review of cases of ophthalmoplegia in the setting of idiopathic intracranial hypertension published last year identified only one case presentation of unilateral oculomotor palsy.

Cefaleia

TL 1106205

ONABOTULINUM TOXIN TYPE A AS AN ADD-ON THERAPY IN REFRACTORY TRIGEMINAL NEURALGIA: PRELIMINARY RESULTS

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Background: Trigeminal neuralgia (TN) is a rare but often debilitating condition, characterized by paroxysmal attacks of sharp pain in one or more branches of the trigeminal nerve. Although several pharmacological options are available for prophylaxis, the treatment of TN is frequently unsatisfactory. Onabotulinum toxin type A (OBTA) is proven safe and effective in migraine and cranial neuralgias such as occipital neuralgia, representing a possible therapeutic option in TN. **Objectives:** This study evaluated the effects of OBTA injections in a cohort of patients with refractory TN. **Methods:** We enrolled patients with confirmed TN referred to our botulinum toxin outpatient clinic, taking standard oral prophylaxis with partial or no response. The Intensity (estimated with the visual analog pain scale) and frequency of pain (estimated by the number of days with pain per month) were noted before the start of prophylaxis, with prophylaxis alone, and 3 months after OBTA injections. OBTA dosage and number of injection points, as well as other demographic and clinical data, were noted. **Results:** Our preliminary sample of 20 patients was marked by a predominance of females ($n = 11$, 55%), with a median age of 70.5 years (Min 44, Max 87), with idiopathic TN ($n = 12$, 60%). Carbamazepine was the most frequent prophylactic (used by 14 patients, 70% of the sample). Patients reported pain with a median intensity of 10 (Min 9, Max 10) and occurring daily (30 days with pain per month in the entire sample) before prophylaxis. Although prophylaxis was effective to reduce pain intensity, lowering an average of 3.5 points ($p < 0.0001$, IC 95% 2 – 5), it was ineffective to reduce the frequency, and the entire sample maintained daily pain. When compared to prophylaxis alone, OBTA + Prophylaxis further reduced intensity on average by 4.9 points ($p < 0.0001$, IC 95% 3 – 5.9), and reduced frequency on average by 28 days with pain per month ($p = 0.002$, IC 95% 23 – 30). In our sample, 7 patients (35% of the sample) reported a full response. The mean duration of effect was 80.75 ± 20.14 days. No side effects were registered in our sample. **Final Comments:** Our results suggest that OBTA is a safe and effective add-on treatment option for patients experiencing TN refractory to oral prophylaxis. Although OBTA was particularly effective in reducing the frequency of painful episodes, further studies are necessary to confirm these findings.

Cefaleia

TL 1106287

SPONTANEOUS PNEUMORRACHIA: A RARE CAUSE OF STROKE MIMICS

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Case Presentation: Female patient, 39 years old, was admitted to the emergency department of a private Hospital in the city of São Paulo, after a severe headache followed by two episodes of syncope and motor deficit in the left upper limb. Neurological examination showed motor deficit in the left upper limb without sensitive impairment. She had previous history of bipolar affective disorder, chronic anemia, and overweight with previous bariatric surgery. The patient was submitted to computed tomography (CT), arterial angiotomography of the skull and cervical neck, which showed rare small gaseous foci in the perivertebral soft tissues and extradural site on the left side in the craniovertebral transition, near the foramen magnum. Small foci of pneumocephalus were found in the posterior fossa, near the left sphenopetrous fissure. Electroneuromyography (ENMG) of four limbs was performed, with normal results, and brain and cervical magnetic resonance imaging (MRI) performed 48 hours after CT showed no more expression of the changes described on CT. She was treated with analgesia and was discharged with referral to rehabilitation service and outpatient follow-up. **Discussion:** Spontaneous pneumorrhachia is rare disease and is characterized by the presence of air in the spinal canal, both in the intradural and extradural compartments. It is usually benign with spontaneous resolution. Our patient was first suspected as having a stroke, due to the thunderclap headache and focal motor deficit. CT revealed the diagnosis **Final Comments:** Spontaneous pneumorrhachia is a rare cause of stroke mimics. Cervical CT should be carried out in patients with thunderclap headache and radicular pain in an upper limb.

Cefaleia

TL 1106296

ULTRAFast DYNAMIC CT MYELOGRAPHY FOR THE IDENTIFICATION OF LEAKAGE LEVEL IN MULTIPLE MENINGEAL DIVERTICULA

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Case Presentation: A 26 year-old woman with clinical and neuroimaging criteria for low cerebrospinal fluid (CSF) pressure headache was submitted to a CT myelography which identified multiple thoracic meningeal diverticula, but specific site of leakage was not possible due to temporal resolution. Ultrafast dynamic CT myelography was then performed and confirmed opacification of only a T8-T9 right-side meningeal diverticula in the initial phases with later opacification of the remaining diverticula and epidural spaces. Targeted surgical treatment with diverticula coagulation and fat grafting with fibrin glue resulted in symptoms resolution. **Discussion:** Spontaneous intracranial hypotension (SIH) seems to be related to the structural fragility of the dura mater, which promotes, even if due to minor traumas (eg, coughing and sneezing), a meningeal rupture. Cases that are refractory to clinical treatment may be submitted to a blood patch or surgical intervention. Only 50% of patients with SIH improve after the first blood patch. In refractory cases, the ideal is to identify the exact point of CSF leakage for surgical correction. Myelotomography is highlighted here, as it can identify meningeal diverticula, which correspond to points of fragility. Ultrafast dynamic CT myelography is an option in cases where there is more than one diverticulum and if you want to identify the one responsible for the leak. This technique involved positioning the patient in Trendelenburg and application of intradural contrast while the images were obtained. **Final Comments:** Ultrafast dynamic CT myelography is a technique that has been employed mainly in ventral dural ruptures related to osteophytes, but can also be applied in SIH, besides the conventional dynamic myelography.

Cefaleia

TL 1106298

SHORT-LASTING UNILATERAL NEURALGIFORM HEADACHE ATTACKS WITH CONJUNCTIVAL INJECTION AND TEARING (SUNCT) RESPONSIVE TO INTRANASAL LIDOCAINE AND NERVE BLOCKAGE: A CASE PRESENTATION

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Case Presentation: A 75-year-old male patient went to the emergency department due to an intense recurrent and refractory daily headache that initiated in the previous two months, was described as a stabbing, was exclusively right-sided and had maximal duration of 10 seconds, with recurrence at least every 10 minutes and no refractory period. It was mostly perceived in the ophthalmic nerve (V1), with ipsilateral allodynia and autonomic symptoms, such as conjunctival injection, lacrimation and ptosis. The pain was unresponsive to acetaminophen and non-steroidal anti-inflammatory drugs. In his neurological examination, we reproduced the headache's episodes when the right V1 region was lightly touched. Based on the elevated frequency, short duration of attacks and presence of ipsilateral autonomic features, SUNCT diagnosis was made. A treatment regimen of trochlear, supra-orbital, major and minor occipital nerve anesthetic blockades using 2% lidocaine without vasoconstrictor was made in association with a preventive treatment with Lamotrigine 100mg twice a day. Also, he was instructed to do a sphenopalatine ganglion block by slowly dripping 2 mL of intranasal 2% lidocaine in his right nose when symptoms recurred. The patient had no pain for at least six months of these treatments, not needing to use intranasal lidocaine anymore. **Discussion:** Trigeminal autonomic cephalalgias comprehend a myriad of headaches which include the short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) and with conjunctival injection and tearing (SUNCT). Intravenous lidocaine has been consistently described as the treatment of choice for acute episodes, but there is a risk of severe cardiac side effects (arrhythmias), mainly for older adults. Therefore, they can benefit from alternate, less harming and secure treatment options such as intranasal lidocaine drip and nerve blockades in the area corresponding to the pain, like in this case. **Final Comments:** Our SUNCT patient had a very satisfactory response of headache after treatment with the simple and relatively safe procedures previously described, which presented efficient acute pain control within a large follow-up period. Similar acute treatments were reported for SUNCT/SUNA with satisfactory results, but data on this topic are still scarce in the literature, being based mostly on case presentation and small studies, so further studies are mandatory in order to corroborate the findings of this case presentation.

Cefaleia

TL 1106301

HEADACHE SECONDARY TO MENINGITIS DUE TO M. TUBERCULOSIS RESPONSIVE TO NERVE BLOCKAGE: A CASE PRESENTATION

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Case Presentation: A 49-year-old male patient, with migraine since 10 years old, sought medical attention with a new and distinct headache pattern that initiated in the previous two months and had a progressive intensity and frequency. It was described as a high intensity continuous pressure in the right frontal-parietal-occipital topography, with worsening periods of recurrent stabbing pain episodes. Initially, a cluster headache diagnosis was suspected due to possible autonomic features associated and a treatment with a right greater occipital nerve block, high-flow oxygen and sumatriptan when headache recurred was offered and preventive treatment with Verapamil was initiated. Also, laboratory and advanced neuroimaging investigation turned out negative. Two weeks later, he reported improvement in the right side pain but described a new headache with the same characteristics as before on the left side that didn't respond to the prescribed treatment and reported a worsening of pain when lying down in his bed. A left greater and lesser occipital nerves block, trigger points injections in bilateral trapezius and left semispinalis were performed and a lumbar puncture was scheduled. Five days later, the patient had a marked headache improvement, presenting only 3 episodes in 2 weeks, with a mild intensity. Lumbar puncture revealed a mildly elevated opening pressure (270 mmH2O) with significant elevations in cell and protein counts. A PCR test for M. tuberculosis was positive and treatment for this condition was initiated. **Discussion:** A minority of headache patients have a secondary headache disorder and the medical literature promotes red flags to direct the clinician to initiate a workup plan. Sometimes, we may not proceed to further investigation even with a red flag present if the patient's headache presents significant improvement with medical treatment. In this case, our patient had significant response to bilateral occipital nerve blocks but, nevertheless, further investigation was warranted because of the presence of recent onset of new headache and positional headache. **Final Comments:** Neurologists worldwide estimate that 18% of patients with a headache have a secondary headache disorder. This case reinforces that even the improvement of headache with occipital nerve blockage or other treatments must not exempt the need to investigate the existence of a secondary cause of headache, which proved to be crucial for this patient.

Cefaleia

TL 1106391

AN UNUSUAL CAUSE OF HEMICRANIAL HEADACHE: A CASE PRESENTATION

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Case Presentation: A 67-years-old woman presents in an outpatient clinic with a 4 month history of new onset left headache and progressive periorbital pain. Few days before the consultation, she noticed discrete binocular diplopia on rightward gaze. Considering red flags (new onset, age > 40 years, persistent hemicrania) a brain magnetic resonance imaging (MRI) was requested. The MRI shows a round expansive lesion at the medial corner of the left orbit, well-defined margins, measuring 1,5x1,1x1,0cm, extraconal, touching the left rectus medialis muscle inferomedially. The lesion had intermediate signal in T1 and high signal in T2 at its internal portion and diffusion restriction and post-contrast heterogeneous enhancement, predominantly peripherally. Hypotheses of Schwannoma or metastasis were considered for the case. Upon the metastasis hypothesis, new imaging tests were performed. A chest CT revealed a 1,8 cm spiky nodule at the right posterior basal segment of the lung and lymphomegalies compatible with secondary implant, neck CT showed multiple lymphomegalies, abdominal CT showed hepatic nodules suggestive of secondary implants. Blastic image at L1 suggestive of secondary implant. The patient was diagnosed with stage 4 non-small cell lung cancer and referred to oncology service. **Discussion:** Although a minority of patients presenting with headache have a secondary cause, screening for these cases by questioning and evaluating some red flags can be useful to reduce costs, false-positives and/or incidental findings provided by routine use of blood tests and neuroimaging. Considering this purpose, a common mnemonic currently used is SNNOOP10. Eye is a rare site of lung cancer metastasis although metastasis to the eye represents by far the most common form of intraocular and orbital cancer. A review of 42 case presentation of ocular metastasis in lung cancer found that the most common lung cancer that metastasizes to the eye is adenocarcinoma. In this study, ocular symptom was the first referred symptom in 83% (35) cases. The choroid was the most common site for ocular metastasis. **Final Comments:** In our case, recent onset, age above 50 years, neurologic dysfunction and progressive nature warranted further investigation, which was of utmost importance to the diagnosis. We therefore encourage further neurological investigation of patients presenting with headache red flags in order to better establish the diagnosis.

Cefaleia

TL 1106423

PERIPHERAL FACIAL PALSY AS A FALSE LOCALIZING SIGN OF INTRACRANIAL HYPERTENSION: A CASE PRESENTATION

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Case Presentation: A 30-year-old female patient presented with acute, pulsatile, progressive, continuous left occipitofrontal headache, nausea and vomiting. One week later, she started feeling a heavy sensation on right hemiface and low vision in the right eye, and after 1 week in the left eye. She denied diplopia. She used combined contraceptives daily. On physical examination, she had low visual acuity of 20/80 in the right eye and 20/60 in the left eye; fundoscopy showed papilledema; she also had right peripheral facial palsy with lagophthalmos and Bell's sign. CT angiography showed filling failure in the left transverse sinus and in the transition between the sigmoid sinus and the jugular bulb, without signs of venous infarction. Cerebrospinal fluid showed an opening pressure of 50cmH₂O and a closing pressure of 25cmH₂O, with no changes in the CSF laboratory analysis. The diagnosis of intracranial hypertension (IH) secondary to cerebral venous thrombosis (CVT) was defined and full anticoagulation and acetazolamide were initiated. Thus, an etiological investigation of peripheral facial paralysis was carried out; however, CT of the sinuses and temporal bones, as well as the MRI of the inner ear, were normal, concluding as a probable cause of a false localizing signal of IH and CVT. **Discussion:** The pathogenesis of false intracranial localization signals usually occurs in the context of IH with herniation syndromes, with VI nerve palsy being more common in 14% of idiopathic intracranial hypertension (IIH) cases. The finding of VII nerve palsy is uncommon in IH, in a case series of 140 patients with IIH, only 2 cases had VII nerve palsy with a peripheral pattern. As a possible pathophysiology, there may be compression or stretching of the facial nerve in its intratemporal segment close to the facial canal, in addition to reduced drainage of the extensive venous plexus that accompanies the nerve in the facial canal. **Final Comments:** Although rare, the finding of peripheral facial paralysis may have intracranial hypertension as one of the possible etiologies.

Cefaleia

TL 1106487

TRANSVERSE SINUS ANGIOPLASTY (TSA) WITH STENT TO REFRACTORY IDIOPATHIC INTRACRANIAL HYPERTENSION

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Case Presentation: Idiopathic intracranial hypertension (IIH) is a relatively uncommon syndrome, but in recent years its prevalence has been increasing. Our group analyzed data from six patients, mean age of 33.5 years, with refractory IIH treated in a tertiary neurology center. They had poor response to pharmacological treatment and were submitted to a transverse sinus angioplasty (TSA). The time from symptoms onset to interventionist treatment varied from 14 days to 4 years. The general technique was transverse sinus stenting but a sigmoid sinus stent was also implanted in one of the cases. Three patients had pain relief varying between 1 to 2 days after the procedure with drugs subsequently withheld. The other three persisted with pain and it was necessary to maintain medications. Two of them had an immediate response, but the pain returned with the same intensity before the procedure. One of them had headache immediately after the procedure. **Discussion:** Although the IIH physiopathology did not have been completely elucidated and multifactorial mechanisms are proposed, there is a clear association with obesity and female gender. Recently, it has been observed that transverse sinus stenosis is a prevalent finding in these patients, but it is not known if it has a causal relationship with elevation of intracranial pressure or if this finding is a consequence of that. The treatment is based on weight loss and drugs that reduce production or increase drainage of cerebrospinal fluid. The aim is to reduce intracranial pressure, relieve headache and avoid visual loss. TSA with stent has been pointed out as an option for refractory cases in which transverse sinus stenosis with hemodynamic repercussion (pressure gradient greater than 7mmHg) is confirmed by venography. **Final Comments:** In refractory IIH with confirmed dural sinus stenosis, angioplasty with stent placement has been suggested as an effective therapy. Published meta-analyses consider significant symptom relief – improvement of up to 90% of papilledema, 80% of visual symptoms, and 80% of headache. Our case series showed symptom benefits in only 50% of patients. However, in patients who responded to therapy, the response was dramatic and allowed to suspend the pharmacological therapy.

Cefaleia

TL 1106580

SMART SYNDROME – A CASE PRESENTATION

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Case Presentation: P.S.P is a 57 years-old male, with a past history of resection of an ependymoblastoma in the thalamic region requiring adjuvant radiotherapy in 1989. In 2005, he initiated a decline in functionality manifested by cognitive complaints, loss of balance and gait disturbances, with slow and progressive worsening over time. In the past 4 years, he began to experience throbbing headaches with moderate to severe intensity and preceded in 2 to 3 days by auto-limited episodes of aphasia and gait instability. In 2021, he was admitted to our Hospital presenting a migraine-like episode, accompanied by aphasia and right-sided hemiparesis, with prolonged duration. Brain MRI showed T2/FLAIR hyperintensity involving the cortex of the temporal and occipital lobes of the left hemisphere, with gyriform gadolinium enhancement, without water diffusion restriction. Videoencephalogram showed rare epileptiform discharges in the left temporal region and left-sided bursts of frontal rhythmic delta waves. Resonance angiography and blood analysis was unremarkable. The patient was diagnosed with Stroke-like migraine attacks after radiation therapy syndrome (SMART syndrome) and treatment with dexamethasone and analgesics was initiated, with complete resolution of the symptoms in 3 weeks. **Discussion:** SMART syndrome is a rare delayed neurological complication of cranial radiotherapy, manifesting years after the initial treatment. The syndrome usually presents and is diagnosed with recurrent and often reversible episodes of migraine-like attacks, seizures and/or focal neurological deficits, representing transitory dysfunction of the irradiated cortical areas, associated with reversible, transient and unilateral cortical gadolinium enhancement associated with abnormal T2/FLAIR signal changes of the affected brain region. Image control after two months can be useful to assess the resolution of the alterations. Differential diagnosis can be mainly tissue necrosis. Treatment consists of managing symptoms with analgesics, anti-seizure medications and corticosteroids. Typically, the course of the disease is self-limiting. **Final Comments:** The case presented illustrates an extremely rare delayed neurological complication of cranial radiotherapy. SMART syndrome is a radiologic and clinical entity of importance, since its recognition as a self-limited disease prevents potentially iatrogenic diagnostic tests pursuing tumor recurrence and also potentially harmful treatments.

Cefaleia

TL 1104886

DUMBBEL-SHAPED INTRA AND EXTRADURAL HYPOGLOSSAL SCHWANNOMA: CLINICAL PRESENTATION AND MICROSURGICAL TREATMENT IN BRAZIL

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Case Presentation: 51-year-old female presented with history of progressive dizziness for the past 2 years, associated with long date left tinnitus. The patient reported no nausea, gait disturbance, phonation or swallow difficulties. On examination, she was alert and oriented; her left hemi-tongue was atrophic and showed deviation to the left side on protrusion. The palate elevation and uvula contraction were normal, with vomiting reflex preserved, as well as hearing acuity and cerebellar function tests. MRI images showed a solid extra-axial lesion, homogeneous, with gadolinium enhancement, occupying the left lateral clivus and petrous bone, with extension to the left hypoglossal canal and jugular foramen, and also caudal extension. A cerebral angiography was also performed, showing left transverse and sigmoid sinuses stenosis, with predominant contralateral drainage and absence of tumour blush. After proper investigation, the patient was submitted to microsurgical resection of the lesion through a far lateral transcondylar approach. During the surgery it was observed that the tumour eroded the left occipital condyle, which was resected; the tumour had also an intradural extension, which could be removed with preservation of the hypoglossal nerve. It was not noticed any dural sinus involvement. The dura was by the end of the procedure sutured with a galeal patch to cover its defect. Intraoperative anatomopathological analysis demonstrated low grade neoplasia suggestive of Schwannoma, later confirmed by definite immunohistochemical analysis. In the post-operative care, there was no development of any new neurological deficits and the patient noticed improvement of dizziness. The tongue symptoms from hypoglossal paresia persisted. During the later 24 months of follow-up, there was no evidence of tumour recurrence on clinical examination and MRI images. The first case of Hypoglossal Schwannoma was described in 1933, with about one hundred new descriptions in the international literature since then. The proper treatment consists of surgical resection of the tumour as curative option, being the far-lateral transcondylar approach – used in this case – of great value. This approach enables the exposition of vertebral arteries and low cranial nerves, what facilitates the tumour resection with lower risk of damage to physiological vascular and nervous structures.

Distúrbio Vestibulares e do Equilíbrio

TL 1105133

REVERSIBLE BITEMPORAL HEMIHYPOKINETIC PUPIL WITHOUT HEMIANOPSIA: A NEW CHIASMAL SIGN

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Case Presentation: A 38-year-old woman presented with a complain of a two-day right retro-orbital pain and right progressive visual loss. Upon examination, she had a right relative afferent pupillary defect (RAPD) and dyschromatopsia, with normal visual field testing (confrontation) and funduscopic findings. The visual acuity was 0.8 in the right eye and 1.0 in the left eye. The automated perimetry test (APT) demonstrated a mild superior temporal reduction in sensitivity in the field of the left eye and a superior temporal scotoma with diffuse loss of sensitivity (pseudoconstriction) in the right eye. A more thorough pupil examination was performed and it was noted that the pupil finding was not simply a RAPD. When hemifield light stimulation of each eye was performed individually (while occluding the non-tested eye) with a beam flashlight, a peculiar pupil response was noted: right nasal hemiretina light stimulation demonstrated feeble pupil contraction, whereas stimulation of the temporal hemiretina resulted in better pupil contraction. In the left eye, the difference was not as significant as contralaterally, but it was also notable. These findings have been reversed after surgical treatment of the lesion. **Discussion:** In this study, we describe a novel chiasmal sign, reporting a patient in which a pupillary bitemporal hemihypokinesia was not accompanied by hemianopsia and the phenomenon was reversible after the surgical treatment. We hypothesizes that there would be a separation between the afferent visual fibres and the pupillomotor fibres in the chiasm, being the last ones responsible for the referred chiasmal sign (like an 'alarm signal') and superficially distributed in this anatomical structure, similarly to the parasympathetic fibers along the third cranial nerve; they would be, therefore, susceptible to extrinsic compressive lesions (and reversible after decompression). **Final Comments:** We hope that other researchers will be able to address the main question of our study and assess the accuracy of the indicated chiasmal sign in the diagnosis and screening of patients with chiasmal diseases of different aetiologies (e.g., neoplastic, demyelinating, vascular, and inflammatory diseases) by comparing the results of automated methods (e.g., pupillometry, pupillary cycle, retinal ganglion cell analysis) and non-automated methods (flashlight, slit lamp).

Distúrbio Vestibulares e do Equilíbrio

TL 1105198

DANGEROUS HINTS AND NORMAL MRI: DON'T FORGET INFERIOR VESTIBULAR NEURITIS

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Case Presentation: We describe two patients complaining of sudden dizziness in the emergency department whose examination revealed dangerous HINTS but normal image studies. A 52-years-old woman presented with acute vertigo associated with vomiting, tinnitus and severe imbalance. She had left torsional spontaneous and positional downbeat and torsional to the left nystagmus and normal vestibulo-ocular reflex (VOR). A 66-years-old man also presented with acute vertigo, vomiting and imbalance. He had positional downbeat and torsional to the left nystagmus, a normal VOR, felt to the right side with eyes closed when standing on foam, subjective visual vertical was abnormal (2,5°) to the right. Both patients had normal brain MRI and abnormal video-head impulse test (vHIT), showing low gain (0.37 and 0.3 respectively) of VOR for the right posterior semicircular canal gain of the posterior semicircular canal and corrective saccades, receiving the diagnosis of vestibular neuritis of the inferior division of the right vestibular nerve. **Discussion:** The HINTS has a high sensitivity (97%) and specificity (95%) in discerning central causes of acute vestibular syndromes. Nonetheless, some peripheral etiologies could mimic central patterns in the first evaluation. The isolated unilateral involvement of the inferior division of the vestibular nerve may manifest as positional downbeat nystagmus with normal horizontal VOR and must be included in the differential diagnosis in a patient with normal MRI. **Final Comments:** An acute onset vertigo, dangerous HINTS, with downbeat nystagmus and normal brain MRI should remind of vestibular neuritis of the inferior division of the vestibular nerve. The vHIT is of great help to confirm the diagnosis.

Distúrbio Vestibulares e do Equilíbrio

TL 1105847

VERTICAL ONE-AND-A-HALF SYNDROME IN A PATIENT WITH PECHERON ARTERY ISCHEMIA: A CASE PRESENTATION

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Case Presentation: A 78-year-old female patient, presented at the Hospital emergency with approximately 12 hours of sudden onset of diplopia, associated with dizziness. Neuro-ophthalmological examination showed no blinking alterations or ptosis. It also revealed torsional nystagmus associated with bilateral upgaze palsy with limitation of infraduction on the left. Pupillary reflexes were hypoactive bilaterally. No facial nerve paresis was observed. Previous pathological history consisted of type 2 diabetes mellitus and systemic arterial hypertension (SAH). Laboratory exams had no apparent abnormalities. The cardiac examination also had no evident alterations. Brain Computerized Tomography and Magnetic Resonance (MR) angiography were normal. Later magnetic resonance imaging (MRI) showed a restriction of the diffusion of water molecules in the thalamic regions and the mesencephalic tegment more evident in the right rostral region. The diagnosis was made with the combination of an ophthalmoparetic syndrome suggesting a midbrain impairment associated with the MRI results. Treatment protocol included dual platelet antiaggregation with acetylsalicylic acid and clopidogrel. Followed by clopidogrel for 21 days with statin association. At follow-up, partial recovery was achieved after 3 months in physical therapy. **Discussion:** An ischemic stroke of the aPe can cause stupor, agitation, change in behavior, aphasia (dominant side), hemineglect (non-dominant side), and diplopia due to the involvement of the midbrain and interstitial nucleus of Cajal (INC), the largest nucleus of the medial longitudinal fasciculus (MLF). The INC is the neural integrator for vertical eye movement and is involved in vertical gaze, both with saccadic generation and the vestibulo-ocular reflex (VOR). Two different types of VOHS have been described in the literature. The first: Consists of bilateral paralysis of the gaze upwards and monocular paresis of the gaze downwards with injury ipsilateral or contralateral. Paretic look down to the left and injury to the right. This variant has been described in thalamomesencephalic lesions, exactly like our patient. The second one: consists of difficulty to look down in association with monocular elevation paralysis, described in bilateral mesodiencephalic infarctions. **Final Comments:** This report describes a rare type of VOHS and brings a new insight on a possible aPe topography possibly causing this clinical presentation.

Distúrbio Vestibulares e do Equilíbrio

TL 1106621

CERVICAL DYSTONIA ASSOCIATED WITH L2-GLUTARIC ACIDURIA (L2HGA)

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Case Presentation: A 38-year-old female patient, daughter of consanguineous parents, seeks neurological care with complaints of worsening gait and seizures. During childhood, she presented learning disabilities and regression in developmental milestones, besides frequent falls and fainting. From 32 years of age on, he reported repeated episodes of seizures and gradual worsening of gait, requiring a wheelchair. Recently, he complains of episodes of behavioral arrest. On physical examination, he presents cervical dystonia in laterocollis, atetotic movements in the left upper limb, and stereotyped open-closed movements in both hands. Magnetic resonance imaging of the skull showed leukodystrophy and ventricular dilatation. He has a previous diagnosis of Dandy-Walker syndrome. Finally, genomic analysis was performed, with identification of homozygosity in L2HGDH gene and diagnosis of L-2-Hydroxyglutaric aciduria. **Development:** L2HGA is an autosomal recessive neurometabolic disorder caused by a deficiency of L-2-hydroxyglutarate dehydrogenase due to homozygous or heterozygous mutations in the L2HGDH gene. The symptoms are developmental and cognitive retardation, seizures, gait problems, as there are cerebellar findings in symptomatic patients such as ataxia, dyskinesia presented by the patient. However, acute metabolic decompensation and rapid neurological deterioration is usually not common. In patients with L2HGA, worsening gait/balance, altered consciousness and seizures, incipient headache may be indicative of brain tumors. Movement disorders are unusual, atetosis is related to loss of neurons and proliferation of astrocytes in the caudate and putamen, as well as markedly reduced gamma-aminobutyric acid and glutamate decarboxylase activity. Dystonia and dyskinesia, on the other hand, occur due to inhibition of neuronal glutamate decarboxylase by glutaric acid with decreased biosynthesis of gamma-aminobutyric acid. Among the neuroradiological findings, cerebral and cerebellar atrophy as the disease progresses. As for treatment, riboflavin to increase FAD levels and carnitine as regulator of DNA damage. **Final Comments:** The authors describe a rare case of toxic accumulation of L-2-hydroxyglutarate causing movement disorder: cervical dystonia in laterocollis and atetosis in the upper limbs, with impaired quality of life.

Distúrbio Vestibulares e do Equilíbrio

TL 1106631

DYSKINESIAS IN A PATIENT WITH CEREBROTENDINOUS XANTHOMATOSIS

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Case Presentation: Male, 48 years old, seen at a neurology outpatient clinic, attendant reports total aphasia. Since childhood he refers to retarded neuropsychomotor development, besides low weight and seizures. Sister has a similar clinical picture. Presents global ataxia, does not assume orthostasis, axial component, dysmetria and decomposition of the upper limbs bilaterally, asymmetrical to the left. Extended arms maneuver: high amplitude and low frequency tremor, associated with ataxia. He has dyskinetic movement of head, trunk and limbs, axial predominance in the neck region and spontaneous nystagmus. He has nodules in joints, suggestive of tendinous xanthomas. Symmetrical reflexes bilaterally in upper limbs and exalted in bilateral patellar, bilateral flexor cutaneous-plantar reflex, unresectable glabellar. Magnetic resonance imaging (MRI) of the skull showed intra-axial, non-expansive, bilateral, symmetrical lesions in the white matter of the cerebellar hemispheres, associated with hyperintense areas on T2/FLAIR in brainstem and basal nuclei, suggestive of cerebrotendinous xanthomatosis (CTX) when associated with clinical. Patient does not use levodopa. **Discussion:** The (CTX) is a rare autosomal recessive disease caused by mutations in the CYP27A1 gene, leading to absence of the mitochondrial sterol 27-hydroxylase, responsible for cholesterol metabolism in the bile synthesis pathway. Thus, there is accumulation of cholesterol in tissues, including the brain, leading to progressive neurological dysfunction marked by dementia, epilepsy, hyperreflexia, spasticity, movement disorders such as parkinsonism, dyskinesias, and cerebellar ataxia. In other tissues it causes tendon xanthomas, atherosclerosis and diarrhea. Cerebellar ataxia and tremor are justified by lesions in the dentate nuclei, extending into the surrounding white matter of the cerebellar hemispheres. Trunk-members dyskinesia can be explained by neurodegenerative theory, neurotoxic mechanisms, such as the production of lipid peroxidation, with excess cholesterol in CTX and free radical formation, leading to a higher rate of dopamine neuron loss due to lipid accumulation in the substantia nigra. Furthermore, CTX is a genetic disease, it can be associated with mutations in the CYP2D6, the dopamine D2 and D3 receptor genes, and the serotonin 2A and 2C receptor genes that cause dyskinesia. **Final Comments:** CTX with dyskinetic and ataxic movements that are unusual in the presentation of this disease.

Distúrbio Vestibulares e do Equilíbrio

TL 1104805

**UNUSUAL MICROBLEEDS CRITICAL ILLNESS
BRAIN MRI OF COVID-19 PATIENT**

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Case Presentation: Covid-19, initially described as a respiratory system's infection, is currently more and more recognized as a multiorgan disease, including neurological manifestations. We are reporting the imaging findings of the rare entity of critical illness-associated cerebral microbleeds in a COVID-19 45 years old woman with hypoxic respiratory failure, who was eventually intubated and ventilated, prolonged coma. She was tested positive for COVID-19 confirmed by reverse transcriptase polymerase chain reaction. The purpose of this study is to describe findings, in the form of cerebral microbleeds affecting different brain structures, observed in MRIs of critically ill patients. Multiple scattered cerebral microhaemorrhages diffusely distributed in the juxtacortical white matter and internal capsule region, sparing the deep and periventricular white matter, basal ganglia, thalami and cortex were seen. Cerebral microbleeds and leukoencephalopathy have been described in COVID-19 patients; although the mechanism remains unknown, possibilities include endotheliitis with thrombotic microangiopathy, excessive inflammation, prolonged respiratory failure, and hypoxemia.

Doença Cerebrovascular

TL 1104809

**CERVICAL PAIN AND HYPOGLOSSAL PALSY:
A CERVICAL ARTERIAL DISSECTION SIGN**

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Case Presentation: A 42 year old man came to the emergency room presenting a sudden neck pain, on the left side, that had started while he was jogging. Soon after, he felt spasms and difficulty moving his tongue inside the mouth, he denied any Other symptoms. On the physical examination there was no sign of lesion in the pyramidal tract. Soon after being admitted, he underwent an AngioCT of the head and neck, that showed a dissection of the internal carotid artery, in its nasofaringeal portion, where it has close contact with the 4th segment of the XII cranial nerve, compressing it. Although it is the most affected cranial nerve in these lesions, 5% of the cases, this presentation is uncommon. For any patient with sudden neck pain, with associated XII nerve palsy it is imperative to investigate the possibility of carotid dissection, considering a dissection in the vertebral arteries as a differential diagnosis.

Doença Cerebrovascular

TL 1104833

**MOYAMOYA DISEASE PRESENTING
AS CEREBRAL HEMORRHAGE: CASE
PRESENTATION**

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Case Presentation: Female, 41 years old, complained of sudden headache, vomiting and posterior neck pain two days ago, evolving with impaired level of consciousness and sphincter release. Past medical history: obesity and hypertension. Physical examination: somnolent, responsive to commands, pupils equal round and reactive to light, hemiplegia and hypoesthesia in the left lower limb, Kernig and Lasègue's signs positive. CT and CT-angiography: newly subarachnoid hemorrhage in the supra and infratentorial cisterns; hematoma in the right thalamus and intern capsule filling all the four ventricles; diffuse parietal irregularity of arteries from carotid system, with multiples areas of stenosis in filiform flow in the distal segments of the middle and anterior cerebral arteries. Cerebral angiography: moyamoya pattern in the carotid systems. **Discussion:** The evidences of the subarachnoid hemorrhage and distal arterial irregularities from carotid system suggested vasculitis or vasospasm, needing complementation with cerebral angiography that showed stenosis of the distal supraclinoid internal carotid arteries with subocclusion/occlusion of the carotid "T" and hypertrophy of the lenticulostriate arteries and of the cisternal piercing branches compatible with 'moyamoya' pattern. **Final Comments:** Only 10 percent of the cases of Moyamoya Disease (MMD) presents as intracerebral hemorrhage. The most often initial presenting is the ischemic stroke. In patients with cerebral hemorrhage the bleeding occurs in deep structures such as the basal ganglia, thalamus and/or the ventricular system. Cortical/subcortical bleeding is less frequent. The patient above presented cerebrovascular syndrome and images with a specific pattern compatible with the description from literature for MMD.

Doença Cerebrovascular

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REVERSIBLE ISCHEMIC NEUROLOGICAL DEFICIT CAUSED BY S-PROTEIN DEFICIENCY ASSOCIATED WITH THE PRESENCE OF PATENT FORAMEN OVALE: A CASE PRESENTATION

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Case Presentation: K.R.G.R., a 36-year-old female patient presented with sudden aphasia, dyslalia and acalculia. She had complete remission of these symptoms in about 10 hours, but remained verbalizing with paraphasia for more than a week, and then evolved completely asymptomatic. The adopted conduct was, in the first two weeks, treatment with acetylsalicylic acid 300mg a day, request for investigation tests (electrocardiogram, Holter, transthoracic echocardiogram, transesophageal echocardiogram, Doppler of the carotid and vertebral arteries, chest X-ray in PA and profile and cranial MRI) and ambulatory monitoring of blood pressure. Serum tests revealed a protein S deficiency and presence of hyperactivated factor VIII, possibly related to the use of oral contraceptives. The conclusion is that the patient had suffered a left parietal cerebral ischemia, with remitted deficit, caused by an embolism originating from a state of transitory hypercoagulability and favoured by the presence of a patent foramen ovale (PFO), confirmed by echocardiography. The patient started oral anticoagulation with warfarin 5mg a day, and started using an IUD, no longer using oral contraceptives, with normalization of protein S and factor VIII. Afterwards the PFO was surgically occluded, and she started taking acetylsalicylic acid 100mg a day. **Discussion:** Protein S is a cofactor for the cleavage of coagulation factors Va and VIIIa and its deficiency predisposes to the formation of venous or arterial thrombi, which when moving lead to the possibility of ischemic strokes. When the phenomenon lasts beyond 24 hours with subsequent improvement it is said to be a reversible ischemic neurological deficit. Another possible etiology for an ischemia is the persistence of communication between the cardiac atria, especially when there is considerable size of this communication, a large amount of shunt between the chambers and the presence of an aneurysm or redundant membrane in the interatrial septum. **Final Comments:** The association of diverse etiological pathways may be understood as being of importance, both for sharing the study, as well as for analysing the evolution of this patient profile and choosing the best conducts to be taken, since treatment ends up necessarily developing on two distinct fronts: anticoagulant drug therapy and surgical intervention.

Doença Cerebrovascular

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PRES OR NOT PRES?: ATYPICAL MRI LESIONS IN A MALE ADULT WITH HYPERTENSIVE ENCEPHALOPATHY

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Case Presentation: A 44-year old man with uncontrolled hypertension presented to the emergency department with severely increased blood pressure and altered mental status in the past two hours. At admission, he had acute hypertension (220/110 mmHg) and impaired kidney function. He was uncooperative, agitated and confused. Head CT without contrast showed only mild effacement of cortical sulci, suggesting edema. A fundoscopic examination showed hypertensive retinopathy with retinal hemorrhage. A presumptive diagnosis of hypertensive encephalopathy was made and high doses of intravenous nitroprusside were immediately initiated for blood pressure control. Cerebrospinal fluid and electroencephalography were completely normal. After five days, the blood pressure was well controlled and the patient recovered without any neurologic deficits. Brain MRI performed one-week after admission symmetric enlargement and increased signal on T2-weighted images of the basal ganglia and base of the pons without restricted diffusion. Contrast was not used due to chronic kidney disease. A control MRI after three-months showed complete resolution of the aforementioned neuroimaging findings. **Discussion:** This patient showed classical symptoms of posterior reversible encephalopathy syndrome (PRES), however, atypical MRI findings led us to reconsider the initial diagnosis and contemplate differential. Typically MRI findings of PRES, seen in almost 98% of cases, include symmetric and bilateral hyperintense foci in the parieto-occipital subcortical regions on FLAIR/T2-weighted images. The imaging findings in this case could be classified as a central variant PRES, in which lesions are predominantly observed in the basal ganglia and brainstem, sparing subcortical and cortical regions. Differential diagnosis need to be considered in this case and infarction, osmotic demyelination, autoimmune diseases, infectious encephalitis and metabolic encephalopathies were discarded because MRI pattern with normal laboratory findings and clinical full recovery without deficits using only antihypertensives. **Final Comments:** PRES is a rare and dangerous condition in which rapid diagnosis and treatment are essential. The diagnosis of atypical PRES may be challenging then the condition could be mistaken for other pathologies because the symptoms are nonspecific and the radiologic findings are rare, especially when lesions are located in central or infratentorial regions.

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OCULAR TILT REACTION AND VERTICAL GAZE PALSY DUE THALAMIC STROKE – A CASE PRESENTATION

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Case Presentation: A 70-year-old-man with uncontrolled hypertension presents at emergency department with sudden diplopia and dysarthria, initiated at home 10 hours late. Neurologic exam showed an upbeat nystagmus, vertical diplopia, vertical supranuclear gaze palsy with normal vestibule-ocular reflex, normal horizontal gaze, mild dysarthria, left head tilt (Bielschowsky sign), right eye with hypertropia, excyclotorsion of the left eye which was in hypotropia and a comitant skew deviation. There was no more neurological deficits. Fundoscopic exam confirmed the contraversive ocular tilt reaction. A brain MRI study showed acute right paramedian-thalamic infarction. The etiology was atheroembolism and patient received aspirin and statin. **Discussion:** Vertical gaze palsy with normal vestibule-ocular reflex suggests supranuclear lesion and probably located at the mesencephalic vertical gaze center. However, this topographic association cannot preserve cranial nerves and at the same time explain a comitant skew deviation, head tilt and the excyclotorsion of the eye in hypotropia, causing a contraversive ocular tilt reaction. There are graviceptives pathways responsible for vestibular tonus (vestibular thalamus) connected with the interstitial nucleus of Cajal, posterior commissure, interstitial nucleus of medial longitudinal fasciculus and Darkschewitsch nucleus (these structures are the vertical gaze center located at mesencephalon). These graviceptives pathways are located at thalamic nuclei of ventral posterolateral nucleus and probably were lesioned by the right paramedian stroke reported in this case. **Final Comments:** There are so many different clinical presentations of neurovascular syndromes of thalamus. The clinical and anatomical correlation is difficult and neurologic exam is crucial to topographic diagnosis. This case presentation aims to get familiar to neurologists a rare presentation of a common disease, for which rapid diagnostic and treatment are important to better patient outcome.

Doença Cerebrovascular

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SIMULTANEOUS BILATERAL HYPERTENSIVE THALAMIC HEMORRHAGIC – A VERY RARE PRESENTATION

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Case Presentation: A 75-year-old woman, hypertensive, diabetic and smoker with irregular treatment, was found unconscious and with signs of sphincter release at home. She was last seen well at 10 am and was admitted at Hospital at 11 pm. In the emergency room she had blood pressure 224/127 mmHg, blood glucose 133 mg/dL, SpO2 98%, 3-points Glasgow Coma Scale, miotic pupils. Her non-contrast computed tomography showed bilateral thalamic hemorrhage. Cranial CT angiography showed no vascular malformations and cranial magnetic resonance imaging showed age-matched bilateral hemorrhage, in addition to deep cerebral microbleeds, bilateral periventricular microangiopathy and ischemic lesions with lacunar aspect of indeterminate date. She was managed conservatively and at the time of publication of this case she is still in a comatose state with a 7-point Glasgow Coma Scale. **Discussion:** Hemorrhagic stroke has an estimated prevalence of 12-15% of all strokes. The most common etiologies include amyloid and hypertensive angiopathy. Considering hypertensive angiopathy, the main sites of hemorrhage are the basal ganglia, thalamus, pons and cerebellum. Hemorrhage in the thalamus represents less than 25% of hemorrhagic strokes and the simultaneous involvement of these structures is even rarer. To date, there are less than 15 case presentations worldwide of bilateral thalamic hemorrhage. The pathophysiology is still uncertain but is related to the rupture of microaneurysms or perforating arteries chronically injured by uncontrolled hypertension. **Final Comments:** Our patient had an atypical presentation of bilateral thalamic hemorrhage. It is necessary to exclude important differential diagnoses such as cerebral venous thrombosis with hemorrhagic infarction, vascular malformations and even ischemic stroke with hemorrhagic transformation for the treatment be performed properly. Some clues that may help in the diagnosis include the spectrum of small vessel disease such as periventricular microangiopathy, deep cerebral microbleeds and previous ischemic lacunar lesions. We hope that this case presentation can help other colleagues in the future who are faced with the same radiological images.

Doença Cerebrovascular

TL 1105263

INTRACRANIAL DURAL ARTERIOVENOUS FISTULA: CASE PRESENTATION

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Case Presentation: Male patient, 40 years old, admitted with a headache for 1 week, progressive, weighty, holocranial, associated with bilateral pulsatile tinnitus, without suggestive ICH. He progressed to a generalized tonic-clonic seizure with sphincter release followed by loss of consciousness in July/2021. He was diagnosed with SAH in the frontal and right temporal regions by CT and CTA, and failure to fill the sagittal venous sinus after contrast administration. He denied any comorbidities or medication use. Referred to ICU with no return of consciousness. A cranial CT was redone one week later revealing the resolution of SAH. Arteriography was indicated to search for an aneurysm, however, due to lack of clinical control, the patient was not submitted. The patient improved and was discharged. A hematologist diagnosed thrombosis and indicated anticoagulant therapy (dabigatran 150mg). After treatment, partial recanalization of the superior sagittal sinus was presented by CTA with thrombolysis resolution and an improvement of headache, but with tinnitus maintenance. The differential diagnosis was cerebral venous thrombosis or intracranial dural arteriovenous fistula (IDAF). New arteriography revealed the presence of IDAF in the torcula and left transverse/sigmoid venous sinuses, Cognard grade I, with an absence of venous reflux; coupled with partial venous thrombosis of the superior sagittal and left transverse/sigmoid sinuses. **Discussion:** IDAFs represent 15% of intracranial vascular malformations, frequently occurring between 50-60 (age). They may present in a diverse symptomatic manner, ranging from less aggressive manifestations (e.g., headache and tinnitus) to more aggressive ones (e.g., progressive cognitive decline) determined by the venous drainage pattern and the location of the fistula. These fistulas mainly occur in the cavernous, transverse, and sigmoid sinuses, and the cerebral tent. The main way of classifying IDAFs is according to Cognard types I and IIa (benign) and IIb, IIa + b, III, IV and V (malignant). The benign ones drain directly into the venous sinuses, while the malignant ones have direct or indirect reflux into the cortical veins. CT, CTA, and MRI are the first-line diagnostic tools, but DSA has established itself as the gold standard for a suspected IDAF. **Final Comments:** The established relationship between IDAF with thrombosis and the possibility of hemorrhagic transformation therefrom obscures the cause-consequence relationship.

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OPALSKI'S SYNDROME AND COVID-19: CASE PRESENTATION

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Case Presentation: A 64-year-old male patient was admitted at the Emergency Department in January 2022 with history of sudden onset severe headache followed by vomiting, right side hemiparesis and ipsilateral sensory loss. He didn't receive thrombolytic therapy and showed progressively improvement of the symptoms. After two days, he developed sudden onset right hemiparesis, dysarthria, dysphagia and gait instability. The patient had a background of hypertension and epilepsy. Furthermore, the patient had symptoms of cough and fever three days previously addition and was diagnosed with COVID-19 at the Hospital based on an antigen test. Neurological examination showed right side facial weakness, ipsilateral miosis and touch sensory loss. Right side arm and leg paresis was present associated with left side pain and temperature loss in trunk and limbs. He also had axial ataxia and gait instability. Characterizing a rare variant of the lateral medullary syndrome: Opalski's syndrome. MRI revealed an acute medullary infarction on the right dorsolateral portion. Routine blood tests were normal, as were the cerebrospinal fluid, electrocardiogram and echocardiogram. CT angiography revealed hypoplasia of the right vertebral artery. The patient was managed conservatively, received secondary prophylaxis with aspirin and statin, physiotherapy and speech therapy at the stroke unit. He was referred for outpatient follow-up. **Discussion:** Opalski's syndrome is a rare variant of Wallenberg's syndrome. It is usually caused by vertebral artery occlusion, compromising blood flow to the lateral region of the medulla, caudal to the pyramids decussation, leading to involvement of corticospinal and spinothalamic tracts, autonomic fibers and cranial nerves. The classical presentation is ipsilateral hemiplegia or hemiparesis, dysarthria, dysphagia, ipsilateral nystagmus, vertigo, hypoesthesia in the ipsilateral hemiface and contralateral sensory loss in trunk and limbs. It's relevant to be familiar with the syndrome as the myriad and bilateral distribution of symptoms could lead to difficulties in topographic localization. It's also worth mention that the recent COVID infection and hypertension were risk factors for the stroke event. **Final Comments:** Thus, in face of sudden ipsilateral hemiparesis and cross-sensitivity alterations, this syndrome should be considered and appropriate treatment should be promptly initiated, considering the impact on survival and prevention of disabling deficits.

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TL 1105338

ALICE IN WONDERLAND SYNDROME DUE TO CHRONIC CEREBRAL VENOUS THROMBOSIS – A RARE CONDITION

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Case Presentation: A 44-year-old man was admitted to the emergency department with sudden seizures and severe headache for 3 days. He had a history of pulsatile tinnitus for 2 years. The patient reported intermittent symptoms of visual illusion lasting about 10 seconds, characterized by self-perceived distortions of his body and people's faces, consistent with metamorphopsia. He also described oscillation in the size of objects suggesting micropsia and macropsia. Physical examination showed proptosis of the right eye. Fundoscopy showed no macular, vascular and optic nerve changes. Cranial computed tomography and angiotomography were performed, which suggested venous infarction and sinus thrombosis, respectively. The electroencephalogram indicated focal base epileptogenesis of the right occipital lobe due to a structural lesion. Treatment with 1500mg/day of valproic acid and 20mg/day of clobazam was initiated with complete resolution of metamorphopsia, including macropsia, and micropsia. Later, cerebral arteriography confirmed thrombosis of the superior sagittal sinus, transverse and sigmoid sinuses on the right. In addition, a tentorial dural arteriovenous fistula nourished by posterior branches of the middle meningeal artery and right occipital artery with drainage to cortical veins was visualized. Embolization of the arteriovenous fistula was performed, and apixaban 10 mg/day was started. The patient evolved with complete remission of headache and pulsatile tinnitus. **Discussion:** Metamorphopsia is a misperception of the size of objects or body parts due to migraine, epilepsy, intoxication, or infectious cerebral diseases. They are referred to as Lilliputian hallucinations and were described as Alice in Wonderland Syndrome (AIWS). In the present case, the patient exhibited this syndrome due to focal epileptogenesis secondary to a venous infarction caused by a chronic cerebral venous thrombosis. To our knowledge, there is only one case of AIWS related to cerebral thrombosis reported in the literature. **Final Comments:** AIWS is a rare condition caused by severe illness. Therefore, the early diagnosis is fundamental, and the treatment also needs to be directed at underlying diseases. This case presentation aims to familiarize neurologists with this uncommon condition.

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EPILEPTIC SEIZURES AS STROKE MIMICS AND CHAMELEONS: A CHALLENGE IN THE EMERGENCY ROOM

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Case Presentation: A – J.C.R., a 58 year-old male, was admitted with sudden onset global aphasia, without no other neurological deficits. An unprecedented focal motor onset seizure, with secondary generalization, was witnessed at home. After admission to the emergency room, he underwent a cranial tomography, which didn't show ischemic or hemorrhagic lesions. Intravenous thrombolysis was performed. After 24 hours, a new tomography still showed no signs of acute ischemic stroke. The patient remained with isolated aphasia and no new seizures were witnessed. An electroencephalogram showed a periodic pattern between 1-1,5 Hertz compatible with continuum interictal-ictal. He completely recovered from his language deficit with antiseizure medication. B – T.M.B.S., a 39 year-old female, was admitted with a history of unprecedented tonic-clonic generalized seizure with subsequent transient left hemiparesis, which lasted about 1 hour, and completely remitted. Initial cranial tomography didn't show any alterations and she was sent back to her original healthcare service for observation. She returned, 48 hours later, with left hemiparesis and dysarthria. A new tomography showed a subacute ischemic lesion in the territory of the right middle cerebral artery. **Discussion:** This poster reports two cases of relevance for the study of acute ischemic stroke and epilepsy. The presence of unprecedented seizures with focal neurologic deficits may cause diagnostic doubt, as well as in the indication of thrombolytic therapy. In the first case, the symptom of aphasia in a patient who was actually in non-convulsive status epilepticus mimicked an acute ischemic stroke. The diagnosis of a "Stroke Mimic" was only made later, with the result of the electroencephalogram. In the second case, a young woman had a transient ischemic attack that was mistaken for Todd's paralysis after a generalized seizure. 48 hours later, her case was found to be a "Stroke Chameleon". **Final Comments:** Stroke mimics and chameleons represent a great challenge in every-day neurological practice. Approximately 5% of ischemic strokes present with seizures. Therefore, it is reasonable to proceed with thrombolytic therapy, in these cases, if evidence suggests that residual impairment is secondary to stroke and not postictal phenomenon. Current evidence shows that, when in doubt about a stroke mimic, intravenous thrombolysis should not be delayed, because the risk of symptomatic intracranial hemorrhage, in these cases, is quite low.

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HYSTOPLASMA CAPSULATUM: AN UNUSUAL ETIOLOGY OF CENTRAL NERVOUS SYSTEM VASCULITIS

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Case Presentation: A 47-year-old woman who had been investigating Systemic Lupus Erythematosus (SLE) was admitted to the Emergency Room with acute onset right hemiparesis and hemihypoesthesia. Computed tomography scan showed a left thalamic hypodensity compatible with a subacute ischemic stroke, along with a lacunar ischemic lesion in the right internal capsule. Cerebrospinal fluid (CSF) showed lymphocytic pleocytosis, consumed glucose and elevated protein levels. The etiologies considered were autoimmune and infectious disorders. Exams to rule out the second one were negative. A cranial magnetic resonance imaging (MRI) showed more ischemic lesions and diffuse arterial narrowing. The rheumatology team considered active arthritis as well, so an autoimmune mechanism was considered the most likely. Due to progression of lesions and the potential gravity of this condition, she was submitted to a course of methylprednisolone and cyclophosphamide. Her initial response was good and she was discharged a few days later. After one week, she started with fever, a novel right-sided oculomotor nerve paresis and mental confusion. Cranial MRI showed recent ischemic events on the midbrain, pons and right cerebellar hemisphere. CSF analysis evidenced worsening of the previous findings with a high opening pressure. A new screening for infectious causes was positive for histoplasmosis, with a counterimmunoelectrophoresis for *Histoplasma capsulatum* titer of 1:16 on CSF and blood, with negative cultures. In view of the hypothesis of CNS histoplasmosis leading to vasculitis and ischemic strokes, antifungal treatment was started. She developed generalized seizures and progressive somnolence, with needing for orotracheal intubation. During her prolonged Hospitalization, she had multiple infectious and clinical complications. Subsequent radiological studies and CSF analysis evidenced poor improvement. Months later, she was discharged with many neurological sequelae, with a plan of prolonged oral antifungal treatment. **Discussion:** CNS vasculitis is rarely caused by SLE. Infectious causes must be remembered due to their prevalence, especially in immunocompromised patients. Our patient had radiologic findings previously described in CNS vasculitis caused by *Histoplasma*, as the main involvement of small vessels of posterior circulation. However, sometimes it is necessary to ponder empirical immunosuppressive treatment because of clinical worsening and sequelae risks, like in our case.

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POST-COVID CEREBRAL VENOUS THROMBOSIS COMPLICATED TO DURAL ARTERIOVENOUS FISTULA ASSOCIATED TO AUDIBLE PULSATILE TINNITUS WITHOUT STETHOSCOPE

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Case Presentation: A.O.S., 36-years-old, attended on 09/2020 with a complaint of intense left hemispheric headache for 01 week associated to nausea, dizziness and blurred vision. On 10/2020, after refractoriness to sintomatic treatment, a neuroimage identified cerebral venous thrombosis (CVT) in the superior sagittal sinus and partial in the transverse and sigmoid sinuses, greater on the left. The patient was treated with enoxaparin full anticoagulation and then converted to warfarin for 6 months. An extensive laboratory etiological investigation was done but it's normal. He had history of Covid-19 infection two months ago. On 04/2021, an image control was performed and showed chronic CVT in the middle and posterior thirds of the superior sagittal sinus, and in the left transverse and sigmoid sinuses. On 07/2021, the case evolved to left pulsatile tinnitus that worsened when lying down, audible on ipsilateral periauricular auscultation and sometimes audible without a stethoscope aid. A new cerebral angiography was performed on 09/2021 and revealed a dural arteriovenous fistula (dAVF) in the sigmoid and left transverse sinuses. It was embolized on 11/2021, with tinnitus resolution. **Discussion:** dAVF can be congenital, idiopathic or acquired. When acquired, its main etiologies are traumatic brain injury, otomastoiditis, CVT, previous craniotomy and pregnancy. It is a rare complication of CVT, evolving in only 1% of cases. It is classified into low and high grade. Low-grade ones (Borden I; Cognard I and IIa) are usually not indicated for surgery, except in cases of secondary glaucoma, refractory orbital pain, and pulsatile tinnitus. High-grade ones (Borden II and III; Cognard IIb, IIa+b, III, IV and V) must be treated early due to of bleeding high risk. This case becomes particularly rare due to the high possibility of it has been triggered by the post-covid prothrombotic state. **Final Comments:** This is a case of CVT probably related to a post-covid prothrombotic state, which evolved after 10 months with dAVF associated to intense pulsatile tinnitus, resolved after embolization. The case deserves to be highlighted due to the rarity of its evolution, as well as the association with an unusual clinical sign of spontaneously audible pulsatile tinnitus, without a stethoscope aid.

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TL 1105421

MARANTIC ENDOCARDITIS LEADING TO STROKE IN AN ONCOLOGIC PATIENT ON FULL ANTICOAGULANT DOSE: WHATS LEFT TO PREVENT NEW EVENTS?

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Case Presentation: We present the case of a 79 year-old woman on treatment for a metastatic lung adenocarcinoma diagnosed upon bilateral pulmonary thromboembolism. She was started on apixaban 5mg BID and osimertinib. Six months later, she developed a severe stroke caused by an occlusive thrombus from right ICA to M1. She was submitted to mechanical thrombectomy, leaving a residual M1 occlusion, with partial recanalization. Transthoracic ecocardiogram showed mobile filamentar wooden images adhered to the right coronary and mitral valves, generating moderate reflux. Laboratory workup showed a hemolytic microangiopathic anemia with marked increase in D-dimers levels. Blood cultures were negative. She was followed with enoxaparin 1mg/kg BID, with no further vascular events. **Discussion:** One in seven ischemic stroke patients can host occult cancers, with up to 20% of these being discharged as Embolic Stroke of Undetermined Source (ESUS). Marantic endocarditis (ME) is a proposed mechanism leading to stroke in oncologic patients. It is a rare condition and is strongly associated with lung adenocarcinoma. It usually damages healthy valves, leading to the formation of high-risk embolic thrombi. Permanent anticoagulation is warranted and surgical approaches are reserved for early disease in younger patients. Secondary prophylaxis with new direct oral anticoagulants (DOACS) lacks substantial evidence, with some studies citing edoxaban 60mg as a potential player. Vitamine K antagonists (VKA) are more accessible, but lack in patient safety due to interactions with chemotherapy plus higher bleeding rates. Enoxaparin is the most recommended by specialists, targeting final pathways in the coagulation cascade, but carries inconvenient logistics and administration. Since no absolute recommendation on the topic has been proposed, choosing an anticoagulation strategy in these patients involves social, economic and technical issues as well as patient education. **Final Comments:** Despite full dose anticoagulation, our patient still developed a major embolic event, highlighting the interface between cancer inflammation and thrombosis. Preventing vascular events in oncologic patients poses a true challenge, since their hyper coagulable state seems to be multifactorial and dynamic. Future studies should approach better stroke prevention strategies, keeping up with the current therapies that are improving cancer mortality and morbidity.

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LOBAR INTRAPARENCHYMATOUS HEMORRHAGE SECONDARY TO AMYLOID ANGIOPATHY IN A YOUNG FEMALE: A CASE PRESENTATION

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Case Presentation: This case refers to a 44-years old female patient, who presented sudden paresis of the right inferior limb (RIL) associated with frontoparietal lobar intraparenchymatous hemorrhage in the left corticospinal region, and also hypodense areas in the white matter. In the propaedeutic, it was performed Brain MRI, it was visualized multiple foci of microbleedings in the cerebral and cerebellar parenchymas, with alteration of white matter sign and recent hematoma in the high left convexity associated with perivascular spaces. With these findings in the imaging, compatible with small vessel disease (SVD), it was widened the investigation to possible inflammatory etiologies (vasculitis, inflammatory amyloid angiopathy) with performing of lumbar puncture to liquor study and it was initiated pulsotherapy with methylprednisolone for five days. It was not detected any liquor alterations. There was partial recovery of the deficits and the patient was discharged with ambulatorial follow-up for etiology investigation. Nearly one year after the first event, she presented a new neurologic deficit with complete right hemiparesis and right hypoesthesia, right homonymous hemianopsia, and aphasia. Brain Tomography evidenced heterogenous lobar intraparenchymatous hemorrhage in the high left frontal convexity, and also demonstrated multiple foci of microbleedings with lobar distribution, characterizing possible cerebral amyloid angiopathy, by the Boston Criteria. **Discussion:** Cerebral Amyloid Angiopathy is a relevant etiology of Lobar Intraparenchymatous Hemorrhage (LIH) and characterizes as deposits of beta-amyloid in small and medium brain blood vessels, being strongly associated with age. Its prevalence is higher in patients older than 65 years old and, in its occurrence in young ones, the medic must hypothesize inflammatory and hereditary causes. **Final Comments:** The investigation of LIH in young patients must be performed, as this is one of the most common causes of cerebral venous thrombosis, arteriovenous malformations, and vasculitis. The occurrence of amyloid angiopathy in these patients must have the inflammatory and hereditary origins investigated and the institution of therapeutic measures, as well as the control of risk factors is of extreme importance to avoid the recurrence of bleeding, which could implicate in the loss of functionality and worsen the individual life quality.

Doença Cerebrovascular

TL 1105455

THE FUNCTIONAL IMPAIRMENT OF GERSTMANN SYNDROME AFTER A SUBACUTE STROKE: A CASE PRESENTATION

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Case Presentation: Female patient, 42 years old, incomplete elementary school, black, obese, with hypertension, type 2 diabetes mellitus and depression. Family history of cardiovascular disease, father with a history of 5 previous ischemic events and deceased from acute myocardial infarction. Followed up at the neurology outpatient clinic due to sequelae of ischemic stroke. She presented the onset of focal symptoms with right hemiparesis, memory loss and dysarthria, however, she was managed as an anxiety crisis in the emergency room. After 3 months, she presented with a sudden severe headache, which was then referred to the tertiary center for imaging tests. Cranial computed tomography showed a hypodense cortical lesion in the frontal and left parietal lobes. Cranial magnetic resonance angiography showed signs of ischemia in the left periorbital cortex. The two ischemic events had an atheroembolic origin. The patient has a deficit in spatial and semantic memory and impairment in instrumental activities of daily living. For functional rehabilitation, she was referred to our institution. On physical examination, patients scored 18 points on the Mini Mental State Examination, with acalculia, agraphia, digital agnosia and right-left disorientation, characteristic findings of Gerstmann Syndrome. In the somatic examination, homonymous hemianopia on the right and paresis in the right lower limb grade IV were observed. **Discussion:** Gerstmann Syndrome is characterized by digital agnosia, right-left disorientation, dysgraphia and acalculia and its development can be caused by different etiologies, such as neoplasms or stroke. The presence of the symptom tetrad of this condition is rare, so having all the associated symptoms is not necessary for the diagnosis. It is noteworthy the persistence of the patient's functional disability, mainly related to cognitive functions, even after the work of the multidisciplinary team for her rehabilitation and motor improvement. **Final Comments:** Recognition of this syndrome is essential to understand which cognitive deficits are impacting the patient's functionality and to design a better rehabilitation strategy.

Doença Cerebrovascular

TL 1105461

ACUTE OCCLUSION OF CENTRAL RETINAL ARTERY DIAGNOSED BY TRANSCRANIAL DOPPLER (TCD): A CASE PRESENTATION

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Case Presentation: 70-years old female, previously with pre-diabetes, systemic arterial hypertension, and hypothyroidism. Presented acute amaurosis in the right eye, being evaluated by an ophthalmologist, who hypothesized retina detachment. On the following day, she was admitted to our service, a tertiary Hospital with an Emergency room and Neurology. In the admission, she presented NIHSS 0. Ophthalmoscopy demonstrated indirect evidence of cherry-red spot and applied retina. Brain Angiotomography was performed, which was normal. Transcranial Doppler (TCD) with transorbital window was performed, demonstrating Right Central Retinal Artery (R-CRA) with a residual pattern, detecting only isolated systolic peaks, findings compatible with occlusion of the central retinal artery (OCRA). It was initiated with secondary prophylaxis with acetylsalicylic acid 100mg per day and simvastatin 40 mg per day. Exams were performed to investigate the stroke etiology, resulting in the atherothrombotic etiology with atheromatosis in the carotid bulbs bilaterally, with stenosis of nearly 50%. **Discussion:** The OCRA is a medical emergency that provokes sudden and painless amaurosis and it must be treated as "Retinian stroke". TCD with transorbital window allows the rapid and reliable detection of several signs which suggest its presence and even the OCRA mechanism, such as the absence of flow in the CRA and the presence of hyperechoic material in the retrolbulbar circulation of the optic nerve. TCD can detect signs of OCRA, including in cases with unremarkable initial ophthalmologic evaluation. It has been evidence high sensibility of this method, and due to this fact, it is an important tool to diagnose OCRA and investigate the stroke etiology, which could guide the clinical treatment. **Final Comments:** TCD is a relevant diagnostic method in the suspect cases of OCRA, which can have no alterations in the angiotomography and ophthalmoscopy. The OCRA diagnosis is fundamental to establishing the acute treatment and definition of secondary prophylaxis. This exam was incorporated into our service in cases and made it possible for the confection of a specific protocol to attend OCRA cases.

Doença Cerebrovascular

TL 1105462

BILATERAL SYMMETRICAL ISCHEMIC STROKE IN A YOUNG PATIENT: CASE PRESENTATION

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Case Presentation: A 30-year-old woman with a past record of resistant systemic arterial hypertension for 15 years, diabetes mellitus type 1 with irregular treatment, heart failure, on therapeutic anticoagulation due to right deep vein thrombosis and seronegative antiphospholipid antibody syndrome, presented to the emergency department with a sudden onset of left hemiplegia, which resolved spontaneously in less than an hour. A head CT was performed and displayed no acute alterations. The patient was discharged without being evaluated by the Neurology team. Later that same day, she exhibited a decreased level of consciousness and bilateral tonic-clonic seizures, requiring orotracheal intubation and mechanical ventilation. This time a head CT angiography was ordered and revealed an occlusion of the left middle cerebral artery and a subocclusion of the right middle cerebral artery. A new head CT confirmed bilateral symmetrical hemisphere infarction. **Discussion:** This is a relevant case since it depicts two rare events taking place simultaneously: a bilateral symmetrical hemisphere infarction and the occurrence of an ischemic stroke in a young patient. The incidence of stroke in young adults is low, however recent studies have shown an increase in the past years. This rise can be attributed to traditional risk factors found in ischemic strokes in older adults, such as hypertension, dyslipidemia, diabetes mellitus and obesity, but also to alcohol abuse, use of illicit drugs, thrombophilias and antiphospholipid antibody syndrome. This case illustrates a catastrophic vascular event in a young woman with multiple comorbidities that could lead both to thrombotic and to embolic events. It is important to acknowledge the occurrence of stroke in the younger population, since its dismissal could lead to tragic outcomes such as the one reported here. If the patient had received proper care when she first presented to the emergency room, with a probable transient ischemic attack, maybe the bilateral hemisphere infarction could have been avoided. **Final Comments:** It's of great importance to control risk factors in young adults, who are in productive and reproductive age, in order to avoid ischemic events and their disabling sequels that affect not only the patients but family and society as well.

Doença Cerebrovascular

TL 1105477

DEEP CEREBRAL VENOUS THROMBOSIS MIMING COVID-19-RELATED ENCEPHALITIS: DISCUSSION OF A CASE IN PANDEMIC TIMES

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Case Presentation: V.Z., 19 years old, female, ballet dancer. Diagnosed by RT-PCR with COVID-19 7 days before arriving at the Hospital, came up with a holocranial headache. One day after, she evolved with apathy and motor aphasia, not witnessed. At Hospital admission, the examination revealed a bedridden patient, Glasgow Coma Scale of 8, with muscular strength overall reduced. Her risk factors for thrombotic events were the use of combined oral contraceptive, current infection and recent vaccination against SARS-CoV-2. The patient was submitted to a Magnetic Nuclear Resonance (MRI) of the brain and lumbar puncture for Cerebrospinal Fluid (CSF) collection. The MRI showed a symmetrical T2/Flair hyperintensity in both thalamus and basal ganglia. Also, sparse foci of diffusion restriction in the supratentorial white matter. In SWI weighting, were hypointense foci in the deep venous structures. With administration of Gadolinium, failure of filling of venous structures and of the straight sinus were evidenced. The CSF did not show abnormalities. Also, D-dimer in blood was 562 ng/mL. Having Cerebral Venous Thrombosis (CVT) and Encephalitis as differential diagnoses, we started anticoagulation with enoxaparin (1mg/kg 12/12h) and pulsotherapy with methylprednisolone (1g for 5 days). Ten days after admission, she was discharged from Hospital walking, with fluent speech and no sequelae, taking dabigatran 150mg 12/12h. After thirty days of discharge, the MRI showed an area of sequela gliosis in both thalami. The patient is clinically well and remains under outpatient follow-up, without neurological sequelae. She has even returned to ballet practice. **Discussion:** COVID-19 is clearly related to thromboembolic events and neurological manifestations, through several mechanisms. CVT is a cerebrovascular disease that can eventually mimic encephalitis, with a non-specific and progressive clinical scenery, mainly due to changes in the level of consciousness, as presented by the patient. The diagnosis was only concluded after a venous Magnetic Resonance Angiography (MRA), performed as soon as the resources were available for this purpose. **Final Comments:** The early approach to the patient and the study of possible differential diagnoses allowed the medical team to quickly start therapies with LMWH and pulsotherapy, which contributed to the good outcome. The cause of CVT was attributed to COVID-19, by the fact that the patient had an active infection, added to the other risk factors.

Doença Cerebrovascular

TL 1105505

REVERSIBLE POSTERIOR ENCEPHALOPATHY SYNDROME IN A YOUNG PATIENT WITH PRIMARY MALIGNANT ARTERIAL HYPERTENSION: CASE PRESENTATION

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Case Presentation: P.S.B, male, 29 years old, low visual acuity 3 months ago, hypertension requiring sodium nitroprusside in an infusion pump, altered level of consciousness, and convulsive crisis. Patient with no comorbidities. In ophthalmological evaluation, there is evidence of bilateral papilledema, increased vascular tortuosity, bilateral microaneurysms and microhemorrhages, and visual acuity of hand movement. Initially, skull tomography (CT) presented diffusely hypodensity in white matter and hydrocephalus. In cerebrospinal fluid, presence of protein increase, and alteration of renal function compatible with chronic kidney disease. This was followed by hypotheses of diseases of the white substance of inflammatory, infectious, neoplastic or vascular origin. In Cranial Magnetic Resonance Imaging (MRI), hypersignal in FLAIR in supra and infratentorial white matter, in optic nerves and ventricular ectasia. Intracranial arterial angiography without alterations. Brain biopsy was also performed, with a normal result. Serological, infectious and rheumatological tests negative, and normal abdominal MRI and renal arteriography. After satisfactory blood pressure control, the patient presented complete improvement of mental confusion, absence of new convulsive crises and partial improvement of visual deficit. On cranial MRI performed after one month of Hospitalization, there was a significant reduction in the lesions. Given the diagnostic exclusions and patient evolution, the diagnosis of Reversible Posterior Encephalopathy Syndrome (PRES) was confirmed. **Discussion:** The PRES course with altered level of consciousness, visual deficits, headache, convulsive crises and focal deficits. There are divergences on the pathophysiology, believing that the main point is endothelial damage with vascular extravasation, resulting in edema. It translates as hypodensity on CT and hypersignal on cranial MRI, predominantly in occipital and parietal lobes, but not exclusively. It may be secondary to severe arterial hypertension, chronic kidney disease, sepsis, and immunosuppression. In the differential diagnoses we found infections, autoimmune diseases, vasculitis, malignant diseases and basilar artery thrombosis. The treatment does not control the primary etiology. **Final Comments:** Atypical and/or infratentorial PRES is less common, but in the face of a classic clinical picture with a typical triggering factor should be part of the differential diagnosis of white matter diseases.

Doença Cerebrovascular

TL 1105511

CEREBRAL VENOUS THROMBOSIS SECONDARY TO MUTATION IN THE GENE PROTHROMBIN IN THE CONTEXT OF VACCINATION AGAINST COVID-19: A CASE PRESENTATION

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Case Presentation: A 15-year-old woman attended the medical office with a headache without improvement for weeks after 30 days of vaccination against coronavirus with the Ad26.COV2.S vaccine. Denied using contraceptives. Evolved with drowsiness, neck stiffness, abducens nerve paresis, low visual acuity, vomiting, ataxia and mild asymmetric tetraparesis. Optical Coherence Tomography (OCT) showed bilateral papilledema. Cerebral Magnetic Resonance (CMR) and CSF examination without alterations. Spinal manometry confirmed the Intracranial hypertension (ICH) (75cmH2O). A relief lumbar puncture was performed and acetazolamide was started. Arteriography showed venous thrombosis of the sigmoid and left transverse sinus. The investigation of the alteration of Leiden V factor, antithrombin III, C and S Protein, antiphospholipid antibodies was all normal. A positive result for the prothrombin gene 20210 mutation in heterozygosity was obtained. Full anticoagulation therapy was performed with 5 mg warfarin (INR between 2 and 3) and enoxaparin bridge, associated with topiramate 25 mg/day. Patient remains in outpatient follow-up, with no signs of disease recurrence. **Discussion:** Cerebral venous thrombosis (CVT) is defined as the presence of a thrombus within a venous sinus, in a superficial or deep intracranial vein. CVT has a higher incidence in young women, with additional risk factors, such as pregnancy, puerperium and use of oral contraceptives². Headache is the most common symptom, but focal neurologic deficits, convulsion episodes and encephalopathies are also evidenced. The most suitable study technique of venous thrombi is CMR venography. The coagulogram may indicate the presence of conditions that help the pathogenesis of CVT and the research of the thrombophilic status and mutations is helpful. Lumbar puncture is important to rule out meningitis cases³. In addition, other conditions such as prior vaccination for COVID-19 as adenovirus vector-based vaccines are related to thrombotic episodes, through a mechanism called Vaccine-Induced Thrombocytopenic Thrombosis (VITT)⁴. **Final Comments:** It is noteworthy that CVT is an uncommon and reversible condition if diagnosed and treated properly. Its association with mutations in the 20210A gene of prothrombin is well established and in relation to vaccination it is important to recognize that the risk of CVT from COVID-19 exceeds that of vaccines.

Doença Cerebrovascular

TL 1105552

VERTICAL AND HORIZONTAL ONE AND A HALF SYNDROME: A CURIOUS STROKE PRESENTATION

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Case Presentation: A 69-year-old female patient with a history of hypertension and heart failure was admitted with a sudden onset of right hemiparesis, dysarthria, and diplopia. On neurological examination, she had right hemiparesis, sparing the face, dysarthria, appendicular ataxia of the four limbs, cutaneous-plantar reflex in extension on the right, moderate bilateral ptosis more prominent on the left. Ocular movements examination demonstrated bilateral upgaze palsy with limitation of infraduction on the left, indicating vertical one and a half syndrome. For horizontal ocular movements, only the right eye could abduct. Also, there was bilateral conjugated leftward palsy, indicating horizontal one and a half syndrome. Doll's eye test (oculocephalic reflex) was absent and the eyes were unable to converge. Brain MRI showed hypersignal in the midbrain and pons to the left on T2-FLAIR, with water diffusion restriction, indicating a recent ischemic event. A small right cerebellar lesion with hypersignal in T2-FLAIR and water diffusion restriction was also present. **Discussion:** The one-and-a-half horizontal syndrome is a well-known and well-described clinical entity, characterized by complete limitation of ipsilateral horizontal eye movement and limitation of adduction of the contralateral eye. It occurs due to injury to the pontine paramedian reticular formation or the sixth cranial nerve nucleus and the ipsilateral medial longitudinal fasciculus. The vertical one-and-a-half syndrome is a less frequent entity, characterized by complete limitation of vertical eye movement in one eye and limitation of upward or downward vertical movement in the contralateral eye. Among its causes, the most common are related to injury to supranuclear structures, such as thalamic nuclei. However, other structures related to the control of the vertical gaze may be involved, such as reticular formation and pretectal area in the rostral midbrain, and rostral interstitial nucleus of the medial longitudinal fasciculus, and interstitial nucleus of Cajal. The most frequent etiology related to both syndromes is cerebrovascular disease, followed by demyelinating diseases and encephalitis. **Final Comments:** The association between the horizontal and vertical one and a half syndromes is rarely described in the literature. In addition to changes in ocular movement, our patient also had other neurological findings that are justified by the topography of the ischaemic lesions.

Doença Cerebrovascular

TL 1105556

ISCHEMIC STROKE AS AN INITIAL MANIFESTATION OF LYMPHOPROLIFERATIVE DISEASE: A CASE PRESENTATION OS POLYCYTHEMIA VERA DIAGNOSIS

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Case Presentation: A 49-year-old female with uterine myomatosis and controlled arterial hypertension. Admitted with right middle cerebral artery syndrome – NIHSS 13 and 45-minute therapeutic window; intravenous thrombolysis was contraindicated due to vaginal bleeding. Past history of nonspecific symptoms of holocranial headache, dizziness, and mild asthenia. CT angiography was not performed to assess the possibility of mechanical thrombectomy. Initial investigation: Electrocardiogram in sinus rhythm, transthoracic echocardiogram without any abnormalities of chambers, valves and walls, as well as systolic and diastolic functions. Control brain CT confirmed Ischemic stroke (IS) showed right temporoparietal cortico-subcortical hypodensity with CT angiography showing absence of stenoses or plaques in carotid and vertebral arteries; internal carotid, anterior, middle and posterior cerebral arteries, as well as vertebral and basilar arteries with preserved path, caliber and enhancement. Dynamic assessment with transcranial doppler evidence means intracranial vessel velocities in the lower limits of normality for the patient's age and normal pulsatility indices. In the laboratory investigation observed: 20.3% hemoglobin and 59.1% hematocrit. Hematologist evaluation performed bone marrow biopsy with characteristic immunophenotyping of Polycythemia Vera (PV). She underwent therapeutic phlebotomy with improvement of systemic symptoms, hematimetric indexes: Hemoglobin of 14.6 and Hematocrit of 42% in addition to NIHSS 10 and modified Rankin of 2, outpatient with a Hematologist at discharge in addition to rehabilitation. **Discussion:** Hematological diseases are responsible for about 1% of IS. Despite being uncommon, IS can be the initial presentation of lymphoproliferative diseases, with its occurrence as the first symptom in 15% or more of patients with this type of condition. **Final Comments:** Despite the fact that many patients do not present common causes of IS even in adults or elderly, we should always pay attention to less frequent etiologies (non-atherosclerotic or non-cardiogenic) since the etiological definition of stroke is the starting point of secondary prophylaxis. and recurrence of brain damage. In this case, the etiological hypothesis was based on a basic laboratory test.

Doença Cerebrovascular

TL 1105574

ANEURISMATIC SUBARACHNOID HEMORRHAGE IN SINGLETON-MERTEN SYNDROME: A MERE COINCIDENCE?

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Case Presentation: A 24-year-old 12-week-pregnant woman was admitted to the emergency room presenting hypertension, a thunderclap headache, neck stiffness, and torpor. She denied trauma. She had a prior medical history of dental and orthopedic anomalies, arthritis, short stature; aortic valvulopathy, and refractory systemic arterial hypertension, in use of four classes of drugs. At the age of nineteen, she was diagnosed with Singleton-Merten Syndrome (SMS) by a molecular test, which evidenced a heterozygous for a c.992C>T p.Thr331Arg variant in the IFIH1 gene. Until this date, she had no evidence of neurologic involvement/disorders. Her cranial computed tomography (CT) imaging showed modified Fisher IV Subarachnoid Hemorrhage (SAH), intraparenchymal hemorrhage, and calcification of bilateral basal ganglia. Arteriography showed an aneurysmal in the perimarginal artery. The aneurysm was successfully treated with coil embolization. **Discussion:** SMS and Aicardi-Goutieres syndrome (AGS) are rare and multisystemic disorders caused by a gain-of-function mutation in the IFIH1 gene. Classically, SMS presents involvement of ectoderm tissues, such as skin and teeth, cardiac involvement, and aortic calcification, but no neurological involvement was reported. On the other hand, AGS classically presents progressive encephalopathy, spastic paraplegia, and calcification of basal ganglia. The variant in the IFIH1 gene identified in this patient was described previously in a family affected by both neurological involvement and features of SMS. An extensive literature review was performed and no report describing intracranial aneurysm was found even when there is AGS overlapping occurrence. **Final Comments:** This case demonstrates SAH in a patient with probable overlapping syndromes (SMS and AGS). Because the diseases are extremely rare, there is still a lack of evidence on whether it is a fortuitous association or intracranial aneurysms are part of the syndrome spectrum.

Doença Cerebrovascular

TL 1105615

CEREBRAL VENOUS THROMBOSIS PRESENTING AS REFRACTORY STATUS EPILEPTICUS IN A PATIENT WITH COMPLEX ARTERIOVENOUS FISTULA AND PTEN MUTATION SPECTRUM – A CASE PRESENTATION

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Case Presentation: A 30 year old patient was admitted to the Hospital with progressive consciousness impairment. She had a previous history of a complex arteriovenous fistula (AVFs), undergoing multiple embolizations. Four hours after admission, she evolved with a refractory status epilepticus, controlled using multiple drugs and with the need of mechanical ventilation. Her admission magnetic resonance imaging showed AVF on transverse and sigmoid sinus, also involving the inferior portion of the sagittal sinus associated with cerebral venous thrombosis on the basal vein of Rosenthal and sphenoparietal sinus. Her complex anatomy made the thrombus inaccessible for mechanical thrombectomy. She was anticoagulated and submitted to four new embolizations, but the venous hypertension persisted. Surgical venous drainage disconnection was made, remaining in a minimal state of consciousness. To elucidate the genetic mechanism behind the clinical phenotype, an exoma was made, showing a variant with loss of function on PTEN. She had no family history of neurological diseases. **Discussion:** AVFs are usually unique. However, the coexistence of refractory and recidivant lesions that progress from the anterior circulation to the posterior circulation, as our case showed, raise the question if there dormant channels between dural arteries and sinuses, which open when the sinus is occluded and venous pressure is increased or that the venous hypertension may cause hypoxia and production of angiogenic factors, that promoted endothelial growth and neoangiogenesis. In our patient, alteration on angiogenic factors like tissue plasminogen activator, may be responsible for the multiple dural fistulas, like on some patients with PTEN mutation. There are few case presentations on literature that show the correlation of patients with Cowden syndrome genotype associated with arteriovenous fistulas without other clinical manifestations but, until now, many phenotypes are correlated with PTEN mutations like Cowden syndrome and Bannayan-Riley-Ruvalcaba syndrome. Some authors suggest predisposition to thrombosis in these patients. **Final Comments:** The FAVD are a subtype of AVFs. Our case displayed an atypical case of complex and large AVFs, manifesting with refractory status epilepticus secondary to a cerebral venous thrombosis that caused intense venous hypertension. The clinical spectrum of PTEN mutations is heterogeneous, and may involve patients without the classical symptomatology.

Doença Cerebrovascular

TL 1105646

RECURRENT ISCHEMIC STROKE DUE TO INTRACRANIAL STENOSIS TREATED WITH BALLOON-EXPANDABLE: A CASE PRESENTATION

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Case Presentation: A 70 year old female with a history of ischemic stroke one year ago in the territory of middle cerebral artery, currently in using of acetylsalicylic acid and clopidogrel, presented with a new stroke in the same territory previously affected. Angiography showed stenosis of 55% of the M1 segment of the right middle cerebral artery. Due to the recurrence of ischemic events despite the double antiplatelet therapy, an endovascular treatment was chosen. The initial planning was a balloon stent, but during the procedure, it was not possible to place the stent, with only the balloon being performed, with good opening of the vessel and evolutionary control exams with transcranial Doppler showing good reperfusion and even improvement of the structural pattern of the wall of the affected vessel. The patient has evolved well, so far without new recurrences of ischemic events, is still under medical treatment. **Discussion:** Intracranial stenosis (ICS) is associated with a higher risk of ischemic stroke. Although the SAMMPRIS trial clearly demonstrated superiority of aggressive risk factor management plus antiplatelet therapy over intracranial stenting, the question remains regarding optimal treatment of those patients who develop recurrent stroke symptoms despite best medical management. A post-hoc analysis of SAMMPRIS evaluated the impact of hemodynamic markers, such as the presence of “water-shed” and impaired collateral flow on baseline imaging, and rates of recurrent stroke in the medical arm. The analysis showed that patients with poor collateral flow and baseline strokes, suggesting a hemodynamic rather than an embolic mechanism, are at most risk for subsequent stroke with medical therapy alone. This finding may help identify patients who are more likely to benefit from endovascular therapy for severe intracranial stenosis. The optimal way to determine which intracranial stenosis is “hemodynamically significant” remains to be determined. **Final Comments:** Intracranial stenosis is an important cause of strokes and therefore needs treatment, especially when symptomatic. Although the overwhelming majority of patients with ICS should be treated with best medical therapy, a subset of patients with recurrent strokes require endovascular revascularization by angioplasty with or without stenting.

Doença Cerebrovascular

TL 1105657

NON-DECUSSATION OF THE PYRAMIDAL TRACT DISCOVERED INCIDENTALLY IN AN ASYMPTOMATIC PATIENT WITH ARTERIOVENOUS MALFORMATION: CASE PRESENTATION

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Case Presentation: A 47-year-old woman with a 5-month headache history underwent neuroimage investigation with a CT scan that suggested an AVM at the right parietal lobe. She had a clinical past of hypothyroidism denying previous seizures or weakness. A neurological examination on admission revealed no focal neurological deficit and the first brain magnetic resonance imaging (MRI) did not reveal any parenchymal lesions. The patient underwent cerebral angiography, which revealed AVM at the right angular gyrus and a 3-stage embolization was indicated. In the postoperative of 3rd embolization session, the patient developed a right motor deficit over the course of a few days. Brain MRI revealed a perilesional cytotoxic edema with restricted-diffusion posteromedially to the AVM in the right parietal lobe – ipsilateral to patient's motor deficit. A 3.0 T MRI Tractography showed solely unilateral non-crossing corticospinal tract with no decussation. **Discussion:** Abnormal pyramidal tract decussation is a very rare event that can be seen in ectodermal dysplasias. Non-decussation of the pyramidal tract is observed in several conditions such as Dandy Walker, Joubert, Kallman syndrome and Ichthyosis. There are only a few case presentations of incidental findings in healthy patients. We describe a patient who exhibited hemiparesis ipsilateral to an arteriovenous malformation (AVM) complication after embolization and whose neuroimaging showed a lack of pyramidal tract decussation. **Final Comments:** Pathologies of the pyramidal tract are rare and are frequently associated with genetic and congenital disorders such as posterior fossa anomalies or congenital mirror movements. However, they can be present in healthy, asymptomatic individuals and observed when a lesion to motor areas causes ipsilateral motor deficits. Only a few cases have been described in neurologic asymptomatic individuals. Our patient presented with right-sided hemiparesis ipsilateral to an ischemic lesion after AVM embolization.

Doença Cerebrovascular

TL 1105702

REVERSIBLE POSTERIOR LEUKOENCEPHALOPATHY SYNDROME (PRES): CASE PRESENTATION

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Case Presentation: A 37-year-old male patient was brought to our Neurologic Emergency Department with acute ataxic neuropathy, cortical blindness, vertigo, gaze palsy, skew deviation, paresthesia associated with tonic-clonic seizures and impaired consciousness. As a result of topographical manifestations, a neurovascular syndrome in the vertebrobasilar arterial territory was hypothesized, consequently undergoing an attempt at chemical reperfusion. However, after the resolution of the acute phase, the patient remained under amaurosis bilaterally, and an etiological evaluation was initiated by requesting MRI Neuroimaging and subsequently lumbar puncture (LP). Radiological findings include confluent areas of T2 hyper signal and posterior cortical lesions when assessed using the FLAIR modality. In addition to DWI sequence, we can distinguish vasogenic edema in the brainstem and cerebellum. Also, cortical laminar necrosis and the corpus callosum are observed, suggesting the diagnosis of Reversible Posterior Leukoencephalopathy Syndrome (PRES). Promptly recognized, we initiated an etiological investigation regarding possible associated comorbidities with severe blood pressure and biochemical management. After 02 weeks, the patient reported regaining visual capacity in low acuity, which he progressively recovered during Hospitalization. Thenceforth, he remains at our service for adequate neurologic rehabilitation control.

Discussion: Reversible Posterior Leukoencephalopathy Syndrome (PRES) is a rare clinical and radiological entity characterized by acute posterior topographical symptoms and mostly reversible characteristics when prompt recognition and treatment occur. The pathophysiology remains unclear, associating endothelial cell dysfunction and dysautonomic changes like arteriolar constriction or dilatation, which may lead to a breakdown of the Blood-Brain Barrier (BBB). Also, as observed in MRI scans, intense vasoconstriction should evolve with local hypoperfusion, cytotoxic edema, and cerebral infarction. The prevention and treatment evolve control of arterial hypertension, reduction of immunosuppressive drugs, and electrolyte disturbances management.

Final Comments: This presentation describes a patient with PRES who fully recovered from his respective Focal Neurologic Deficits (FND), such as cortical blindness. Most cases in this syndrome result from case presentation and case series, requiring multicenter trials to define a reliable conclusion.

Doença Cerebrovascular

TL 1105703

CAROTID WEB: A RARE FINDING INVOLVED IN ISCHEMIC STROKE

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Case Presentation: A 70-year-old male with medical history of arterial hypertension, type 2 diabetes, hyperlipidemia and metastatic prostate cancer, presented with acute onset right-side weakness and global aphasia which had started 2 days before the admission in the Hospital facility. Patient did not receive IV alteplase. CT revealed an acute infarct on the left temporoparietal lobe. Computed tomography angiography (CTA) described a linear structure in the posterior portion of the carotid bulb on the left side, suggestive of carotid web, partial occlusion of M1 segment of the left medial cerebral artery and in M2 e M3 segments as well, without other signs of atheromatosis or stenosis. No evidence of embolism was shown on the further investigation. The patient received aspirin 100mg and high-intensity statin therapy associated with strict control of risk factors, without intervention on the AFMD. In short-term follow-up, patient maintained the deficits.

Discussion: Carotid web is considered an atypical variant of fibromuscular dysplasia (AFMD) and presents itself as an abnormal shelf-like intra-luminal projection of hyperplasia intima in the posterior wall of the proximal internal carotid artery, best visualized on CTA or carotid angiography. This finding has potential to cause blood stagnation, thromboembolism and distal embolization, with oncoming evidence of stroke association, specially in younger patients, but also on hypercoagulable state, such as malignancy. Previous studies have shown that carotid web is found in up to 9.5% of patients <65 years of age with anterior circulation stroke of unknown cause. The incidence of AFMD in older patients with ischemic stroke is unknown. In patients with symptomatic carotid web, without other attributable causes of stroke, antiplatelet therapy is recommended to prevent recurrence. Carotid stenting or endarterectomy may be considered in refractory cases, based on limited data. Nevertheless, optimal management (medical or interventional) is unknown.

Final Comments: The carotid web is a proposed stroke mechanism particularly in younger patients without vascular risk factors. However, it may have an important roll in older patients. Future studies involving diagnosis and secondary prevention of symptomatic AFMD should focus on all age spectrum and on the association with other risk factors.

Doença Cerebrovascular

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CAROTID WEB: A SUBDIAGNOSED ISCHEMIC STROKE CAUSE IN YOUNG ADULTS

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Case Presentation: 40-year old male was admitted in the emergency room with a sudden onset left hemiparesis and asymmetry of the smile 2.5 hours ago. Previously hypertensive, alcoholic and ex-smoker. The primary neurologic examination showed a right middle cerebral artery (RMCA) syndrome with a NIHSS (National Institutes of Health Stroke Scale) of 14. A cranial computerized tomography (CT) showed a hyperdensity of the RMCA and an ASPECTS of 10. Intravenous thrombolytic therapy with Alteplase 0.9mg/kg was administered, since no contraindications were found, with a subsequent NIHSS of 11. Laboratory investigation, electrocardiogram and trans-thoracic echocardiogram showed no relevant findings. An arterial angiotomography (AT) of the cervical vessels found a minor irregularity with a linear aspect in the posterior wall of the right carotid bulb, suggesting a carotid membrane, which was confirmed with an angiography. An angioplasty with stent of the right internal carotid artery was opted as treatment.

Discussion: The carotid web is a cause of Ischemic Stroke in young adults with no cardiovascular history and no positive investigation for other cerebrovascular diseases. It is shown as a projection of linear aspect inside the bulbous portion of the carotid artery, and is better visualized in the sagittal cuts of the CT. It is an acquired vasculopathy, pathologically defined as an intimate fibromuscular dysplasia. The ischemic stroke pathogeny resides in the stagnation of flow and remote embolization of fibrin-based clots. It can cause recurrent hemispheric ischemic strokes. Although easily seen in ATs, the carotid web can be missed or misinterpreted, as it doesn't cause a significant hemodynamic stenosis and can mimic an arterial dissection, non-calcified arteriosclerotic plaque or an intraluminal thrombus. The diagnostic golden standard is the angiography, with a non-invasive option being the AT of the neck. Treatment options include antiplatelet and anticoagulant therapies, both with a high recurrency rate (40-50%). Both carotid endarterectomy and angioplasty with stent in the internal carotid are definitive treatments.

Final Comments: The therapeutic decision making in the secondary prevention of the ischemic stroke depends on the cause of said stroke, being the vascular study a primary one in the diagnostic investigation. The carotid web is an undiagnosed and sub treated entity, making it an important point of discussion among neurological cases.

Doença Cerebrovascular

TL 1105773

AN UNUSUAL PRESENTATION OF THORACIC SPINAL CORD ATROPHY DUE TO SUSAC SYNDROME

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Case Presentation: A 35-year-old man presented with 3 years history of progressive crural paraparesis and urinary incontinence. He had a previous Hospitalization for cognitive symptoms, headache and seizure. Was observed in MRI increased T2 signal in the corpus callosum and lumbar puncture showed an increase protein (86 mg/dL) and White cells (9 with 88% neutrophils). Because of these findings, at this moment, the main hypothesis was multiple sclerosis (MS). A treatment for presumed primary demyelination disease with methylprednisolone was performed with improvement of the mental confusion. Three years later was performed another MRI, showing increased T2/FLAIR signal in the central corpus callosum, assuming snowball appearance. At this time he reported urinary incontinence, erectile dysfunction, sensory symptoms and weakness. Physical exam showed a crural paraparesis and a spastic gait. Moreover he had an ophthalmological examination showing areas of ischaemia demonstrating branch retinal artery occlusion and the MRI images had involvement of central corpus callosum and deep grey matter lesions in the basal ganglia. The image of spinal cord showed a thoracic atrophy with no sign enhance and cerebrospinal fluid analysis was normal. Under these circumstances another acute treatment was not performed. The presumptive diagnosis of Susac Syndrome was made. **Discussion:** Susac's syndrome is a rare autoimmune disease that affects the microvasculature. It's an endotelopathy that can be manifest with neurological, visual or auditive symptoms. (1) The classic clinical presentation was described as the triad of encephalopathy, sensorineural hearing loss and branch retinal artery occlusion. (2) Our patient had bilateral and multiple retinal ischaemia, but no involvement of the optic nerve. Although his spinal cord image showed a thoracic atrophy, spinal cord involvement was extremely rare in Susac's syndrome. (4) **Final Comments:** Susac syndrome is a rare disease and the differential diagnosis includes others autoimmune diseases, infectious diseases and paraneoplastic syndromes. The ocular and vestibulo-cochlear involvement, along with neurological symptoms were cardinal features of the syndrome, but it could not be seen at the same time. Presentation with spinal cord involvement is rare, but have been described and it's important to be known or could lead to a misdiagnosed with other demyelination disease.

Doença Cerebrovascular

TL 1105807

THROMBOLYSIS PERFORMANCE IN A BRAZILIAN PUBLIC SERVICE

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Background: Stroke is the second most common cause of morbimortality worldwide and one of the most important causes of death in Brazil. The ischemic stroke is the most common type and its treatment in the acute phase aims to restore the cerebral blood supply. This treatment can be performed by intravenous thrombolysis with alteplase and/or mechanical thrombectomy, the latter not being available in most of Brazilian public Hospitals. Thrombolysis is currently one of the first line treatments in patients with acute ischemic stroke within 4,5 hours of symptom onset. Available data shows functional improvement of patients 3 to 6 months after treatment. The most dreaded complication of thrombolytic therapy is symptomatic intracerebral hemorrhage which is associated with risk factors such as high blood pressure, diabetes mellitus, advanced age, microangiopathy, atrial fibrillation and stroke severity. **Objective:** Reveal data about the benefits and possible complications of the patients treated with IV thrombolysis in a real life scenario in a Brazilian public Hospital, in comparison with the data in the literature, focusing on hemorrhagic complications. **Method:** A prospective observational study was conducted between November/2019 and April/2020. The sample was calculated to compare outcomes with the studied population and the data of the available RCTs. The study included 53 sequential patients diagnosed with acute stroke treated with thrombolytic therapy. **Results:** Most of the patients were female and the mean age was 70 years old. The symptomatic hemorrhagic transformation rate was slightly higher in our study, as was the rate of worsened NIHSS after 24h of treatment, compared with data from the RCTs ECASS II and ECASS III. Moreover, 24,5% of the patients had other complications such as angioedema, hypertension and seizures and 15,09% of the patients died. **Final Comments:** It was observed that the population in our study was older and had at baseline more comorbidities, which are probably the features associated with higher odds of complications. In addition, despite the higher rates of hemorrhagic transformation, the patients in the study had an improvement in the functional status similar to the comparative studies. All in all, although in real life settings the rate of complications appears to be higher than that of RCTs, thrombolysis still leads to better outcomes, and therefore should be performed following proper protocols.

Doença Cerebrovascular

TL 1105810

EIGHT AND A HALF SYNDROME: CASE PRESENTATION

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Case Presentation: Female patient, 52 years old, hypertensive, was admitted to the service with a complaint of blurred vision and difficulty closing her left eye suddenly 24 hours ago. She denied other symptoms or previous episodes. On examination, she had medial rectus muscle paresis in the right eye and lateral and medial rectus muscle paresis in the left eye, facial paralysis of a peripheral pattern on the left, without further changes. The following exams were performed: cranial tomography without alterations, skull magnetic resonance imaging (MRI) with recent lacunar ischemia in the left pons, normal cranial magnetic resonance angiography, neck magnetic resonance angiography with tapering and parietal irregularities in the distal portion of the foramen segment (V2) of the left vertebral artery at the level of the axis tooth, without defining dissection areas to the method, normal echocardiogram, normal 24-hour Holter. Dual antiplatelet therapy, statin and blood pressure control were started. **Discussion:** Eight-and-a-half syndrome (EHS) is rare, with few cases described in the world literature. Its early recognition is of great practical importance, as it has varied and potentially serious etiologies, with different therapeutic implications. It is characterized by conjugated horizontal gaze palsy, ipsilateral internuclear ophthalmoplegia, and peripheral facial palsy without impairment of taste, as taste fibers for the anterior 2/3 of the tongue travel through the intermediate nerve, which has its nucleus in the medulla. The syndrome occurs due to a lesion that affects the medial longitudinal fasciculus, the paramedian pontine reticular formation and the fasciculus of the facial nerve on the same side at the level of the pons. The diagnosis is easily missed as it needs a detailed eye movement examination. It is mainly caused by stroke, multiple sclerosis, gliomas, metastases and vascular malformations. Less frequent etiologies include iatrogenesis related to tumor resection, neurocysticercosis, systemic lupus erythematosus and myasthenia gravis if isolated ocular involvement. **Final Comments:** The clinical recognition of EHS is essential for an accurate topographical diagnosis of pontine tegmentum lesions. Vascular disease and multiple sclerosis are the main differential diagnoses, although rarer causes should always be considered. MRI is the exam of choice for topographic and etiologic diagnosis. Treatment depends on the cause.

Doença Cerebrovascular

TL 1105836

HIV ASSOCIATED MOYAMOYA SYNDROME MANIFESTATION AS AN ACUTE HEMICHOREA

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Case Presentation: A 24 year-old presented to our Hospital clinic complaining about spontaneous movements in the left side of her body which began approximately 6 months earlier. Her medical history was notable for congenital HIV infection (undetectable viral load and CD4 count of 506), well controlled epilepsy and migraine. She has been on antiretroviral therapy (ART), amitriptyline, valproate and low dose chlorpromazine. She denied any other neurological or psychiatric symptoms, use of other drugs or seizures recurrence in the past ten years. Her level of consciousness was normal. There were random involuntary movements in the left side of her face, arm and leg in a choreoathetoid phenomenology and incorporated to voluntary movements. Distraction had no influence on the pattern of her movements. Her left limbs were hypotonic. CSF profile, bacterial and fungi cultures, erythrocyte sedimentation rate, thyroid function tests, glycosylated hemoglobin A1c were unremarkable. Brain magnetic resonance imaging (MRI) showed no acute or chronic stroke lesions, nor evidence of neuroinfection. The time-of-flight angiogram was notable for absence of flow in both middle cerebral arteries, suggesting Moyamoya syndrome (MMS). Arterial angiography located the occlusion on both supraclinoid segments of internal carotid arteries with blood flow redirected to a wide collateral circulation and posterior communicating arteries, confirming MMS pattern. Reviewing older exams a contrast CT scan done 4 years before had no abnormalities, including arterial circulation. We found no other etiology to justify her chorea besides the MMS. The patient received daily risperidone 2mg with complete resolution of the symptoms. **Discussion:** MMS is a rare cause of chorea with a frequency of 3 to 6%, as well incidence of MMD in HIV patients. The dilated collateral vessels compress the basal ganglia or can cause local ischemia, which can lead to a disruption of normal striatal activity. HIV infection has been reported as one of many conditions implicated in the etiopathogenesis of MMS. HIV can cause endothelial damage and also induce proinflammatory states that later manifest as stenosis or dilation of the vessel wall that can occur despite effective ART and stable CD4 counts. **Final Comments:** Since HIV infection has a well recognised relation to cerebrovascular disease as well as to movement disorders, we believe that it was a major factor to the development of this secondary form of chorea.

Doença Cerebrovascular

TL 1105876

MOYAMOYA'S SYNDROME RELATED TO PULMONARY TUBERCULOSIS – CASE PRESENTATION

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Case Presentation: Female, 19 years old, admitted to the ER with acute desaturation, and a one month's history of fever and cough, in use of rifaroxaban for a 3 weeks diagnosis of pulmonary embolism; has shown worsening for a week. Neurology has been called to evaluate for an acute confusional state, lower limb weakness and respiratory symptoms. Previously rigid, the patient had domiciliary Pulmonary Tuberculosis contact. To the neurologic examination: sleepiness, lentified, disoriented in time and space, no language impairment, mild dysarthria, muscle grade 1 on left and 3 on the right, and heightened deep reflexes. Previous Hospital admission review: Tomography showed ischemic areas in the vascular frontier territory, the new CT has shown new hypodensifying cortico-subcortical areas in the frontal and parietal high convexities of both hemispheres, determining amplitude reduction of some adjacent liquor spaces. AngioCT has shown no thrombus, but an unspecified narrowing in the internal carotid. Cerebrospinal Fluid: protein elevated (62mg/mL), undetectable Mycobacterium tuberculosis PCR. Angiography: occlusions and "smoke" aspect collateral circulation, suggesting Moyamoya's disease. Laboratory investigation for thrombophilias, falciform anemia and other infections has turned negative. One month of treatment, no cough or dyspnea, with progressive improvement of the neurologic symptoms. In 60 days, she was evaluated in the ambulatory, alert, oriented and muscular strength grade 4 to the left, still in rehab. The investigation still awaits anticoagulation cessation for complete neurovascular evaluation. **Discussion:** Moyamoya in the terminology that refer to the progressive narrowing of a big intracranial artery in parallel to multiple collaterals that confer the "smoke" to the angiography. It's called Moyamoya's disease when it has genetic association, and Moyamoya's Syndrome when related to other clinical entities. A few case presentation relate the syndrome to standard resistant tuberculosis treatment and Neurotuberculosis, cases with long infection periods, necessary to the disease's progress and collateral circulation development. **Final Comments:** Despite the findings of Moyamoya Syndrome and its association with tuberculosis being described in few cases in the literature, it is possible that the chronic infectious condition plays a role in the pathophysiology of the neurovascular condition.

Doença Cerebrovascular

TL 1105881

HEMOPERICARDIUM: AN UNCOMMON COMPLICATION OF THROMBOLYSIS IN STROKE

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Case Presentation: Male 72-year-old patient, with hypertension and a previous hearing loss, started with sudden confusion and aphasia. He was sequentially referred to the tertiary service due to a condition compatible with stroke, presenting a 9 NIHSS on admission. After a brain CT scan without any evidence of hypodensity and ASPECTS of 10, thrombolysis was performed. The procedure was uneventful and there was a complete improvement of the deficit. After 2 days of Hospitalization, while waiting the etiologic investigations, the patient started with tachycardia (110 bpm) and respiratory distress requiring oxygen supplementation. A transthoracic echocardiogram was performed, which showed significant pericardial effusion with mild hemodynamic repercussion with right atrial collapse during systole. The Cardiac Surgery's team quickly attempted an ultrasound-guided subxiphoid drainage, however, it was unsuccessful. Progressively the patient starts with sings of cardiac tamponade, jugular swelling, drowsiness and hypotension (85x55mmHg), and urgent surgical approach was performed with pericardial drainage of 400mL of serohematic fluid. After the procedure, the patient evolved with hemodynamic stability, and underwent a new echocardiogram, with complete improvement of the pericardial effusion. The basic investigation was unremarkable and not show an etiological cause and the patient was discharged for outpatient follow-up. **Discussion:** Spontaneous pericardial effusion is a rare complication of thrombolysis in stroke. It is more common when thrombolysis is performed in acute myocardial infarction, in which the patients probably had as a pre-existence cardiomyopathy or pericardiopathy. Mortality is usually very high, due to rapid evolution to cardiac tamponade, but in this case the patient did not have an immediate complication. However, any sings of cardiovascular complications should promptly undergo a cardiac investigation. **Final Comments:** As the diagnosis is echocardiographic, in any case of hypotension in the first hours or days of Hospitalization, with no other possible clinical causes, an urgent echocardiogram should be performed. And the existence of a cardiac surgery team at the Hospital to perform pericardiocentesis or pericardiostomy could be what would save the patient.

Doença Cerebrovascular

TL 1105905

STROKE DUE TO SPONTANEOUS SUBCLAVIAN AND VERTEBRAL ARTERIES DISSECTION

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Case Presentation: A 39-year-old woman suddenly presented rotational vertigo, loss of balance with fall from standing height, without head trauma, and intense left temporoparietal pulsatile headache. She used simple analgesics and didn't search for medical service at that moment. On the next day, due to the continuation of the symptoms, she looked for medical service. She didn't have previous cervical trauma or physical activity before the installation of the symptoms and she referred that it all started while she was taking a shower. The patient has systemic arterial hypertension, dyslipidemia and diabetes. She didn't have any smoking or etilism history and in her family there was no history of previous stroke. At the admission she presented incoordination on the left side of her body and an ataxic gait without other findings. The CT showed hypoattenuation on the posterior and inferior cerebellar vermis and on the left cerebellar hemisphere compatible with an acute stroke. The CTA demonstrated a hypodense semicircular formation partially obliterating the flow of contrast on the left subclavian artery determining a stenosis greater than 50% with extension to the ipsilateral vertebral artery. The diagnosis of stroke due to spontaneous subclavian and vertebral arteries dissection was given. The patient was discharged in use of warfarin and returned in consultation with almost full resolution of the symptoms after a few months. Arterial dissection is when a tear occurs in the inner layer of the artery, causing an intramural hematoma. The stroke due to arterial dissection is a consequence of the embolism originating on the inner layer of the vessel or a consequence of local hemodynamic compromise due to reduction of the lumen of the vessel. The arteries most commonly affected are the cervical and this type of lesion is generally due to traumatic events, surgical positioning or endovascular procedures. However, a spontaneous subclavian arterial dissection causing a stroke is a very rare event, with only a few cases described in medical literature, and can be associated with migraine, pregnancy and use of illegal substances. The incidence is higher in women between 40 and 50 years associated with systemic arterial hypertension. Although rare, the spontaneous subclavian arterial dissection is a relevant cause of stroke, specially in young adults, thus clinical suspicion is necessary for a proper management of the case. Anyway it has a good prognosis even with conservative therapy.

Doença Cerebrovascular

TL 1105908

COENZYME Q10 DEFICIENCY LEUKOENCEPHALOPATHY – CASE PRESENTATION

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Case Presentation: Female patient, 42 years old, history of gait incoordination for 03 years and progressive worsening, in addition to nocturnal enuresis and clonus in the lower limbs, requiring support to walk. There were no reports of vertigo, diplopia, changes in motricity or sensitivity. Personal history: Depressive disorder for 10 years; Bilateral cataract; Consanguinity between parents; sibling with psychiatric disorder, bradykinesia, bradypsychism and ataxia. On examination: alert, oriented, mild dysarthria and bradypsychism; isochoric and photoreactive pupils, with bilateral central opacification; decreased speed of ocular saccades; hypopalesthesia to iliac crests bilaterally; muscle strength preserved with global bradykinesia; global hyperreflexia, Hoffman and Trommer present bilaterally; inexhaustible bilateral achilles clonus; broad-based and bouncing gait; positive romberg. Minimal with a score within the expected level of education (29/30). Cranial MRI with bilateral and symmetrical signal alteration of the supratentorial white matter, with periventricular predominance, with intermingled cystic formations, compatible with leukoencephalopathy; it also has T2/FLAIR hypersignal in the caudate nucleus and putamen bilaterally. **Discussion:** The association of the progressive clinical findings of ataxia and extrapyramidal manifestations with the aspects evidenced in the magnetic resonance suggests a neurodegenerative picture with predominant involvement of the white matter. The history of consanguinity between the parents, in addition to a similar evolutionary picture in the sibling, point to a genetic etiology. During Hospitalization, a genetic panel for leukoencephalopathies was performed on the patient, which showed probably pathogenic homozygosity in the COQ8A gene and an increase in copies (03) of the EIF2B1 gene, with uncertain significance. **Final Comments:** The COQ8A gene plays a key role in the biosynthesis of coenzyme Q10, which acts as an electron carrier in the mitochondrial respiratory chain for increased ATP production and also acts as an antioxidant and membrane stabilizer. The phenotypic characteristics reported in a study with patients with a biallelic alteration in the COQ8A gene include cerebellar ataxia (100% of cases), movement disorders, cognitive impairment, epilepsy and neuropsychiatric symptoms (depression, anxiety, psychosis or impulsive-aggressive behavior). The main therapeutic approach has been coenzyme Q10 supplementation.

Doença Cerebrovascular

TL 1106019

MULTIPLE ISCHEMIC STROKES DUE TO ADULT-ONSET STILL'S DISEASE: CASE PRESENTATION

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Case Presentation: A 44 year old Woman reports that in february os 2022 presented sudden change of visual field with a homonymous right hemianopia. She had previous diagnosis of schizophrenia, a stroke in 2019, and Adult-onset still's disease (AOSD) diagnosed in 2020. In the Hospital admission, the CT scan revealed a recent onset ischemic stroke in the left thalamus, occipitotemporal gyrus and cuneo, besides a stroke sequel on the right frontal, temporal, parietal, insula, adjacent to the lateral fissure. The head and neck angio tomography revealed a occlusion of the communicating segment of the right internal carotid as well as the A1 segment of the right anterior cerebral artery, the right middle cerebral artery and the P2 segment of the left posterior cerebral artery (PCA). Cardiac investigation was done with electrocardiogram, 24 hour holter, transthoracic and transesophageal echocardiogram resulting normal. It was then performed the cerebral arteriography resulting intracranial vascular occlusive disease in two distinct vascular territories (a cronic feature occlusion on the top of the right ICA and a subocclusion on the P2 segment of the left PCA), possibly due to an inflammatory component. Pulse therapy was then performed with 1 g per day of methylprednisolone for 5 days, and a posterior introduction of prednisolone 60 mg per day. **Discussion:** AOSD is an inflammatory disorder, characterized by daily fever, arthritis and evanescent rash that occurs in patients older than 16 years old. The etiology is unknown with an annual incidence of approximately 0,16 cases per 100.000 people. The AOSD is a diagnosis of exclusion and can be done on base on the clinical features and the typical laboratorial findings following the yamaguchi diagnostic criteria. Ischemic stroke is a rare complication of this disease, and the explanation for this event is the vasculitis pathophysiology. The corticosteroids are the first line drugs for treatment. **Final Comments:** We reported an uncommon case of a young 44 years old woman with previous diagnose of AOSD, that had a stroke in different territories, with involvement of the anterior and posterior circulation, visualized by the head and neck angio tomography and arteriography corroborating with typical finding of vasculitis. Thus, it's importante to be alert that one of the possible complications of the AOSD is the occurrence of stroke events due to vasculitis inflammatory process.

Doença Cerebrovascular

TL 1106059

COMPLETE BLOOD COUNT IN ANEURYSMAL SUBARACHNOID HEMORRHAGE SCENARIO: POSSIBLE USES OF BLOOD ELEMENTS FOR PREDICTING OUTCOMES

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Background: Aneurysmal subarachnoid hemorrhage (SAH) is a worldwide health burden with high fatality and permanent disability rates, and variable prognosis. Recent evidence suggests that some biomarkers play a critical role in predicting outcomes. **Objectives:** We propose a systematic review of literature on recent advances in blood element biomarkers for aneurysmal SAH, translating from a physiological point of view to their potential usefulness. **Methods:** Pubmed, Embase, Scopus, Cochrane, and Web of Science databases were systematically searched to identify studies assessing red blood cell element biomarkers for aneurysmal subarachnoid hemorrhage, following sensibility, and specificity results found in each paper published. **Results:** Six studies were included. Delayed Cerebral Ischemia (DCI) was predicted by Neutrophil/Lymphocyte ratio (87.3%/48.4%, and 63%/53%), and Platelet/Lymphocyte ratio (55.3%/78.5%). Vasospasm was seen by Platelets (40%/90.5%, and 78%/85%), Lymphocytes (57.9%/83.3%), Neutrophils (40%/100%), Leukocytes (50%/100%), and Combined Platelet and Leukocytes high values (60%/89.5%). Rebleeding in Neutrophil/Lymphocyte ratio (72.3%/63.3%), and Neutrophil/Lymphocyte ratio combined with Fisher Grade (39.94%/100%). Poor outcome in Neutrophil/Lymphocyte ratio (74.5%/69.3%), and Platelet/Lymphocyte ratio (55.3%/78.5%). Mortality in 3-months appears predicted by RDW value (80.6%/91.6%). **Final Comments:** Biomarkers could help in predicting complications in aneurysmal subarachnoid hemorrhage patients. We listed some of the recent advances in this translational, intriguing, fast-growing field. Although the knowledge gaps are still significant, some markers are demonstrating promising results and could be helping patients in the near future.

Doença Cerebrovascular

TL 1106085

COMPLETE RESOLUTION OF A HUGE CAROTID THROMBUS AFTER ANTICOAGULATION IN 48-YEAR-OLD WOMAN WITH STROKE

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Case Presentation: A 48-year-old woman, without previous comorbidities, admitted by complete right hemiparesis, mixed transcortical aphasia and right homonymous hemianopsia 7 hours after admission, NIHSS 7. Imaging of the skull at the arrival showed no established ischemic lesion, however, arterial angiography of the neck showed the presence of a subtraction image inside the left common carotid artery with irregular contours reducing the lumen of the vessel by up to 90%, suggesting the presence of intra-arterial thrombus. Started full anticoagulation with soredic enoxapara at a total dose of 120mg/d. He presented a transesophageal echocardiography (ETE) demonstrating patent foramen of high conductance. Maintained anticoagulation with Apixaban at Hospital discharge at a total dose of 10mg and after 15 days of treatment, a new control image was performed now with vessel wall protocol with evidence of complete resolution of the thrombus and absence of dissection of the vessel wall. **Discussion:** common carotid artery occlusion (AOCC) is a rare disease and accounts for 1%-5% of all cerebrovascular events. May present as stroke, transient ischemic attack (ITA), or with features of cerebral hypoperfusion. Chronic arterial occlusions are often attributed to atherosclerosis, while other less frequent causes include cardiac embolism, Takayasu's arteritis, post-radiation arteriopathy, syphilis, trauma. To date, there is no clear consensus on the best treatment in the face of a AOCC and the decisions for treatment of these patients are made according to each case and experience of the service. **Final Comments:** Occlusion of the common carotid artery although rare and acute clinical presentations vary from mild to devastating, the acute management of these patients remains a major challenge. In our service we report the case of a young patient with probable cardioembolic etiology who obtained benefit in the resolution of the thrombus with the use of direct oral anticoagulants.

Doença Cerebrovascular

TL 1106105

CONSECUTIVE HEMORRHAGIC STROKES POSSIBLY CAUSED BY CEREBRAL AMYLOID ANGIOPATHY RELATED TO DURAPLASTY

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Case Presentation: A 44-year-old male patient was admitted to our Hospital in March 2022 complaining of headache and left homonymous hemianopsia. In the past two years, he had two intraparenchymal hemorrhages, both in the right parietooccipital lobe. Computed tomography showed right parietooccipital intraparenchymal hematoma, left parietooccipital encephalomalacia and signs of a previous left occipital craniectomy. Magnetic resonance imaging confirmed the above findings, also showing several corticosubcortical punctate foci of low signal on SWI sequence predominating on bilateral occipital lobes indicating microbleeds. Angioresonance did not show any abnormalities. These findings support the possible diagnosis of cerebral amyloid angiopathy (CAA) related hemorrhage according to the modified Boston criteria, excluding the age criteria. The patient had a past medical history of head trauma at the age of 7 years, caused by a reaping hook, for which he underwent a neurosurgical procedure including craniectomy and duraplasty. He had no relevant familiar history of cerebrovascular disease or dementia. **Discussion:** CAA is commonly found in older people and in patients with Alzheimer's disease accompanying cerebrovascular disorders and dementia. Cerebral blood vessels affected by CAA are associated with functional and pathological changes, leading to hemorrhagic disorders, such as lobar intracerebral hemorrhage and cortical micro-hemorrhage. It is hypothesized that amyloid- β (A β), a pathological hallmark of this condition, is transmissible by a similar mechanism to acquired prion diseases. Furthermore, besides genetic inheritance, it has been described the occurrence of early onset A β -CAA associated cerebral hemorrhage, several decades after neurosurgical procedures performed in children involving cadaveric dura mater grafts or neurovascular embolization using material derived from cadaveric dura mater. Due to the early age, temporal association, no family history of stroke, we hypothesize that the duraplasty may be associated with the occurrence of amyloid angiopathy in this patient. **Final Comments:** This case illustrates a possible cause of early onset CAA and emphasizes that A β -CAA should be considered in the differential diagnosis even for young patients with intracerebral hemorrhage and history of neurosurgery. Furthermore, the exact physiopathology of this condition needs more elucidation due to the possibility of iatrogenic human transmission.

Doença Cerebrovascular

TL 1106121

DIAGNOSTIC CHALLENGE IN A YOUNG MAN: DENGUE ENCEPHALITIS OR POST-DENGUE ACUTE DISSEMINATED ENCEPHALOMYELITIS (ADEM)?

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Case Presentation: A 25-year-old male patient presented a clinical course suggestive of dengue, with a positive NS1 test. With less than 24 hours of symptoms, he began progressive mental confusion, focal seizures, and subsequent sensorium lowering. Brain magnetic resonance imaging was performed, which showed foci of diffusion restriction and hyper signal on T2/FLAIR bilaterally in the brainstem, hippocampus, and thalamus topographies, a pattern suggestive of encephalitis. An electroencephalogram was performed, which showed bursts of slow waves predominantly frontally bilaterally and rare epileptiform paroxysms in the parietal topography bilaterally. A lumbar puncture was performed, which showed only an increase in protein (100mg/dL). Treatment of seizures was performed with Phenytoin and Levetiracetam. Discussed as a team and opted to perform Immunoglobulin for 5 days, with subsequent significant improvement of the condition. At Hospital discharge, the patient had improved orientation and had no focal deficits. **Discussion:** Dengue is the main cause of encephalitis with normal cellularity in the cerebrospinal fluid in endemic regions. Although there are no characteristic lesions for dengue encephalitis, the patient had lesions characteristic for encephalitis in general, that is, involvement of the limbic system, in addition to hyperproteinorrachia and a suggestive clinical course. Therefore, dengue encephalitis was our main diagnostic hypothesis. However, there was also a differential diagnosis with ADEM, which is a post-infectious autoimmune encephalitis. Despite differences in pathogenesis and histological findings, their clinical course and signs in imaging tests can be very similar. Therefore, since it was not possible to rule out ADEM and given the patient's clinical severity at the time, we chose to perform an Immunoglobulin infusion for 5 days. **Final Comments:** There is no specific treatment for dengue encephalitis, with most patients having a good prognosis. ADEM can be treated with immunosuppressive medications. Given the diagnostic doubt, we chose to use Immunoglobulin, and the patient had a good response and recovery.

Doença Cerebrovascular

TL 1106124

BILATERAL SIMULTANEOUS NON-ARTERITIC ANTERIOR ISCHEMIC OPTIC NEUROPATHY IN A ELDERLY AFTER SPINE SURGERY: A CASE PRESENTATION

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Case Presentation: A 78 years-old woman, caucasian, a retiree, residing in Belo Horizonte, Minas Gerais State, Brazil. Patient with systemic arterial hypertension and right peripheral facial nerve palsy. Prior cataract surgery. Due to spinal stenosis, she was submitted to a thoracic lumbar spinal decompression procedure on March 31th, 2022. Performed for nine hours in a prone position without intercurrents. After eight hours, she awoke from anesthesia with psychomotor agitation and complaining of severe pain at the surgical site and reduced bilateral visual acuity. Progressed to loss of color vision and ultimately bilateral amaurosis. She denied other focal symptoms or headaches. No reports of mental confusion by assistant staff. Without a history of visual impairment. The patient presented to the neurological examination with a regular general condition, good spatial orientation, alertness, Glasgow Coma Scale 15, right corectopia, normal extrinsic ocular motricity, hand movement perception half a yard away, right peripheral facial nerve palsy, without meningeal irritation signs. Ophthalmoscopy revealed bilateral papilledema more evident on the left and retinal pigment epithelium atrophy, without signs of vasculitis. Magnetic Resonance Angiography of the skull revealed moderate stenosis of the left anterior cerebral artery, hypoplasia of the left vertebral artery, and moderate stenosis of the supraclinoid segment internal carotid artery, but with the good distal flow. Magnetic Resonance Imaging of skull without restriction on diffusion-weighted imaging or hyperintense signal on T2-weighted FLAIR. There was a gradual spontaneous improvement of the visual deficit. On April 04th, she reported a return to baseline visual acuity. She had uncorrected visual acuity in both eyes: 20/70 vision. **Discussion:** Postoperative visual loss following prone spine surgery occurs in from 0.013% to 1% of cases, and the most frequently quoted risk is 0.2%. It is attributed mainly to ischemic optic neuropathy. The multiple factors contributing to POVL may include; prolonged operative times, long-segment spinal instrumentation, anemia, intraoperative hypotension, greater estimated blood loss (EBL), microvascular pathology, and decreased percent colloid administration. **Final Comments:** Laboratory tests revealed no hematimetric abnormalities and no excessive bleeding or hemodynamic instability was reported during surgery, however, the operative time was prolonged.

Doença Cerebrovascular

TL 1106125

WYBURN-MASON SYNDROME: A RARE PHACOMATOSIS WITH MULTIPLE ARTERIOVENOUS MALFORMATIONS

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Case Presentation: An 18-year-old male patient, initially evaluated at the age of 2 for proptosis and visual impairment of the right eye. A red supraorbital skin lesion was perceived at that moment. There was no family history of neurological disorders and his parents were nonconsanguineous. Magnetic resonance imaging (MRI) revealed an arteriovenous malformation (AVM) involving the right cavernous sinus, which was treated with embolization. Despite the procedure, proptosis and edema of periorbital tissue continued to worsen and at the age of 12 he underwent a biopsy of frontal, infra-orbital and periorbital subcutaneous tissue which was compatible with a cavernous angioma. At the age of 18 he was evaluated at our institution. On neurologic examination there was marked proptosis with impairment on abduction, adduction and elevation as well as amaurosis of the right eye. Edema of periorbital, frontal and zygomatic regions on the right was also seen. Preserved strength and tone, but deep tendon reflexes were brisk, and Hoffman and Babinski signs were present. MRI revealed an expansive formation centered in the right orbit causing proptosis and compromising conal spaces with distortion of eye muscles and optic nerve. The lesion extended through the superior orbital fissure into the right cavernous sinus and to the contralateral orbit. **Discussion:** Wyburn-Mason syndrome is a rare, non-hereditary congenital disease, belonging to the group of neurocutaneous syndromes, with fewer than 100 cases reported since its first description in 1937. It usually affects the orbit, retina and brain, with predominant involvement of the midbrain. Lesions may also affect the skin, maxilla, jaw, pharynx, oral and nasal cavities. Headache, seizures and hemiparesis are some other neurological manifestations of the syndrome. Despite an uncertain etiology, an embryonic defect is believed to result in the dissemination of vascular lesions originating from the cephalic mesoderm along the migratory route, before the actual migration of vascular cells to their destination. **Final Comments:** Treatment of AVMs with endovascular techniques, surgery and radiosurgery are possibilities, but conservative treatment may be preferred due to the high risk of recurrence. However, bevacizumab, an anti-VEGF agent, has been shown to contain macular edema and improve visual loss in a short follow-up. Therefore, given the rarity of the pathology in question, the present report seeks to add to the literature.

Doença Cerebrovascular

TL 1106145

RECURRENCE OF STROKE IN A PATIENT WITH FIBROMUSCULAR DYSPLASIA

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Case Presentation: A 64-year-old woman, previously hypertensive, woke up with a sudden loss of strength on the right and dysmetria on the left. She was rescued, but with a time longer than 4.5 hours, which contraindicated the use of thrombolytics for stroke. Basic investigation was performed with cranial tomography at admission and after 24 hours, transthoracic echo, carotid and vertebral Doppler. After normal findings in all exams, the investigation continued with cranial magnetic resonance, which showed T2/FLAIR hypersignal in bridge and left cerebellum with restriction of water molecule to diffusion. Patient progressed using secondary prophylaxis with dual antiplatelet therapy. However, 17 days later, she was Hospitalized again, with a worsening of the same symptoms. A new CT scan 1 day after the new condition showed subacute hypodensity in a capsule on the left. Cerebral angiography showed occlusion of vertebral arteries and a pattern of multiple parietal irregularities, extending throughout its course in the "string of beads" appearance. Secondary prophylaxis was changed to anticoagulation with apixaban, in addition to motor rehabilitation and outpatient follow-up. **Discussion:** Fibromuscular dysplasia (FMD) is a non-inflammatory and non-atherosclerotic disease that courses with arterial stenosis, occlusion, aneurysm, dissection and arterial tortuosity, more frequent in young adult women, the internal carotid artery is more frequently affected, histological form more common is fibroelastic dysplasia with thickening of the media and muscular layer with destruction of the elastic lamina. Diagnosis is by vessel study. Genetic predisposition, chronic trauma, stretching of the involved vessel and hormonal factors are believed to have been suggested as etiologic agents. The clinical presentations are determined by the affected artery. In this case, the patient presented a recurrent and relapsing symptoms in a short period of time, in two different territories, first in the anterior inferior cerebellar artery territory, and later in the lenticostriate artery territory. **Final Comments:** FMD associated with neurovascular events can be treated with antiplatelet therapy, blood pressure control, or treatment with angioplasty with or without stent implantation. However, in this case, antiplatelet therapy was not enough for prevention in a short period of time, opting for anticoagulation with apixaban, which kept the patient stable without new neurovascular events.

Doença Cerebrovascular

TL 1106157

SEPTIC THROMBOSIS OF THE TRANSVERSE AND SIGMOID SINUS, DISCUSSION ABOUT THE INTRODUCTION OF FULL ANTICOAGULATION: CASE PRESENTATION

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Case Presentation: Septic sigmoid sinus thrombosis is a rare disease and is commonly caused by middle ear infections, with *Staphylococcus aureus* being the main etiologic agent. Clinical manifestations are diverse and headache is the most frequent complaint followed by focal deficits such as cranial nerve involvement. Radiological investigation and lumbar puncture are important in aiding the diagnosis. To discuss the controversial use of full anticoagulation in cases of septic thrombosis, describing a patient treated at a tertiary Hospital in which it was decided not to introduce this therapy. **Discussion:** G.S.P.L, 15 years old, male, admitted with a report of otalgia due to refractory oral antibiotic therapy. Evolved with vomiting, loss of appetite, persistent fever and periods of sleepiness. A neurological examination showed right appendicular ataxia and purulent discharge from the external auditory canal directly to otoscopy. Performed cranial tomography with diffuse veiling of the cells of the mastoid and the right middle ear and hypodensity of the right cerebellar hemisphere with a nonspecific appearance. Cerebrospinal fluid was collected with pleocytosis and hyperproteinorraquia. During Hospitalization, a Cranial Magnetic Resonance with Angioresonance was performed, which showed opacification of the mastoid cells and tympanic cleft on the right and filling failure in the sigmoid and adjacent transverse sinus, inferring venous thrombosis. Broad-spectrum antibiotic therapy was started for a long time and a surgical approach was scheduled with the otorhinolaryngology team. **Final Comments:** When questioned about the use of anticoagulants, the benefit was not superior to the risks related to this therapy, the use prevents the persistence of the thrombus, but it can facilitate the release of septic emboli and increase the risk of bleeding. Thus, in the case presented, a conservative approach was chosen, based on antibiotic therapy and neurological surveillance without the introduction of anticoagulation.

Doença Cerebrovascular

TL 1106162

PERIMESENCEPHALIC HEMORRHAGE: A RARE ENTITY WITH POSSIBLE COMPLICATIONS

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Case Presentation: A 37-year-old woman, previously healthy, was admitted with a 5-day history of occipital and nuchal headache with irradiation to the back, inedit, progressing to worst intensity in 5 to 10 minutes, with nausea associated. It was also evidenced by high blood pressure (220x130 mmHg). 3 days after the initial symptoms, she had mental confusion and irritability. The neurological examination revealed neck stiffness and parisis of the right lateral rectus. A brain non contrast computerized tomography (NCCT) was performed - we found hyperdense images near the mesencephalic cisterns. Then we performed a digital subtraction angiography (DSA), which was normal. The following day, the patient presented worse of mental confusional, papilledema, complete ophthalmoplegia in the right eye and parisis of the lateral rectus in the left eye. A brain magnetic resonance imaging (MRI) showed multiple areas of restricted diffusion in the occipital lobe, bilateral cerebellar and right front. A MRI angiography with vessel wall was performed and showed multiple stenotic focus in large vessels (ACID, ACMD, ACPD), and fine enhancement still on their walls. Based on these findings, and considering the severity and aggressiveness of this disease, we treated her for central nervous system (CNS) vasculitis with pulse therapy (methylprednisolone 1 gram for 5 days). After the third day, the patient improved her mental confusion and her ophthalmoplegia. In order to evaluate if the patient's improvement was related to radiologic findings, we therefore submitted her to another DSA. The findings were related to cerebral vasospasm than to CNS vasculitis. 25 days after the initial symptoms, we performed a new DSA, which was normal. Her final diagnosis was delayed cerebral ischemia (DCI) due to a perimesencephalic hemorrhage (PMH). The PMH's clinical features described in some reviews were headache with instantaneous or more gradual onset, accompanied by nausea and vomiting. It's rare, but can also present with loss of consciousness, transient focal symptoms or epileptic seizure. Complications are not common, but can occur hydrocephalus, DCI, rebleeding, amnesia and cranial nerve palsies. The diagnoses of PMH can be done with a NCCT. PMH with short term complications are rare and long-term outcome is excellent. Although it is a condition that remains underdiagnosed. We understand very few aspects of this condition. But knowledge about this disease is still under development.

Doença Cerebrovascular

TL 1106179

SPONTANEOUS DISSECTION OF THE BASILLAR ARTERY: CASE PRESENTATION

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Case Presentation: Female, 16 years old, history of migraine, without other comorbidities. She presented intense pain in the cephalic region, evolving immediately with loss of consciousness, without previous traumatic brain injury. Therefore, she was admitted to an intensive care unit. Brain computed tomography with contrast performed in the acute phase showed hyperdensity in the basilar artery with filling failure after contrast, suggesting a thrombus inside it, without hemorrhagic transformation. The patient was admitted by neurology after 14 days ofictus presenting tetrasegmental hypotrophy, reduced tone and complete quadriplegia provided. Tetrasegmental hyperreflexia with pyramidal signs. Tromner and Hoffman are present. Plantar cutaneous reflex in extension to the left and indifferent to the right. Absence of meningeal signs. Isophotoreactive pupils. Slowed ocular motricity. Facial mimicry without alterations, paralysis of the conjugated gaze to the left, nausea reflex absent. She underwent cranial magnetic resonance angiography 25 days after the event, showing a caliber reduction of about 50% in the middle third of the basilar artery. During Hospitalization, a negative investigation was performed for vasculitis and thrombophilia. Dissection is the loss of structural integrity of the arterial wall, allowing blood to pool between the arterial layers. Occlusive disease of the basilar artery most often presents as pons ischemia. The paramedian pontine base contains the descending long motor tract and the cerebellar fibers that cross it. The paramedian tegmentum contains mainly oculomotor fibers. Altered level of consciousness is an important sign in patients with basilar artery occlusion. They may present with coma when the bilateral medial pontine tegmentum suffers ischemic insult. In the case presentation, a cerebrovascular accident was evidenced after spontaneous dissection of the basilar artery, compromising the brainstem, generating Basilar Artery Syndrome characterized by motor and oculomotor signs and alteration in the level of consciousness.

Doença Cerebrovascular

TL 1106191

UNUSUAL PRESENTATION OF ISCHEMIC STROKE DUE TO STENOSIS OF PERSISTENT PRIMITIVE HYPOGLOSSAL ARTERY: A CASE PRESENTATION

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Case Presentation: A 74-year-old man only treated for benign prostatic hyperplasia underwent elective magnetic resonance imaging (MRI) to investigate left hypoacusis with months of evolution. An area of bilateral parieto-occipital diffusion restriction was detected with hypersignal in fluid-attenuated inversion recovery and hyposignal in the apparent diffusion coefficient. The patient was admitted without any neurological deficits, but reported a recent episode of vertigo lasting seconds in addition to reduced visual acuity. He had a blood pressure of 129/74mmHg and capillary blood glucose level of 97mg/dL. Therefore, a dual antiplatelet therapy was initiated. During the investigation, carotid and vertebral Doppler revealed non-significant carotid stenosis. Electrocardiography revealed sinus rhythm and Holter monitoring detected minimal changes. Transthoracic echocardiography identified an enlarged left atrium and ventricle with concentric remodeling. Computed tomography angiography revealed bilateral vertebral artery (VA) hypoplasia and a persistent primitive hypoglossal artery (PPHA) originating from the right internal carotid artery, giving rise to the basilar artery, in addition to a hypodense plaque during its course in the hypoglossal canal, reinforced by vessel wall MRI. Craniocervical junction computed tomography did not reveal any extrinsic compression. **Discussion:** PPHA is a rare vascular anomaly that is usually detected incidentally during cerebral angiography and is generally asymptomatic. It is mainly associated with aneurysms, probably because of the anomalous structure of the vessel wall, which can expose the basilar trunk to excessive hemodynamic stress. As most patients have hypoplastic VA, the blood supply to the posterior circulation travels through PPHA. Therefore, in ischemic stroke patients with PPHA, both the anterior and posterior circulations are often involved. We present the case of a patient with PPHA and bilateral hypoplasia of the VA, who presented with a bilateral oligosymptomatic stroke with a hemodynamic component caused by a plaque found in the hypoglossal artery, which is an uncommon location of stenosis. **Final Comments:** We demonstrate a case of rare anatomic variation communicating the anterior and posterior circulations. In this way, we show the importance of knowing the anatomical variations in cerebral circulation to identify possible unusual presentations of stroke and to choose the best secondary prevention.

Doença Cerebrovascular

TL 1106228

STROKE IN A YOUNG PERSON: SLE AND FAS ASSOCIATED WITH MULTIPLE EVENTS: A CASE PRESENTATION

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Case Presentation: Patient 25 years old, with a history of diagnosed idiopathic dilated cardiomyopathy and a stroke in July 2021, was admitted to the service with a decreased level of consciousness, mental confusion and right heparesis, preceded by headache with onset of the previous three days. Cranial CT showed acute cerebellar hypodensity on the right, with chronic hypodensity in the midbrain and right parietal lobe. In other exams, normocytic normochromic anemia, lymphopenia, hypoalbuminemia, proteinuria and microscopic hematuria were evidenced. During complementary investigation, right vertebral dissection was evidenced in AngioMR. Taking into account the set of findings, an evaluation of Rheumatology was requested. Rheumatologic screening was positive for Lupus with associated Antiphospholipid Antibody Syndrome (FAN 1: 1280; Anti-SM: positive; anti-cardiolipin IgM and IgG: positive, lupus anticoagulant positive). The patient evolved with pre-renal failure, due to probable class 3/4 lupus nephritis, acute respiratory failure of cardiogenic origin, with bilateral transudative pleural effusion. **Discussion:** About 10% of stroke cases occur in people under 45 years, with acquired thrombophilia and autoimmune diseases being the main precipitating factors. SLE is an important cause, as it is a chronic systemic inflammatory disease. About 36% of SLE cases are associated with Antiphospholipid Antibody Syndrome (APS), responsible for more than 20% of strokes in young people and the most frequent cause of acquired thrombophilia. The mechanism of injury is mainly due to alterations in coagulation, generating a prothrombotic state of hypercoagulability, predisposing to multiple events. **Final Comments:** In the case presented, the patient had SLE and APS associated with a high level of disease activity, even in the presence of adequate treatment. A fact that predisposed the patient to present several thrombotic events, with evidence of three strokes and bilateral deep vein thrombosis of the lower limbs, even in the presence of full anticoagulation. Evidencing that in these patients the main decisive factor is the autoimmune disorder.

Doença Cerebrovascular

TL 1106229

STROKE ASSOCIATION WITH EXTENSIVE CENTRAL VENOUS THROMBOSIS IN THE CONTEXT OF NEUROINFECTION

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Case Presentation: A 48 old female patient was admitted to the emergency department due to several episodes of seizures without recovery of consciousness lasting more than 5 minutes. There was no record of previous diseases besides one Hospitalization due to status epilepticus four months earlier, from which she was discharged with valproate 500 mg once a day. Nonetheless, the patient was known to live in precarious situation and to be a frequent user of alcohol, tabaco, crack and cocaine. At the admission, the patient presented comatose, however without vital signs alterations. She was stabilized after pharmacological measures and was performed intubation for clinical support. Physical examination revealed left hemiparesis, worse at the upper limb and right eye mydriasis. The Neurological Team was required due to symptoms being presented within time for thrombolytic use in case of acute stroke. A head computed tomography (CT) scan revealed intense cerebral oedema and hyperdensity of the cerebellar tentorium. At CT angiography it was observed absent flow in inferior sagittal sinus, straight sinus and bilateral transverse sinus. Patient was conducted as a case of Central Venous Thrombosis and transferred to the intensive care unit. Patient recovered partially with persistence of right eye mydriasis and after waking up, presented right ptosis. Further investigation revealed on Magnetic Resonance Restricted diffusion within the right lateral portion of the pons and midbrain, identifying compatible ischemia with clinical findings and pachymeningeal enhancement on cerebellar tentorium too. It was also performed an angiography that observed absent flow in sinuses and diffuse small venous engorgement leading to the hypothesis of ischemia associated with low venous flow. Fluid analysis showed discrete rise of cellular count (06 cells with 70% lymphocyte) hyperproteinorachia (66 mg/mL) and hypoglycorrhachia (CSF to serum glucose ratio 0,45). Granulomatous disease such as tuberculosis was raised as a suspicion. On CT scan, the patient had a typical diffuse "tree-in-bud" pattern, consolidations and cavitations. This report intention is to contribute to the differential diagnosis and relevance of neuroinfection causing CVT which can also be responsible for arterial ischemia. Association of epidemiological features with unusual differential diagnosis were vital to deliver the right treatment for the patient and could be used as an example of a good clinical practice.

Doença Cerebrovascular

TL 1106233

SPONTANEOUS BILATERAL CAROTID DISSECTION: A RARE PRESENTATION OF ACUTE ISCHEMIC STROKE

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Case Presentation: A 57-year-old male patient was referred from an Emergency Care Unit to our Hospital due to a sudden loss of consciousness associated with headache and high blood pressure at 10:55 on 15/04/2022. There was no history of neck pain, trauma, fall or substance use or abuse. The patient was admitted intubated due to Glasgow Coma Scale < 9. Computed tomography was performed, which showed slight left basal nuclei and bilateral frontoparietal subcortical hypodensity, with no bleeding. A new computed tomography was performed the next day, which showed progress of the initial lesion and a small right frontal cortical hypodensity. The patient underwent magnetic resonance imaging that showed DWI increased signal and reduced ADC values involving the frontal cortex, frontoparietal white matter, basal nuclei and internal capsule of both hemispheres with hyperintense signal on T2/FLAIR, suggesting ischemia. Furthermore it showed T2 and DWI high signal crescent sign indicating intramural hematoma of both internal carotid arteries and angiography was performed the next day which also suggested bilateral carotid dissection and ruled out other causes. **Discussion:** Dissection of vertebral and carotid arteries are known causes of ischemic stroke and transient ischemic attack. Dissection can occur due to an external factor, such as trauma, neck manipulation, fall, or due to an intrinsic vessel disease, such as fibromuscular dysplasia, or spontaneously. Bilateral carotid dissection is a rare presentation of the phenomenon. In the literature, these cases were associated with blunt trauma, Eagle syndrome, medication use, Ehlers-Danlos syndrome, and others. Spontaneous bilateral carotid dissection is an even rarer presentation of neck artery dissection, especially in a patient without previous history of vessel disease. **Final Comments:** Spontaneous bilateral carotid dissection is a rare presentation of acute ischemic stroke and can cause sudden loss of consciousness due to bilateral infarcts in the brain. This case illustrates this rare presentation and the importance of performing a vessel study in a patient with a history suggestive of stroke for the evaluation of cervical and intracranial vessels.

Doença Cerebrovascular

TL 1106234

PACHYMEINGITIS: A CASE PRESENTATION

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Case Presentation: A 48 years old female, with history of headache with phenotype of migraine with visual aura. In 2005 she developed headache with pattern change, suggestive of intracranial hypertension, and targeted propedeutics was performed. On clinical and imaging investigation, indirect signs of intracranial hypertension were found on skull MRI such as: partially empty sella turcica, enlargement of the perioptic liquoric spaces and of the cavum of meckel, in addition to regular and diffuse pachymeningeal thickening at the level of the cerebellar tentorium, mainly on the right, as well as of the inter-hemispheric sickle, in its posterior aspect without a clear nodular aspect, a nonspecific finding from the etiological point of view, but may be associated with diverse conditions, such as pachymeningitis. The CSF was altered, with increased opening pressure (29 and 30); and inflammatory features (cells: 11 (leucocytes 97%), no macrophages, glucose: 58, prot: 119 ldt: 23). Ophthalmology evaluation with campimetry was without significant changes. Rheumatological and serological tests were negative, except for a high serum IgG 4 value (260.1 - VR: 4-86). Thus, he started treatment for IgG4 hypertrophic pachymeningitis with 1mg/kg/day Prednisone and Azathioprine 2-3mg/kg/day. After starting the medication, there was a significant improvement in symptoms, but with a side effect attributed to the corticosteroid, which is currently still being weaned. **Discussion:** IgG4-related hypertrophic pachymeningitis is only one of many possible IgG4-related diseases, it denotes inflammation and thickening of the dura mater and often comes associated with systemic symptoms such as fever, weight loss or abdominal pain. Neurological disorders in these patients are often associated with nerve or vascular compression, leading to focal deficits, but diffuse inflammation of the dura can lead to more diffuse presentations such as headache, cervical stiffness, and seizures. Its diagnosis remains a great challenge because of the compendium of possible presentations and its indolent character, which ends up delaying the search for medical assistance by patients. **Final Comments:** The early diagnosis of hypertrophic pachymeningitis is essential to stop disease progression and brain damage. The management of the disease still is a challenging due to its recent description in 2003, with a pathophysiological picture not yet fully understood.

Doença Cerebrovascular

TL 1106244

AN UNUSUAL PRESENTATION OF AN ACUTE ISCHEMIC STROKE DUE TO LARGE VESSEL OCCLUSION

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Case Presentation: A 58-year-old male with unremarkable medical history, was admitted in the emergency department presenting sudden abnormal movements in right side limbs. At initial neurologic evaluation, the patient showed only hemichorea on the right side, and his National Institute of Health Stroke Scale (NIHSS) score was zero. After excluding cerebral hemorrhages in brain CT scan and no contraindications for chemical thrombolysis, Alteplase was administered. Cranial and cervical CT angiography showed tandem occlusion in the left internal carotid artery and middle cerebral artery. Brain MRI displayed, an area of restricted diffusion in the left insula. He was admitted at Stroke Unit for etiologic evaluation, in addition to acute phase care. There was a fluctuation of deficits in the first 24 hours, nonetheless the patient was discharged after 4 days of Hospitalization with complete improvement of abnormal movements. The mechanism was defined as athero-embolic with internal carotid artery occlusion, probably chronic due to flow distribution pattern evidenced in carotid, vertebral and transcranial doppler scan. **Discussion:** Although hemichorea is an uncommon symptom of AVC, studies reveal that vascular etiology is the most prevalent non-genetic cause among the various diseases that induce chorea. The results of a study published in Neurology showed lesion network mapping to anatomically heterogeneous lesions that can cause hemichorea-hemiballismus, and while the findings failed to localize a single region, it did show a 92 percent connectivity to the posterolateral putamen, which plays a central anatomical and functional role within the basal ganglia-thalamocortical motor loop, emphasizing its importance in this unknown pathophysiology. Research indicates that most patients experience complete remission of hemichorea-hemiballismus within a few days after stroke vascularization. The underlying cause of the clinical finding must be treated, as in the case of the highlighted patient, which even without thrombectomy, presented full remission post thrombolysis. **Final Comments:** A sudden onset hemichorea should be treated as an acute stroke until proven differently. This case supports the idea that the cortico subcortical transition of the insular region has a role in the genesis of aberrant hyperkinetic movements, as well as a probable functional linkage with the basal ganglia

Doença Cerebrovascular

TL 1106245

HYPERKINETIC MOVEMENTS OF HYPERACUTE ONSET: A CASE PRESENTATION TREATED WITH RTPA

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Case Presentation: 64-year-old woman, hypertensive, previously functional (Rankin 0). She had sudden dysarthria, central facial palsy (CFP), and left hemiparesis. There was the resolution of the deficits and a clinic picture of hyperkinetic movements began. She was admitted to the neurological emergency with a stroke in the therapeutic window for thrombolytic treatment and on physical examination, chorea movements were observed, characterized by brief, random and irregular involuntary muscle contractions affecting the left side, associated with a slight effacement of the ipsilateral nasolabial fold and slurred speech. NIHSS: 2 (dysarthria and CFP). Cranial computed tomography: 10 aspects and cranial computed tomography angiography: occlusion in the proximal segment of the right middle cerebral artery. No dysglycemia. After ruling out exclusion criteria, thrombolysis was performed. A few minutes after the rTPA, she had a resolution of the movement disorder. Control image delimited lesion in Lentiform Nucleus and Insula section on the right. The investigation of the mechanism evidenced Atrial Fibrillation as, a probable cardioembolic mechanism. The deficits were discharged stable and with outpatient follow-up. **Discussion:** Movement changes can be primary (genetic) or secondary to cerebrovascular disease, autoimmune conditions, metabolic causes, neoplasms (mass effect or paraneoplastic syndromes), iatrogenic (drug-induced), or trauma. Movement disorders secondary to stroke are a rare manifestation of cerebrovascular disease. They account for up to 22% of secondary movement disorders and develop in 1-4% of stroke patients. The main structures affected are the basal ganglia (44%) and thalamus (37%), although there are case studies that also affect the cerebral cortex. Vascular chorea is rarely progressive and may regress spontaneously in about half of patients, but others may persist with symptoms and need to be treated. **Final Comments:** Although rare, the initial presentation of acute cerebral ischemia may present with hyperkinetic movement disorders and should be an important differential diagnosis for acute-onset chorea. Thus, rapid and decisive action must be performed with an acute phase protocol.

Doença Cerebrovascular

TL 1106246

SPONTANEOUS SPINAL EPIDURAL HEMATOMA AS A STROKE MIMIC IN TIME WINDOW FOR THROMBOLYSIS

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Case Presentation: A 73-year-old woman presented to the Emergency Department with right hemiparesis and ipsilateral neck pain with sudden onset three hours before arrival. Her past medical history included hypertension and diabetes, but no remarkable history of neurological disease. Code Stroke was activated. At admission, her blood pressure was 220/105 mmHg and her blood glucose level was 213 mg/dL. Her National Institutes of Health Stroke Scale score was 8 due to a right-sided hemiplegia; there was no facial palsy. CT scan showed no signs of recent ischemic infarction or intracranial hemorrhage. CT angiography showed no sign of arterial occlusion, critical stenosis or dissection. As the patient was taken back to the emergency room, her motor deficit improved substantially spontaneously, but then got worse again. The team decided to proceed with thrombolytic therapy, but the time window ran out due to poor blood pressure control despite increasing doses of intravenous vasodilator drug. The patient was then started on antiplatelet therapy for secondary prophylaxis. 24 hour follow-up CT scan showed no changes from the initial exam. The patient performed an MR angiography for persisting neck pain, which revealed a posterolateral right-sided cervical epidural hematoma. Because her motor deficit had remained stable over the last day, a conservative approach was chosen. **Discussion:** Spontaneous Spinal Epidural Hematoma (SSEH) represents a rare cause of spinal cord compression and neurological emergency. Paraplegia or tetraplegia are the most common presentation, while hemiparesis is considered to be a rare feature, more commonly associated with cervical epidural hematoma. In these rare stroke mimicking presentations, thrombolytic therapy, anticoagulation or treatment with an antiplatelet agent can lead to hematoma expansion and need for a prompt surgical evacuation. Although clinical presentation may be similar to a stroke, previous studies have reported persistent neck or back pain and absence of facial weakness as a high index of suspicion of SSEH, and may help to distinguish from cerebral etiologies. **Final Comments:** This case represents a rare stroke mimic with potential harm associated with thrombolytic therapy, so SSEH should be always suspected as a differential diagnosis of ischemic stroke to avoid fatal outcomes.

Doença Cerebrovascular

TL 1106251

A CASE OF STROKE IN A PATIENT WITH DOWN SYNDROME ASSOCIATED WITH MOYAMOYA SYNDROME

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Case Presentation: A patient with a previous diagnosis of Down Syndrome (DS) was sent to a stroke referral Hospital ten days after a stroke. At the initial neurological examination, he manifested a state of alertness, in addition to being uncooperative, aphasic, and unresponsive to commands. He presented facial paralysis, hemiparesis, and hypoesthesia on the left, indicating a suspected cerebral infarction. A Cranial Computed Tomography (CT) showed hypodensity in the right middle cerebral artery (MCA) and Ecass 1 transformations. Due to the ictus having occurred ten days before admission, the approach remained conservative at the subacute stroke division. On CT angiography, there were occlusions in M1 segments of MCA and Moyamoya pattern, manifested by stenosis of the left internal carotid artery and apparent hypertrophy of the collateral lenticulostriate arteries. The patient evolved clinically and hemodynamically stable, but still alert, uncooperative, and aphasic. After discussion with a neurologist, the patient was discharged from the Hospital with follow-up care in health services. **Discussion:** Moyamoya disease represents an arteriopathy with stenosis in segments of the internal carotid arteries, with hypertrophy of collateral vessels. Former characterized as an idiopathic condition, it's now associated with genetic disorders and a higher risk of cerebral infarction, which is relevant in this case presentation, due to the aggravating factor related to DS. Such an etiological correlation for arteriopathy resides in the fact that the gene for the alpha chain of collagen VI is located on chromosome 21, as well as the genes of other proteins that interfere with arterial physiology. Moyamoya diagnosis in patients with DS occurs at an average age of 9.7 years. The previous diagnosis helps avoid the commonly observed course of recurrent stroke. Anticoagulant preventive measures can also be implemented to reduce complications. In the report, the patient with DS did not receive follow-up care by health professionals until the age of 31, when he presented with the acute condition of arteriopathy that culminated in stroke, revealing the risks of late observation of the disease. **Final Comments:** Due the evidence of severe conditions after the correlation between Down Syndrome, Moyamoya disease and the reported stroke, the greater dissemination of knowledge associated with the case is essential to health professionals is necessary to define the appropriate conduct and treatment.

Doença Cerebrovascular

TL 1106252

INTRAVASCULAR LYMPHOMA PRESENTING STROKE AND ISCHEMIC MYELOPATHY

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Case Presentation: A 78-year-old female presented with subacute and progressive lower limb weakness and numbness, urinary retention, confusion and weight loss. Her past medical history included hypertension, trigeminal neuralgia and meningioma resection eleven years before presentation. Upon admission, neurological examination showed impaired attention and executive dysfunction, along with asymmetric spastic paraparesis (worse on the right leg) and a dermatomal sensory deficit level at T4. Laboratory findings were noticeable for pancytopenia. Patient underwent neuroimaging. Brain MRI revealed multiple areas of T2/FLAIR hyperintensities in addition with restricted diffusion in the parasagittal gray and white matter bilaterally, overlapping with internal and external watershed zones of the frontal, parietal, and occipital lobes, indicating an ischemic etiology. Spine MRI showed hyperintensities on T2-weighted sequences, predominantly in the central and upper segments of thoracic spinal cord, also consistent vascular insult. CSF examination was normal. Because of pancytopenia, a bone marrow biopsy was obtained, which confirmed an intravascular non-Hodgkin B cell lymphoma. Chemotherapy was initiated afterwards and patient is showing gradual neurologic improvement during follow-up. **Discussion:** Intravascular large B cell lymphoma (IVLBCL) is a rare subtype of extranodal large B-cell lymphoma with an aggressive course, which may affect multiples organs, including central nervous system. Its main feature resides on the fact that tumor cells have a confined proliferation within the lumen of small and medium-sized vessels, differentiating IVLBCL from other forms of lymphoma. Even though any organ may be involved, skin and CNS are the most commonly affected sites. Most frequent neurological symptoms are cognitive impairment (encephalopathy and dementia) and myelopathy with neuroimaging showing infarct-like lesions due to vessel occlusion by tumor cells. IVLBCL poses a diagnostic challenge because of its protean manifestation and lack of specific laboratory and imaging findings. In addition, PET-CT is usually negative because this type of tumor tends to remain intravascular. **Final Comments:** The importance of this case lies in the fact that the combination of concomitant brain and spinal cord scattered infarction with systemic findings (pancytopenia and constitutional symptoms) must include this rare, underrecognized disease in the differential diagnosis.

Doença Cerebrovascular

TL 1106262

MANIC EPISODE ASSOCIATED WITH ISCHEMIC STROKE: CASE PRESENTATION

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Case Presentation: A 61-year-old man was admitted to a psychiatric inpatient center with a manic episode. He had a diagnosis of hypertension and a past history of a short-lived mild depressive episode 4 years ago with spontaneous resolution. The patient had been in his usual state of health until 15 days before the admission, when he presented mood alteration, labile affect, disinhibition, grandiose ideas and persecutory delusions, noted by family and coworkers. At that point, the patient was admitted to the inpatient setting. The neurological exam showed preserved strength, sensitivity, coordination and gait. Laboratory screening tests with serologies showed no alterations. The brain magnetic resonance imaging showed signs of subacute ischemic event in the posterior branch of the left internal capsule and mild microangiopathy. A cerebral magnetic resonance angiography showed no hemodynamically significant stenosis. A treatment with lithium 900mg and olanzapine 5mg was started with great improvement. When the patient was discharged after 3 weeks, he returned to work and kept follow-up on an outpatient basis. **Discussion:** Mania is characterized by affective disturbances and can manifest with various symptoms, including increased talkativeness, flight of ideas, racing thoughts, abnormal distractibility and increased energy. It is reported to be an uncommon manifestation after stroke. In this setting, its frequency is < 1%, compared with depression and other mood disorders. Post-stroke mania should be considered in any manic patient who has concomitant neurological deficits and is older than expected for the onset of primary mania. This presentation is most common when lesions occur in the right cerebral hemisphere, specially in the thalamus and caudate nucleus, although left-sided lesions have also been reported. From a review of the literature, it was noted that most of the data about this topic focuses on case presentation. **Final Comments:** The case presented is a rare episode of mania after ischemic stroke with unusual involvement of the left internal capsule which emphasizes the importance of neuroimaging in the first episode of mania. A longer follow-up is needed to assess the recurrence of symptoms and, in this case, distinguish between causality and correlation. Further studies are required to better understand this association of events.

Doença Cerebrovascular

TL 1106319

SUBARACHNOID HEMORRHAGE DUE TO SPONTANEOUS RUPTURE OF AN ARACHNOID CYST: CASE PRESENTATION

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Case Presentation: A 37-year-old female patient seeks medical care at a private Hospital in São Paulo. She was referring sudden, pulsating headache, with photophobia and phonophobia. The neurological examination was unremarkable. As the only previous medical history, she had deep venous thrombosis in 2014, since then she was irregularly using of Warfarin. She underwent CT and MRI of the skull which showed enlargement of the bilateral retrovermian and retrocerebellar cerebrospinal fluid space, with thin membranes/septations inside, and heterogeneous content, with deposition of hemoglobin degradation products. Such findings are consistent with a retrovermian arachnoid cyst with hemorrhagic content. No abnormalities were found in cervical and intracranial arteries with angiotomography. The prothrombin time was normal and international normalized ratio (INR) was 1. The patient was discharged after 2 days of Hospitalization with improvement in the headache. **Discussion:** Arachnoid cysts are collections of fluid located between the meningeal membranes. They are congenital and are formed due to a valve defect in the arachnoid membranes, which facilitate the passage of cerebrospinal fluid into its interior, leading to the formation of the collection. Most arachnoid cysts are an incidental finding. About 1 to 2% of the population has a subarachnoid cyst and symptoms can be seen when the cyst increases in size or bleeds inside the cyst. Thus, the case presented is a rare condition. **Final Comments:** The patient presented a headache with a warning sign – thunderclap headache – and was investigated aiming to rule out a subarachnoid hemorrhage which is frequently associated with aneurysm rupture. The neuroimaging findings disclosed a rare complication of the arachnoid cyst. The most important risk factor for arachnoid cyst hemorrhage is trauma. In the case described, there was no history of trauma. Another possibility would be the use of an anticoagulant; however, this association was ruled out with normal prothrombin time. The risk of intracystic hemorrhage in people using anticoagulants is unknown and so far there is no contraindication to their use in this population.

Doença Cerebrovascular

TL 1106329

BLUE TOE SYNDROME AND STROKE: A CASE PRESENTATION ON SIMULTANEOUS CARDIOEMBOLISM

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Case Presentation: A 76 years old male, with chronic atrial fibrillation (AF), CHA₂DS₂-VASc: 8, was admitted with sudden homonymous left hemianopsia and mild left hemiparesis (NIHSS 2) associated with fixed cyanosis of the first left pododactile. A diagnosis of acute right posterior cerebral artery occlusion was made concomitance with acute occlusion of the dorsal digital artery. The patient had previously suspended warfarin treatment 30 days before the event. Brain magnetic resonance showed a definite ischemic area in the right occipital, left medial occipital, and left cerebellar region. A transesophageal echocardiogram (TEE) showed a mobile thrombus in the left atrial appendage (LAAT) measuring approximately 1.1 cm x 0.8 cm. The vascular surgery team recognized systemic embolization to the distal arterial bed, and only medical therapy with anticoagulants was recommended. Abdominal ultrasound revealed discrete atheroma plaques in the abdominal aorta without any other signs of embolic events at different sites. The warfarin anticoagulation was resumed, but the patient presented a labile INR, so anticoagulation was switched to dabigatran with good evolution. **Discussion:** AF is a significant cause of systemic and especially brain embolization. Dislodgment of thrombus from the left atrium into the circulation is one possible mechanism that explains embolic episodes in atrial fibrillation patients. Although blue toe syndrome was initially described in patients with microemboli from proximal atherosclerotic lesions, an annual rate of 0,4%/yr was documented in atrial fibrillation patients in antiaggregation therapy. Prevention of stroke and systemic embolic events had been successfully demonstrated with DOACs use in AF patients, but in some series, the prevalence of LAAT in patients anticoagulated for at least three weeks with DOACs can be as high as 5,8% depending on the TEE screening strategy used a similar rate in anticoagulation with warfarin. **Final Comments:** We describe a simultaneous blue toe syndrome and ischemic stroke secondary to multi-source embolic source treated with anticoagulation with dabigatran. DOACs use can be an option in cases like these, particularly in patients with labile INR or contraindications to warfarin use.

Doença Cerebrovascular

TL 1106395

CASE PRESENTATION MEDULLARY VASCULAR ACCIDENT DUE TO DISSECTION OF THE DESCENDING BRANCH OF THE ABDOMINAL AORTA ARTERY HOLY HOUSE OF MERCY OF SANTOS

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Case Presentation: A 65 years-old man, previously diabetic and hypertensive. Admitted to the intensive care unit, with sudden back pain, associated with weakness and paresthesia of the lower limbs. During Hospitalization, it evolves with urinary retention, paraplegia and lower limb anesthesia. On admission, an echocardiogram was performed in atrial fibrillation rhythm with diffuse hypokinesia. Magnetic resonance imaging of the skull with areas of restriction to diffusion in the occipital, periventricular and corona radiata regions and of the thoracic spine with signal alteration in the anterior and central situation of the spinal cord, in the T4-T7 region, which correspond to spinal infarction. To proceed with the investigation, a chest angiotomography was requested, which showed thoracoabdominal aortic dissection (type IIIB), with thrombus in the descending aorta and bilateral pericardial and pleural effusion. After the examination findings, the patient was transferred to a referral Hospital and surgical correction of the dissection. **Discussion:** Aortic artery dissection is the rupture of the aortic intima that allows blood to penetrate the aortic wall and promote the discontinuity of vascular layers. The event may be secondary to the presence of an aortic aneurysm, characterized by irreversible dilation of the vessel wall, whose predisposing factors are smoking, hypertension, and atherosclerosis, congenital, genetic and infectious diseases. When dissection occurs, there is a risk of complications, with ischemic phenomena being observed upstream and downstream of the initial orifice, such as spinal cord ischemia, a rare complication with significant neurological symptoms and a high risk of sequel. To report a case of medullary vascular event secondary to abdominal aortic dissection, as a differential diagnosis of acute flaccid paralysis using data from medical records. **Final Comments:** The case in question raises the discussion about a rare type of complication resulting from abdominal aortic dissection, spinal ischemia, which must be remembered and investigated in cases of acute flaccid paralysis, aiming at early diagnosis and treatment making it possible to reduce neurological sequel.

Doença Cerebrovascular

TL 1106397

BOTULINUM TOXIN AS TREATMENT FOR VASCULAR HEMICHOREA–HEMIBALLISM: A CASE PRESENTATION

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Case Presentation: A 55-year-old male patient started with altered sensitivity in the right hemibody seven days before admission, in addition to abnormal movements in the right arm, starting after waking up, with no apparent triggers and which improved during sleep. He had a previous history of unspecified cardiac arrhythmia, with irregular use of propafenone, acetylsalicylic acid and rosuvastatin. Physical examination showed abnormal movements on the right side, sparing the face and worse in the proximal part of the right upper limb, with low frequency, high amplitude, arrhythmic and erratic, characteristics of hemichorea-hemiballismus. Biochemical and serological tests were normal. Brain MRI showed ischemic stroke with areas of subacute ischemia in the occipital lobe and left occipitotemporal transition. Drug treatment was performed with high doses of Haloperidol, Valproic Acid and Clonazepam without satisfactory clinical control. Botulinum toxin type A was applied with 300UI to the right trapezius muscle and 200UI to the right latissimus dorsi muscle, with considerable improvement in movements after thirty days and with preservation of strength in all four limbs. **Discussion:** Hemichorea-hemiballismus syndrome is one of the main movement disorders caused after a stroke. The main treatments described are typical and atypical antipsychotics, antiepileptics and presynaptic dopamine depletors. The use of botulinum toxin is well established in some hyperkinetic movement disorders such as dystonia, myoclonus and tics; its use in hemichorea-hemiballismus is still poorly studied, with little evidence found in the literature. Our case presented a good response similar to the few cases described in the literature. **Final Comments:** The use of botulinum toxin in the treatment of hemichorea-hemiballismus can be effective in patients who are refractory to drug treatment.

Doença Cerebrovascular

TL 1106403

A CASE OF STROKE CORRELATED WITH THREE TERRITORY SIGN IN A PATIENT WITH TROUSSEAU SYNDROME

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Case Presentation: A 78-year-old male patient, hypertensive and former smoker, was admitted to a tertiary Hospital with suspected ischemic stroke, presented with left upper limb plegia and left lower limb paresis for three days before the admission, with mild dyslalia and responsive to verbal commands. Cranial Computed Tomography (CT) revealed gliosis and myelin rarefactions, and Cranial Magnetic Resonance Imaging (MRI) showed randomly distributed foci of ischemic insult in three brain regions. Four days after admission, the patient reported worsening epigastric pain with irradiation to the spine. A CT scan of the abdomen/pelvis indicated a primary solid-cystic lesion, involving the body of the pancreas and bilateral renal cysts. USG indicated secondary neoplastic liver implants. The presence of cerebrovascular lesions in three regions suggested a diagnosis of Trousseau Syndrome (TS) associated with a neoplastic condition. The patient remained clinically stable and it was decided to maintain the clinical treatment of the stroke, and he was assigned for a follow-up at the oncology department. **Discussion:** TS is characterized as a spectrum of pathophysiological disorders associated with neoplastic conditions that can cause microthrombi by mucin, P-selectin, and L-selectin and thrombi induced by the action of tissue factors. Thrombotic events may precede the diagnosis of visceral malignancy or arise concomitantly with the tumor, indicating a hypercoagulable state with a tendency to multiple thromboembolisms. Ischemic stroke is characterized by the occlusion of blood vessels that carry out the vascularization of the Nervous System (NS), with active cancer being a risk factor for this clinical condition. In TS, the excess of embolic signals in patients with active neoplasia explains the lesion in three territories associated with stroke. In the report, the 78-year-old patient was diagnosed with solid-cystic lesions in the pancreas with an MRI indicating multiple foci of ischemic insult and pattern in three territories of cerebrovascular compromise. **Final Comments:** Due to the association of TS with a clinical condition of hypercoagulability that favors thromboembolic occurrences, it is necessary for health professionals to identify the etiology of ischemic stroke episodes of neoplastic origin more quickly and accurately. Familiarization with such vascular injuries may allow conduct that leads to a better prognosis for patients.

Doença Cerebrovascular

TL 1106427

IMPACT OF COVID-19 PANDEMIC ON THE NUMBER OF THROMBOLYSIS PERFORMED DURING THE CLOSURE OF THE STROKE UNIT IN A NEUROLOGICAL REFERRAL CENTER

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Case Presentation: With the Covid-19 pandemic, a new scenario and a new demand has become a priority in health services globally, causing impact, restructuring and even closing of stroke units around the world. (1) All these measures had an impact on the care of patients with acute stroke, both in developed and developing countries. (2) The neurology emergency unit at Hospital da Restauração in Recife/Pernambuco, one of the services with the highest flow of care for patients with acute stroke in the Northeast, also had to readjust to the new scenario imposed. **Objective:** To observe the impact of the closure of the stroke unit at Hospital da Restauração in the period from May 2020 to October 2021 and to compare the difference in the numbers of thrombolysis performed before closing (January 2020 to April 2020) and after the reopening, from November 2021 to April 2022, with the numbers of thrombolysis performed in the 18 months in which the unit remained closed. **Methods:** An observational, retrospective, cross-sectional study was performed by analyzing the medical records of patients undergoing chemical thrombolysis, in a period of 28 months, which corresponded to the period from January 2020 to April 2022 in the neurological emergency of the Hospital da Restauração – Recife. **Results:** The total number of patients undergoing chemical thrombolysis from January 2020 to April 2022 was 153. In 2020, 79 patients were thrombolysed and in 2021 there were 42 patients. In these 28 months of study, 10 months corresponded to the period of operation of the unit, with closure in May 2020 and reopening in November 2021. The period of operation of the stroke unit corresponded to 35% of the period analyzed. While the stroke unit was open, 60% of the Hospital's thrombolysis were performed throughout the analyzed period. With the reopening of the stroke unit in November 2021, 48 patients have already undergone chemical thrombolysis in the first 4 months of 2022, corresponding to an absolute increase of 342.8% over the previous year. **Final Comments:** Stroke units around the world guarantee better care for patients with acute stroke phase. (3) They promote better functionality outcome in all acute-phase patients, regardless of whether they were undergoing thrombolytic therapy. (4) In this study, we observed the negative impact on the amount of chemical thrombolysis performed, at a time when the Covid 19 pandemic reached the worst rates in the state Pernambuco.

Doença Cerebrovascular

TL 1106439

STROKE MIMIC THROMBOLYSIS IN A PATIENT WITH PARTIAL ABDOMINAL ANEURYSM THROMBOSIS

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Case Presentation: MGFM, 73 years old, female, with normal pressure hydrocephalus (NPH) using a ventriculo-peritoneal shunt (PVD), was admitted to the Hospital due to spontaneous exteriorization of an abdominal catheter. During the investigation, a partially thrombosed fusiform aneurysm was observed in the infrarenal segment of the abdominal aorta, with an extension of 10 cm and a diameter of 8.0x6.2 cm, with characteristics of instability. Indicated aortic endoprosthesis. While waiting for the procedure, the patient developed vomiting and sudden aphasia, presenting a score of 11 on the National Institutes of Health Stroke Scale (NIHSS). Cranial tomography presented an ASPECTS of 10. Intravenous chemical thrombolysis with alteplase was performed, the post-procedure NIHSS score was 8, with no other complications. Ultrasound study of the cervical arteries and echocardiogram were normal. Magnetic Resonance Imaging of the Skull did not show an area of previous ischemia, configuring, therefore, a mimic stroke of a patient with hydrocephalus of undetermined origin. The patient was discharged with a NIHSS of 2. Three months later, the endoprosthesis implantation surgery was successfully performed. **Discussion:** Intravenous thrombolysis is associated with an increased risk of intracranial aneurysm rupture, resulting in intracranial hemorrhage. The 2018 American Heart Association (AHA) guidelines suggest that thrombolysis is safe in unruptured intracranial aneurysms less than 10 mm in diameter. There is no recommendation regarding extracranial aneurysms. Experience with endovascular thrombectomy in patients with thrombosed intracranial aneurysms causing acute stroke is also limited. Proposed mechanisms of stroke related to unruptured aneurysms include thrombosis with distal embolization, extension of the thrombosis to the parent artery, or compression of the parent artery by the aneurysm. The risk of symptomatic intracranial hemorrhage in people with stroke mimic is low. **Final Comments:** It is unclear whether thrombolysis is safe in extracranial aneurysm cases, future case series may be useful to answer the question. Furthermore, in cases of ischemic stroke within the therapeutic window, intravenous thrombolysis should be performed, except in known contraindications, because even in cases of stroke mimics the risk of intracranial bleeding is low.

Doença Cerebrovascular

TL 1106444

OPALSKI SYNDROME IN LEFT BULBAR ISCHEMIC STROKE: CASE PRESENTATION

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Case Presentation: A 50-year-old male patient started ten days ago with intense headache in the left frontal region, tightness, with nausea and vomiting. Symptoms worsened after four days, with loss of left lower limb strength and vertical diplopia. On physical examination, Horner syndrome on the left, with miosis and ptosis; skew deviation with hypotropia in the left eye; complete left spastic hemiparesis predominantly in the left lower limb; alternating thermal and painful hemihypoesthesia was also observed, alternating with involvement of the left hemiface and right limbs. Brain MRI showed an ischemic stroke in the lateral region of the left medulla. **Discussion:** Opalski syndrome is a rare variant of lateral bulbar infarction, characterized by the triad of ipsilateral hemiparesis, Babinski sign and contralateral hypothermalgia. Although our patient did not have Babinski's sign, he did have spasticity, which corroborates the involvement of the pyramidal tract. Lateral bulbar lesions may present with diplopia, nystagmus, and bias deviation due to involvement of the vestibular pathways. **Final Comments:** In cases of stroke with hemiparesis, diplopia and asymmetric deviation, with contralateral sensory changes, Opalski's syndrome due to lateral bulbar involvement should be considered.

Doença Cerebrovascular

TL 1106447

THROMBOLYSIS IN A LARGE UNRUPTURED INTRACRANIAL ANEURYSM: A CASE PRESENTATION

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Case Presentation: A 17-year-old patient, with a previous diagnosis of aneurysm in the right middle cerebral artery (4.7x2.4x1.9 cm), presented to our Hospital complaining of sudden onset of left sided hemiparesis and hemihypoesthesia (NIHSS 14). On admission, computed tomography hasn't showed any ischemic parenchymal injury or intracranial bleeding. Computed tomographic angiography has confirmed diffusely irregular aneurysmal dilatation throughout its M1 segment of right middle cerebral artery associated with occlusion of the same vessel and the A1 segment of anterior cerebral artery. He underwent chemical reperfusion therapy with Tenecteplase 0.25mg/kg, with excellent outcome, improving the NIHSS to 5 in a few hours. Arteriography confirmed a dissecting aneurysm of right MCA. Patient has maintained neurological improvement being discharged with NIHSS 3 and an elective evaluation of aneurysm approach. **Discussion:** The major concern with intravenous thrombolytic therapy is the presence of symptomatic intracranial bleeding. The presence of aneurysmal formation is not an absolute contraindication for thrombolysis, however, due to the increased risk of bleeding, there is a tendency to choose not to proceed with the therapy. **Final Comments:** Most guidelines and studies point to a reasonable safety of thrombolysis in aneurysms < 10 mm, but there is still little information about aneurysms with larger dimensions.

Doença Cerebrovascular

TL 1106465

INTERNAL CAROTID ARTERY DISSECTION IN THE POSTPARTUM PERIOD

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Case Presentation: Caso 1 – A 36-year-old woman, on the 21st day after vaginal delivery, presenting a sudden episode of complete left hemiplegia, hemi-hypoesthesia and tactile and auditory hemineglect. She had moderate-intensity right hemicranial headache started 12 hours before the event. She was admitted to the emergency department 1 hour and 17 minutes after the ictus (NIHSS 11), undergoing a CT scan of the skull with ASPECT 10 and then submitted to venous thrombolysis, with no neurological improvement. Subsequently, she performed a new cranial CT showing right frontoparietotemporal hypodensity and extracranial CT angiography showing signs of dissection of the right internal carotid artery and irregular proximal third on the left internal carotid artery, with focal ectasia. Case 2 – A 33-year-old woman, on the 14th day after cesarean delivery, had a sudden episode of complete left hemiplegia, hemi-hypoesthesia and hemineglect for all modalities. She had a mild headache with progressive onset. She was admitted to the emergency department with 5 hours after the ictus (NIHSS 8), with CT of the head was normal. Under investigation, she performed a new head CT 48 hours after the event, with right frontoparietotemporal hypoattenuation and extracranial CT angiography with signs of bilateral internal carotid artery dissection. **Discussion:** Cervicocerebral arterial dissection is one of the most frequent causes of ischaemic stroke in young and middle-aged adults. (1) However, postpartum cervicocerebral arterial dissections are much rarer, accounting for only 6% of spontaneous arterial dissection in women under the age of 50. Headache and neck pain are the most common symptoms in postpartum cervical carotid and vertebral artery dissections, accounting for 60% to 90% of cases. (2) The pathophysiology of cervical artery dissection appears multifactorial, with evidence suggesting environmental and genetic contributions. (3) **Final Comments:** We describe two cases of ischaemic stroke secondary to carotid artery dissection in the puerperium. Therefore, severe headache and neck pain during this period should be rigorously evaluated in view of the possibility of a poor prognosis mainly related to misdiagnosis and delay in treatment.

Doença Cerebrovascular

TL 1106480

LEFT ATRIAL APPENDAGE MORPHOLOGY PRESENTING AS A RISK FACTOR FOR STROKE IN A YOUNG PATIENT IN THE ABSENCE OF ATRIAL FIBRILLATION: A CASE PRESENTATION

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Case Presentation: A 43-year-old woman treated with losartan, aspirin, and simvastatin had a medical history of stroke two years ago without etiological investigation, in addition to using a pacemaker for total atrioventricular block since the age of 20, without atrial fibrillation (AF) documented in previous exams. She arrived at the emergency department with sudden hemiparesis and hypoesthesia on the left side of her body at 10:30pm. On examination, she had a National Institutes of Health Stroke Scale (NIHSS) score of five, a blood pressure of 172/85mmHg and a capillary blood glucose level of 98mg/dL. Cranial computed tomography (CT) performed at 1:00am was normal, and thrombolytic therapy was initiated at 1:30am with full recovery of deficits. On subsequent examination, she had an elevated N-terminal pro B-type natriuretic peptide (NT-proBNP) of 1379.9pg/mL. Carotid and vertebral Doppler findings were unremarkable. Electrocardiography revealed sinus rhythm, and pacemaker telemetry showed no evidence of arrhythmia in the previous four months. Transthoracic echocardiography detected an enlarged left atrial appendage (LAA) and an enlarged left atrium (LA). Transesophageal echocardiography did not reveal any intracavitary thrombi or vegetation. Cardiac CT angiography confirmed the increased LA and LAA, the latter with a non-chicken-wing morphology and without evidence of thrombi inside. The patient was discharged after full anticoagulation at the Hospital with prescription of dabigatran, atorvastatin, amlodipine and outpatient follow-up in the neurology and cardiology departments. **Discussion:** The LAA originates from the main body of the LA and represents one of the major sources of cardiac thrombus formation. Based on imaging findings, LAA morphology can be classified into simple (chicken-wing morphology – most common and with lower risk of thrombi) and complex (non-chicken-wing). Studies have suggested that complex LAA morphology by itself may lead to stroke, even in the absence of AF, probably because of lower flow velocities compared to simple LAA morphology. In addition, there is evidence that ischemic stroke patients without known AF and with elevated NT-proBNP levels may benefit more from anticoagulants than antiplatelet agents, explaining this patient treatment. **Final Comments:** We present the case of a patient without documented AF and with two strokes at a young age, of probable cardioembolic etiology due to an LAA of complex morphology.

Doença Cerebrovascular

TL 1106516

“CAROTID WEB” AN IMPORTANT ETIOLOGY OF STROKE IN YOUNG PEOPLE: CASE PRESENTATION

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Case Presentation: J.A.T.T., male, 44 years old, has prothrombin II mutation. Attended at Albert Einstein Hospital on April 21, 2022 with dysarthria lasting 5 minutes one day before admission. He denies motor or sensory deficits. On admission, cranial CT and AngioCT identified areas of recent infarction in the subcortical and right inferior frontal gyrus associated with linear and irregular filling failure adhered to bulb wall/origin of right internal carotid artery (ICA), suggestive of carotid membrane with associated thrombus, typical of “carotid web”. The findings were confirmed with MRI and AngioMRI with vessel studies that showed occlusion of the M4 segment of the right middle cerebral artery. Angioplasty with stent was performed on April 24, without complications. The patient evolved without changes in the neurological examination, using full anticoagulation. **Discussion:** “Carotid web” (CA-Web) is defined as thin, linear, membrane that extends from the posterior aspect of the internal carotid artery bulb into lumen, located just beyond the carotid bifurcation. Histopathologically is characterized by fibroelastic thickening of the arterial intima. Prevalence is estimated ranging from 9.4% to 37% in patients with cryptogenic stroke. Studies showed average age of 41 years and an NIH of 10.5 in patients with this vascular disorder. Thus, symptomatic CA-Web patients are younger and likely to have more severe strokes compared to ischemic stroke patients in general. Among the non-invasive diagnostic methods, there is AngioCT which CA-Web is usually seen as a thin, smooth, membrane-like intraluminal filling defect along the posterior wall of the ICA bulb on oblique sagittal images. Treatment is based on secondary prevention of stroke, as well as an individualized anticoagulation decision, as the risk of recurrent ischemic events is likely to be higher in antiplatelet therapies. Surgical resection can be considered, especially for patients with therapeutic failure, and carotid artery stents are alternative to surgical resection, with a lower risk of complications. **Final Comments:** CA-Web is rare variant of fibromuscular dysplasia that affects the carotid artery intima. Recent studies have associated this change as important etiology in young patients with cryptogenic ischemic stroke. The knowledge about radiographic findings, as well as management in the treatment of this etiology can prevent recurrence of ischemic vascular accidents, especially in the young population.

Doença Cerebrovascular

TL 1106522

CLINICAL CASE PRESENTATION DISSECTING ANEURYSM OF THE MIDDLE CEREBRAL ARTERY IN A THREE-MONTH-OLD INFANT: CASE PRESENTATION

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Case Presentation: A 3-month-old female infant with a report of her first seizure in her life. On examination, he was in good general condition, normotensive anterior fontanelle, without neck stiffness and afebrile. Normal cardiopulmonary auscultation and abdomen without a palpable mass. Previous history: Late premature (34 weeks and 2 days), vaginal delivery with meconium amniotic fluid. There was a prolonged Hospital stay due to proximal jejunal atresia, and she underwent corrective surgery on her 10th day of life. No history of infection or previous trauma. Cerebral aneurysm embolization with detachable platinum microcoils was chosen, using a total of 10 units. Control angiography showed aneurysmal occlusion with preserved filling of distal branches of the right MCA. The postoperative evolution was satisfactory, and the patient was discharged on the 3rd postoperative day. **Discussion:** Cerebral aneurysms are rare in pediatric patients. In relation to the adult population, its presentation differs in terms of incidence, size and location. Despite its low incidence, 95% of the cases have subarachnoid hemorrhage as the initial manifestation, giving high morbidity and mortality to patients. Among the risk factors of the pediatric population, trauma and mycotic aneurysms, in terms of acquired pathologies, deserve to be highlighted; and family history and collagen diseases (Elher-Danlos, Marfan syndrome, Type 1 neurofibromatosis and polycystic kidney disease) and congenital pathologies. In the presented, we did not identify associated risk factors. **Final Comments:** Due to the lower incidence of intracranial aneurysms in the pediatric population, the scientific literature lacks robust evidence pointing to the best therapeutic option. Our case aims to contribute to the scientific literary scope as evidence of a successful endovascular approach.

Doença Cerebrovascular

TL 1106528

STURGE WEBER SYNDROME TYPE 3: A CHALLENGING DIAGNOSIS

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Case Presentation: 19 years-old female with obesity, hypothyroidism, insulin resistance, and precocious puberty presented, when at age of 5 years-old, a first episode of self-limited left hemiparesis following a new-onset headache. At the same age, she presented the first episode of seizure. Since then, she has persisted with a chronic headache. Evaluation with brain MRI showed extensive right parenchymal calcification and venous-capillary angioma with ependymal drainage. Although the patient has had typical radiological findings for Sturge-Weber Syndrome (SWS), she imposed a diagnosis challenge since she did not have the typical cutaneous port-wine nevus. Annual follow-up MRI demonstrated progression of cortical calcifications and increasing venous stasis due to venous-capillary malformation. Despite progression, the patient maintains clinical and neurological stability. **Discussion:** Sturge-Weber syndrome is a neurocutaneous syndrome characterized by a facial port-wine birthmark (capillary malformation) with abnormal blood vessels in the brain and the eye. It is a sporadic developmental disorder caused by somatic mosaic mutations in the GNAQ gene. This syndrome is the third most common neurocutaneous disorder after neurofibromatosis (NF) and tuberous sclerosis complex (TSC), but it is not associated with a greatly increased risk of tumors. SWS exists as a spectrum ranging from the isolated brain involvement seen in about 10% of the cases, classified as type 3 SWS, to facial birthmark associated with brain and eye involvement, type 1. Seizure is usually the first neurological manifestation in the first or two years of life. Diagnosis of Sturge-Weber syndrome is based on typical clinical symptoms and brain magnetic resonance imaging (MRI) findings characterized by enhancing dilated leptomeningeal vessels associated with enlarged deep draining vessels and a choroid plexus glomus. **Final Comments:** This case exemplifies the challenge of neurological diagnosis of SWS in the absence of port-wine stain facial angioma. In a patient with typical symptoms such as focal seizure, cognitive impairment, and metabolic disturbance presenting with radiological findings, SWS type 3 should always be considered as a possibility.

Doença Cerebrovascular

TL 1106553

STROKE IN POSTERIOR CIRCULATION DUE TO ANTERIOR CIRCULATION STENOSIS: A CASE PRESENTATION OF VERTEBRAL ARTERY ORIGIN IN INTERNAL CAROTID

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Case Presentation: A 57 year old female admitted in the emergency department presenting sudden onset of left side weakness, drowsiness and vomiting. At neurological evaluation, hemiplegia and hemianopia were observed. Angiographic study showed right common carotid artery stenosis, internal carotid origin of right vertebral artery and right posterior cerebral artery occlusion. Because of the delayed time between symptoms onset and Hospital admission, patient was not submitted to reperfusion therapy with alteplase or endovascular treatment. In 24 hours, massive edema caused by posterior ischemia was shown in a new CT scan. Then, the neurosurgical team performed a craniectomy. Patient was discharged 1 month after Hospitalization, with several disabilities. Etiologic investigation was negative for cardiologic sources. **Discussion:** The anatomical variations of cranial and extra cranial arteries are well established in literature, and its current implication as risk factor for ischemic stroke. In this presentation, the patient had a stenosis in anterior circulation origin that had implications in posterior circulation, not having a fetal pattern of posterior cerebral artery, causing a stroke. The right vertebral artery had its origin at distal portion of extracranial internal carotid artery. **Final Comments:** It is very important for the clinical practice of physicians to understand the variability of the human anatomy, as they help with the accurate diagnosis process. Rare variations can have implications in the setting of cerebrovascular diseases.

Doença Cerebrovascular

TL 1106570

ATYPICAL DISCOVERY OF A CEREBRAL DURAL ARTERIOVENOUS FISTULA

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Case Presentation: A 35-year-old male with an acute history of recurrent seizures and decreased level of consciousness associated with high fever of unknown onset. He arrived at the emergency department with a Glasgow Coma Scale of 3 alongside automatism. Past medical history was remarkable for traumatic brain injury ten years prior to these events and a seven-months history of progressive impaired gait. He was submitted to a brain computed tomography (CT) which showed cortical hypodensities, periventricular and symmetrical bilateral basal ganglia calcifications, venous engorgement due to an extensive arteriovenous fistula with brainstem involvement. Lumbar puncture showed a high open pressure, normal glucose levels, elevated white blood cell and protein counts. Cerebrospinal fluid cultures were positive for gram negative cocci. Prolonged EEG showed moderate to severe diffuse symmetrical baseline activity disorganization without epileptiform activity. The patient evolved with refractory non-epileptic seizures and healthcare-associated infections and was then submitted to palliative care. Intracranial dural arteriovenous fistulas (DAVFs) are abnormal connections between dural arteries and dural venous sinuses, meningeal veins or cortical veins and are responsible for 10-15% of all intracranial arteriovenous malformations. It can be related to trauma, dural sinus thrombosis, venous hypertension, previous craniotomy or mostly idiopathic aetiology. It is diagnosed preferably through conventional angiography, but CT and Magnetic Resonance (MR) can also be used, although they confer lower accuracy. After suspicions of DAVFs, additional evaluation with CT or MR angiography is needed to confirm. Most common locations in adults are at the cavernous, sigmoid and transverse sinuses. Most symptoms start around the fifth and sixth decades of life. The pattern and location of the DAVF venous drainage is associated with its clinical presentation and severity. DAVFs are not a risk factor for meningitis or other central nervous system (CNS) infections. Gait impairment is more associated with spinal DAVF or brainstem DAVF. Herein we present a case with a past history of gait impairment and a sudden onset of status epilepticus due to meningitis. Although CNS infections are not facilitated by DAVFs, it led to the discovery of an extensive one in our patient which already compromised cortical regions, which contributed partially to severity of the case despite treatment.

Doença Cerebrovascular

TL 1106587

SPONTANEOUS RUPTURED ARTERIOVENOUS MALFORMATION IN A YOUNG PATIENT: A CASE-BASED UPDATE

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Case Presentation: A 17-year-old female has presented a history of severe frontoparietal headache for 6 days, refractory to analgesics. After admission, the patient evolved with worsening headache associated with drowsiness, nausea, persistent vomiting and inappetence. There are no reports of comorbidities in the family and her parents are not consanguineous. Neurological exam revealed mild paresis in left limbs. A skull Computed Tomography (CT) showed the presence of an extensive frontoparietal hemorrhagic infarction with brain mass effect, indicative of intracranial hypertension. In addition, a brain Digital Subtraction Angiography (DSA) was performed and showed lobar intraparenchymal hemorrhage in the right temporal opercular region, with a estimated volume of 48cm³, presenting circulating vasogenic edema and Spetzler-Martin grade 1. A right temporal arteriovenous malformation was diagnosed. **Discussion:** Arteriovenous malformations (AVMs) are the most dangerous of the cerebrovascular malformations with the potential to cause intracranial hemorrhages and even epilepsy. The most common clinical presentations are intracranial hemorrhages, most commonly intraparenchymal, but they can also occur in the intraventricular or subarachnoid space. Bleeding into the subarachnoid space is common in superficial AVMs. In addition, seizures, focal deficits and headache are common. The Spetzler-Martin scale (1 to 5) classifies the risk of surgical removal of the AVM according to its size, location in eloquent areas or not, and whether deep venous drainage is present. High scores correlate with greater risks of surgical morbidity and neurological deficits. Patient age is an important factor in the decision to treat brain AVMs, as patients with a longer life expectancy appear to accumulate a higher lifetime risk of hemorrhage. Thus, therapy is best suited for children and young adults. Brain DSA is essential for treatment planning and postoperative follow-up, as it is possible to better obtain the configuration of the nidus, its relationship with the circulating vessels, and the location of the draining or efferent portion of the cerebral AVM. **Final Comments:** Spontaneous rupture of arteriovenous malformations in young patients is associated with the possibility of performing immediate surgical interventions, which allow lower risks of acute complications. However, these patients with longer life expectancy appear to have a higher lifetime risk of hemorrhage.

Doença Cerebrovascular

TL 1106612

ACUTE ISCHEMIC STROKE IN A YOUNG PATIENT CAUSED BY BINGE DRINKING: A CASE PRESENTATION

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Case Presentation: A previously healthy 40-year-old man from Recife, with a history of occasional binge drinking, presented sudden hemiparesis with hypoesthesia in the right side of the body after binge drinking. Associated with the condition, he presented transient gait incapacity for two minutes. He arrived at the emergency department in 2 hours with a NIHSS score of 2 points and a normal cranial CT scan, with no indication for thrombolysis. A double antiplatelet therapy was started with acetylsalicylic acid and clopidogrel. After 24 hours, the weakness in the right side remained, although of lesser intensity, evolving with hyperreflexia and superficial hypoesthesia in the entire right hemibody. The brain MRI revealed an area of restriction to water diffusion in the left parietal cortico-subcortical region associated with signal hyperintensity on T2 and FLAIR, compatible with subacute ischemic vascular insult. Spectroscopy showed a reduction in the levels of N-Acetyl aspartate (neuronal marker) and a slight increase in the peak of lipids/ lactate with features suggestive of necrosis. Autoantibody research revealed ANA 1:640 (fine speckled). Serology for HIV, syphilis and hepatitis were non-reactive. Magnetic resonance angiography showed no evidence of atherosclerosis or other changes. New screening tests were performed for lupus, thrombophilia and vasculitis, including a new ANA, with normal results. The echocardiogram and 24h Holter did not show heart disease. The patient was discharged clinically stable after one month of Hospital investigation with complete recovery of strength and sensitivity. **Discussion:** Ischemic stroke mainly affects people over 65 years of age, with atherosclerosis as the main etiopathogenesis. Ischemic stroke in young individuals, below 45 years of age, should lead to the investigation of secondary causes such as infections, autoimmune diseases, genetic conditions, neoplasms, heart diseases and drug abuse, including alcohol and smoking. In this case, the patient started the symptoms within 24 hours after the binge drinking episode, with no other etiopathogenic causes being evidenced. **Final Comments:** In the context of stroke in a young patient, an adequate investigation of secondary causes should be carried out. In this case, alcohol abuse were notorious as a possible cause of stroke in young people, and alcohol consumption should be considered in the primary and secondary prevention of stroke.

Doença Cerebrovascular

TL 1106616

CEREBRAL VASOSPASM: HOW FAR DO THE SEQUELS OF SARS-COV-2 INFECTION GO?

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Case Presentation: Female, 22 years-old, referred by SAMU, was admitted to the neurological emergency service after an episode of generalized tonic-clonic epileptic seizure, lasting about 20 minutes, and drowsiness after the crisis. She denied sphincter release. On admission, the patient was feverish, pale, drowsy, GCS 14, without focal deficits. No neck stiffness. Complementary investigation showing hypokalemia and leukocytosis. Brain Computed Tomography (CT) did not show any changes. Before, she reported Hospitalization for a month due to a SARS-Cov-2 infection, requiring mechanical ventilation for 17 days. Considering the hypothesis of infectious encephalitis, the patient received empirical treatment with Acyclovir and Meropenem. After two days of Hospitalization, she presented a new episode of generalized tonic-clonic epileptic seizure. The investigation proceeded: negative serology, no hypovitaminosis, negative COVID swab, electroencephalogram without changes. Magnetic resonance imaging (MRI) of the skull with hyperintense lesions in the occipital region. Positive Cerebrospinal Fluid (CSF) Analysis for herpes simplex virus 2, with negative bacterioscopy. Electroneuromyography without significant changes. The patient evolved with better clinical improvement. New CSF study: red blood cells: 120, leukocytes: 1 cell (neutrophils: 2%, lymphocytes: 93% monocytes: 5%) proteins: 28. glucose: 57, LDH: 27 ADA: 0.1, VDRL: negative. A new MRI was performed, which showed an increased signal at the cortico-subcortical T2 and Flair sequences in bilateral parieto-occipital lobes and right frontal lobe with contrast enhancement. Vessel wall with microconstrictions inferring a reversible cerebral vasospasm pattern. **Discussion:** Here we present a case of cerebral vasospasm in a young patient with no previous comorbidities. It is known that COVID-19 has been associated with immune dysregulation of the host that can increase the risk of infections such as herpes virus, but this mechanism is still not well understood. In addition, Sars-cov-2 has been showing a vast amount of sequelae in those affected, including cerebral vasospasm. **Final Comments:** The literature shows to an endothelial inflammation caused by the coronavirus as a contributor to the development of cerebrovascular disease, which includes, in addition to vasospasm, stroke, headaches and other changes in the central nervous system, and it may even aggravate other infections.

Doença Cerebrovascular

TL 1106620

STROKE MIMIC SECONDARY TO VACICELA ZOSTER VIRUS ENCEPHALITIS

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Case Presentation: Female, 77 years old, hypertensive and coronariopathy, searched our service after being found sleepy, disordered and with alteration of speech. on admission, it was evaluated by the neurology team, which confirmed aphasia. a ct scan of the skull was performed, which showed an asymmetry of density between the insulae, which, together with the sudden focal deficit, strengthened the diagnostic hypothesis of hyperacute ischemic brain vascular accident, being thrombolytic treatment with alteplase and the patient referred to the ischemic stroke unit. the following day, the patient was sleepy and encephalopathy, when ulcerated lesions covered by serohematical crusts and lesional hyperemia were noticed, located in the front region on the right, in the territory of the ophthalmic branch of the trigeminal nerve, the diagnostic hypothesis of herpes-zoster being proposed. lumbar puncture performed which evidenced cellularity of 201 (66% lymphocytes and 19% monocytes), protein 31 mg/dl and glucose 52 mg/dl, in addition to multiplex pcr confirmed varicella zoster virus infection. the mri and angio-mri brain showed lesion with restriction to left parietal diffusion. treatment with acyclovir started and in 48 hours the patient evolved with improvement of symptoms. **Discussion:** The varicella zoster virus causes chickenpox as a primary infection, after which the virus becomes latent in the cranial nerves and dorsal root ganglia along the neuroaxis. when cell mediated immunity decreases, the virus may reactivate, travel peripherally to cause a skin eruption and peripheral neuropathy (herpes zoster) or, less commonly, it may move centrally and cause a variety of neurological complications, including vasculopathy and encephalitis, these can cause irreversible brain damage. in the case in question, the apparent ischemic lesions on mri did not justify the patient's initial symptoms, being treated as stroke mimic secondary to encephalitis framework by vzv. this agent is susceptible to various antiviral drugs, but acyclovir is the drug of choice and its early initiation has contributed to avoiding serious complications in the case in question. **Final Comments:** There is a large quantity of diseases that can behave like stroke mimic, vzv encephalitis is one of them, that should be suspected, especially, when there is clinical acute encephalopathy associated with suggestive skin lesions. timely treatment can prevent serious sequelae.

Doença Cerebrovascular

TL 1104832

MAN IN THE BARREL SYNDROME AS A PRESENTATION OF MOTOR NEURON DISEASE: CASE PRESENTATION

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Case Presentation: Male, 52 years old, history of fasciculations and sudden biparesis of the limbs, asymmetric, for 3 years, evolving with mild paresis of the LLL with progression to RLL in 10 days, without prejudice on gait, associated with dysphonia and intermittent dysphagia. The condition followed with a gradual worsening of the tetraparesis, with a severe predominance in the upper limbs, with no complaints of sensitivity. Personal background: systemic arterial hypertension. On examination: alert, oriented, dysarthria and dysphonia, isochoric and photoreactive pupils, plegia and bilateral hypotrophy of the upper limbs, generalized hypotonia, with patellar hyperreflexia, Babinski's sign on the right, Hoffman's sign on the left. Neuroaxis imaging exams without alterations. Electroneuromyography (ENMG): findings consistent with motor neuron disease. **Discussion:** Clinical signs of upper and lower motor neuron lesions, with a clear predominance of involvement in the upper limbs, with a characteristic phenotype of man-in-the-barrel syndrome, required the need for an electroneuromyographic study, which showed alterations compatible with sensory pre-ganglionic involvement, showing signs of chronic denervation and reinnervation in at least two body segments, corroborating the hypothesis of motor neuron disease / amyotrophic lateral sclerosis (ALS). **Final Comments:** The nomenclature "man in the barrel syndrome" (MBS) was first used in 1983, since then, many cases have been reported, although the vast majority of motor neuron pathologies have a classically different etiopathogenesis from this description. Still, MBS is not exclusive to ALS, as a considerable part of the reports are caused by Progressive Spinal Amyotrophy, differential diagnosis that requires neurophysiological evaluation for clarification, given its lower severity compared to ALS. The patient above presented an atypical clinic of upper limb biplegia, and the ENMG was consistent with the diagnosis of ALS manifested with MBS.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1106095

WHEN OCCAM'S RAZOR ISN'T SHARP: ASSOCIATION BETWEEN PROGRESSIVE MUSCULAR ATROPHY, PIRIDOXIN NEUROTOXICITY AND LATE ONSET MYASTHENIA GRAVIS

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Case Presentation: A 71 years old male, Swiss, started recurrent episodes of cramps on both feet in the last 3 years, progressing to the hands in the next several months. The symptoms progressed insidiously to distal weakness first in the right foot, then to the left one, within 12 months, causing stumbling when walking. He started complex B vitamins supplementation by a neurologist prescription. He brought an electromyography (EMG) with signs of acute and chronic axonal cervical and lumbosacral motor degeneration. There was also a sensory axonal polyneuropathy (SAP) in the four limbs. He took gabapentin for chronic low back pain due to L5 herniated vertebral disc. He was told that he had motor neuron disease spectrum (MNDE) after extensive complementary exams, so he asked for a second opinion. Positive neurological findings on examination at this moment were: nasal voice, Romberg sign, deltoid myofasciculations (<https://www.youtube.com/shorts/KSv9yZS4y0Y>), absent Achilles reflex, hypoesthesia on the lateral aspect of the right leg with ipsilateral weakness on hallux dorsiflexion. There were no signs of upper motor neuron (UMN) disease. Curiously, patient state fatigue during the physical examination. Due to muscular fatigability, screening dosage of nicotinic acetylcholine (AChR)-binding antibody was requested and showed a titer of 0,56 mmol/L (positive when greater than 0,40). Piridoxin seric levels were high (154,8 µg/L, normal range between 2,8-74,6 µg/L), but lowered spontaneously to 68,6 µg/L when vitamin B6 supplementation was stopped. Patient started prednisone 40mg/dia and pyridostigmine 360mg/dia. **Discussion:** The association between myasthenia gravis (MG) and amyotrophic lateral sclerosis (ALS) is strong, but its association with the lower motor neuron (LMN) sporadic phenotype (Progressive Muscular Atrophy, PMA) is unknown. PMA is a controversial entity and represents 3% of all cases of MNDE. One third of patients with PMA develop UMN features within 2 years during disease course. The patient had not palpebral ptosis or diplopia, but fatigability and nasal speech pointed to MG and not a bulbar UMN sign. Sensory ataxia due to B6 toxicity was a bias in the context of interpretation of EMG, since sensory nerve action potentials (SNAP) are normal in MND. **Final Comments:** Neurologists should be aware since muscular weakness may be either the common final path symptom or a coexisting condition in both neuromuscular junction disease and motor neuron disease.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1104860

MUSK-RELATED CONGENITAL MYASTHENIC SYNDROME: FIRST REPORT OF ADULT-ONSET CASE IN BRAZIL

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Case Presentation: A 33-year-old man presented with severe muscular fatigue for more than a year, more pronounced in the evening, associated with double vision and eyelid ptosis, both initially intermittent, then continuous after six months. Neurologic examination revealed dysphonia, bilateral eyelid ptosis with fatigue during maneuvers, discrete global ophthalmoparesis, bilateral facial nerve palsy, and discrete and symmetric palatal numbness. MRC muscle strength was 4/5 in the proximal lower and upper limbs and global hyporeflexia. Needle EMG showed a myopathic pattern and a deltoid muscle biopsy revealed nonspecific mild and chronic myopathic findings. Initially, we tried pyridostigmine and prednisone. LF-RNS showed a pathological decrement of 18%. His condition progressively worsened until he suffered six episodes of myasthenic crisis within a year, requiring prolonged mechanical ventilation, IGIV, PLEX, and remained refractory even with rituximab. Both anti-AChR and anti-MUSK were negative. His 29-year-old sister had a history of eyelid ptosis, neonatal hypotonia, and an isolated episode of GTC seizure associated with hypoxia. At age 22, she began to experience intense and slowly progressive muscular fatigue, particularly in the lower limbs, associated with eyelid ptosis. Neurologic examination revealed appendicular fatigue, eyelid ptosis, and discrete ophthalmoparesis without dysphonia or dysphagia. Genetic testing on both siblings confirmed the diagnosis of MUSK-related autosomal recessive congenital myasthenic syndrome. NGS-based gene panel testing disclosed the presence of compound heterozygous variants c.2125G>A (p.Val709Met) and a deletion of exons 3-4 in the MUSK gene. **Discussion:** Congenital myasthenic syndrome (CMS) is a clinic and genetically heterogeneous group of inherited neuromuscular disorders, which share impaired neuromuscular transmission due to distinct genetic mechanisms. Autosomal recessive pattern of inheritance is observed in the MUSK (9q31.3) gene-related CMS. MUSK encodes a post-synaptic protein involved in endplate maintenance and maturation, proper function of Rapsyn and AChR, thus acting in the AChR clustering pathway. Treatment of CMS is not standardized because of the small number of patients and the lack of specific therapeutic trials. **Final Comments:** Our cases expand the current knowledge about CMS and provide original information regarding the first genetically confirmed adult-onset MUSK-related CMS case in Brazilian population.

Doenças Neuromusculares

TL 1104862

EXPANDING THE SPECTRUM OF TBK1-RELATED DISORDERS: ADULT-ONSET NON-5Q SPINAL MUSCULAR ATROPHY

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Case Presentation: A 36-yr man presented with a slowly progressive history of weakness and painful cramps in his lower limbs since age 22 years. After 8 years, he noted mild amyotrophy involving the distal portion of the thighs and calves, slow gait speed, and waddling gait. Medical and family history were unremarkable. Parental consanguinity was not present. Examination disclosed crural flaccid paraparesis with proximal predominance, absent tendon reflexes and bilateral pes cavus. Neurophysiological studies disclosed symmetric chronic denervation pattern with reinnervation in the lumbosacral myotomes (L3-S1). Brain and spine MRI and cerebrospinal fluid analysis were unremarkable. Muscle MRI disclosed mild fatty replacement of the vastus medialis, adductor and focal fatty replacement bilaterally in the medial head of the gastrocnemius muscle. As clinical and neurophysiological features were suggestive of non-5q spinal muscular atrophy (SMA), especially SMA with lower extremity predominance (SMALED), a genetic testing with whole-exome sequencing (WES) was performed. WES evidenced the heterozygous pathogenic variant c.1960-1G>T (a splice acceptor variant) in intron 18 of the TBK1 gene, leading to a loss of function effect due to protein truncation and activation of nonsense-mediated decay. Thus, we concluded that our patient presented with a new MND phenotype associated with TBK1 gene. Additional testing for 5q SMA (including MLPA and Sanger sequencing) and a wide NGS-based gene panel test were performed and their results were unremarkable. **Discussion:** TBK1 (TANK binding kinase-1) gene (12q14.2) is associated with modulation of autophagy and inflammatory function in neuronal and glial cell types. TBK1 variants are related to Amyotrophic Lateral Sclerosis (ALS), Frontotemporal Dementia, and susceptibility to acute infection-induced herpes simplex-specific encephalopathy type 8. Our patient presented with clinical and neurophysiological findings suggestive of long-standing lower MND, mimicking features of SMALED syndrome and atypical ALS variants. SMALED phenotypes have been previously associated with DYNC1H1 and BICD2 heterozygous pathogenic variants, however with distinct muscle imaging findings when compared to our patient. **Final Comments:** Clinicians must be aware about the possibility of TBK1 variants as new monogenic basis of pure non-5q SMA and SMALED-like phenotypes with specific muscle imaging pattern of involvement.

Doenças Neuromusculares

TL 1105097

HYPERTHYROIDISM PRESENTING WITH HYPOKALEMIC PERIODIC PARALYSIS AND CLUSTER HEADACHE: A CASE PRESENTATION

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Case Presentation: A 26-year-old Brazilian male, without known comorbidities, presented with acute flaccid tetraparesis. He reported an abusive carbohydrate intake the previous night. He denied strenuous physical exercise, fever, recent trauma, loss of sphincter control, use of medications, drugs or alcohol. On neurological examination, he presented flaccid tetraparesis with proximal weakness predominance. Laboratory investigation showed serum potassium 2.89 mEq/L and there was no alteration on neuroimaging. After treatment with intravenous potassium, the patient completely recovered his motor strength in four hours. During outpatient follow-up, he reported episodes of headache described as a left-sided excruciating pain, originated in the orbital region, irradiated to supra-orbital and temporal region, lasting 15 minutes. The pain was associated with ipsilateral ptosis. Results of analysis collected during Hospitalization showed TSH (<0.01 UI/mL), T4 (6.11 ng/dL), TRAB (4.7 UI/L). The patient was diagnosed with Thyrotoxic Hypokalemic Periodic Paralysis (TPP) and met the criteria for cluster headache. He was treated with tiamazol and atenolol for hyperthyroidism. He started prophylactic and symptomatic treatment for cluster headache. Five months after Hospital discharge, the patient denied new episodes of acute tetraparesis and episodes of cluster headache. **Discussion:** TPP is a very rare complication of thyrotoxicosis, more frequent in the Asian population. It must be considered as a differential diagnosis of acute tetraparesis. The role of thyroid dysfunction in the etiology of headache remains uncertain. In literature, there are cases of patients with newly diagnosed hyperthyroidism whose headaches remitted after treatment of hyperthyroidism. A study found reduced TSH response to thyrotropin-releasing hormone (TRH) in patients with cluster headache during cluster period, hypothesizing a hypothalamic involvement in the pathogenesis. **Final Comments:** We report this case to emphasize the importance of the evaluation of thyroid function in patients with acute flaccid tetraparesis. Timely recognition and treatment of TPP can reduce the risk of severe complications such as respiratory muscle weakness and arrhythmias. Further studies are needed to clarify the association between hyperthyroidism and cluster headache, but physicians should be aware that there may be a correlation and consider a thyroid function assessment.

Doenças Neuromusculares

TL 1105102

NECROTIZING MYOPATHY RELATED TO ARBOVIROSIS – CASE PRESENTATION

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Case Presentation: A 35-year-old man, 3 weeks after a febrile condition with polyarthralgia, evolved with proximal lower limb weakness and subsequent progression to upper limbs. He denied myalgia, cramps, myoglobinuria, falls, respiratory complaints, dysphagia or dysphonia. Neurological examination revealed scapular and pelvic muscle wasting, proximal weakness, hypoactive deep tendon reflexes and waddling gait. Laboratory investigation revealed high levels of creatine phosphokinase (CPK: 11,591 UI/L), lactate dehydrogenase (DHL: 1420 U/L) and positive IgG serology for chikungunya (Elisa). Electroneuromyography showed a myopathic pattern. Muscle biopsy, performed at right biceps brachialis, revealed several necrotic and regenerating myofibers, with scarce perivascular inflammatory infiltrate, several CD68+ cells, rare CD8+ cells and no CD4+ cells, consistent with necrotizing myopathy. The patient was then treated with prednisone 1mg/kg/day. However, due to increasing CPK levels and persistence of weakness, human intravenous immunoglobulin (2g/kg in 5 days) was added to treatment. The patient was discharged with prednisone 1mg/kg/day and methotrexate 10mg per week. After 45 days, in follow up visit, patient showed significant improvement in muscle strength, normal gait and normalization of CPK and DHL levels. Prednisone was tapered and methotrexate was maintained. **Discussion:** Immunemediated necrotizing myopathy is a inflammatory myopathy, characterized by proximal muscle weakness, high CPK levels, necrotic and regenerating fibers and macrophagy in muscle biopsy, as well as good response to immune therapy. Necrotizing myopathy is related to various etiologies, such as use of statins, neoplasms and viral infections. One of the viral causes, the chikungunya virus, an alphavirus present throughout the Brazilian territory, presents with a febrile disease with intense arthralgia. Neurological complications include encephalitis and myelitis; there are reports of cases with peripheral nervous system involvement, including skeletal muscle. **Final Comments:** Arbovirois-related necrotizing myopathy is a rarely reported entity, and there is still discussion about the best therapeutic approach. We reinforce the clinical response with the use of intravenous intravenous immunoglobulin; however, long-term immunosuppressive therapy remains a recommendation in the follow-up of these patients.

Doenças Neuromusculares

TL 1105154

GUILLAIN-BARRÉ SYNDROME AFTER DENGUE VIRUS INFECTION, A RARE NEUROLOGICAL COMPLICATION

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Case Presentation: A 39-year-old man with a history of 10 days of fever, arthralgia and rash that evolved with nausea, vomiting and abdominal pain, admitted at the Hospital because of worsening abdominal pain, liver dysfunction, increase in hematocrit and thrombocytopenia with positive serology for dengue (IgM). At the third day of Hospitalization, patient started with paresthesia of both legs that rapidly progressed, distal weakness, areflexia, bilateral facial paralysis, dysarthrophonia and dysphagia. The analysis of cerebrospinal fluid showed cytological protein dissociation (1 cell and 58 proteins). Considering the clinical and laboratory features the diagnosis was Guillain-Barré Syndrome (GBS). Then, we referred him to the intensive care unit and intravenous immunoglobulin (IgIV) for five days was the treatment. At the second day of IgIV he evolved with dysautonomia (heart rate instability and urinary retention) associated with worsening of the bulbar symptoms with necessity to orotracheal intubation. He had clinical complications such as ventilator-associated pneumonia receiving broad-spectrum antibiotics and evolved with pseudomembranous colitis because of the antibiotics, but presenting a good treatment response. Given the slow recovery and prolonged Hospitalization, he underwent tracheostomy and gastrostomy, being discharged after 60 days of Hospitalization. **Discussion:** GBS is a rare presentation of dengue infection. The most common neurologic manifestations associated with dengue virus (DENV) infection include headache, encephalopathy and seizures. Other neurologic syndromes reported include acute pure motor weakness, mononeuropathies, polyneuropathies, stroke and transverse myelitis. A cohort study evaluated the increasingly incidence of neurologic complications of DENV and demonstrated that clinical and laboratory parameters like higher body temperature, liver dysfunction, increase in hematocrit and thrombocytopenia are independent predictors of neurologic complications, those changes were found in our patient what is in agreement with the literature. **Final Comments:** The incidence of GBS associated with dengue fever is still unknown and in the literature, there are just few isolated case presentation. In Brazil, dengue fever is endemic and early recognition of neurological conditions that may be associated with this infection and being attention to predictors that increases the risk of these complications is very important in order to prevent bad outcomes.

Doenças Neuromusculares

TL 1105158

DISTAL PRESENTATION IN CENTRAL CORE MYOPATHY CAUSED BY RYR1 MUTATIONS: A CMT-LIKE PHENOTYPE

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Case Presentation: A 16-year-old patient, with no consanguinity in the family, presented with frequent falls and tip-toe gait, at the age 2. She developed a distal leg weakness with significant deformities. On examination, she presented global hyporeflexia, MRC grade 4 in the proximal upper and lower limbs. She had a more intense distal lower limbs weakness (MRC 3), with deformities in pes cavus, hammer fingers and aquileus retractions. Due to the marked presentation of distal weakness with the described deformities, she was initially diagnosed with Charcot-Marie-Tooth (CMT) disease and referred to our center. On investigation, we found normal levels of creatine phosphokinase (CK); no evidence of polyneuropathy was seen in neurophysiological studies. Biopsy of brachial biceps muscle revealed well delimited cores by the staining of NADH-TR. MRI showed involvement of paravertebral and posterior compartment of the leg muscles. The whole exome sequencing showed two missense likely pathogenic variants in RYR1 (p.Phe41Ser and p.Gly4686Asp). **Discussion:** The RYR1 gene encodes the ryanodine 1 receptor channel and is the main cause of susceptibility to malignant hyperthermia and a wide spectrum of congenital myopathies, including central core myopathy (CCM). CCM is characterized by central cores on muscle biopsy and clinical features of a congenital myopathy with autosomal dominant (AD) and recessive (AR) inheritance patterns. The patients present predominantly proximal weakness pronounced in the hip girdle with orthopedic complications. On the other hand, classic CMT phenotype reflects length-dependent axonal degeneration represented by distal sensory loss and weakness, deep tendon reflex abnormalities, and skeletal deformities. According to an international cohort of 21 AR RYR1-related CCM patients, all of them had predominantly proximal muscle weakness. Although most had some involvement of distal muscle groups, distal weakness was as severe as proximal weakness in only 4. An early exclusively distal phenotype has not been reported in the main studies of CCM. **Final Comments:** Our patient presented an atypical phenotype for RYR1-related CCM, with predominantly distal weakness and feet deformities that resembled the CMT phenotype. RYR1 must be considered as a cause of distal myopathies. Pes cavus and hammer fingers are not specific for CMT.

Doenças Neuromusculares

TL 1105194

A CASE PRESENTATION OF RECESSIVE MYOTONIA CONGENITA (BECKER'S DISEASE) AND EPILEPSY. IS IT CAUSAL OR CASUAL LINK?

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Case Presentation: Male, with normal neuropsychomotor development in early childhood, started at 7 years of age with generalized epileptic seizures. He was treated with carbamazepine and phenytoin, evolving with complete remission of the attacks in 1 year. At 10 years of age, he developed stiffness, progressive distal and proximal weakness of the upper and lower limbs, initially with stumbling and difficulty running. At the age of 15, he developed the need for a wheelchair to walk distances of more than 100 meters. At 18 years of age, he presents fatigue and tiredness to chew solid foods. Past history: Pregnancy and delivery without complications. Family history: no neurological disease. Neurological examination shows: Myotonic phenomena in hands, mouth and tongue, with Warm up sign. Gowers sign present. Hypermobility of the interphalangeal joints of the hands, swan-necked fingers. Biceps, deltoids and gastrocnemius with pseudohypertrophy. Presents facial and masticatory muscle weakness, mild weakness for neck flexion. Muscle strength grade 4 proximal and distal lower limbs, grade 4 proximal and 3 distal upper limbs. Patellar reflexes 3/4 and Achilles 3/4. Sensitivity preserved. Myopathic gait. Laboratory tests with a slight increase in CK levels (341 U/L). Electroencephalogram (2012): epileptic activity characterized by polyspikes. MRI of the brain (2012): no changes. Electromyography (2022): diffuse myotonic discharges. Genetic Test (2022): heterozygous variants of autosomal recessive pattern, CLCN1 chr7: 143,351,593 G>A c.2596-1G>A and Gene CLCN1 chr7: 143,342,461 T>C. Confirming the diagnosis of Myotonia Congenita (MC) recessive disorder (Becker) that is associated with moderate to severe myotonia. Studies show evidence that CLCN1 may be a gene that can also cause generalized genetic epilepsy. This case illustrates a patient diagnosed with Becker's Autosomal Recessive Myotonia Congenita, confirmed by genetic testing, associated with epilepsy and hypermobility of the phalanges. Epilepsy is rarely described in patients with CM but some studies suggest a role of CLCN1 in regulating brain excitability, it may be that the disorders are linked by this mutation in the chloride channel.

Doenças Neuromusculares

TL 1105212

TUMOUR NECROSIS FACTOR RECEPTOR-ASSOCIATED PERIODIC FEVER SYNDROME (TRAPS) IN TWO HYPOTONIC SIBLINGS WITH AXONAL POLYNEUROPATHY AND RECURRENT ABDOMINAL PAIN

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Case Presentation: Two siblings, a 10-years-old girl and a 7-years-old boy, hypotonic since birth with delayed motor milestones. Gait was unbalanced with frequent falls with aggravated after yellow fever vaccination. Family complained of progressive worsening of ambulation. Both had recurrent episodes of vomiting and abdominal pain associated with bloody diarrhea. Fever without known source occurred frequently. Both complained of chronic burning pain in lower limbs. During neurological examination both had signs of a symmetrical sensory motor polyneuropathy with areflexia, ataxic gait intensified when closing eyes and global hypotonia. C-reactive protein and erythrocyte sedimentation ratio were highly elevated. MRI was unremarkable. Electroneuromyography depicted a chronic axonal symmetrical distal sensory motor polyneuropathy. Whole exome sequencing showed a heterozygous variant in the TNFRSF1A gene, confirming the diagnosis of Tumour Necrosis Factor Receptor-Associated Periodic Fever Syndrome (TRAPS). Canakinumab was initiated with complete resolution of the gastrointestinal symptoms, lower limb pain and marked improvement in gait. **Discussion:** TRAPS is an autosomal dominant autoinflammatory syndrome caused by variants in the TNFRSF1A gene. Recurrent episodes of periodic fever, myalgia, arthralgia, abdominal pain, periorbital edema and migratory rashes are well known features. Episodes occur either spontaneously of following a trigger. The neurological manifestation still need further characterization. Headache, aseptic meningitis, myositis and neuropathy are described but hypotonia and delayed motor milestones has never been reported to our knowledge. Inflammatory biomarkers are often elevated. Electroneuromyography should be ordered in all cases to better distinguish the features of myopathy and neuropathy that may be present in this group. Early diagnosis is essential since patients experience significant improvement with treatment. The knowledge and early recognition is needed for a better depiction of the neurological manifestations. **Final Comments:** TRAPS is an autosomal dominant hereditary autoinflammatory syndrome, associated with an expanding neurological picture in the literature. Our case shows two siblings with marked hypotonia, delayed motor milestones, axonal sensory motor polyneuropathy and a marked recovery after

Doenças Neuromusculares

TL 1105213

FUKUTIN-RELATED LIMB-GIRDLE DYSTROPHY PRESENTING AS PSEUDOMETABOLIC MYOPATHY

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Case Presentation: A 35-years-old woman, born from non-consanguineous parents with a Japanese ancestry, presented with an episode of myalgia, dark colored urine, high CK levels after volleyball training at age 15- years-old. During this episode, CK level was 15.450 U/L. She had a spontaneously recovery. She then developed progressive exercise intolerance, mild proximal weakness, and CK levels remained four times upper limit value. Electrodiagnostic studies suggested a myopathic process with normal repetitive stimulation. A muscle biopsy of biceps brachii revealed a mild myopathic pattern with mitochondrial and glycogen accumulation, suggesting a metabolic myopathy. On a recent visit, physical examination showed MRC 4 in proximal muscles, axial weakness, and winged scapula. Cervical weakness was also noted. She was unable to get up from lying in the examination table. Neuromuscular ultrasound indicated mild hyper echogenicity (Heckmatt 2) in brachialis and vastus intermedius, and moderate hyper echogenicity (Heckmatt 3) in hip adductors and rectus abdominal muscle. Diaphragmatic muscle involvement was also demonstrated. No tongue involvement was seen. A next generation panel for neuromuscular genes revealed compound heterozygous pathogenic variants in the Fukutin gene (FKTN). DNA segregation confirm the recessive pattern. **Discussion:** FKTN biallelic mutations usually present as a congenital myopathy, mainly in Japan and is considered quite rare in other populations. Presentation of limb-girdle manifesting during late childhood is even rarer. Only one case of FKTN related congenital muscular dystrophy was reported in a Japanese patient living in Brazil. In a recent national limb-girdle muscle dystrophy cohort, this gene was not included. As muscle panels are becoming more widespread and accessible, FKTN could be an answer of previous undiagnosed muscle dystrophy in people with a Japanese ancestry. **Final Comments:** Brazil has the larger Japanese community outside Japan, therefore FKTN-related myopathies should be considered not only when facing a congenital myopathy or a limb-girdle phenotype, but also when a pseudometabolic myopathy is present. More studies are needed to identify real frequencies of this rare disorder in our country.

Doenças Neuromusculares

TL 1105221

ANTI-AQUAPORIN 4 ANTIBODY-RELATED MYOPATHY

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Case Presentation: 47-years-old Woman, presented bilateral optic neuritis in the third decade of life associated with longitudinal extensive transverse myelitis. Anti-aquaporin 4 antibody was requested and came back positive. Due to clinical picture and positive anti-aquaporin 4 the diagnosis of neuromyelitis optica spectrum disorder (NMOSD) was made. Chronically complained of xerostomia and xerofthalmia and had positive anti-Ro antibody. One year ago, noticed pain in lower limb muscles, being worse in the proximal muscles and associated with important fatigue, aggravating the gait that was already compromised by the demyelinating outbreaks. Creatinephosphokinase and aldose were highly elevated. Lower limbs MRI depicted liposubstitution on the T1 sequencing and edema on STIR sequencing, in the anterior compartment of the thigh bilaterally and in the medial gastrocnemius on the right, an image pattern suggestive of myopathy. **Discussion:** Aquaporin 4 is the most prevalent aquaporin channel in the central nervous system (CNS). Its function is to provide rapid transport of water, maintaining balance and cellular homeostasis. The detection of anti-aquaporin 4 antibody is an important hallmark in the diagnosis of NMOSD, a well-characterized disease of the CNS. However, aquaporin 4 is also expressed outside the CNS, as in the striated skeletal muscles, and can cause hyperCKemia, myositis with muscle lesions evidenced in muscle biopsy and MRI. We present a case of a patient with NMOSD, with worsening fatigue and significant myalgia in the lower limbs, elevation of creatinephosphokinase and aldolase, with myopathic image pattern on MRI of the lower limbs. The condition described is an extra-CNS manifestation of anti-aquaporin 4 positive NMOSD and should be screened for in cases of myalgia, worsening of fatigue and gait without evidence of a new demyelinating outbreak. **Final Comments:** The association of anti-aquaporin 4 antibody with NMOSD is well established. Currently recognized, but less explored in clinical practice, is its association with myopathy. HyperCKemia associated with anti-aquaporin 4 antibody should be considered a sign of muscular manifestation of NMOSD. Through this case presentation we hope to increase knowledge and interest about muscle involvement in anti-aquaporin 4 positive NMOSD.

Doenças Neuromusculares

TL 1105226

CASE PRESENTATION DYSAUTONOMIA IN SPASTIC PARAPLEGIA TYPE 5A (SPG5A)

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Case Presentation: A 26-years-old woman presented with slowly progressive weakness and stiffness in her legs, followed by urinary incontinence. She also reported dizziness when standing with frequent falls and changes in the color of her toes associated with swelling when the weather is cold. Physical examination revealed persistent painless toes cyanosis and neurological examination showed crural spastic paraparesis, patellar and ankle hyperreflexia, bilateral Babinski sign and distal impairment of all sensory modalities in lower limbs. Neuroimaging studies were unremarkable. Tilt test demonstrated postural hypotension and urodynamic study reveals detrusor overactivity and stress urinary incontinence. Genetic testing identified the pathogenic variant c.825T>A (p.Tyr275*) in homozygosity in the CYP7B1 gene associated with hereditary spastic paraplegia 5A (SPG5A). **Discussion:** The chronic phase of spinal cord injury may present with dysautonomia, including urinary dysfunction, orthostatic hypotension, and even impaired regulation of the body extremities due to loss of vasomotor tone in lower limbs. Neurogenic bladder, postural hypotension, sexual dysfunction, and decreased sweat output have been associated with different SPG subtypes. However, to our best knowledge, we did not find any case presentation with an association between these symptoms and SPG5A. In this case, the presence of postural hypotension, urinary incontinence and cold-induced acrocyanosis suggest impairment of the autonomic nervous system. **Final Comments:** Our case expands current knowledge about the association between SPG5A and autonomic disturbances. We conclude that autonomic involvement may be underrecognized in the different SPG subtypes.

Doenças Neuromusculares

TL 1105283

A COMPLEX ASSOCIATION OF SENSORIMOTOR AXONAL NEUROPATHY, DEAFNESS, EPISODIC WORSENESS, AND RETINOPATHY: MITCHELL SYNDROME

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Case Presentation: A 28-year-old Brazilian woman presented with a long-standing clinical course of lower limb weakness starting at age 15 years, associated with recurrent and prolonged episodes of numbness and neuropathic pain. Her symptoms evolved and after five years she presented with worseness of motor symptoms and sensory loss in the lower limbs, mild motor compromise of the upper limbs, and bilateral sensorineural hearing loss. After almost one decade of symptom-onset, she noted marked worseness of motor symptoms, urinary disturbances, severe hearing loss (with the need of bilateral cochlear implants), and progressive unilateral visual loss of the left eye. Medical history disclosed short stature of unknown etiology. No parental consanguinity was observed. Examination disclosed distal dominant quadriplegia with brisk global reflexes and absent Achilles tendon reflexes, sensory ataxia, steppage gait, and marked hypoaesthesia and hypopalesthesia in the lower limbs. Neurophysiological studies disclosed axonal sensorimotor polyneuropathy. Brain MR imaging showed mild periventricular leukoencephalopathy. Optic coherence tomography showed bilateral thinning of the neurosensory retina. As riboflavin transporter defect was suspected, oral riboflavin was started, 20 mg per kg, with moderate improvement of motor, visual and hearing complaints. NGS-based gene panel testing showed the heterozygous variant c.1384C>T (p.Arg462Cys) in the ACOX1 gene involved with Mitchell syndrome. **Discussion:** Palmitoyl acyl-CoA oxidase 1 represents the rate-limiting peroxisomal enzyme involved in the beta-oxidation of fatty acids, which is related both to loss-of-function mechanisms with enzyme deficiency due to biallelic variants and to gain-of-function with neurotoxic mechanisms due to heterozygous variant. Mitchell syndrome is an extremely rare progressive inherited neurometabolic and neurodegenerative disorder associated with sensorimotor polyneuropathy, deafness, and episodes of clinical worseness with central or peripheral demyelination. Our case description disclosed progressive visual loss and moderate response to riboflavin therapy. **Final Comments:** Clinicians must be aware about the possibility of facing new clinical presentations of inherited peroxisomal disorders, such as Mitchell syndrome, in the clinical context of progressive hearing loss, polyneuropathy, sensory ataxia, visual loss, and acute or subacute episodes of motor and sensory worseness.

Doenças Neuromusculares

TL 1105285

CONGENITAL MYASTHENIC SYNDROME DUE TO GMPPB VARIANTS

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Case Presentation: A 40-year-old Brazilian woman presented with a long-standing slowly progressive clinical course of muscle fatigue and exercise intolerance since age 22 years. After two years of symptom-onset, she also noted eyelid ptosis without diplopia and dysgraphia. There was also worsening of muscle weakness at the end of the day and after physical activity. By the age of 36, weakness worsened, and she could not walk for more than 10 minutes. At that time, serum creatine kinase levels were evaluated and disclosed mild elevation (900; 3 times the upper limit value). One year ago, she started diplopia, dyspnea, and dysphagia. Medical history disclosed dyslipidemia. Parental consanguinity was present. Family history disclosed her sister with similar symptoms. Examination disclosed mild proximal weakness in the upper and lower limbs, mild axial muscle weakness. Her nerve conduction studies were unremarkable. Needle electromyography disclosed typical chronic myopathic features. Muscle biopsy showed nonspecific myopathic findings. A next-generation sequencing-based multigene panel for inherited neuromuscular disorders disclosed the presence of the pathogenic variant c.95C>T (p.Pro32Leu) and the variant of uncertain significance c.478G>A (p.Val160Met) in compound heterozygosity in the gene GMPPB, thus enabling the diagnosis of GMPPB-related Congenital Myasthenic Syndrome (CMS) associated with myopathic involvement. Oral salbutamol and pyridostigmine were started with clinical stability of symptoms. **Discussion:** GMPPB-related neuromuscular disorders represent Congenital Disorders of Glycosylation and occur due to variable deficiency of mannanose-1-phosphate beta-guanidyltransferase, which is involved with GDP-mannose biosynthesis related to multiple glycosylation pathways. It has been previously associated with autosomal recessive limb-girdle muscular dystrophy type 19 (formerly LGMD2T), Congenital Muscular Dystrophy-Dystroglycanopathy type 14 and, more recently, with late-onset CMS phenotypes. Some of the previously described GMPPB-associated myopathies had dysfunction of the neuromuscular junction and mildly raised serum creatine kinase levels. **Final Comments:** Clinicians must be aware about the possibility of late-onset presentations of CMS due to GMPPB variants, which should be considered in cases with myopathic findings in neurophysiological studies and raised serum creatine kinase levels.

Doenças Neuromusculares

TL 1105293

FACIAL ONSET SENSORY AND MOTOR NEURONOPATHY (FOSMN SYNDROME): A CASE PRESENTATION

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Case Presentation: A.M.M., male, 60 years old. This patient was referred for neurological evaluation due to an insidious complaint of left arm paresthesia, weakness and atrophy since 2003. Electroneuromyography was performed, demonstrating cervical radiculopathy. He didn't continue the medical investigation and returned in 2017 complaining that the weakness had progressed to his ipsilateral face and leg. On neurological examination, upper and lower motor neuron signs were found. In addition, midface hypoesthesia ("onion bulb") and blink reflex abnormality were observed. Electroneuromyography was performed, demonstrating signs of denervation in bulbar segments and alteration of the blink reflex, suggesting the diagnosis of FOSMN. **Discussion:** FOSMN is a rare neurologic syndrome and pathogenesis is uncertain, but studies suggest that the pathophysiology is more neurodegenerative than autoimmune. Apparently, FOSMN is most likely to be a TDP-43 proteinopathy within the ALS-FTD spectrum. The mean age at onset is approximately 55 years with progressive course involving sensory disturbances in trigeminal nerve pathway, bulbar symptoms, upper extremities and, in some cases, to the lower extremities, as in this case. The diagnostic marker is the absence of the blink reflex on EMG in addition to the presence of denervation that can be found in the bulbar, cervical and thoracic preganglionic segments, although there are no specific criteria, which makes the diagnosis difficult. **Final Comments:** This case presentation emphasizes that it is very important to recognize rare diseases within the spectrum of neuromuscular disorders, such as FOSMN, in the context of progressive pain and weakness with cranial-caudal progression. However, the clinical pictures and pathophysiology of FOSMN have not been well established.

Doenças Neuromusculares

TL 1105306

TUBULAR AGGREGATE MYOPATHY WITH RHABDOMYOLYSIS: AN UNCOMMON PRESENTATION IN ELDERLY MALE

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Case Presentation: A 76-year-old male with acute symmetrical inferior limbs weakness and pain. No previous history of muscle weakness. Denied alcohol intake, prior exercise intolerance, fatigue, myoglobinuria, ptosis, or pigmenturia, as well as worsening after periods of fasting. History of systemic arterial hypertension and dyslipidemia. He had been treated with the same dose of simvastatin and antihypertensives. Furthermore, no relevant familial history of consanguinity. On examination, the strength of proximal inferior limbs graded MRC grade IV. Neoplastic, infectious, rheumatological, and endocrinology diseases were ruled out. Initial CK = 6.719U/L. Muscle MRI revealed a heterogeneous edema pattern, with predominant proximal impairment. No evidence of atrophy was detected. Statins were promptly withdrawn. After 5 to 7 days, CPK lowered to normal level and muscle biopsy confirmed Tubular Aggregate Myopathy. There was a complete improvement of muscle weakness after 30 days. **Discussion:** Tubular Aggregate Myopathy is a rare disorder of unknown prevalence. The symptoms and severity are widely heterogeneous, and the diagnosis is based on a history of progressive loss of strength and/or muscle pains, muscle biopsy revealing tubular aggregates, and absence of another neuromuscular disease. The Tubular Aggregate Myopathy can present as a pseudo metabolic feature, without progressive weakness, mimicking the metabolic myopathies. **Final Comments:** Tubular Aggregate Myopathy is a rare and an inherited disorder and, differently from metabolic myopathies, is characterized by slowly progressive proximal muscle weakness, cramps and myalgia. Muscle biopsy usually points to tubular aggregates of variable morphology and type II fibers atrophy.

Doenças Neuromusculares

TL 1105309

CLINICAL IMPROVEMENT IN ANTI-MUSK MYASTHENIA GRAVIS PATIENT AFTER THYMECTOMY – A CASE PRESENTATION

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Case Presentation: Female patient, 32 years old, presented eyelid ptosis to the right, diplopia, dysphonia and proximal appendicular weakness in superior members with floating characteristic. She was submitted to an electroneuromyography with repetitive stimulation (decremental pattern). The tomography of the chest showed thymic hyperplasia associated to the antireceptors antibody dosage of acetylcholine (AChR) negative, but anti-kinase antibody muscle-specific (MuSK) positive. She initiated the usage of prednisone up to 80mg/day and pyridostigmine reaching 360mg/day with partial response. Continued with the symptoms worsening, tachypnea and Hospitalization in an intensive care unit. Medication adjustment was attempted without any improvement, besides developing Cushing's syndrome secondary to the corticotherapy. Then a thymectomy was chosen. The patient presented diaphragmatic paralysis after the thymectomy, and a diaphragmatic plication was carried out with clinical improvement. The weaning of the corticotherapy was initiated. After six months she is found using exclusively prednisone 5mg/day and without myasthenic symptoms. **Discussion:** Myasthenia gravis (MG) is an autoimmune disorder of the neuromuscular junction hallmarked by motor fluctuations (fatigability). Treatment options are acetylcholinesterase inhibitors, glucocorticoids, immunoglobulin, plasmapheresis, immunosuppressives and monoclonal antibodies (eculizumab). The antibodies for the diagnosis are anti-AChR, anti-MuSK and anti-protein related to the lipoprotein 4 (LRP4). The thymectomy is indicated based on the flaw of the thymic recognition function and negative selection of autoreactive T cells – physiopathology implied mechanism of mediated MG AChR (MG-AChR). The thymectomy is indicated on thymic persistence or on thymoma presence, when is performed the mediastinal toilet. Not long ago thymectomy was not indicated in anti-MuSK positive cases, but the last updates do not define thymectomy as a resource or contraindicate. Interestingly, in this case, despite thymectomy's possible inefficacy for anti-MuSK positive patients, an unequivocally improvement and corticoids/pyridostigmine dosage reduction was observed after the thymectomy. **Final Comments:** The success of the therapeutic response after the thymectomy on an anti-MuSK positive patient puts in discussion the debate about thymectomy as a therapeutic weapon in myasthenic anti-MuSK positive patients with persistent thymus/ thymoma.

Doenças Neuromusculares

TL 1105342

INCREASING GENETIC AND PHENOTYPIC VARIABILITY OF PRPS1 GENE MUTATIONS: A NEW MUTATION ASSOCIATED TO A COMPLEX HEREDITARY MOTOR NEUROPATHY

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Case Presentation: Two male siblings from non-consanguineous parents presented in their first decade of life with a complex phenotype including delayed motor milestones, dysarthria and deafness. Ataxic gait was noted at age of three in both. Symptoms worsened over the years, mainly gait, with frequent falls. Their mother presented hypoacusia by the age of 54, with no other symptoms. Brain MRI revealed cerebellar atrophy. Nerve conduction studies revealed motor axonal neuropathy in both siblings. WES revealed a class 5 variant on the PRPS1 gene: (NM_002764.4) c.894A>C. Hence, a diagnosis within the spectrum of Arts syndrome and PRPS1-related disorders was made. **Discussion:** Arts syndrome is a rare entity described in 1993. Twelve young boys of a family presented sensorineural hypoacusia, ataxia, delayed motor milestones and tendency to upper respiratory tract infections. Later, an autopsy showed absence of myelinated axons in the posterior column of the spinal cord. It is an X-linked condition caused by a loss-of-function mutation on the PRPS1 gene leading to decreased enzyme function. Besides Arts syndrome, mutations in PRPS1 have been associated with a large spectrum of manifestations, as X-linked recessive Charcot-Marie-Tooth disease-5 (CMTX5) and X-linked deafness-1 (DFNX1). Adult female carriers can present isolated hypoacusia, as did the mother of our patients. We found description of 22 pathogenic variants in this gene, apart from the aforementioned new variant. Clinical features of this case showed the presence of motor neuropathy, sensorineural hypoacusia and delayed motor milestones, which have been described in some loss-of-function mutations. Gain-of-function mutations described so far are mostly related to hyperuricemia and gout arthritis. This is of great importance as a minor study has shown benefit from providing S-adenosylmethionine (SAM) to some patients. Description of a novel variant in the PRPS1 gene may support the impression that according to the mutation and residual enzyme function, different clinical features are seen. **Final Comments:** We present a new variant in the PRPS1 gene related to Arts syndrome, and a pure motor neuropathy could be a new finding that had not been described so far. Segregation analysis suggests an X-linked inheritance. PRPS1 gene is related to nucleotide metabolism and the synthesis of purines, and identification of this mutation is of practical relevance as SAM supplementation may be beneficial to these patients.

Doenças Neuromusculares

TL 1105358

RHABDOMYOLYSIS AND MUSCLE ADAPTATION AFTER WHOLE-BODY ELECTROSTIMULATION (WB-EMS)

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Case Presentation: A 25-year-old woman (BMI 22kg/m²), without comorbidities, presented with an incidental high creatine kinase (CK) level (29.068 U/L) after starting whole-body electrostimulation (WB-EMS). She had been previously untrained and identified mild myalgia believing it to be expected due to training. There was no urinary complaint or muscle weakness. Her neurological exam was normal. Her renal function was normal and urine sample one week after the event showed no changes. Muscle MRI demonstrated small foci of edema in gluteus maximus, vastus lateralis and intermedii muscles. We proceeded with a complete investigation including full rheumatological panel, thyroid function, serologies (HIV, hepatitis B and C, dengue, leptospirosis, toxoplasmosis) and whole-exome sequencing which resulted all negative. Most plausible explanation was exertional rhabdomyolysis and we suggested a return to WB-EMS trainings with weekly serum CK measurement. The high CK level seen after individual WB-EMS session was progressively blunted and normalized after 2 months of regular training. A control muscle MRI five months after regular training showed resolution of muscular edema. **Discussion:** WB-EMS uses an electrical stimulator with several channels that can recruit more muscle fibers than normal exercises and attracts people for the belief of being more time-effective. The case illustrates the risks of WB-EMS related to muscle injury, especially in physically deconditioned people, and how to manage training-related CK level increase. There are not enough data regarding the prevalence of rhabdomyolysis associated with WB-EMS. However, it has been previously reported especially after the first workout session and studies show a drop in CK levels after regular training. Our case reinforces that maintenance of WB-EMS training may be safe even after a very high CK level. Muscle ability to adapt through exercise training is the most probable explanation to justify serum CK increase attenuation. **Final Comments:** Patients and health care professionals must be alert to intense muscle pain and changes in urine color, especially at the beginning of training. Clearer protocols are needed to guide physicians and neuromuscular practitioners to manage these patients as WB-EMS will become even more popular. Serial CK measurement may be necessary in subjects beginning WB-EMS training and an evaluation prior to WB-EMS is advised to identify potential individual risks.

Doenças Neuromusculares

TL 1105370

CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY (CIPD) WITH UNILATERAL FACIAL AND HYPOGLOSSAL INVOLVEMENT: A CASE PRESENTATION

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Case Presentation: A 31-year-old female with a two-months history of progressive weakness and paresthesia in both upper limbs, mainly distal, followed by a two-day right foot numbness. She referred transient and recovered weakness in the right hand three years prior to admission, with normal exams and no follow-up needed. She had no comorbidities, antecedent infection or recent immunization. She underwent hospitalization and extensive laboratory and radiological work-up. Cerebrospinal fluid analysis revealed high protein content (>90,8mg/dL), normal cell count (WBC 1/mm³, 100% lymphocytes) and normal glucose level (54 mg/dL). Needle electromyography (EMG) and nerve conduction velocity (NCV) study showed diffuse, symmetric, sensorimotor, predominantly primary segmental demyelinating polyradiculoneuropathy. No other relevant findings were found. Initial treatment included intravenous methylprednisolone and immunoglobulin that shown immediate clinical improvement. Outpatient management identified facial asymmetry two months later, besides recurrency of incipient weakness and numbness in extremities. A new course of medication was carried out also with improvement in symptoms and bimonthly sessions were scheduled. On her next return, she complained of difficulty speaking and eating. Neurological exam evidenced right hypoglossal palsy that determined dysarthria and impaired mobility of the bolus within the mouth. No atrophy or tongue fasciculation were found. New laboratory and radiological exams did not show any abnormalities and 45-day interval course of medication was restarted with no further significant neurological deterioration. **Discussion:** The usual phenotype of CIPD is a sensorimotor disorder with symmetric proximal and distal affectation, areflexia and conduction slowing, temporal dispersion and/or conduction block. Cranial nerve findings are not common: facial muscle weakness may be present in up to 13% and hypoglossal involvement has been reported in only 0–3.8%. **Final Comments:** CIPD is an immune-mediated chronic neuropathy with a wide spectrum of clinical manifestations, lasting for more than two months. Although cranial nerve manifestation is infrequent (7%–16%), we describe this uncommon finding in a patient with facial and hypoglossal involvement. Therefore, therapeutic strategies and prognostic implications of the presence of cranial neuropathy have yet to be determined and active examination to search this affection must be carefully accomplished.

Doenças Neuromusculares

TL 1105398

JAG2-RELATED LIMB-GIRDLE MUSCULAR DYSTROPHY: FIRST REPORT IN LATIN AMERICA AND DESCRIPTION OF A NOVEL VARIANT

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Case Presentation: Two mixed-race siblings, non-consanguineous parents, Rio Grande do Sul, Brazil, referred to our outpatient clinic this year at ages 35 (case 1) and 24 (case 2). Both had normal neuropsychomotor development up to 24 months and 12 months, respectively, when they developed progressive proximal weakness on 4 limbs and lost the ability to walk at ages 8 and 7. Evolution of motor disability with increased frailty and muscle atrophy in 4 limbs for the following 10 years, predominantly proximal in the lower limbs, associated with contractures with joint deformities and limited extension. Both presented dorsal scoliosis and mild cervical weakness. Case 1, at age 25 showed significant bulbar weakness, with dysphagia and dyspnea at rest with the need for intermittent bilevel positive airway pressure. Case 2 does not present dysphagia or symptomatic dyspnea. Both maintain support therapy with motor physiotherapy. The initial investigation included: normal CK, EMG showing myopathic pattern and muscle biopsy at ages 4 and 8 (case 1) and at age 3 (case 2) with no conclusive results. The molecular confirmation of the diagnosis was only reached in 2022, by means of a next-generation multigene sequencing (NGS) panel, which revealed a heterozygous pathogenic variant in the JAG2 gene [T>TGGCCP. His1171ArgFS*9] that promotes amino acid substitution histidine in code 1171 by Arginine and change in the reading matrix from this point promoting early interruption of protein translation in case 2. In addition, case 1 was also tested with another NGS panel for neuromuscular conditions (which did not include JAG2), but it resulted in no relevant variants. **Discussion:** Autosomal recessive limb-girdle muscular dystrophy-27 (LGMDR27) is caused by pathogenic variants in the Jagged Canonical Notch Ligand 2 (JAG2). It was first described in 2021 in a single international cohort with no reported cases in Latin America. This variant is classified as pathogenic due to early interruption of protein translation that contributes to protein dysfunction in the notch signaling pathway, which is important in intercellular signaling. **Final Comments:** This report expands the geographical distribution of LGMDR27 to Latin America, describes the clinical features of two new cases of this rare condition and contributes a novel variant affecting the JAG2 gene.

Doenças Neuromusculares

TL 1105409

A DANCER WITH CRAMPS: MOERSCH-WOLTMAN SYNDROME OR STIFF-PERSON SYNDROME

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Case Presentation: A 30 yo, female, psychologist and dancer, previously healthy, non-consanguineous parents, without family history of neurological diseases. In 2018, she noticed limited movement of her left leg and hip, especially when dancing and performing “pilates”, activities that she previously performed without complaints. She had severe painful cramps, associated with intense stiffness that later migrated to her right leg up to the lumbar column. She started physiotherapy with partial improvement of the condition, but it was discontinued in August 2020, which led to progressive worsening, now compromising her upper limbs and leading to fixed cervical hyperextension and hyperlordosis, confining her to the wheelchair. She also developed solid-liquid dysphagia, dysphonia, and dyspnea on minor exertion. Neurological examination revealed a fixed semi-flexed position of her lower limbs, lumbar hyperlordosis, and a semi-flexed trunk posture. Strength was grade 2 of the iliopsoas, thigh abductors, and adductors; grade 4+ of the biceps and quadriceps femoris, and grade 5 on the other muscle groups. Generalized grade 3+ deep tendon reflexes with bilateral plantar flexion reflexes. Postural instability and inability to walk. Baclofen and Pregabalin were administered up to maximal doses, with no response. Clinical improvement was observed only with Clonazepam 2 mg/d, with muscle relaxation, resuming independent gait. An investigation was carried out with serology, rheumatological profile, skull MRI and screening for neoplasms, with all negative results. Normal CPK dosage. Serum dosage of Anti-GAD with result > 2000 IU/ml. Therefore, clinical, laboratory, and therapeutic response led to the final diagnosis of Stiff-Person syndrome, which is why electroneuromyography was not performed at the time. Therapy continued with Clonazepam, Prednisone 60 mg/day, Azathioprine 150 mg/day, and due to partial response to therapy, she is now awaiting infusion of Human Immunoglobulin. **Discussion:** Due to its often prolonged evolution and rarity, the diagnosis of Stiff-Person syndrome is still challenging, and its diagnostic criteria are not yet well established. **Final Comments:** This hypothesis should be considered in all patients presenting muscle stiffness associated with pain and muscle spasms, mainly in the lower back. There are several differential diagnoses, and anti-GAD dosage should be requested for diagnostic aid.

Doenças Neuromusculares

TL 1105431

NUSINERSEN AND MULTIDISCIPLINARY APPROACH IN IMPROVEMENT OF MOTOR FUNCTION IN SPINAL MUSCULAR ATROPHY TYPE 3: A 2 YEARS FOLLOW-UP CASE PRESENTATION

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Case Presentation: Male patient initiated progressive weakness at 2-years-old. He was able to walk, but began to have frequent falls and difficulty climbing stairs. At the age of 14 he was referred to genetics due to a slow and progressive worsening. Denied dysphagia, dyspnea, speech or sphincter alterations. Upon examination, he presented with proximal and symmetric weakness, associated with lower limb atrophy and 4-membered areflexia. In 2018, at the age of 23, he was diagnosed with Spinal Muscular Atrophy (SMA) type 3 due to electroneuromyography and genetic examination. He began treatment with nusinersen in 2019, along with multidisciplinary follow-up. Motor function data were assessed by trained evaluators using Hammersmith Functional Rating Scale Expanded (HFMRSE) and Ten-Meter Walking Test (10 MWT), according to the international recommendations on SMA standard of care, at the onset of treatment and reassessed every six months. After 2 years, the patient increased 5 points in the HFMRSE and decreased 7s in the 10 MWT, which, according to the literature, defines clinically meaningful changes. **Discussion:** SMA is an autosomal-recessive lower motor neuron disease causing progressive muscular atrophy and weakness. It is divided into four subtypes (0-4) according to age of symptoms onset and achieved motor landmarks. Nusinersen is an antisense oligonucleotide administered intrathecally, able to increase functional SMN protein levels, modifying SMA natural history. Although approved for any 5q SMA, most studies focus on nusinersen efficacy in infants and children, with data on adults limited to few observational studies and case series. This case shows clinical improvement in motor function of a SMA type 3 patient, even with the onset of treatment being twenty years after the onset of symptoms. This supports nusinersen efficacy in a long disease duration setting. Furthermore, multidisciplinary management is a well established therapy designed to address the psychosocial challenges of patients with SMA and cannot be excluded as an adjuvant in this patient's enhancement. **Final Comments:** In adult SMA, outcome measures have not been as thoroughly standardized and validated. Although it is proven that a longer disease duration has a negative impact on treatment efficacy, this report indicates a satisfactory enhancement in unfavorable clinical conditions. Also, a multidisciplinary approach acts as an adjuvant for Nusinersen treatment in the improvement of motor function.

Doenças Neuromusculares

TL 1105448

URBANORUM SPP AND GUILLAIN-BARRÉ SYNDROME, A POSSIBLE ASSOCIATION?

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Case Presentation: A 57-year-old, white, male patient, previously without comorbidities, presented with diarrhea lasting four days, with aqueous content, without pathological products and with improvement after the use of symptomatic. After eight days, he started paresis in feet and legs evolving with paraparesis and needing support to walk. On admission, the neurological examination revealed asymmetric tetraparesis of proximal predominance and areflexia with Medical Research Council (MRC) with a total score of 24/60 and proteinocytological dissociation in cerebrospinal fluid. Due to the neurological clinical condition and laboratory findings, the diagnosis of Guillain Barre syndrome was suggested. Electroneuromyography was performed with signs of acute motor neuropathy, of axonal pattern, compatible with motor axonal form (AMAN). Treatment with intravenous human immunoglobulin was then started on the third day of Hospitalization at a dose of 0.4kg/kg/day for 05 days. During Hospitalization, abdominal distension associated with watery diarrhea was observed. Positive stool parasitological examination and visualized urbanorum spp. with a positive fecal leukocyte test. In addition to the therapeutic approach of the syndrome, treatment for diarrhea secondary to urbanorum spp was also instituted with nitazoxanide for 07 days with improvement of abdominal distention and reduction of evacuation frequency. **Discussion:** The high prevalence of parasites are sources of constant infections for the Brazilian population due to its territorial extension and precariousness in terms of basic sanitation. knowing the new incident parasites, their pathophysiology, virulence and possible associated diseases means that there are ways to combat and minimize the damage. A more detailed study of Urbanorum spp, its morphological structure and mainly, which drug therapy to use will bring uniformity of information and conduct. Awareness measures and basic hygiene care must be carried out to reduce transmissibility and reduce the incidence of the parasite. **Final Comments:** We report the first case of clinical suspicion of a temporal association between Guillain-Barré syndrome and gastrointestinal tract disease caused by Urbanorum spp. Thus, seeking to alert about the probable relation of the parasite and the risk of development of GBS, as well as the pathophysiological mechanisms and treatment.

Doenças Neuromusculares

TL 1105495

CHILDHOOD-ONSET MYOTONIC DYSTROPHY TYPE 1: A CASE PRESENTATION

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Case Presentation: Thirteen year old girl with cramps and pain on both hands that initiated during early childhood and got worse after coronavirus disease 2019 (COVID-19) in november of 2020. She had myotonic phenomena at both hands and tongue, mild proximal weakness on both hip flexors and hip extensors, and a myopathic gait. No history of neurologic or genetic disorders in the family was reported. Laboratory investigation of nutritional deficiencies, endocrine and other systemic involvement was negative. Electromyography showed myotonic myopathy. Genetic testing helped conclude the diagnosis of Myotonic Dystrophy type 1 (DM1). **Discussion:** DM1 is the result of a CTG repeat expansion in the 3' untranslated region of the DM1 protein kinase (DMPK) gene on chromosome 19q13.3. The clinical triad associated with DM1 is myotonia, distal weakness and early-onset cataracts. Patients may refer to myotonia as cramps, muscle stiffness or delayed muscle relaxation. Proeminent pain and proximal weakness are features most associated with Myotonic Dystrophy type 2 (DM2), although case presentation with this presentation in DM1, such as our patient, have been previously reported. Childhood-onset of DM1 is characterized by symptoms initiated between after the completion of the first year of life to ten years old. A childhood-onset is more linked to intellectual impairment and gastrointestinal symptoms, such as diarrhea, constipation and fecal incontinence. Common adult features of myotonia do not present until early adolescence. Our patient describes that pain and cramps got worse after coronavirus infection in 2020 and that was what motivated the family to search for medical counseling. At our best knowledge, there are no reports of worsening Myotonic Dystrophies after coronavirus infection. But patients with DM1 may be at higher risk for presenting severe acute respiratory syndrome (SARS) associated with COVID-19. **Final Comments:** DM1 is a genetic disorder with onset described between birth and early adulthood associated with three cardinal features (myotonia, weakness and early-onset cataracts). Proximal weakness and pain are not the most prominent symptoms in patients with DM1, although previously reported. Patients with childhood-onset are expected to have a more severe course than those with adult onset, and an early recognition of symptoms may lead to a better quality of life in these patients.

Doenças Neuromusculares

TL 1105559

FAMILY WITH HSP 75 PHENOTYPE DUE TO MAG GENE VARIANT

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Case Presentation: A 47-years-old man presented with a slowly progressive history of weakness in the left lower limb since age 35 years. After 1 year, he noticed involvement of the right lower limb, with gradual worsening of gait. Subsequently, onset of paresthesia in the feet associated with episodes of urge urinary incontinence. It has alpha-1 antitrypsin deficiency. In the family history, the father had a history of progressive spastic paraparesis, starting at age 60 years, with recurrent falls associated with cognitive decline over 7 years and urinary incontinence. He later progressed with dysarthrophonia and dysphagia, and death occurred 14 years after the onset of motor symptoms. Parental consanguinity was not related. The examination disclosed crural paraparesis, global hyperreflexia, spasticity in the dorsiflexion of the feet, hypoaesthesia in the left foot and bilateral positive Babinski sign. Biopsy of muscle is unremarkable. A genetic panel was collected, showing a genetic variant of uncertain significance (with "in silico" prediction of pathogenicity) heterozygous mutation in MAG (Myelin Associated Glycoprotein). Thus, we concluded that our family presented hereditary spastic paraplegia 75 (HSP - 75) with cognitive involvement by the MAG gene. **Discussion:** Hereditary spastic paraplegias (HSP) are a heterogeneous group of neurodegenerative diseases and more than 70 genes have been implicated. Although most variant contexts occur in an autosomal recessive scenario in HSP75, previous descriptions (adult-onset spastic paraparesis, action tremor in the hands, cognitive decline) with an autosomal dominant inheritance pattern have already been reported and justify the presented clinical context by patients. **Final Comments:** Clinicians should be aware of the possibility of HSP75 presentation by the MAG gene in the context of autosomal dominant inheritance.

Doenças Neuromusculares

TL 1105565

LATE ONSET ASYMMETRIC MYOPATHY: A CASE OF NEUTRAL LIPID STORAGE DISEASE

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Case Presentation: We present a male patient, 42 years-old, active smoker, without comorbidities. At 38 years of age, he developed an asymmetrical proximal weakness in the upper limbs. Over the next 04 years, there was slowly progressive worsening of weakness in the upper limbs and evolution to the lower limbs. On neurological examination, the patient had asymmetric tetraparesis, with proximal predominance in the upper limbs and distal predominance in the lower limbs, with grade 2 strength at arm abduction bilaterally, grade 2 for forearm flexion and 4 for extension bilaterally, but more prominent on the right side. Dorsiflexion of the right feet was grade 2, and 4 on the left. Selective atrophy of the biceps brachii muscle with sparing of the triceps brachii were remarkable. Investigation showed serum creatine kinase of 3.033. Electromyography had a pattern of asymmetric myopathy with proximal predominance, nerve conduction study was normal. Biceps brachii muscle biopsy showed muscle fibers with intracytoplasmic vacuoles and intracytoplasmic masses, alterations suggestive of metabolic myopathy. A genetic panel was performed and showed a homozygous likely pathogenic variant in PNPLA2, (c.421-1G>A) consistent with neutral lipid storage disease with myopathy (NLSDM). **Discussion:** Neutral lipid storage disease with myopathy (NLSDM) is a metabolic myopathy, characterized by the presence of triglyceride-containing cytoplasmic droplets in leukocytes and in other tissues, including bone marrow, skin, and muscle. NLSDM is an autosomal recessive muscle disorder caused by mutations in the PNPLA2 gene. Usually presents with juvenile-onset (after the first decade) of slowly progressive proximal muscle weakness with increased serum creatine kinase; distal muscle weakness may also occur. Multisystem involvement with cardiomyopathy, dyslipidemia, hepatomegaly, and/or diabetes is present in 20 percent or more of cases. Our patient had a late onset of weakness and absence of multisystem involvement, with neurological examination and complementary work up compatible with metabolic myopathy due to lipid accumulation. **Final Comments:** Until the present date of submission of this report, no registry was found for the genetic variant c.421-1G>A on PNPLA2, making this a probably novel pathogenic variant. Our case study highlights the importance of considering neutral lipid storage disease for patients with late onset asymmetric myopathies.

Doenças Neuromusculares

TL 1105569

CHARCOT MARIE-TOOTH DISEASE (CMT) SUPERIMPOSED ON CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY (CIPD) IN A PATIENT WITH A HISTORY OF CHILDHOOD POLIOMYELITIS: A CASE PRESENTATION

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Case Presentation: A 68-year-old male patient presented weakness in lower limbs, myalgia, paresthesia in the feet and hands, initiated 9 months ago, with progressive worsening. He denied loss of sphincter control, dyspnea, dysphagia and infections preceding the onset of the condition. History of childhood poliomyelitis with left lower limb atrophy. Neurological examination showed bilateral pes cavus, hammer toes and lower limbs in a characteristic “inverted champagne bottle” appearance. The cranial nerves were all intact. Motor examination showed reduced muscle strength (MRC grades were 4 for lower limbs and 5 for upper limbs) and hyporeflexia in upper and lower limbs. Sensory examination revealed a decreased vibratory sense in the distal lower and in upper limbs. The coordination test assessment showed ataxic gait aggravated by closing eyes and dysmetria on the left side. Cerebrospinal fluid (CSF) revealed albuminocytological dissociation (leukocytes: 1,56 cell/mm³ and proteins: 116,4 mg/dL). Electroneuromyography demonstrated a chronic demyelinating sensory-motor polyradiculoneuritis associated with a bilateral chronic neuropathic process, without current denervative activity, in bilateral myotomes from L2 to S1, consistent with the history of childhood poliomyelitis. The patient has been submitted to monthly pulse therapy with methylprednisolone 1g/day for 5 days, with progressive improvement of the condition.

Discussion: In the first evaluation, the patient has lower limbs deformities characteristic of CMT, but due to the presence of progressive loss of muscle strength, the CIPD hypothesis was raised and corroborated by protein-cytological dissociation. Although the coexistence of Charcot-Marie-Tooth disease with CIPD is already established in the literature, the inflammatory diagnosis superimposed on the hereditary neuromuscular process is not always easy. This case was even more challenging from a clinical and electrophysiological perspective because of the history of childhood poliomyelitis. **Final Comments:** This case presentation highlights the importance of considering clinical exacerbation of CMT due to the possibility of superimposed CIPD. The recognition of superimposed inflammatory neuropathy, although difficult, is of high clinical importance since with specific treatment the patient's clinical condition can be improved.

Doenças Neuromusculares

TL 1105578

DEMYELINATING NEUROPHYSIOLOGICAL FINDINGS IN FAMILY AMYLOID POLYNEUROPATHY

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Case Presentation: A 69-year-old man presented with paraesthesias, significant weight loss, fasciculations and proximal and distal weakness in the four limbs. His clinical picture had begun less than a year ago. Physical examination of the patient showed diffuse areflexia, thermoalgic hypoaesthesia with dependent length pattern and flaccid tetraparesis. There was no cranial nerve dysfunction. Electroneuromyography showed polyradiculoneuropathy with mixed axonal/demyelinating pattern that met criteria for chronic inflammatory demyelinating polyradiculoneuropathy (CIPD). Cerebral spinal fluid did not show significant albuminocytological dissociation. The patient underwent intravenous corticosteroid therapy with methylprednisolone, without satisfactory clinical response. He was then submitted to a genetic panel of neuromuscular diseases, which identified the Ile127Val mutation, known to be pathogenic for transthyretin family amyloid polyneuropathy (TTR-FAP). **Discussion:** Although the typical neurophysiological finding of TTR-FAP is axonal sensory-motor polyneuropathy, it is common to observe heterogeneous patterns of presentation in electroneuromyography. There are reports in the literature of clinical and neurophysiological presentations that mimic CIPD. Demyelination findings are probably secondary to axonal degeneration. **Final Comments:** The variability of TTR-FAP manifestations may delay its diagnosis, which may directly compromise the indication of treatment for these patients. In these cases, knowledge about the neurophysiological findings of FAP can be used as an auxiliary tool in the differential diagnosis.

Doenças Neuromusculares

TL 1105580

REFRACTORY RE-THYMECTOMY AND IMMUNOTHERAPY IN MYASTHENIA GRAVIS: CASE PRESENTATION

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Case Presentation: A 41-year-old patient, female, previously healthy, presented to this service in 2017 with a myasthenic syndrome, referring progressive bulbar musculature weakness associated with bilateral ptosis without fatigability maneuver, being later diagnosed with myasthenia gravis (Osserman Classification IIIa). In clinical-radiological evaluation (CT), a mass with 60 cm³ of volume was identified in the anterior mediastinum, and subsequent histopathological analysis of Thymoma (Masaoka I) was associated with the positivity of Anti-AChR (1,28). She has been refractory to traditional therapy methods, like using glucocorticoids, Intravenous Immunoglobulin (IVIG), and acetylcholinesterase inhibitors in optimized dosage, keeping the progression of symptoms and being at risk of hypercapnic respiratory failure. As a result of recent scientific evidence regarding the surgical benefit of thymectomy in these select patients, she underwent a Video-assisted Thoracoscopic Surgery (VATS) to remove the mass. However, despite IV pulse therapy and the measures previously adopted, the patient continued to experience progressive clinical worsening, requiring a definitive airway proper to hypoxemia and carbon dioxide narcosis. Due to the recurrence of the disease, it required immunotherapy with Rituximab, Cyclophosphamide, and a second surgical re-approach for a new removal of the recurrent mass. Since then, there has been partial improvement in fatigability control through this second investigation of ectopic secretion of anti-AChR. **Discussion:** Myasthenia gravis is a neuromuscular transmission disorder resulting from the binding of autoantibodies to components of the neuromuscular junction (NMJ), most commonly the acetylcholine receptor (AChR). These autoantibodies are directly responsible for the pathological change in the endplate – being dependent on CD4+ T and B cells. It is characterized by fatigable muscle weakness, with a variable course with a tendency to progress to distal muscles. A particular subpopulation is restricted to ocular and facial manifestations. Other autoantibodies involved are anti-MuSK and anti-LRP4, acting on proteins responsible for maintaining the NMJ and on the ability to emit action potentials at the endplate, respectively. **Final Comments:** In this presentation, we describe a patient with refractory Myasthenia gravis, leaving the usual pattern of clinical control and suggesting the importance of the thymus in the pathophysiology.

Doenças Neuromusculares

TL 1105585

RESISTANT MYASTHENIA GRAVIS WITH RITUXIMAB AND RE-THYMECTOMY APPROACH: CASE PRESENTATION

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Case Presentation: A 32-year-old patient, female, previously healthy, presented to this service in 2016, implying bulbar musculature weakness with dysphagia and dysarthria. Due to the fatigability pattern of bulbar musculature, there was a diagnostic suspicion for Myasthenia gravis, which was later confirmed by humoral dosage of anti-AChR (61) with negative anti-MusK. Subsequently, despite optimizing measures adopted, such as Intravenous Methylprednisolone (IVMP) pulse therapy, acetylcholinesterase inhibitors, and Intravenous Immunoglobulin (IVIG), the patient presented progressive clinical worsening with consequent respiratory failure suitably to diaphragmatic loss (Osserman Classification V). Therefore, properly to refractoriness of previous measures, specialists recommended a Video-assisted Thoracoscopic Surgery (VATS) excision and histopathological analysis of the thymus, as it is associated with possible humoral hypersecretion and consequent development of cross-reactions and autoimmune lesions. As a result, the Immunohistochemical Profile (IP) evidenced thymic hyperplasia without atypia. However, after the procedure, the patient remained with muscle fatigability requiring clinical control through Azathioprine and immunotherapy with Rituximab – since there is no availability of the monoclonal antibody Eculizumab at our disposal. Then, we proposed a second approach to investigate ectopic tissue with complete resection of pericardial and mediastinal fat. The analysis showed a small aggregate of lymphoid tissue in re-thymectomy. Since then, the patient has had adequate clinical therapy, maintaining optimized doses according to guidelines for fatigability control. **Discussion:** Myasthenia gravis is a neuromuscular transmission disorder resulting from the binding of autoantibodies to components of the neuromuscular junction (NMJ), most commonly the acetylcholine receptor (AChR). These autoantibodies are directly responsible for the pathological change in the endplate – being dependent on CD4+ T and B cells. Multicentric trials recommend a sternotomy approach with en bloc resection of all mediastinal tissue. This occurs due to microscopic thymic components that are not macroscopically identifiable. A focused and conservative approach remains in question. **Final Comments:** In this presentation, we describe a patient with resistant Myasthenia gravis, leaving the usual pattern of clinical control and suggesting the importance of the thymus in the pathophysiology.

Doenças Neuromusculares

TL 1105586

RITUXIMAB THERAPY IN THE TREATMENT OF STIFF-PERSON SYNDROME: A CASE PRESENTATIONS SERIES

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Case Presentation: Stiff-person syndrome (SPS) is characterized by a huge axial and proximal members stiffness associated with spasms. Treatment is basically symptomatic, based on the use of benzodiazepines and baclofen as the main therapeutic interventions. However, there are few studies that describe the benefit of using immunosuppressants in this condition. **Case 1:** A 30-year-old female patient complained of low back pain radiating to the lower limbs and frequent falling episodes. She presented a claudicating gait, hypertonia of the paravertebral, abdominal and lower limb muscles. After using benzodiazepines, the patient reported a partial improvement of symptoms, but maintained hypertonia and mild pain. Anti-GAD serology showed high titers. With immunoglobulin therapy, ambulation has improved, but falls and painful stiffness persisted. She was then treated with infusions of rituximab every 6 months. After 2 years, she reports improvement of symptoms and life quality, resuming the practices of their activities of daily living. **Case 2:** A 43-year-old female patient complained of pain in the lower limbs, limiting stiffness and recurrent seizures. She presented hypertonia of paravertebral musculature and high titers of anti-GAD. She had already been unsuccessfully treated with immunoglobulin. During 2 years she was treated with benzodiazepines and presented clinical instability with sporadic seizures and no important recovery from the symptoms. After rituximab therapy with infusions every 6 months, there was a significant clinical and life quality improvement. **Case 3:** A 46-year-old male patient complained of lumbar and lower limbs pain, limitation of movement, frequent epileptic seizures. He presented hypertonia of the paravertebral muscles and lumbar lordosis. Anti-GAD titers showed elevated values. After unsuccessful immunoglobulin therapy, he underwent infusions of rituximab interspersed over 6 months. There was a recovery of motricity and stiffness and complete cessation of pain, contributing to functional improvement and resumption in the practice of their work activities. **Discussion:** Most common type of SPS is associated with the presence of anti-GAD impairing the GABA function. Rituximab is a monoclonal antibody against CD20, expressed on the surface of B cells. Its use in SPS is still a recent approach. **Final Comments:** Rituximab may be a benefit drug option in SPS treatment. However, these reports show partial results, requiring long-term follow-up.

Doenças Neuromusculares

TL 1105590

RECIDIVED THYMOLIPOMA APPROACH FOR MYASTHENIA GRAVIS FATIGABILITY CONTROL: CASE PRESENTATION

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Case Presentation: A 26-year-old woman presented to our Neurologic Emergency Department, referring unilateral ptosis and dysphagia associated with fatigability of shoulder girdle muscle compartment as a myasthenic syndrome was hypothesized. Lately, neurophysiologic study with Electromyoneurography (EMNG) and humoral analysis of autoantibodies has been positive (anti-AChR 1,0 nmol/L), suggesting autoimmune Myasthenia gravis diagnosis. Despite using Immunomodulators such as Azathioprine, Glucocorticoid therapy, Therapeutic Plasma Exchange (TPE) and Intravenous Immunoglobulin (IVIG) in addition to pyridostigmine, the patient remained with the breakthrough symptoms (Osserman Classification IV) and specialists proposed a thymectomy while an attempt to control the ectopic secretion of antibodies. The patient underwent the mediastinal mass removal (12 x 10 x 2.5 cm) with posterior histopathological analysis of disclosed thymic gland tissues with an abundant mature adipose tissue, suggesting a rare neoplasm thymolipoma, considered a possible factor with humoral discharge. After surgical excision, there was partial maintenance of symptoms for a certain period, with recurrence of the tumor evidenced by a CT scan approximately 04 years after the procedure, performing a re-thymectomy proposal through thoracoscopy approach for clinical control. Thenceforth, she has moderate fatigability with immunotherapy, avoiding clinical decompensation such as diaphragmatic loss in myasthenic crisis. **Discussion:** Myasthenia gravis (MG) is a neuromuscular transmission disorder resulting from the binding of autoantibodies to components of the neuromuscular junction (NMJ), most commonly the acetylcholine receptor (AChR). These autoantibodies are directly responsible for the pathological change in the endplate – being dependent on CD4+ T and B cells. The incidence of Thymolipoma is around 0.12 out of 100.000 cases per year, including 34 documented cases reporting the association between Myasthenia gravis and this specific tumor. The exact mechanisms remain unclear; however, there is guidance for en bloc resection of all anterior mediastinal tissue in order to symptomatic control. **Final Comments:** In this presentation, we describe a patient with refractory Myasthenia gravis, leaving the usual pattern of clinical control and suggesting the importance of the thymus in the pathophysiology. However, the complete remission rate of symptoms needs further studies for a reliable conclusion.

Doenças Neuromusculares

TL 1105602

VAMP1-RELATED PRESYNAPTIC CONGENITAL MYASTHENIC SYNDROME

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Case Presentation: 21-year-old male, resident in a long-stay institution for patients with cerebral palsy since the age of 5 months. Abandoned by his mother and with no information about his father or family history. There are no reports of problems during pregnancy or childbirth. At 5 months of age, clinical evaluation showed microcephaly (-2.5 SD), global hypotonia, with decreased spontaneous movement of limbs, proximal and distal, with semiflexion of the wrists. CK was normal. The patient needed nocturnal BiPAP and exclusive G-tube feeding. Initial diagnosis was congenital myopathy. Muscle biopsy was performed at 3 years of age and showed increased variation in fiber size, type I and type II fiber atrophy, with rounded fibers and slight increase in endomysial connective tissue. In an evaluation at 11 years of age, hypotonia and global muscle hypotrophy were observed, with proximal and distal muscle weakness (MRC grade 3 and grade 2, respectively). Deep tendon reflexes were absent. There were contractures on the knees, wrists, fourth and fifth fingers, with thoracic kyphosis, everted ankles, and flat feet. The face had a myopathic appearance with a high-arched palate. Ophthalmoparesis was predominant in vertical gaze but became global with the progression of the condition. Dysarthria was severe with barely intelligible speech. Molecular panel for neuromuscular diseases was performed at 19 years of age which demonstrated a homozygous nonsense variant in the VAMP1 gene (c.97C>T; p.Arg33*). Electromyography (EMG) demonstrated increased insertional activity, with fibrillations and positive sharp waves, as well as markedly polyphasic motor unit action potentials, with reduced recruitment and interference pattern. The above findings suggested a severe motor neuropathy, but a muscle ultrasound showed a hyperechogenic pattern, without marked muscle atrophy, suggesting that a severe neuromuscular dysfunction may be responsible for the loss of CMAP amplitude and EMG changes. Pyridostigmine was then prescribed up to a dose of 300 mg/day. Improvement after the drug treatment appeared to be slight, with a mild improvement in the movement of the upper limbs and voice turned out a little louder. VAMP1-associated presynaptic CMS is a rare form of disease from a group that is already uncommon. All cases appear to be consistent with severe congenital muscle weakness and hypotonia, with facial and bulbar involvement, and early feeding difficulties. Some patients responds to pyridostigmine.

Doenças Neuromusculares

TL 1105642

CENTRONUCLEAR MYOPATHY: CASE PRESENTATION

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Case Presentation: Female, 47-year-old, previously healthy, started at age 23 with a condition of distal, progressive and mild weakness in the lower limbs, with stumbling and falls, but without limitations in activities of daily living. After the age of 40, she began to have difficulty climbing stairs and getting up from chairs, in addition to being unable to carry bags or purses anymore. No report of bulbar symptoms. Past history: no comorbidities. Family history: mother and younger brother (40 years old) with similar neurological picture. Neurological examination shows mild eyelid ptosis, especially on the right. facial diparesis. Divergent strabismus evident in the right eye. Muscle strength by the Medical Research Council Scale demonstrating grade 4 in the upper and lower limbs distally, Grade 3 in foot dorsiflexion and grade 4 in flexion. Grade 4 quadriceps and bilateral gluteus medius. Muscle hypotonia. Grade 1 deep reflexes globally except for abolished Achilles reflexes, bilateral Achilles tendon retraction, bilateral cutaneous-plantar flexion reflex. No winged scapula. Absence of myotonic phenomena. Sensitivity preserved. Myopathic gait, not walking on tiptoes and heels. Laboratory tests demonstrate CPK at normal levels. ENMG (2021): signs of primary muscle fiber involvement, associated with signs of active denervation in most motor units examined, predominantly proximal and lower limbs. Genetic Testing: Identified heterozygous pathogenic variant DNM2 c.1393C>T (p. Arg465Trp). **Discussion:** Mutations in the DNM2 gene lead to congenital centronuclear myopathy (CCM) and two forms of Charcot-Marie-Tooth (CMT) in the autosomal dominant form. NCM manifests as a disorder of progressive deterioration and delays in motor milestones, facial and generalized muscle weakness, ptosis, and ophthalmoplegia. Some patients also develop pes cavus before experiencing symptomatic muscle weakness. CNM patients with DNM2 mutations had mild axonal neuropathy, which may or may not have reduced motor conduction velocities. **Final Comments:** This case illustrates a patient with late-onset and slowly progressive Centronuclear Myopathy, associated with pes cavus and global hyporeflexia, with normal levels of CPK, ENMG with myopathic pattern and confirmed by genetic testing.

Doenças Neuromusculares

TL 1105750

PROGRESSIVE ENCEPHALOMYELITIS WITH RIGIDITY AND MYOCLONUS (PERM) MIMICKING SEPSIS IN STIFF-PERSON SYNDROME

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Case Presentation: A 65-year-old female patient, previously diagnosed with anti-GAD positive Stiff Person Syndrome (SPS), on treatment with monthly corticosteroid cycles, was seen due to a complaint of pustular skin lesions after completion of the last methylprednisolone infusion. She was treated with topical corticosteroid and anti-histaminic agents, but with no improvement was observed. About three weeks later, she developed high fever, confusion, prostration, and worsening of generalized muscle stiffness. Physical examination upon admission evinced significant limb stiffness and to diffuse papular lesions in the pelvic region, dorsum, and buttocks, with associated hyperemia. She was admitted with a hypothesis of sepsis of cutaneous focus, and empirical antibiotic therapy was initiated. Laboratory tests showed worsening of the renal function and mild leukocytosis. The patient evolved with generalized tonic-clonic seizures, worsening of hyperthermia, presenting a temperature of 41 Celsius degree, despite the use of high dose antipyretics, in parallel with decreased level of consciousness, requiring orotracheal intubation. Antibiotic therapy was optimized to cover central nervous system infection, but all infectious screening with blood cultures, cerebrospinal fluid analysis, and chest and abdominal imaging tests were negative. White blood cell count and C-reactive protein were within normal limits, although he persisted with hyperthermia maintained between 39 and 40 degrees. Considering infectious causes excluded as the etiology of the decompensation, it was decided to schedule immunotherapy treatment considering the diagnosis of Progressive Encephalomyelitis with Rigidity and Myoclonus (PERM), and the patient underwent plasmapheresis with marked and clinical improvement. **Final Comments:** PERM is a rare SPS spectrum disorder characterized by severe prognosis and high mortality. Clinical manifestation include level of consciousness impairment, dysautonomia and brainstem dysfunction along with typical SPS phenomenon. Diagnosing PERM might be challenging, since some of these manifestations may overlap with other more common diseases, such as infections. Neurologists should be aware of PERM as it is a potentially treatable condition; patients often improve with aggressive immunotherapy.

Doenças Neuromusculares

TL 1105751

CASE PRESENTATION BILATERAL GASTROCNEMIUS MYOSITIS ASSOCIATED WITH INFLUENZA-LIKE SYMPTOMS

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Case Presentation: The patient is a 34 year old male of previous good health. It was his second day of symptoms when he went to the doctor complaining of headache, sore throat, body ache and fever. The oropharynx had a diffuse hyperemia and exudates at the tonsillar crypts. The patient was discharged with Amoxicillin for 7 days, symptomatic medication and diagnosed with acute tonsillitis. Three days later he returned with loss of strength in arms and legs, also feeling better from theodynophagia and body pains. At the neurological examination nothing pathological was found just as the cranial CT. The patient was hospitalized and a lumbar puncture was performed, the liquor analysis had non pathological signs. All the laboratorial routine exams didn't show any abnormalities. On the second day of Hospitalization an increase of the pain was noticed on bilateral calf muscles. Prednisone was initiated, 60mg/day, more exams were requested, and the discrepancy was on CPK levels (1.166,4) and ANF (anti-nuclear factor) titulation 1/160. The day after the patient was dismissed without any neurological deficits and complaints, the final diagnosis was bilateral gastrocnemius myositis. **Discussion:** Our case was probably an IAM (Influenza-Associated Myositis), that disease was defined as follows: Virologically proven influenza or influenza-like illness plus clinical evidence for localized myalgia plus elevation of serum creatine phosphokinase (CPK) or abnormal muscle biopsy. Acute myositis has been reported as an pediatric exclusive disease, normally with symptoms like painful oedema of leg muscles in the days following a viral illness. Calf muscles were the only muscles affected in the cases reported in the past. The incidence is higher in boys, muscle symptoms usually begin 3 days following influenza symptom onset, with pain in one or both calves and difficulty walking being the most common complaints. Among cases of IAM, in ? of cases, the calf muscles were affected alone, and ? of cases occurred in multiple muscular groups in addition to the calf muscles. Treatment is symptomatic, as myositis is self-limited, usually resolving within a mean of 3 days. **Final Comments:** Our case of IAM behaved differently from previous published reports, specially because of the age of the patient reported. So, it is a study that expands the definition for the general population, being not an exclusive pediatric disease and should be on the list of differential diagnosis on muscular disorders.

Doenças Neuromusculares

TL 1105756

PROGRESSIVE MUSCLE WEAKNESS AND DYSPHAGIA IN AN ELDERLY PATIENT: CHLOROQUINE MYOPATHY

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Case Presentation: A 70 years-old woman sought medical care due to unintentional weight loss of 10 to 15kg in 2 months. There was progressive weakness, dysphagia and dysphonia. She had been treated for gastric cancer and arthritis and was under the use of chloroquine, nortriptyline, desvenlafaxine and a combined formula of indomethacin, prednisone and famotidine. Neurological examination disclosed generalized muscle wasting and weakness, with marked involvement of proximal, axial and bulbar muscles. CK was elevated. Investigation for paraneoplastic syndrome was normal. She was admitted to our Hospital to receive clinical and nutritional care and complete the investigation. The EMG showed increased spontaneous activity at rest and findings consistent with a myopathic pattern. The muscle biopsy showed a vacuolar myopathy with autophagic vacuoles. A hypothesis of toxic myopathy due to chronic chloroquine use was made. Medication was discontinued and, although in intensive care, the patient evolved with multiple clinical complications and death. **Discussion:** Chloroquine is a drug used worldwide for treatment of malaria, and as an immunomodulator to treat a variety of inflammatory disorders. It has a high affinity to some cellular compartments, resulting in abnormalities of lysosomal function and autophagy, as chloroquine remains sequestered into those organelles after chemical modifications due to the acid pH. The cumulative effect can be deleterious to skeletal muscle, which can have a spectrum of clinical presentations. There is a predilection for axial, proximal limb, bulbar, respiratory, and cardiac muscles. The morbidity is high, even after drug discontinuation, although some patients may improve. In our case, there was a rapid evolution to multiple organ dysfunction with respiratory failure secondary to her severe clinical condition. **Final Comments:** The skeletal muscle is a highly vascularized tissue and, thus, susceptible to the toxic effects of various substances, including a variety of therapeutic drugs. Although rare, chloroquine myopathy can result in marked morbidity and potential mortality if not accurately and promptly diagnosed. The nonspecific clinical manifestations, specially when occurring in the elderly population, may contribute to a misleading diagnosis. In our patient, the muscle biopsy was a crucial tool to characterize the skeletal muscle involvement and attribute chloroquine toxicity as a preponderant factor in her muscle disease.

Doenças Neuromusculares

TL 1105789

INCLUSION BODY MYOPATHY AND FRONTOTEMPORAL DEMENTIA DUE TO VCP GENE MUTATION: CASE PRESENTATION

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Case Presentation: A 51-years-old female presented with progressive muscular weakness for 10 years. She got worse from weakness in the past year and started complaining of forgetfulness, anomia and periphrases. She developed apraxia for daily activities, frequent falls and joint pain. Her sister was diagnosed with Paget's disease of bone at age 20, and has language and behavioral changes. Her father died at advanced age with proximal weakness and behavioral changes without confirmed diagnosis. On physical examination, the patient was thin, bald and disoriented in time and place. There was wasting of trapezius and deltoid muscles, proximal quadriparesis (MRC IV), normal tonus and reflexes, abolished plantar responses, difficulty in arising from chair, waddling gait and brisk facial reflexes. Coordination and sensibility were normal. MMSE: 10 points (11 years/schooling), poor reading and figure interpretation, hesitant and reduced fluency, anomia, digital agnosia, acalculia, apathy, disinhibition and executive changes. Laboratory data showed normal CPK, aldolase and cardiologic screening. EMG showed proximal myopathic pattern in all limbs; brain MRI demonstrated brain atrophy, mostly in left temporal lobe. In muscle biopsy, there was marked variability in fibers size, increased connective endomysial and perimysial tissue, endomysial inflammatory reaction, necrotic fibers and rimmed vacuoles. Immunohistochemistry was positive for CD4 and CD8. The Next-Generation Sequencing panel showed a pathogenic heterozygous mutation in exon 5 of VCP gene [c.463C>A; p.(Arg155Ser)], also present in her sister. Thus, it was consistent with inclusion body myositis (IBM) with frontotemporal dementia. **Discussion:** The mutation on VCP gene (valosin containing protein) includes a group of hereditary diseases that can cause myopathy, frontotemporal dementia and bone disease, as well as others neurodegenerative diseases. It shares the same pathophysiology of protein aggregates with ubiquitin-proteasome system dysfunction and autophagy. This hereditary type of IBM can be associated with the presence of inflammatory infiltrate in muscle biopsy, being the presence of rimmed vacuoles of great diagnostic contribution. **Final Comments:** The recognition of the particularities of this multisystemic proteinopathy, a group of autosomal dominant diseases with variable penetrance and phenotypes, allow improving the treatment and follow-up of patients and promoting genetic guidance to their relatives.

Doenças Neuromusculares

TL 1105820

PARANEOPLASTIC MYOPATHY WITH PHENOTYPIC MANIFESTATION OF OCULOPHARYNGEAL MUSCULAR DYSTROPHY (OPMD): A CASE PRESENTATION

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Case Presentation: A 53-year-old male, dyslipidemic, former smoker and drinker patient presented a walking difficulty and left ptosis, which progressed bilaterally, dysphonia, dysphagia for liquids and fever. Physical examination revealed paralysis of the lateral rectus and medial rectus muscles; paresis of the oblique muscles; grade 3 strength for cervical flexion and grade 4 for lateralization; dysmetria in the lower limbs; hyporeflexia; absent cutaneous-plantar reflex on the right side and in flexion on the left side; grade 2 gynecomastia. ENMG showed neurophysiological findings of myopathy. Thigh MRI revealed mild liposuction of the quadriceps femoris and gluteus maximus muscles. Skull MRI showed marked atrophy of the masticatory muscles. Lumbar puncture showed a CSF without alteration. The rheumatology team evaluated him and ruled out the hypothesis of inflammatory origin because the clinical (no response to corticosteroids) and imaging exams (no inflammatory signs on thigh MRI) were not compatible. After weeks, the patient started to manifest lymphadenomegaly: bilateral cervical and left supraclavicular. According to the neck CT, the cervical one was suggestive of neoplasia, and according to the pathological examination, the supraclavicular one was a metastasis of an epidermoid carcinoma. Based on the picture evolution, the main diagnostic hypothesis was an occult primary site epidermoid carcinoma and paraneoplastic myopathy syndrome. **Discussion:** Paraneoplastic syndromes are rare disorders triggered by an altered immune system response to a non-metastatic neoplasm and most commonly occur in patients who are not known to have cancer. The symptoms of neuromuscular paraneoplastic syndromes may mimic common neurological conditions, such as Myasthenia gravis syndrome. But, the patient of this case showed an oculopharyngeal muscular dystrophy (OPMD), which is a myopathy characterized by slowly progressive ptosis, dysphagia, and proximal limb weakness with myopathic involvement of other muscles, particularly of the limb girdles, starting in the legs. **Final Comments:** OPMD is a very rare phenotypic manifestation of paraneoplastic syndromes, even in cases of neuromuscular paraneoplastic syndrome with myopathy. Once the patient had a family history of similar condition and lung cancer, he was kept under investigation for some weeks until the diagnosis of neoplasia. After that, treatment with Carboplatin and Paclitaxel was instituted with good response.

Doenças Neuromusculares

TL 1106066

INCLUSION BODY MYOSITIS ASSOCIATED WITH SJOGREN'S SYNDROME: A NEW MYOPATHY DESCRIPTION?

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Case Presentation: A 47-year-old man presented with polyarthralgia associated with knee and ankle joints swelling 4 years prior to evaluation. A year later, he noticed progressive weakness of upper and lower limbs, with difficulty raising the arms and climbing stairs. He also referred trouble in writing, reaching out for things and picking up objects. Neurological examination revealed myopathic gait, atrophy of the deltoids and pectorals, facial weakness, proximal and distal muscle weakness in the upper and lower limbs. Deep tendon reflexes were decreased in the upper limbs and normal in the lower limbs. Cranial nerve, sensitivity, cerebellum and cognition tests were normal. The creatine kinase (CK) was 3,077 U/L and serum aldolase level was 32 (normal, < 7,6 U/L). Erythrocyte sedimentation rate was elevated. Anti-cN1A and Anti-SSA/Ro antibodies were positive. Complement levels and serum levels of thyroid hormones were within normal ranges. Negative autoantibodies included: anti-nuclear antibodies, anti-SSB/La, anti-Sm, anti-DNAh, anti-SRP, anti-RNP, anti-SCL-70, anti-RNA polymerase III. Lower limb MRI showed muscle edema with fatty partial replacement and atrophy. Electromyography demonstrated a pattern of proximal and distal weakness in both upper and down extremities associated with spontaneous activity. Repetitive stimulation test was normal. Muscle biopsy was consistent with Inclusion Body Myositis (IBM). An association between IBM and Sjogren's Syndrome (SS) was considered and the patient was started on oral prednisone (1mg/kg daily). After two weeks with corticosteroid, he showed partial clinical improvement thus we indicated methylprednisolone pulse therapy (1g IV/ daily for 5 days). At 1-month follow-up, the patient showed improvement in muscle strength, specially of proximal upper extremities. **Discussion:** Inclusion Body Myositis and its histopathological features were first described in 1967. An association between IBM and SS was proposed fifteen years later, in 1982. The pathogenesis of this association is unclear. Recent studies suggest for disease concurrence a shared association with T-cell granular lymphocyte leukemia. Even though sporadic IBM is considered treatment-refractory, studies have reported response to immunotherapy when this combination is present, likewise our case. **Final Comments:** Faced with the diagnosis of IBM we suggest that SS should be investigated, as patients with this association may benefit from immunosuppressive therapy.

Doenças Neuromusculares

TL 1106092

MOTOR FUNCTIONAL FOLLOW-UP OF TWO SPINAL MUSCULAR ATROPHY PATIENTS TREATED WITH GENE THERAPY

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Case Presentation: The first case is a male, with SMA type II receiving GT at age of 7 years and 11 months old, and with 8 years and 3 months old at time of last evaluation. The patient had already been using nusinersen for 5 years. Before treatments, the patient was evolving with loss of motor functions such as rolling. At the beginning of nusinersen treatment he scored 14 points on the HFMSE scale, and after 5 years of treatment (before receiving GT) he scored 24 points. The child continued to roll over and became able to move from a sitting to a lying position with the use of nusinersen. After 3 months of GT he increased the score to 26 points and started to assume the crawling posture and became more agile in the motor functions that he had already acquired. He had gains in basic activities of daily living such as dressing. The second case is one of the first Brazilian children, a female with SMA type I, to receive GT at 1 year and 11 months old, and is currently 3 years and 8 months old. With the previous use of nusinersen, she acquired head control, ability to sit without support and to roll over. Regarding motor assessments in scales, the patient scored 08 to 16 points on HFMSE after 3 months of GT, and scored 25 points after 20 months of treatment. Currently, she walks with hip support and stands with support for a few minutes. **Discussion:** Children with 5q-Spinal Muscular Atrophy (SMA) have predominantly axial muscle weakness, which directly impacts the acquisition of motor milestones. To demonstrate motor improvement of two children with SMA, one was among the first patients to receive a single dose of gene therapy (GT) in Brazil and another was one of the oldest in the world to receive the therapy. Assessment of motor functions using the CHOP-INTEND (Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders) and HFMSE (Hammersmith Functional Motor Scale Expanded for SMA) motor scales before GT and after 3 and 20 months of treatment, respectively. **Final Comments:** patients treated with GT showed improvement in motor function even if they were treated previously with nusinersen.

Doenças Neuromusculares

TL 1106093

MOTOR IMPROVEMENTS OF AN ADULT SMA TYPE 3 PATIENT TREATED WITH RISDIPLAM

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Background: Spinal Muscular Atrophy (SMA) type 3 in adulthood is still a challenge in terms of treatment response due to disease duration, previous spine fusion, and some cases may benefit from oral drug treatment. **Objective:** To describe motor improvements of a patient with SMA type 3 treated with Risdiplam for 6 months. **Methods:** Evaluation of motor function using Hammersmith Functional Motor Scale expanded for SMA (HFMSE) before started treatment and after 6 months of drug use. Results and **Discussion:** The patient is a 38 year old female with SMA type 3 (homozygous deletion in SMN1 gene and with 3 SMN2 copies), who lost her gait at 11 years of age and was submitted to surgery for scoliosis when she was 13. Before treatment she was able to sit unsupported, transfer independently, and is able to roll over. She has shortened hip extension, knee extension and dorsal flexion and she never used orthotics. She started using Risdiplam since November 27, 2021 and scored 26 pontis in HFMSE. After 6 months treatment the patient gained 7 points in the same scale. The gains were as follows: item 3, is now able to take both upper limbs above the ear line; items 6, 7, 8 and 9, which assess rolling, and now she is able to roll without the help of the upper limbs; item 11, she is able to remain in the prone position, with support on the forearms and cervical extension; item 12, is now able to, in prone, perform cervical extension with upper limbs in an intermediate position (elbows at shoulder height). After using started treatment, patient also presented an improvement in fatigue, more agility in everyday life, such as for transfers, and is now able to pack her body in the car without help. **Final Comments:** In this specific case, even though the patient was an adult and had a previous spine surgery, treatment with Risdiplam was effective in improving motor function, such as transfers and fatigue.

Doenças Neuromusculares

TL 1106122

LMOD3-RELATED NEMALINE MYOPATHY (NEM10): NEW INSIGHTS BASED IN TWO BRAZILIAN CASES

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Case Presentation: A – Male, 5 months old, polyhydramnios and reduced fetal movement, born with generalized hypotonia, cyanosis, respiratory distress, and irregular breathing. Submitted to orotracheal intubation, tracheostomy and gastrostomy. Inspection: elongated and hypotonic face, bilateral palpebral ptosis, adducted thumb and fifth finger, flaccid and areflex tetraparesis. Received pyridostigmine (20mg/kg), evolving with increased mobility in the shoulder girdle, trunk, and extremities of the upper limbs. No side effects were observed. B – male, 19 years old, son of consanguineous parents, hypotonia since birth, delay in motor milestones, mild dysarthria, elongated and hypotonic face, bilateral palpebral ptosis, generalized amyotrophy and muscle weakness with distal predominance, adducted thumb, and steppage gait. Muscle Biopsy showed nemaline bodies. EMG in both cases revealed a myopathic pattern. Repetitive stimulation identified a decrement of 18.8% in case B. Received pyridostigmine with a subjective improvement. Molecular analysis identified in both cases 2 different variants (homozygosity) in LMOD3 gene. **Discussion:** Nemaline myopathy (NM) is a congenital myopathy defined by the presence of rod-like structures, called nemaline bodies, in myofibers, caused by 13 genes. The LMOD3 is associated with a severe birth-onset form of NM (NEM10). We describe the first two Brazilian cases of NEM10. Out of 34 cases reported in the literature, 5 reached adulthood. Our Case A has a severe form with a non-sense variant (c.154del), previously described in a Portuguese family. Case 2 has a mild form, with a missense variant (c.1194G>C) not previously described. There seems to be a genotype-phenotype correlation in NEM10, based on the type of consequence of the variant and the position of the amino acid in the protein, determining a more severe (case A) or milder (case B) phenotype. The presence of decrement is a classic finding of neuromuscular junction disease (NMJD), however, it has also been described in subtypes of congenital myopathy. So, these patients may have a good response to acetylcholinesterase inhibitors. **Final Comments:** Our cases are the first NEM10 cases in the literature with a partial response to pyridostigmine. The use of pyridostigmine in NM was reported in one case related to KLHL40, with significant improvement. Therefore, we should consider using pyridostigmine in NM as an alternative therapy.

Doenças Neuromusculares

TL 1106172

HEART FAILURE IN LMNA-RELATED EMERY DREIFUSS MUSCULAR DYSTROPHY

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Case Presentation: A 26-year-old female patient was admitted to the Cardiovascular Intensive Care Unit due to acute shortness of breath and hypoxia at rest, reporting exertional dyspnea over the past years. She was accompanied at the Neuromuscular Disorders outpatient clinic due to a past medical history of worsening muscle weakness and contractures beginning at the age of 3. These complaints were associated progressively with inability to walk without support and dysphagia. Her birth and family history, as well as her developmental milestones, were unremarkable. Examination revealed weakness affecting the proximal arms and distal legs with sparing of facial muscles, associated with scoliosis and contractures involving elbow and heel. Cardiorespiratory exam showed bilateral jugular vein distension and an irregular pulse. A thorough diagnostic work-up was conducted and a diagnosis of acute decompensated heart failure (HF) with reduced ejection fraction was made, associated with atrial fibrillation and bilateral pleural effusion. Laboratory assessment disclosed a serum creatine kinase level of 478 IU/L. Previous investigation was remarkable for a muscle biopsy with myopathic features and immunohistochemistry revealing deficiency of every tested protein (dystrophin, sarcoglycan, dysferlin, dystroglycan and emerin). A genetic analysis was performed, disclosing one pathogenic variant in LMNA gene [c.83G>A (p.Arg28Gln)] shortly after the patient's death due to refractory respiratory insufficiency, confirming a diagnosis of autosomal dominant Emery-Dreifuss muscular dystrophy (EDMD). **Discussion:** EDMD commonly results from a defect of one or more proteins comprising the cell nuclear envelope. The classic clinical triad consists of early contractures, progressive muscle weakness and cardiac abnormalities. The two most common associated genes are EMD and LMNA. Mutations in LMNA most typically display an autosomal dominant inheritance pattern and de novo mutations are common. Mostly, these patients have a more severe disease course, and the incidence of cardiac involvement increases in an age-dependent manner. LMNA-related cardiomyopathy also carries a substantial risk of end-stage HF development, as seen in our patient. **Final Comments:** Despite its rareness, early diagnosis of LMNA-related muscular dystrophy is important for the detection of potentially fatal cardiac abnormalities. Thus, routine cardiac surveillance may prevent mortality in these young patients.

Doenças Neuromusculares

TL 1106198

**IMMUNE-MEDIATED ANTI-SRP
NECROTIZING MYOPATHY – A CASE
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Case Presentation: A 51 years old female started 6 months ago with muscle weakness in the upper and lower limbs and difficulty to raise the arms. She had dysphagia, alopecia, paresthesias in hands, and lost weight in the last 3 months. Physical examination showed proximal muscle strength grade of 3 in upper and lower limbs. The laboratory tests revealed abnormal high levels of CK (4.309), LDH (948) and aldolase (13). ANA test, anti-JO1, anti-SSA, anti-SSB, anti-SM and anti-RNP were negative. The autoantibody recognizing the SRP was positive. The electromyography showed myopathic pattern and the MRI of the thighs presented signs of myopathy with mild edema of the semitendinosus, hamstring, biceps femoris, tensor muscles of the fascia lata, diffuse muscle hypotrophy. Mild peripheral uptake by contrast medium and mild edema. The muscle biopsy revealed necrotic fibers, sometimes surrounded by myophagocytes. A significant variation in the size of muscle fibers was also seen, with several regenerated fibers and capillaries with mural thickening. Necrotic fibers and a few lymphocytes dispersed throughout the endomysium, in addition to the scarce sarcolemmal expression of MHC-I. She was treated initially with prednisone 60mg/day, but after a bad response to the glucocorticoid she received immunoglobulin human (IGIV) monthly during one year and rituximab each 6 months. **Discussion:** This report aims to expose a rare case of inflammatory myopathy, characterized by typical laboratory and clinical features including rapidly progressive muscle weakness, elevated muscle enzymes and characteristic histopathology of muscle biopsies. The immune-mediated necrotizing myopathy (IMNM) represents 20 to 30% of cases of inflammatory myopathy and is distinguished from other subtypes by the existence of necrosis of muscle fibers and absence of inflammatory infiltrate. **Final Comments:** IMNM, a type of autoimmune myopathy that has three different serologically subtypes: Anti-SRP, anti-HMGCR and antibody-negative IMNM myopathy. In histology these diseases have similar characteristics: myofiber necrosis in muscle biopsy, but the different subtypes have different risk factors, cancer risks, extramuscular manifestations and prognosis. These differences indicate that those are three different diseases, such as distinct pathological mechanisms and may respond to a variety of therapeutic modalities. More data on disease pathogenesis and optimal treatment of each subtype of IMNM are needed.

Doenças Neuromusculares

TL 1106206

**STIFF PERSON SYNDROME, PARTIAL
VARIANT, IN A 12-YEAR-OLD PATIENT –A
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Case Presentation: A.S.S, 12 years old healthy male, reports burning back pain and lack of control of the lower limbs, unable to move them. Evolved with sustained muscle contraction of the left hemiabdomen and left lower limb. He denies urinary or gastrointestinal alterations or concomitant and previous infectious symptoms. He also denies recent vaccinations. On physical examination, spastic paresis of the left abdomen and left lower limb, pronator deviation of the left foot, and a left hemiparetic spastic gait. Magnetic resonance imaging of all central nervous system was not remarkable except for the dubious enhancement of some lumbar roots inside the dural sac, which may be related to radiculitis. Non-reactive antiGAD, non-reactive serology, unavailable anti-MOG and aldolase 13.3. Electroneuromyography (ENMG) showed persistent and continuous muscle action potentials in the rectus abdominis muscles, and the muscles of the left lower limb without relaxation. The patient was treated with muscle relaxants, benzodiazepines, and physical therapy. **Discussion:** Stiff person syndrome (SPS) is a rare entity characterized by painful muscle stiffness associated with muscle spasms¹. It is a rare neuromuscular disorder that affects about one to two cases per million, primarily affecting adults aged 20 to 50 years, with women being affected about two to three times more than men. It is infrequent to occur during childhood^{1,2}. The association of autoantibodies against glutamic acid decarboxylase (anti-GAD) is found in about 60 to 80% of cases. It can be subdivided into three clinical forms: Classic, Partial and Paraneoplastic Variant. **Comments:** The exposed case shows us a rare syndrome in a patient with atypical epidemiology and an even more unusual presentation. Furthermore, the lack of association with autoantibodies makes this case even more unusual, making the diagnosis possible only with ENMG.

Doenças Neuromusculares

TL 1106218

**GUILLAIN-BARRÉ SYNDROME FOLLOWING
ACUTE DENGUE INFECTION: A CASE
PRESENTATION**Sarah Diógenes Alencar, Sara Arcanjo Bringel,
Ana Sílvia Sobreira Lima Verde, Alessandra Braga
Cruz Guedes de Moraes, Matheus Costa Bessa,
Karla Rafaela Silva Vasconcelos, João Gabriel
Dias Brasiliense Frota, Karoline Ferreira Mororó
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Case Presentation: In February 2022, a 20 year old woman was admitted with a 19-day history of ascending tetraparesis and paresthesia. 30 days after admission, she presented with cough, nasal congestion, odinophagia, and tested negative for Covid-19. Upon Hospital admission, the patient was unable to walk without help. She was conscious, well oriented and had normal vital signs. Neurological examination revealed global hypotonia, areflexia and severe reduction of strength in the bilateral lower limbs. In upper limbs, she had hyporeflexia, moderate lower limb weakness bilaterally, and global involvement of deep sensitivity, her neck muscle strength was reduced and she had right peripheral facial palsy. Initial laboratory exams were unremarkable. Cerebrospinal fluid study (CSF) showed protein-cytological dissociation and Electroneuromyography (ENMG) revealed a demyelinating polyradiculoneuropathy pattern. The patient was treated with 5 cycles of plasmapheresis. By the end of the cycles she was able to walk without help. This patient was diagnosed with Guillain Barre Syndrome (GBS) in its acute inflammatory demyelinating polyneuropathy form (AIDP). Additionally, she was tested for dengue-specific IgM in the CSF, providing the association of GBS AIDP with dengue fever (DF). Symptoms of GBS may begin a few weeks after a febrile infection. Though there are well established preceding infective agents, arbovirus as aetiological factors for GBS are uncommon. Studies show that the annual incidence of GBS after arbovirus infection is 0.24 cases per 100 thousand inhabitants. Arbovirus infections are frequent, but they rarely involve the nervous system. In this patient case, the presence of a positive dengue-specific IgM in CSF, following neurological manifestations, patterns of ENMG and the typical CSF findings, supported the association of GBS and DF. Although GBS associated with DF is a rare condition, it's a serious neurological complication that remains underdiagnosed. Strategies for notification and investigation should be implemented. Our findings highlight the importance of further studies to better understand this condition and raise awareness amongst clinicians.

Doenças Neuromusculares

TL 1106227

CERVICAL SPONDYLOTIC AMYOTROPHY: A RARE CAUSE OF FOCAL PERIPHERAL NERVE HYPEREXCITABILITY SYNDROME

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Case Presentation: A 47 year-old man presented to the emergency department reporting involuntary movements and weakness in the right upper limb in the previous 28 days. These symptoms were associated with paresthesia and neuropathic pain. Neurological examination showed myokymia, fasciculation and distal weakness in the right upper limb, a claw hand, atrophy of the hypothenar eminence, and asymmetrical response of the right finger flexion reflex. A magnetic resonance imaging of the cervical spine showed a C5-C6 spondylotic compression of the spinal cord with a "snake eye sign". Electroneuromyography showed an axonal compromise of the preganglionic portion of the C7-C8 myotomes. Myokymia, fasciculation and pain had a significant improvement with carbamazepine. **Discussion:** Cervical spondylotic amyotrophy is characterized by predominantly motor symptoms related to compression of the ventral neural root or injury to the anterior horn due to cervical spondylosis. The intrinsic injury to the anterior horn can be related to insufficient blood supply, which was demonstrated by high intensity signal in the spinal cord, the so-called snake-eye sign. Peripheral nerve hyperexcitability syndrome is characterized by cramps, fasciculations, neuromyotonia and myokymia. PNHS is usually associated with autoimmune disorders such as in the presence of anti-CASPR2 autoantibodies, but can also occur in channelopathies, amyotrophic lateral sclerosis, and, rarely, cervical spondylotic amyotrophy. **Final Comments:** Focal PNHS should include cervical spondylotic amyotrophy in its differential diagnosis.

Doenças Neuromusculares

TL 1106257

ADENOSINE DEAMINASE 2 DEFICIENCY SYNDROME (DADA2): A CASE PRESENTATION OF A RARE AUTOINFLAMMATORY NEUROLOGICAL DISEASE

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Case Presentation: 35-year-old woman, born in southern Brazil, presented with livedo reticularis, reactive C protein (RCP) constantly elevated (30-50mg/dL), and mononeuritis multiplex. In addition, she had recurrent unexplained fever, and anemia since childhood. At 22 years old, she was Hospital admitted due to a third cranial nerve impairment, probably caused by an ischemic event. She received anticoagulants for a couple of months, which were discontinued by her neurologist due to a normal cranial MRI. In her family history, she had a 5-year-old sister, who also had livedo reticularis, and died due to cerebral aneurysms, and alveolar hemorrhage. The complementary investigation showed low levels of IgM, and negative tests for ANA, rheumatoid factor, ANCA, cryoglobulin, and antiphospholipid antibody. The serological tests for infectious diseases were also negative (VDRL, HIV, hepatitis B, and C testing). The thoracic and abdominal magnetic resonance angiography discarded vessel stenosis or aneurysms. Clinical and complementary exam data were compatible with medium vessel vasculitis, suggesting a polyarteritis nodosa phenotype. She was treated with corticosteroids, methotrexate, and tocilizumab. Even under treatment, she presented with an acute ischemic stroke located in the mesencephalon. Her clinical syndrome, associated with a positive familial history, makes the diagnosis of DADA2 plausible, and the next step would be the molecular test. Considering this hypothesis, anti-TNF drug adalimumab was prescribed, and oral anticoagulant warfarin was immediately stopped. **Discussion:** Adenosine deaminase-2 deficiency (DADA-2) is described as an autoimmune disease caused by mutations in the CECR1 (ADA-2) gene. Vasculopathy involving small and medium-sized vessels is one of the main clinical manifestations of DADA-2. It is one of the rare causes of stroke in young people, associated with varied systemic manifestations, such as polyarteritis nodosa (PAN), livedo reticularis, ulcerations of the extremities, arthritis, recurrent infections, and peripheral neuropathy. Immunosuppressive drugs can prevent further attacks and morbidity; on the other hand, anticoagulants and antiplatelets are not recommended. **Final Comments:** The reported case highlights the importance of considering DADA-2 as a differential diagnosis in patients with recurrent neurological deficits at a young age, associated with systemic manifestations, especially regarding prompt treatment.

Doenças Neuromusculares

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Doenças Neuromusculares

TL 1106274

LAMINOPATHY PRESENTING AUTOPHAGIC VACUOLAR HISTOPATHOLOGY

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Case Presentation: A 17-year-old patient, with no consanguinity and normal motor development in childhood presented with difficulty in walking at the age 10, with gait on tiptoe. On examination, he had distal weakness MRC grade 3 for anterior tibialis and finger extensors and MRC 4 for gastrocnemius. Deep tendon reflexes were hypoactive and Achilles abolished. Contractures were observed in cervical, elbows and Achilles tendons, characterizing Emery-Dreifuss phenotype. On investigation, we found elevated CK levels (1901 U/L). EMG showed myopathic changes on distal muscles. Muscle biopsy revealed dystrophic myopathic histological pattern and abundant rimmed vacuoles with autophagic characteristics. Additional evaluation revealed no cardiomyopathy nor respiratory damage. The whole-exome sequencing showed a likely pathogenic variant in LMNA (c.946A>C;p.Lys316Gln). **Discussion:** Variants in heterozygosity in the LMNA can cause a wide spectrum of clinical manifestations known as laminopathy. There may be distal muscle weakness, with phenotype similar to Emery-Dreifuss muscular dystrophy, or due to peripheral nerve involvement, resembling Charcot-Marie-Tooth phenotype. Other patients present a congenital muscular dystrophy with severe axial involvement or, even, isolated cardiomyopathy. Described myopathological findings in skeletal muscle laminopathies typically manifest as nonspecific myopathic changes or dystrophic features. Altered autophagy accompanied by rimmed vacuoles is a common denominator of many muscle diseases. When it is combined with protein aggregates the definition is of myofibrillar myopathy (MFM). Classical MFM pathology in skeletal muscle laminopathies has been reported in a few patients, however isolated autophagic vacuolar myopathy has not been described associated to LMNA. It was speculated that the mutated lamin A/C may disrupt nuclear lamina integrity, unanchor desmin from the nuclear envelope and cause disorganization of myofibrils linked to desmin. This process could lead to MFM pathology. **Final Comments:** Vacuolar myopathies remain diagnostically challenging. Our case highlights the occurrence of this pathologic finding in patients with LMNA-related myopathy. We suggest including pathogenic variants in LMNA as a causative gene in the groups of vacuolar and myofibrillar myopathies. Identification of this condition means a high risk of ventricular arrhythmias in these patients and its importance is to prevent lethal cardiac events.

Doenças Neuromusculares

TL 1106302

CONGENITAL FIBER TYPE DISPROPORTION DUE TO HOMOZYGOUS HACD1 MUTATION: A CASE PRESENTATION

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Case Presentation: Congenital myopathies are a broad group of pathologies whose symptoms include hypotonia, proximal weakness, distinct phenotypic features, and wide variation in severity. Its histological findings are important and helped in its classification. Currently, however, with the advent and easier access to genetic testing by next generation sequencing (NGS), knowledge about the pathophysiological bases of these diseases has expanded greatly. We report the case of a 66-year-old male patient whose symptoms started at birth with hypotonia and, subsequently, gait delay (walked independently at 7 years of age). After childhood there was stability of symptoms, with slight worsening after the fifth decade, when we started to follow him up. In the initial assessment proximal weakness, mainly in the lower limbs, winged scapula and myopathic gait were noticed. There were frequent falls, and difficulty climbing stairs. There were no bulbar symptoms, sensibility or coordination impairment, myotonic phenomenon, joint hypermobility, cicatrization impairment or skin lesions. The patient's parents were consanguineous (first degree cousins), and there was a history of premature death of a sister from an uninvestigated neuromuscular disease. Aldolase and creatine phosphokinase levels were normal. Further investigation with nerve conduction study and electromyography showed a proximal myopathic pattern and muscle biopsy sampling the left biceps brachii showed congenital fiber type disproportion. No necrosis, interstitial fibrosis or inflammatory infiltrate were found. Echocardiogram and electrocardiogram showed no abnormalities. The patient was subsequently tested with a broad genetic NGS panel for neuromuscular diseases, and a homozygous mutation in the HACD1 gene (c.373_375+2del) was found (classified as probably pathogenic according to ACMG criteria). Additionally, a heterozygous pathogenic mutation in the COL6A2 gene was identified. As mentioned above, however, the patient did not have phenotypic characteristics compatible with mutations in collagen 6 gene. To date, few cases of myopathy caused by HACD1 mutations have been described worldwide and, as far as we are aware, this is the first case of HACD1 mutation related myopathy reported in Brazil. By reporting this case, we expect to contribute to expanding the knowledge of this congenital myopathy and bring it to attention as a possible cause of congenital myopathy in our country.

Doenças Neuromusculares

TL 1106347

A RARE CASE OF EARLY-ONSET INCLUSION BODY MYOSITIS (IBM): A CASE PRESENTATION

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Case Presentation: Female, 37 years old, from Salvador/Bahia, with crural paraparesis for 10 years. She perceived difficulty in climbing stairs and today she walks with bilateral support. In the last two years, she also presented upper limbs weakness, distal, more evident when trying to open pots. Hypotrophy of the quadriceps and on the ventral surface of the forearms was identified, as well as reduced muscle strength in leg extension and foot dorsiflexion and also weakness in the fingers. Myopathic gait pattern. A CPK (Creatine Phosphokinase) of 164 U/L (normal < 145) was measured. Electroneuromyography of the lower limbs with myopathic pattern. Muscle biopsy revealed rimmed vacuoles, expression of the MHC-I complex in sarcoplasmic membranes, immunophenotypic staining of membrane attack complex in sarcoplasmic membranes and capillaries, presence of CD8+ lymphocytes and presence of P62 aggregates in focal areas of the endomysium and sarcoplasm. The clinical suspicion was of Inclusion Body Myositis. The treatment started with prednisone 1mg/kg/day for 90 days, with no clinical response. The choice of not introducing any other immunosuppressants was made. **Discussion:** Inclusion Body Myositis (IBM), an inflammatory myopathy, is a rare, slowly evolving disease common in males over 50 years of age 1,2,3. Clinical manifestations include weakness in the quadriceps femoris muscle and ankle dorsiflexors, resulting in difficulty in walking and climbing stairs, as well as in the wrist and finger flexors 3,4,7,8,9. CPK (Creatine Phosphokinase) can be normal or increased. Anti-cN1A antibody dosage provides good specificity. On electroneuromyography, it is characteristic the presence of fibrillation and positive acute waves and increased polyphasic motor unit potentials 6,7,8,9. On muscle biopsy, the gold standard exam, it was verified the presence of endomysial infiltrate of mononuclear cells: CD8+ T lymphocytes and macrophages. A small number of myofibrils presented margined vacuoles, a very characteristic finding in IBM 3,6,8,9. Evidence indicates that patients with IBM respond poorly to corticosteroid therapy. Other immunosuppressants can be tried in those patients who respond to corticosteroids 10,11,12,13,14. **Final Comments:** IBM remains a rare and challenging entity. This is the case presentation of a patient who, despite being an unusual age group, had clinical and pathological findings suggestive of IBM and unresponsiveness to corticosteroid therapy.

Doenças Neuromusculares

TL 1106362

BRUNS-GARLAND SYNDROME: CASE PRESENTATION

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Case Presentation: Female patient, 72 years old, hypertensive, dyslipidemic, obese, diabetic, seeks care for pain that started in the left ankle, 20 days of evolution, with burning, shock and paresthesia, with ascending progression and that reached the lumbar spine on the fifth day of evolution. She developed paresis in the entire left lower limb, initially with leg extension, but culminating in an inability to flex, abduct, and adduct the thigh, grade 2 leg strength, and grade 1 thigh strength. Bilateral hyporeflexia. Initial suspicion of spondylosis neuropathy neurosurgery. Lumbosacral spine CT showing circumferential disc bulges between L3-L4 and L4-L5, with epidural fat effacement and determining impressions on the ventral surface of the thecal sac, with narrowing of the conjugation foramina in L4-L5 and possible involvement of the L4 roots. Cervicothoracic myelotomography without obstruction to contrast passage. Absence of spinal cord or radicular conflicts on MRI of the lumbar and thoracic spine. She evolved to partial improvement and, on the seventh day of Hospitalization, she had grade 3 thigh strength and neuropathic pain relieved with gabapentin and nortriptyline. The neurology team was called, which found recent involuntary weight loss. Report of improved median mononeuropathy. Head CT without changes. Lumbar puncture with 113 proteins, 100 nucleated cells (100% lymphocytes), normal glucose, no pathogens. Electroneuromyography: left lumbar radiculoplexoneuropathy, marked recent axonal sensory-motor involvement and active signs of denervation. Glycated hemoglobin 8.5% (poor glycemic control). Effective insulin therapy was introduced. Discharged after 21 days, complete resolution of pain, walking with support. **Discussion:** Diabetic amyotrophy has an annual incidence of 4.2/100,000 and affects 1% of diabetics. Called Bruns-Garland Syndrome or proximal diabetic neuropathy, it is installed, probably, due to an ischemic lesion caused by microvasculitis. It appears as acute pain, proximal or distal, asymmetric and focal, followed by weakness in the proximal lower limb, autonomic dysfunction and weight loss. **Final Comments:** No treatment is proven to be effective. Data on immunosuppressive therapies are limited and controversial.

Doenças Neuromusculares

TL 1106363

PREVIOUSLY UNPUBLISHED HOMOZYGOUS CHRNA1 C.997C>T VARIANT ASSOCIATED WITH RELATIVELY MILD CONGENITAL MYASTHENIC SYNDROME (CMS): A CASE-REPORT

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Case Presentation: We present a 51-year-old male patient, referred to the neuromuscular unit with history of mild transient neonatal bulbar weakness, presenting with worsening limb and bulbar weakness after 30 years of age. Family and past medical history were unremarkable except for consanguineous parents. Previous NCS and needle EMG revealed some evidence of post-synaptic neuromuscular junction dysfunction with diffuse myopathic motor unit potentials. Anti-AChR and anti-MuSK results were negative. The patient was initially diagnosed with seronegative auto-immune myasthenia gravis and received various immunosuppressant therapies and pyridostigmine with partial response. Nevertheless, from 30-50 years of age, the patient underwent multiple Hospitalizations due to respiratory infections, with successful recovery. Genetic testing revealed homozygous c.997C>T variant resulting in a change p.Arg333Trp. In silico prediction tools showed potentially deleterious value, and was reported as probably pathogenic based on previous fast-channel CMS description associated with compound heterozygosity for that variant. After CMS diagnosis, the patient was kept in rehabilitation and waned immunotherapy. By the time of this report, the patient partially recovered motor functions, walks unaided and sustains improvement with pyridostigmine and ephedrine. **Discussion:** CMS englobes a variety of genetically inherited neuromuscular junction disorders. The most common form of CMS is post-synaptic and can be primarily due to decrease in its subunit's expression or by altering the channel's kinetic. CHRNA1 mutations causing CMS are rare, and generally severe at early onset. The present report describes a patient with relatively mild clinical features harboring a c.997C>T homozygous variant in CHRNA1 which was only previously reported in a patient with compound heterozygosity. This variant prevalence is approximately 5 in 105 and is probably associated with autosomal recessive CMS. In summary, this case expands our knowledge of the complex genotype-phenotype correlation of CMS with demonstration of a homozygous mutation expected to cause a very severe phenotype but instead originating a mild adult-onset phenotype. **Final Comments:** Since the only previous report of the same variant was compound heterozygous, and definition of kinetic disturbances and receptor deficiency requires functional proof, we acknowledge the benefit of further histological and advanced neurophysiological testing.

Doenças Neuromusculares

TL 1106377

NEW TREATMENTS, COMBINATION THERAPY AND VENTILATORY CHANGES: REPORT OF TWO CASES OF SPINAL MUSCULAR ATROPHY UNDER TREATMENT

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Background: Spinal muscular atrophy (SMA) is characterized by progressive muscle weakness and respiratory involvement. Recently, disease-modifying drugs in association with multidisciplinary management have increased survival. A small group of patients have received therapies in combination, such as the use of gene therapy (GT) followed by nusinersen. However, the effects of this approach in terms of efficacy and safety are not well established. **Objective:** The aim of this study is to present the ventilatory changes in two patients with SMA type 1. The respiratory functional measures collected were forced vital capacity (FVC) and thoracic development was measured by chest circumference (CC). Case 1 - Male patient, SMA type 1, who started nusinersen at 5 months and received GT at 1 year and 7 months old. He continued using oligonucleotide antisense after GT. The patient underwent 18 assessments at 30-day intervals. In the first evaluation the child was 1 year and 7 months old, he presented FVC of 300 ml and CC of 48 cm of axillary line and 50 cm of xiphoid process. After 6 months, FVC was 240 ml and CC was 49cm of axillary line and 50cm of xiphoid process. After 12 months, FVC was 250 ml and CC was 51 cm of axillary line and 52 cm of xiphoid process, and after 18 months, FVC was 290ml and CC was 55 cm of axillary line and 56 cm of xiphoid process. Case 2 - Male patient, SMA type 1 who started nusinersen at 4 months and received GT at 1 year 4 months, then nusinersen was stopped. The patient underwent 10 assessments at 30-day intervals. In the first evaluation the child was 1 year and 7 months old, he presented FVC of 150ml and CC of 44cm of axillary line and 48 cm of xiphoid process. After 3 months, FVC was 200 ml and CC was 48cm of axillary line and 50 cm of xiphoid process. After 06 months, FVC was 220ml and CC was 49 cm of axillary line and 51 cm of xiphoid process, and after 09 months, FVC was 220 ml and CC was 46 cm of axillary line and 50 cm of xiphoid process. **Final Comments:** CC increased throughout the evaluations, demonstrating satisfactory growth of the thoracic cage. During the evaluation period, there were no serious respiratory complications. Ventilatory support has been maintained only at night. The two cases maintained ventilatory management through air stacking techniques, and mechanical assistance for cough. Thus, specific therapies for SMA, either alone or in combination, have been shown to be effective in improving respiratory function in SMA.

Doenças Neuromusculares

TL 1106388

MOLECULAR CHARACTERIZATION OF CONGENITAL MYASTHENIC SYNDROMES IN REFERENCE OUTPATIENT CLINIC, IN SALVADOR, BAHIA

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Case Presentation: JGL, fem., 6y, frequent falls since 2y, evolved with difficulty to walk and to stand up. MS 4/5 distal and 3/5 proximal, axial hypotonia and areflexia. Gait with adducted feet and tendency to fall. Genetic testing (GT): DOK7 gene mutation. Improvement with Albuterol. RSS, fem., 47y, ptosis, diplopia, fluctuating weakness of distal and cervical predominance. EMG: mild neuromuscular junction (NMJ) disorder. GT: mutation in the CHRND, DOK7 and COL1 genes. Improvement with Albuterol and Fluoxetine. AJS, fem., 36y, weakness in both LL with frequent falls since birth. Between 11-27y used an unremembered medication with improvement. At 27y, evolved with fluctuating weakness in both UL, cramps and fasciculations after cesarean section. MS 2/5 proximal and 4/5 distal. EMG: decremental pattern and postsynaptic impairment of NMJ. Reduced vital capacity. GT: mutation in the DOK7 gene. Improvement with Albuterol and Fluoxetine. JMS, fem., 34y, fatiguing global weakness, bilateral ptosis and NPMD delay since birth. Consanguineous parents and three family generations with intellectual deficit and epilepsy. Presents facial dysmorphism, bilateral semiptosis, paresis of VI CN, MS 3/5 proximal and 4/5 distal. EMG: postsynaptic involvement of JNM. Suspected of congenital myasthenia (CM), with improvement with Albuterol but not Mestinom. GT: mutation in the CHRNE gene. **Discussion:** CM is a rare and difficult to diagnose entity usually seen in childhood, but may be insidious/present late. Common symptoms include fluctuating muscle weakness, bulbar symptoms, ophthalmoparesis and ptosis. The most common associated gene mutations are: CHRN, DOK7, RAPSN. CHRN encodes the subunits of the ACh receptor and the mutation deforms it, being able to cause two separate syndromes depending on the affected subunit, as seen in RSS's and JMS's cases. DOK7 is an adaptive protein bound to MuSK. Its mutation can affect breathing since birth and cause weakness and ptosis throughout life. There is marked improvement with Albuterol or Ephedrine, as seen in the cases. **Final Comments:** Despite rare, CM should be suspected when the clinical history and therapeutic response isn't similar to the usual response for Myasthenia Gravis. Genetic testing helps to guide the treatment and dictate the prognosis, modifying the quality and lifestyle of the patient and their relatives. It also helps to identify the most common mutated genes and new mutations that can contribute to future diagnoses.

Doenças Neuromusculares

TL 1106404

POLYRADICULONEURITIS BY VARICELLA ZOSTER VIRUS (VZV) WITHOUT CUTANEOUS INVOLVEMENT IN A IMMUNOCOMPETENT MAN: A CASE-REPORT

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Case Presentation: A 62-year-old healthy man presented with a 3-week history of progressive severe low back pain and weakness. One week after pain onset, he noted progressive proximal weakness in lower limbs and left facial palsy. He had no fever, weight loss, choking or recent travel history. He was evaluated twenty days after the symptoms onset and his neurologic examination showed an MRC grading system (right/left) of: hip flexion 2/2, hip extension 2/3, knee flexion 4-/4, knee extension 4/4, ankle dorsiflexion 5/5 and plantar flexion 5/5, along with mild weakness in the distal upper limbs (UL). Deep tendon reflexes were absent in the LL and normal in the UL. Sensory examination revealed abnormal response to pain and temperature sensations in the medial aspects of both legs. Vibration was absent on the ankles. There was a left-sided facial palsy House-Brackman (HB) grade III. No skin lesions were seen. Sensory and motor nerve conduction studies were normal in both UL and LL. Needle examination showed diffuse acute denervation affecting the lumbosacral segment bilaterally. Brain MRI showed bilateral facial neuritis, worse on the left. CSF revealed lymphocytic pleocytosis, elevated protein and a CSF/blood glucose ratio of 53%. Spinal MRI had diffuse leptomeningeal thickening and enhancement of thoracic and lumbar nerve roots. He was treated with high doses of oral steroids, with improvement of weakness and pain in the lower limbs. Laboratory results evidenced elevated levels of IgG for Varicella Zoster Virus (VZV) in blood and a 1:8 titer for IgG VZV in CSF. **Discussion:** We present a rare manifestation of VZV infection with no skin lesions in an immunocompetent patient. VZV remains latent in the sensory ganglia of the peripheral nervous system after a primary infection and can reactivate. Usually, during reactivation, there are papulovesicular skin lesions and neuropathic pain. Some patients show signs of segmental radiculopathy, but polyradiculitis as this patient presented is a very rare manifestation. VZV is also a relevant cause of cranial neuropathy, and the trigeminal and facial nerves are the most frequently affected. Patients often recover from motor deficits. **Final Comments:** The presence of polyradiculoneuritis in immunocompromised patients should include VZV infection as a differential diagnosis. In the absence of immunosuppression and a characteristic skin lesion, diagnosis is challenging and laboratory tests are essential.

Doenças Neuromusculares

TL 1106412

SPINAL MUSCULAR ATROPHY WITH PREDOMINANT LOWER EXTREMITY TYPE I (SMALED1) WITH NO SIGNS OTHER THAN PURE MOTOR SYMPTOMS

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Case Presentation: 5-year-old boy, with a history of delayed motor development. It evolved with proximal weakness in the lower limbs (Gowers signal positive), hyperreflexia, atrophy in the legs and lordosis. Electroneuromyography showed chronic signs of denervation with normal sensory conduction. The father, not genetically tested, has atrophy in his legs. A large DNA panel of neuromuscular diseases (NGS) detected the heterozygous pathogenic variant, chr14: 101.986.017 C>T (or alternatively c.1792C>T – ENST00000360184) in the DYNC1H1 gene. Considering the clinical and electrophysiological data and the pathogenic variant, this is a case of SMALED1 due to a pathogenic variant of the DYNC1H1 gene. **Discussion:** Spinal Muscular Atrophies (SMA) correspond to a group of hereditary diseases caused by loss or abnormal development of lower motor neurons. The major cause is SMA associated with homozygous deletion of exons 7 and 8 of the SMN1 gene on chromosome 5q. Other causes, called non-5q SMA, are less frequent and poorly recognized. One of these is SMALED1 (Spinal muscular atrophy lower extremity predominant; OMIM #158600), of autosomal dominant inheritance, associated with the DYNC1H1 gene (OMIM #600112), with a less aggressive evolution than 5q SMA. This gene lead to two human motor neuropathies, SMALED and axonal Charcot-Marie-Tooth (CMT) disease. **Final Comments:** The phenotype SMALED1 show childhood onset, lower limb weakness, associated with arthrogryposis and cognitive impairment. There is great clinical variability, with cases of adult onset (after 18 years) and purely motor and proximal cases, simulating a myopathy, as in the case described.

Doenças Neuromusculares

TL 1106419

CENTRONUCLEAR MYOPATHY: A CASE PRESENTATION

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Case Presentation: M. S, 48 years old, female, living in João Pessoa, admitted with a complaint of muscle weakness with progressive worsening since the age of 20. She reported difficulty walking, running and climbing stairs associated with falls, fatigue, significant dysphagia for solids and difficulty holding objects, with progression of symptoms over the years. The patient reports that limb weakness intensified during and after pregnancy at age 32. She is apathetic, and reports nocturnal awakenings and frequent choking. On physical examination, she has proximal tetraparesis, grade 3 muscle strength in iliopsoas, deltoids and biceps, and grade 4 in the other muscle groups; global areflexia; positive Gowers sign, myopathic lifting and falling, limiting limb movement. In October 2020, she underwent an echocardiogram, which showed mild tricuspid valve regurgitation. In addition, she has a family history of muscle weakness, a great-grandmother and two sisters, and the biopsy of a sister is compatible with an autosomal dominant centronuclear myopathy. She denies consanguinity between her parents. Polysomnography, electroneuromyography (ENMG) and muscle biopsy were requested. ENMG showed a predominantly proximal myopathic pattern. The histological result was compatible with autosomal dominant centronuclear myopathy. Polysomnography showed 1 episode of desaturation. The patient was referred for neurorhabilitation in order to receive multidisciplinary care, in addition to receiving genetic counseling. **Discussion:** Centronuclear myopathy is a congenital muscle disorder with a heterogeneous clinical presentation, which usually involves muscle fatigue, muscle atrophy and varied cardiorespiratory involvement, characterized histologically by the prominence of muscle fibers with centralized nuclei. The patient in the present case presentation a progressive worsening of symptoms, emphasizing a decline in strength after pregnancy. Furthermore, the presence of dysphagia, with episodes of postprandial choking, was also reported. Even with this situation, she is completely independent for daily activities, with maintenance of quality of life, with multidisciplinary follow-up. **Final Comments:** Finally, the present case has its relevance in reporting a case of centronuclear myopathy, a rare disease with an incidence of 1: 100,000 live births, with the presence of atypical findings, as well as the association of the patient with worsening of symptoms after pregnancy.

Doenças Neuromusculares

TL 1106460

USE OF INSPIRATORY MUSCLE TRAINING IN A PATIENT WITH AMYOTROPHIC LATERAL SCLEROSIS: A CASE PRESENTATION

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Case Presentation: Amyotrophic lateral sclerosis (ALS) is a disease of the central nervous system characterized by a progressive degeneration of upper and lower motor neurons. Respiratory muscle weakness is commonly observed in these patients, as well as a progressive reduction in lung function, leading to complications such as alveolar hypoventilation and impairment of the cough reflex. Despite the known effects of inspiratory muscle training (IMT), there are few studies on the benefits of this therapy in neuromuscular diseases. Therefore, this work aims to demonstrate the results obtained through a pulmonary rehabilitation program with IMT protocol, associated with a multidisciplinary approach in patients with ALS. **Methods:** 38 years old patient, with a clinical diagnosis of ALS for 2 years, followed in the same period in a pulmonary rehabilitation program twice a week, period: 30 to 40 min, protocol: IMT with Powerbreathe® Plus Medic in interval exercises, with load 30% of MIP; Expiratory exercises with EMST 150° load of 30cmH₂O; Pulmonary hyperinflation exercises with ambu®. Non-invasive ventilation (NIV) was not used during pulmonary rehabilitation, only used once a day and during the night. **Results:** For a year, the patient increased inspiratory strength by 30 cmH₂O, reaching MIP -140cmH₂O in this period and maintained the expiratory strength without losses or gains at +110cmH₂O. Regarding pulmonary function during this period, the patient showed a slight decrease in FEV₁ (99 - 93%) and FVC (94 - 86%) with no pattern of pulmonary restriction or obstruction. There was an improvement in the subjective sensation of exertion, where the resting BORG reduced from 3 to 0. It is possible to conclude that IMT can be a feasible therapy and bring benefits to the patient with neuromuscular disease, however, more studies in the area are necessary.

Doenças Neuromusculares

TL 1106554

CONGENITAL MYASTHENIC SYNDROME WITH DISTAL MYOPATHIC PHENOTYPE

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Case Presentation: A 50-year-old male with bipolar disorder was evaluated due to a progressively-worsening weakness that began 34 years ago. Weakness was predominantly distal and asymmetrical (right > left), involving his upper-limbs and causing fine motor activities impairment. He evolved with neck extensor weakness and dysphagia. Physical examination revealed predominantly distal and right-sided weakness in his upper-limbs with bilateral atrophy of the thenar, hypothenar and interosseous muscles. EMG showed proximal and distal post-synaptic neuromuscular junction disease associated with distal upper-limb myopathy. Family history revealed a similar condition in his paternal grandmother, father and two daughters. Genetic test yielded Congenital Myasthenic Syndrome (CMS) related to a mutation in the CHRNA1 gene. **Discussion:** CMSs include a group of neuromuscular diseases of genetic origin with variable genotype and phenotype. Clinically, patients present fatigability, transient or permanent weakness of the extrinsic ocular, facial, bulbar, trunk, respiratory or limb muscles. Symptoms usually start in childhood and may be present from birth or more rarely in adolescence. The CMS 1A is related to a mutation in the CHRNA1 gene and has an autosomal dominant character, but with variable penetrance and expression, explaining the different phenotypes. Mutations occur in the acetylcholine receptor (AChR) subunits causing a delay of the opening time of the acetylcholine ion channel that are located at the neuromuscular junction. CMS 1A should be suspected in patients of any age who present weakness and fatigability of the scapular, cervical and dorsal forearm (wrist and finger extensors) muscle groups in which there is no response to the use of cholinesterase inhibitors or the worsening of symptoms. The diagnosis is based on the symptomatology and on the electroneuromyography findings that demonstrate a decrease in the repetitive stimulus; the dosage of anti-AChR antibody can also be performed. **Final Comments:** CMS with myopathic phenotype is a rare presentation, with only a few cases described in the literature with different phenotypes in the same family. Our patient had the distal myopathic phenotype while his two daughters had CMS of the slow channel type with mutation in the same gene.

Doenças Neuromusculares

TL 1106561

GUILLÁIN BARRÉ SYNDROME: AN ATYPICAL PRESENTATION

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Case Presentation: A 50-year-old man, hypertensive and pre-diabetic, reported polyarthralgia, low-grade fever, adynamia and prostration with gradual worsening for 9 days. On the fifth day of evolution, he presented voiding effort and burning pain on the extremities, followed by clear urinary retention, requiring relief probing, intestinal constipation, in addition to progressive ascending and symmetrical paraparesis. The neurological examination showed mild facial diparesis and dysphonia, grade 4 paraparesis and hyporeflexia in the lower limbs, without sensory level. 24 hours later, he already had grade 2 paraparesis. CSF with no cells and 76 mg/dL of protein. Arboviruses serologies were non-reactive, as was PCR COVID. Performed treatment with immunoglobulin 400 mg/kg/day for 5 days (starting on the 11th day) in an intensive care unit, without the need for mechanical ventilation, where he developed non-dialytic acute kidney injury, but completed the proposed regimen. Electroneuromyography revealed axonal active sensorimotor polyradiculoneuritis (AMSAN), predominantly in the lower limbs. For differential diagnosis with myelodysplasia, spinal MRIs with contrast were requested, which showed no relevant changes. **Discussion:** GBS is an acute autoimmune polyradiculoneuritis that primarily affects the myelin of the proximal portion of peripheral nerves in an acute/subacute manner. Its most common etiology is post-infectious, but less important precipitating factors are surgery, immunization and pregnancy. The diagnosis is clinical, complemented by laboratory and electrophysiological tests, and two criteria are essential, such as progressive weakness of more than one limb or cranial muscles of varying degrees, and hyporeflexia and distal areflexia with varying degrees of hyporeflexia proximal. In this case, the patient demonstrated atypical manifestations of GBS, with urinary retention/constipation and fever at the beginning of the condition, leading to the imperative exclusion of other differential diagnoses, which did not delay the start of treatment. Given the rapid evolution, and late search for specialized medical care, the prognosis was unfavorable. **Final Comments:** GBS is a disease whose incidence has increased dramatically in the COVID era, and its complications can lead to high degrees of disability and, in some cases, death. Identification of GBS and early initiation of specific treatment are essential to reduce recovery time and minimize sequelae.

Doenças Neuromusculares

TL 1106572

THE IMPORTANCE OF DIAGNOSING POMPE DISEASE AMONG NEUROMUSCULAR DISEASES: A CASE PRESENTATION

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Case Presentation: TRA, male, 3 years and 7 months old. At 2, he started having abdominal pain and alteration in the transaminases. After an investigation with a gastro-pediatrician, without a clear diagnosis, a neuromuscular condition was suspected, and the patient was referred to a neuropediatrician. The mother reported that, from 1 year and 6 months old, the child began to show difficulty to climb stairs, symptoms that were not valued. At 2 and 3 months, he began to have difficulty walking, dysphagia, fatigue on moderate and big efforts and nocturnal hypoventilation. The CPK level was 2175, and molecular tests for spinal muscular atrophy, Steinert's myotonic dystrophy and facioscapulohumeral muscular dystrophy were negative. Electroneuromyography revealed altered motor response. Neuromuscular Disease Panel was performed: 2 acid alpha-glucosidase (GAA) variants in heterozygous pathogenic were found, which confirms the diagnosis of Pompe disease, with an alpha-glucosidase dosage of 0.26. In addition, the patient has Autistic Spectrum Disorder. **Discussion:** Pompe disease has an incidence of 1: 40,000 live births, being a lysosomal storage disease caused by a mutation in the gene encoding GAA, leading to intracellular glycogen accumulation. The disease is characterized by muscle weakness, hypotonia, dysphagia, and respiratory failure, cardiomyopathy, among other nonspecific symptoms. It can be classified in Infantile Onset Pompe Disease (IOPD) and Late Onset Pompe Disease (LOPD). IOPD symptoms appear up to 12 months of age, with associated heart disease. In the late form, symptoms appear after 12 months, but are less severe. The diagnosis is made by the GAA enzyme dosage, followed by a molecular test. The treatment is carried out by the enzyme replacement. The introduction of enzyme replacement in patients diagnosed with Pompe, in 2006, changed the prognosis of the disease, as it stabilized the damage caused by the intracellular accumulation of glycogen. Therefore, with early diagnosis, it is possible to quickly institute the appropriate therapeutic measures, ensuring a better quality of life and preventing irreversible lesions. **Final Comments:** Pompe disease, although rare, should be considered as a differential diagnosis in pediatric patients with neuromuscular symptoms. Since it has a disease-modifying therapy, an early diagnosis can significantly change the patient's prognosis. There is no evidence of a relationship between Pompe disease and Autism.

Doenças Neuromusculares

TL 1106585

CONGENITAL MYOPATHY WITH MYBPC1 GENE MUTATION ASSOCIATED WITH MUSCULAR ACTIVITY TREMOR-LIKE

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Case Presentation: A 10-year-old boy, born with cleft palate that was corrected at age 1.5, with a history of delayed language development, presented for assessment of tremor. Relatives inform the presence of postural and action tremor since birth. Patient complained of tremor interfering with his daily living activities associated with myalgia affecting his lower-limbs, particularly when running. On physical examination, it was noted proximally-predominant tetraparesis and postural and action tremor. EMG showed notably proximal myopathy without complex repetitive or myotonic discharges. Surface electromyographic study of tremor, with a channel recording the flexor and another one recording the extensor muscles of forearms, showed continuous muscular activity in the extensor muscles during wrist extension, with no bursts (typical sign of tremor). Brain MRI was normal. There was no family history of neurological diseases. Given the clinical picture suggestive of myopathy, a neuromuscular panel was ordered and the genetic test confirmed MYBPC1 gene mutation, thus, a congenital myopathy. **Discussion:** Congenital myopathies are a group of inherited muscle disorders clinically characterized by hypotonia and weakness. It usually begins at birth and has a static or slowly progressive clinical course. Diagnosis and classification of congenital myopathies has historically been based on clinical features and histopathology. Nevertheless, recent advances in genetics have changed diagnostic practice. The gene MYBPC1 encodes a member of the myosin-binding protein C Family (sMyBP-C), a modular sarcomeric protein that exerts structural and regulatory roles through dynamic interactions with actin and myosin filaments of the muscle. This protein has three isoforms: cardiac, fast skeletal and slow skeletal. The main roles attributed to the MyBP-C family are the stabilization of thick filaments and the regulation of the cross-bridge cycle. Recent studies demonstrate that the genesis of tremor may be associated with mutation in some gene positions; this hypothesis is supported by the formation of cross-bridges by the sarcomeric protein. **Final Comments:** Constant advances in the field of genetics have led to a significant increase in knowledge of the broad phenotypic spectrum, including overlapping phenotypes, of congenital myopathies.

Doenças Neuromusculares

TL 1106596

PURELY BULBAR MYASTHENIA GRAVIS: A CASE PRESENTATION

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Case Presentation: A 69-year-old female, previously hypertensive, was evaluated for sudden dysphagia with mild dysarthria and dysphonia. Neurological examination also showed hypoesthesia of the right hemiface. She spontaneously recovered 24 hours before admission to Hospital. There was no alcohol or drug use history. Laboratory tests and cranial CT were normal, but electrocardiogram showed an atrial fibrillation rhythm. Within 48 hours of admission, the symptoms returned, with sudden severe dysphagia, which worsened throughout the day, requiring the use of a nasogastric tube. She was urgently submitted to a new cranial CT with intracranial and cervical CT angiography, but both were normal. Brain MRI was then performed, but was also normal. We ended up ruling out a vascular cause and pursued a therapeutic test with pyridostigmine 60 mg and oral corticosteroid. A positive response was rapidly seen during the first hours after administration, evolving with progressive dysphagia improvement. She tested negative for muscle-specific thyrokinase and acetylcholine receptor binding antibodies. Electroneuromyography with repetitive stimulation and chest tomography came out normal. **Discussion:** Myasthenia gravis is an autoimmune disease of the post-synaptic neuromuscular junction characterized by fluctuating weakness involving ocular, bulbar, limb and/or respiratory muscles. The disease occurs at any age, with an early peak in the 2nd and 3rd decades and a late peak between the 6th and 8th decades. Diagnosis is based on investigation of the clinical history and typical examination findings. More than 50% of patients have isolated ocular symptoms of ptosis and/or diplopia and only about 15% of patients have bulbar symptoms. **Final Comments:** This is a case presentation of a 69-year-old hypertensive woman who suddenly started the condition with isolated bulbar symptoms, without involvement of other muscle groups, mimicking a posterior circulation stroke/transient ischemic attack. Although the initial hypothesis was a vascular condition, the fatigability presented led to the suspicion of myasthenia. The case highlights the importance of differential diagnosis of sudden neurological events, particularly involving bulbar symptoms, which should raise suspicion for myasthenia gravis.

Doenças Neuromusculares

TL 1106597

RECURRENCE OF BREAST NEOPLASM IN MULTIPLE SCLEROSIS: CASE PRESENTATION

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Case Presentation: Female, 54 years old, with Relapsing-Remitting Multiple Sclerosis, diagnosed in February 2013, she used Betainferon 1a and is currently using Glatiramer Acetate. In 2017, she presented a breast nodule in the left breast, identified as Invasive Ductal Carcinoma, undergoing neoadjuvant treatment with chemotherapy and subsequent excision, which occurred without intercurrents or complications. Later, in 2022, she presented a nodule in the right breast, and a biopsy was performed and identified as Invasive Ductal Carcinoma, with estrogen and progesterone receptor positive and HER2 negative, being in clinical follow-up and therapeutic management with hormone therapy. **Discussion:** Multiple Sclerosis (MS) is an autoimmune disease of a degenerative nature, which leads to demyelination of the Central Nervous System. Regarding the link with neoplasms, in a cohort study, it was found that the presence of MS is associated with an increase in all-cause mortality, but it was not associated with changes in neoplasm specific survival. In patients with MS, longer diagnostic intervals may occur if the symptoms of neoplasms were wrongly attributed to the underlying disease or if the cancer symptoms were less relevant than other health problems. In addition, MS related disability, which increases with age, as well as the occurrence of neoplasms, can make access to diagnostic tests, such as mammography, difficult. Thus, the lower rate of cancer detection through screening in these patients compromises disease control, negatively affecting survival. **Final Comments:** Studies show that patients with MS do not appear to have an increased risk of developing neoplasms. However, some have suggested an increased risk not only of breast cancer, but also of the CNS, urinary tract and nasopharynx. Commonly, these observations are based on a small number of cases, which makes these estimates inaccurate. Thus, understanding the pathways to early detection of breast cancer is critical to developing and planning interventions to improve outcomes for people with MS and cancer.

Doenças Neuromusculares

TL 1106610

CASE PRESENTATION ATIPICAL LATE ONSET POMPE DISEASE IN A 36 YEAR PATIENT WITH SPASTIC PARAPARESIS

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Case Presentation: Male, 36 years old, smoker 1 pack of cigarette per day for 20 years, without previous comorbidities, with consanguineous parents (cousins). Patient refers lower limb paresis, initially on the right distal, with progressive worsening and spasticity, started three years ago. Refers 8 kg loss of weight in the period. He denies alteration of upper limbs. Report six months ago, beginning of dysphagia for liquids and dysphonia. Neurological physical examination shows dysphonic speech, muscle strength grade IV- in lower limbs with spastic rigidity, exalted reflex in lower limbs without signs of pyramidal release and a spastic gait. Serological tests for HIV and syphilis are negative. Brain Magnetic resonance imaging (MRI) without alterations and cervical, thoracic and lumbar spine MRI with normal spinal Cord sign. Electromyoneurography (ENMG) with Carpal tunnel syndrome. Positive genetic test for Pompe disease. **Discussion:** Pompe disease (PD), or glycogen storage disease type II, is a rare inherited autosomal recessive disease caused by a deficiency in acid alpha-glucosidase (GAA) enzyme. The late-onset phenotype results from an incomplete GAA deficiency, which appears later in childhood, in adolescence or in adulthood. It is characterized by a slowly progressive myopathy. The diagnosis of late-onset Pompe disease (LOPD) is still challenging and often quite delayed. It is not unusual for patients to go undiagnosed for many years as screening for PD is often overlooked or delayed in adults. LOPD it is usually milder than the infantile-onset forms of this disorder and is less likely to involve the heart. Most individuals experience progressive muscle weakness, especially in the legs and the trunk, including the muscles that control breathing. The treatment includes enzyme replacement therapy. A drug called alglucosidase alfa is given intravenously. It may help reduce the buildup of glycogen inside cells, and slow the progression of the disease. **Final Comments:** The present case of a patient with adult-onset progressive spastic paraparesis with a positive genetic test for pompe's disease. A clinical manifestation not commonly seen in late-onset Pompe disease.

Doenças Neuromusculares

TL 1106615

FAMILY AMYLOIDOTIC POLYNEUROPATHY: A RARE DISEASE INCREASINGLY COMMON

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Case Presentation: A 76-year-old man, from Guanambi-BA, born in Cansanção-BA, with kidney disease and heart disease, with an implantable cardioverter-defibrillator, with symmetrical paresthesia and dysesthesia in gloves and boots, in addition to dysautonomia with episodes of syncope, which had been worsening for about 1 year. On neurologic examination, moderate symmetrical distal hypopalesthesia and gait of small steps with wide base without support. Four-limb ENMG (26/04/2022) showed chronic polyneuropathy with axonal predominance in the lower limbs, distal and proximal involvement, symmetrical, moderate to severe. The transthoracic echocardiogram showed significant biatrial enlargement, absence of intracavitary thrombi, diastolic dysfunction, significant left ventricular hypertrophy, minimal pericardial effusion and mild to moderate mitral regurgitation. Myocardial pyrophosphate scintigraphy (26/04/22): strongly suggestive of amyloidosis. The panel for transthyretin-linked hereditary amyloidosis revealed susceptibility to amyloidosis, variant val142Ile. The Fabry test was negative. Tafamidis meglumine 80 mg/day was requested for his treatment. The investigation of first-degree relatives (ongoing) has so far revealed two asymptomatic susceptible cases. **Discussion:** Familial Amyloid Polyneuropathy (FAP) is a rare multisystem disease characterized by an autosomal dominant transmission, in which the mutant form of transthyretin (TTR) is synthesized in the liver. More commonly, FAP presents in clinical conditions of varying forms and intensities that mix familial amyloidotic polyneuropathy and familial amyloidotic cardiomyopathy. The diagnosis is clinical and based on the presence of the genetic mutation in TTR. In the case in question, the changes were compatible with FAP, whose variant val142Ile is predominantly associated with cardiomyopathy, renal dysfunction and polyneuropathy. **Final Comments:** The diagnosis of FAP at a pre-clinical stage has an important epidemiological value, aiming at mapping the mutation in the community. In addition, the early initiation of treatment is related to the symptomatic improvement of the patient, minimizing the evolution of the disease. The dissemination of information and accessibility to genetic tests has led to an increase in the incidence of FAP, which becomes a possible etiology in the face of polyneuropathies without a specific cause.

Doenças Neuromusculares

TL 1104889

OPHTHALMIC HERPES-ZOSTER AFTER VACCINATION AGAINST COVID-19: CASE PRESENTATION

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Case Presentation: The patient: Female, 59 years old. No noteworthy medical history, including denying Varicella infection during childhood. On 04/13/2021, she received the first dose of the vaccine against COVID-19 (Coronavac) without post-vaccination reactions. On 05/07/2021, she received the second dose of the Coronavac vaccine, with no adverse reactions until 05/14/2021, when she noticed the appearance of vesicobullous lesions with an erythematous base associated with ocular edema, electric shock-like pain and local hyperesthesia, evidenced in the course of the V1 branch of the trigeminal nerve, being clinically diagnosed as ophthalmic herpes-zoster. Treatment with Acyclovir was started, but hyperesthesia persisted. **Discussion:** Trigeminal neuralgia is characterized by paroxysms of excruciating pain in the V nerve. Among the infectious causes, herpes zoster (HZ) should be considered. After primary infection with Varicella, the virus becomes quiescent in cranial sensory nerve ganglia and spinal dorsal. Regarding vaccination against COVID-19 until the present writing of this article, case presentation were observed in Europe. No case presentation were found on the South American continent. A systematic review entitled "Can SARS-CoV-2 vaccine increase the risk of reactivation of Varicella zoster?" showed a relationship between HZ development and mRNA vaccination against COVID-19. A total of 54 cases were analyzed. The median period between the development of HZ and vaccination against COVID-19 was 7 days. 68.27% of HZ cases were reported by mRNA vaccine. Thirty-six patients 36/45 developed HZ after the initial dose of COVID-19 vaccine among those who received the mRNA vaccine. This led to the hypothesis that the disease, with physical stress, could trigger HZ. Functional impairment of T lymphocytes could reactivate varicella zoster virus. Another study titled "Phase I/II study of COVID-19 RNA vaccine BNT162b1 in adults" on Nature, showed a transient decrease in lymphocyte counts observed within a few days after vaccination and return to baseline within 6-8 days on all participants. Thus, associating this post-vaccination lymphocytopenia and the Varicella virus reactivation, precisely when there is a drop in CD4+ and CD8+ levels to be a plausible justification. **Final Comments:** The report a case in which HZ 7 developed days after vaccination against COVID-19 in a patient without comorbidities. Consider the report of importance to the scientific community.

Dor

TL 1105393

USE OF SPINAL ELECTRODE IN A PATIENT WITH VASCULITIC NEUROPATHY

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Case Presentation: Female patient, 23-year-old, reports sudden onset of petechiae on the lower limbs, from the knee to the foot, which bilateral and apparently symmetrical character. The lesions disappeared after a month, when pain appeared in the same location. After consultation with specialists, the patient underwent electroneuromyography, which revealed a distal symmetrical pattern and received a multidisciplinary diagnosis of vasculitic neuropathy. The rheumatological treatment partially improved the condition, but the intensity and frequency of pain increased after a while. An evaluation with a doctor, pain specialist, was requested to perform a spinal electrode test in the thoracolumbar region. There was a good response with the test, decreasing more than 50% on the pain scale, without intolerable side effects, and so it was decided to implant the electrode and the generator permanently. The patient is in the seventh month of follow-up, with pain intensity around 3, on the Visual Analogue Scale (VAS), with improved quality of life. **Discussion:** Vasculitic neuropathy is characterized by the submission of a vessel by an infiltrate of cells due to inflammation, generating ischemia and injury to peripheral nerves. The diagnosis is made by biopsy of the suspected nerve and its treatment is done with immunosuppressants. However, this condition is still poorly documented, since only 31 results were found in the search for MeSH Terms (vasculitic) AND (neuropathy) in the Pubmed database, making this issue a real challenge. The importance of spinal electrical stimulation in the treatment of neuropathic pain refractory to pharmacotherapy has become clear. Its mechanism of action works according to the pain gate theory, in which the posterior horn of the spinal cord controls the passage of stimuli by activating larger-diameter fibers and inhibiting the passage of stimuli from smaller-diameter ones. **Final Comments:** Therefore, although vasculitic neuropathy is not among the most evident pains, this work addresses the issue between it and the treatment of chronic pain through the use of spinal electrode, having in this report a good response to treatment. Considering that the electroneuromyography of the disease reveals axonal loss, It is possible that just the rheumatologic treatment, alone, is not sufficient for sensory recovery. Therefore, it is necessary to resort to complementary therapies to alleviate the patient's suffering and generate greater functionality.

Dor

TL 1105783

SUCCESSFUL TREATMENT WITH SPINAL CORD STIMULATION IN A PATIENT WITH TESTICULAR TORSION: CASE PRESENTATION

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Case Presentation: Chronic testicular pain is defined as constant or intermittent pain in the testicle lasting three months or more, which generates impairment in patients' daily activities. For those who do not respond to conservative treatment and nerve blocks in the spinal, a good option is to use electrode implants. We can state that peripheral neuropathic pain is a type of chronic pain that occurs when there is injury or damage to the somato-sensory system, which persists for longer than 3 months or remains for more than 1 month after the resolution of an acute tissue injury. In this case presentation, we will describe a young patient who presented testicular torsion and evolved with peripheral neuropathic pain, undergoing treatment with implantation of a spinal cord stimulation electrode. With this report, we want to describe the qualitative outcome of the patient with chronic testicular pain treated with a spinal electrode.

Dor

TL 1105869

ACUTE PAINFUL DIABETIC NEUROPATHY: PRESENTATION OF A TREATMENT-INDUCED CASE

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Case Presentation: Female, 19 years old, presented in October 2021 to the neurology service with a picture of burning pain in the lower limbs. History of DM1, diagnosed when he developed diabetic ketoacidosis in 2018; Treated with insulin and oriented on the management of the disease. Follow-up was irregular in the first two years, with no attendance at follow-up appointments. Readmitted to the endocrinology department in March 2021; She weighed 70kg and had an HbA1C index of 15.6%. She was oriented and showed no changes in the level of consciousness. Returns in September 2021, with 55kg, after strict metabolic control. HbA1C of 6.9%, with intense pain, characterized by numbness and a burning sensation involving bilateral feet. Her thyroid profile was within normal limits, as was vitamin B12. She was medicated with 60mg/day of morphine and referred to the neurology department for investigation. A nerve conduction study suggested bilateral axonal predominant sensory polyneuropathy of the lower limbs. Upper limb nerve conduction studies were normal. The cause of the neuropathy was diagnosed as treatment-induced neuropathy (TIN). Treatment was started with opioid dose reduction and gradual introduction of amitriptyline and gabapentin. In the control consultations, he showed good evolution, despite gradually lower doses of medication. **Discussion:** Acute painful diabetic neuropathy (APDN) is rare and seen in both type 1 and type 2. The incidence of APDN is not known and is probably less than 1 in 1,000 diabetic patients. APDN can manifest as treatment-induced neuropathy (TIN) and diabetic neuropathic cachexia (DNC). Studies have shown that it can occur after rapid glycemic control with insulin, as in the case presented here, and in patients using oral antidiabetic drugs. A set of diagnostic criteria for TIN is proposed, including: (1) acute onset of neuropathic pain or autonomic symptoms, (2) a decrease in HbA1c level of more than 2% at 3 months, and (3) onset of neuropathic pain and /or autonomic symptoms within 8 weeks after the decrease in HbA1c level. Pharmacological treatment should be performed in conjunction with the holistic management of the patient. **Final Comments:** Physicians' awareness of this rare and completely reversible condition is necessary to ensure proper treatment and adherence to glycemic control.

Dor

TL 1105895

PULSED RADIOFREQUENCY FOR POST-COVID OCCIPITAL NEURALGIA: A CASE PRESENTATION

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Case Presentation: Patient, 45 years old, male, history of infection by Covid-19, was attended at an outpatient neurosurgery service complaining of pain in the occipital and cervical region, of strong intensity, paroxysmal, burning type. On physical examination, he had more marked paresthesia on the right and a positive Tinel's sign on the right. Test blockade of the greater and lesser occipital nerves with anesthetic solution showed improvement in the condition for 4 days. From the diagnosis of post-Covid-19 Occipital Neuralgia, he underwent neuromodulation therapy with pulsed radiofrequency in the C2 dorsal root ganglion, at a temperature of 42 °C, for 3 minutes, with repetition after an interval of 3 minutes. The procedure was uneventful, with complete resolution of pain and Hospital discharge on the same day. **Discussion:** In regard to chronic neuropathic pain, there are reports of it being triggered by psychological stressors, viral infection or as a result of Hospitalization in the intensive care unit. There are post-viral immunological reactions capable of promoting an environment conducive to the establishment of neurological complications. Neural virulence would likely be linked to violations of the blood-brain barrier, either by endothelial invasion through the binding of the virus's Spike protein to the angiotensin-2-converting enzyme, or trans-synaptically through the invasion of the olfactory nerves, or even an increase in the permeability of the barrier caused by the release of cytokines resulting from the systemic inflammatory process as a reactional response. Pulsed radiofrequency consists of radiofrequency emission by a catheter through an electrode, positioned close to a nociceptive pathway, in short pulses, leaving intervals between emissions in order to allow the affected tissue to cool. The area affected can be well defined, being restricted to the nerves responsible for transmitting and modulating painful sensations. In the case of occipital neuralgia, the electrode implant is placed in the C2 ganglion, where greater and lesser occipital nerves emerge, thus allowing pulsed and non-continuous waves to induce a low-intensity electric field around these nerves, aiming to modulate and regulate the activity of A-delta fibers and C fibers. **Final Comments:** Occipital neuralgia implies a careful investigation and carries a vast possibility of treatments, among them, pulsed radiofrequency neuromodulation acquires notoriety along with its promising results.

Dor

TL 1105913

NASOPHARYNGEAL POLYP: AN UNUSUAL PRESENTATION

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Case Presentation: J.I.R.B., a 17-year-old male student with a history of pharyngeal pain, nocturnal nasal obstruction for 2 months, associated with a complaint of throat swelling for 1 month. He denied dysphagia, rhinorrhea, headache and allergic symptoms. He was not asthmatic. Oroscore revealed a large polypoid lesion protruding from the oropharynx. On dynamic inspection, the soft, but not friable, lesion invaded the oral cavity, occupying almost the entire oropharynx. Magnetic resonance imaging revealed a bulky lesion with cystic components, hyperintense on T1-weighted and hypointense on T2-weighted images, of approximately 8.0 cm in its largest diameter on the sagittal section, extending from the nasopharynx to the oropharynx, reaching the region of the epiglottis. Endonasal and oral endoscopic resection was indicated, and the polyp was dissected and removed from the nasopharynx, which, because of its size, was removed through the oral cavity. The procedure was uneventful during or after surgery, and he was discharged after 48 hours. The anatomopathological study revealed an inflammatory polyp, with intense interstitial edema and an inflammatory infiltrate rich in eosinophils, with no signs of malignancy. The patient has been followed up for ten months with no recurrence and no pharyngeal, nasal or oral complaints.

Discussion: Primary non-neoplastic inflammatory polyps of the nasopharynx are rare, representing 4 to 6% of nasal polyps. There is still no evidence about its pathogenesis, but it is believed that both the adaptive and innate immune response could be involved. The present case is of great importance because of its rarity, the site of involvement of the benign inflammatory polyp, as well as the size of the lesion. This rare pathology should be added to the list of differential diagnoses of nasopharyngeal mass lesions between nasoangiofibroma and nasopharyngeal carcinoma. Clinically, there is a benign course of this lesion despite the extensive volume. Therefore, regarding neurological manifestations, literature confirms that acute sinus inflammatory damages can possibly result in nerve hypersensitivity. **Final Comments:** In the approach of nasopharyngeal and oropharyngeal masses in young and adult patients, primary inflammatory polyps are among the diagnoses to be considered. Despite the small number of cases, the radiological and pathological features of this lesion should be remembered, minimizing the risk of being misdiagnosed.

Dor

TL 1106479

CANNABIDIOL: NEW PERSPECTIVES FOR TREATMENT OF POST-MASTECTOMY PAIN SYNDROME, A CASE-REPORT

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Case Presentation: J.C.A female, 46 years old, presents to the neurology outpatient clinic with a complaint of disabling pain in the hemithorax and right upper limbs after mastectomy and worsening with subsequent ipsilateral radiotherapy in 2018. The patient reports that the pain interferes with the quality of her sleep and limits her daily activities. It is characterized as arrowheads, without irradiation, constant, but with frequent exacerbations, 10 out of 10 on the pain scale, accompanied by paresthesia and loss of strength in the affected limb. A directed physical examination was not performed because the patient was intolerant to touch and movement. She takes several medications, including vitamin B and D, clonazepam, dipyrone, pregabalin, amitriptyline, and methadone, without improvement. She has a history of intestinal, rectal, and breast cancer, the latter still active. She was diagnosed with post-mastectomy pain syndrome (PMPS), maintained on previously prescribed medications, and introduced cannabidiol 250mg per serving. After 7 days using 3 drops a day, the patient reported improvement, with pain 8 out of 10. In the following 7 days he started taking 7 drops a day with the same concentration, with significant improvement, reporting pain 5 out of 10, which allowed the reduction of the methadone dose.

Discussion: PMPS is a chronic neuropathic pain, resulting from direct injury or scar tissue formation of plexuses and nerves ipsilateral to the mastectomy, and may be aggravated by local radiation-based therapies such as radiotherapy. Nerve injury results in inflammatory processes that favor ectopic impulses and neuronal hyperexcitability, and thus neuropathic pain. Thus, due to its anti-inflammatory potential and ability to modulate pain pathways and dopamine release, which interferes with the response to pain, it is possible to understand the choice of cannabidiol as a therapeutic measure for the condition, considered refractory to conventional therapy, as well as the response presented by the patient after years without any improvement. **Final Comments:** In this perspective, despite the findings that speak in favor of the therapeutic potential of cannabidiol, it is important to emphasize the need for more studies to deepen the knowledge on the subject in question.

Dor

TL 1105320

PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA: THE CHALLENGES OF DIAGNOSTIC WORKUP – CASE PRESENTATION

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Case Presentation: A 16-year-old male patient developed hypersomnia, dysarthria, left hemiparesis, loss of sphincter control and seizures for 4 months. He received acyclovir, followed by pulse therapy with methylprednisolone, showing clinical improvement. After 2 months, he evolved with cognitive deficit, axial and appendicular ataxia, and diparesis. He received another pulse therapy, with no response and showed progressive impairment of ambulation. During the Immunoglobulin infusion, he developed clinical complications. Under intensive care, he performed plasmapheresis, with no improvement. Autoimmune markers were negative. The search for neoplastic cells in the cerebrospinal fluid (CSF) was inconclusive. Infectious screening was negative, as was extraneural neoplastic screening. Spinal resonance was normal and brain resonance showed multiple supra and infratentorial lesions, with moderate mass effect, post-contrast enhancement and moderate water diffusion restriction, with a nodular lesion in the pineal; proton spectroscopy suggested cell proliferation. Brain biopsy favored the diagnosis of lymphoproliferative disease. Immunohistochemistry described high-grade B-cell lymphoma of the central nervous system. With the diagnostic confirmation, she underwent systemic chemotherapy, showing slight clinical and radiological improvement, but it was interrupted by infectious complications, which led to death. **Discussion:** Primary central nervous system lymphoma is a rare and aggressive non-Hodgkin lymphoma that may involve the brain, leptomeninges, spinal cord or eyes. Approximately 90% are of the diffuse large B-cell lymphoma type. Congenital or acquired immunodeficiency is the main risk factor. In immunocompetent patients, the mean age at diagnosis is 53 to 57 years. Neurological symptoms tend to overlap with systemic ones. Diagnosis requires neuroimaging, CSF analysis and brain biopsy. Treatment involves radiotherapy and chemotherapy. **Final Comments:** The purpose of this report is to describe a case of Primary central nervous system lymphoma, a rare disease in a young and immunocompetent patient, emphasizing the importance of its recognition among the differential diagnoses, and the consequent search for timely therapy, providing greater survival.

Educação Médica

TL 1105366

CENTRAL RETINAL ARTERY OCCLUSION: POINT OF CARE TECHNOLOGY, TELESTROKE AND INDIRECT OPHTHALMOSCOPY TECHNIQUES

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Case Presentation: An 83-year-old man presented to the Emergency Department (ED) due to sudden non-painful left eye monocular vision loss 15h after he was last known well. His neurological examination was notable for a left eye relative afferent pupillary defect and a left eye visual acuity of light perception at 30 cm with no other focal neurological deficits. He was immediately taken to Radiology on stroke code and, during transport, mydriatic drops were instilled on his eyes. Approximately 20 minutes after first contact with Neurology, an indirect ophthalmoscopy done after neuroimaging with a 20 diopter lens and a smartphone camera confirmed a left eye central retinal artery occlusion (CRAO). **Discussion:** Most Hospitals do not have an ophthalmologist on call 24h a day and formal specialized consultations can result in treatment delays in time sensitive situations such as stroke. It is essential neurologists in EDs are capable of performing good quality fundoscopies. Although extensively advocated by the neurology education community as mandatory part of training, many neurologists are uncomfortable with the use of direct ophthalmoscopes. In addition, the 15° fundus view amplitude of direct ophthalmoscopy is very useful for optic disc pathology detection, but could potentially miss CRAO, in which optic discs are spared. Our proposed technique circumvents these drawbacks with a much wider, 360° view of the eye fundus, without adding a significant amount of time to the care of patients on acute stroke codes, since mydriatic drops can be used during the neurological exam or while transporting patients to imaging. Furthermore, when the on call neurologist is not sure of the correct interpretation of the eye fundus, images can be sent via secure instantaneous communication channels, such as mobile phone applications, for discussion with fellow and more experienced colleagues, ensuring tailored care delivery as in telestroke. As limitations of our work, we emphasize some patients have absolute contra-indications to the use of mydriatic drops, the 15 minute delay it takes for pupillary dilation, the price of indirect ophthalmoscopy lens and the need for staff training. **Final Comments:** CRAO diagnosis can only be established by good eye fundus exam. Indirect ophthalmoscopy with smartphone camera is a powerful mean of rapidly making this diagnosis in the ED and broadening access to emergency stroke reperfusion therapy in select eligible patients.

Educação Médica

TL 1105818

GROUP ACTIVITIES FOR PARKINSON'S PATIENTS IN THE COUNTRYSIDE OF BRAZIL – A SUCCESSFUL EXPERIENCE

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Case Presentation: The present work intends to describe the creation and strengthening of an association of patients with Parkinson disease (PD) in a medium-sized city in Brazil. In September 2021, it started a partnership with a local college with 6 patients and the neurologist who led the cases. The initial objective of the partnership was to encourage the socialization and practice of physical activities in those patients, in addition to reducing the stigmas that accompany the disease. In May 2022, the group had grown to 50 participants, activities twice a week, conducted by students from the 9th and 10th periods of physiotherapy. The strategies are set up with stimuli for gait training, cognitive strategies to improve transfers, exercises for balance, mobility training joints and muscle strength to implement physical capacity. Low cost materials used are dumbbells, balls, shin sticks, cones, walking tracks with obstacles. In addition to routine activities, the group counts on the participation of guests in lectures that explore other aspects of the parkinsonian's life. **Discussion:** Parkinson disease is a neurodegenerative disease characterized by bradykinesia and hypokinesia combined with rest tremor and/or rigidity. In addition to these typical motor features, patients with PD may experience nonmotor symptoms related to the disease itself or to the medications used to treat it. Those patients require broad-based management including patient and family education, support group services, general wellness maintenance, exercise, and nutrition. Education is essential in order to provide the patient and family with some understanding and control over the disorder. Education can combat the stigma and misinformation that often surround the disorder. Regular exercise is very important in those patients, because it promotes a feeling of physical and mental wellbeing. Exercise may help to slow motor progression, improve nonmotor symptoms, and alleviate some secondary orthopedic effects of rigidity and flexed posture. Patients are at risk for social isolation, depression is one of the most common psychiatric disturbances seen. **Final Comments:** With a low-value investment, it is possible to offer quality of life and dignity to patients with Parkinson's Disease in medium-sized cities. We believe that centers like this one should be stimulated by the interior of Brazil, improving the quality of life of patients and allowing higher quality studies by uniting patients.

Educação Médica

TL 1105047

STROKE OR SEIZURE: A CHALLENGING CASE OF PROLONGED TODD PARESIS

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Case Presentation: A 32-year-old male with a history of remittent epilepsy without current use of antiepileptic drugs, traumatic brain injury with amaurosis in the left eye, smoking, and abstinence from cocaine presented to our ER with left-sided weakness, mild dysarthria and loss of nasolabial fold at the left with 30 min of onset. CT scan at the admission presented no signs of bleeding. His initial NIHSS was 21. He had no contraindications to thrombolysis and alteplase was administered without complications. After thrombolysis, NIHSS remained the same. On the next day, the patient remained with hemiparesis and a motor examination showed weakness on his left side (2/5, Medical Research Council [MRC] scale). Brain MRI was performed and showed no alterations. Transthoracic echocardiogram and doppler ultrasound imaging of the carotids and vertebral arteries were normal. Blood count, autoimmune disease biomarkers, and metabolic panel were without alterations. By day 5 of admission, the patient had an improvement in his symptoms but remained with weakness on his left side (4/5 on his left leg and 2/5 on his left arm). Prolonged Todd's paresis was considered and an EEG showed disorganization, asymmetry, hyporeactivity, and slowing of background activity, compatible with mild-moderate toxic-metabolic encephalopathy, with a background asymmetry possibly related to a post-ictal seizure state. On day 11 of admission, the patient was asymptomatic and was started on valproic acid 250 mg t.i.d., discharged home, and referred to the Neurology outpatient clinic. **Discussion:** Todd paresis, also known as Todd paralysis or postictal hemiparesis, was initially described in 1849 and is associated with weakness or paralysis in part or all of the body after a seizure. Since stroke is the main differential diagnosis in patients presenting with weakness in the ER, this may lead to misdiagnosis and unnecessary treatments. In fact, in one series, this was the most common non-stroke cause of referral to a stroke unit. However, Todd paresis tends to be a regressive deficit with symptoms that do not exceed 36h, and only rarely do symptoms last beyond this period of time. **Final Comments:** In this way, this is a crucial differential diagnosis in the ER and a thorough evaluation involving medical history, physical examination, clinical parameters, neuroimaging, and EEG is often necessary to establish the diagnosis, particularly when atypical presentations such as prolonged Todd paresis take place.

Epilepsia

TL 1105087

STATUS EPILEPTICUS IN A PATIENT WITH ANOREXIA NERVOSA-RELATED WERNICKE-KORSAKOFF SYNDROME: CASE PRESENTATION

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Case Presentation: A 42-year-old female underwent exploratory laparotomy due to acute intestinal obstruction, and nineteen days later lost consciousness. She had been severely depressed and anorexic for over four months by that time. There was no report of previous seizures, other neurological disorders, smoking or regular alcohol use. Physical examination revealed pupillary hippus, vertical and horizontal nistagmus, tachyarrhythmia, irregular breathing pattern and bilateral focal neurological signs - particularly on the left - findings suggestive of focal status epilepticus, which was successfully treated with IV midazolam. The hypothesis of status epilepticus complicating Wernicke-korsakoff syndrome (WKS) was raised. MRI displayed several T2/Flair hyperintense areas and "faint" hyperintense DWI areas in the mammillary bodies, thalamus, parieto-occipital zones and cerebellum, as well as gyriform oedema within the telencephalic sites. In addition, a medium right-sided parieto-occipital area of DWI hyperintensity (with corresponding ADC map hypointensity) was found. The radiological findings were suggestive of peri-ictal epileptic state and WKS + secondary parieto-occipital stroke. EEG, performed three days after seizure control was obtained with phenytoin and levetiracetam, was compatible with toxic-metabolic encephalopathy and focal slowing on the right posterior cerebral quadrant due to stroke. **Discussion:** WKS is a disorder caused by thiamine deficiency, usually related to binge alcohol drinking, or, alternatively, to anorexia nervosa, hepatic dysfunction, hyperthyroidism or hyperemesis gravidarum. The recommended daily thiamine intake is supplied by a normal healthy diet. However, malnourished people undergoing medium-large surgeries (which demand a good storage of B1), are especially prone to this condition. Nevertheless, associated focal cortical lesions are rather rare, and among these cases 70% are non-alcohol-related and 30% present seizures at some point of clinical follow-up. **Final Comments:** we have reported an atypical evolution of anorexia nervosa-related WKS, which led to permanent multifocal encephalic damage following status epilepticus. Although its atypical course here, WKS should be considered - as a standard of care - in every single patient at the emergency room with reduced conscious level. WKS must always be a "don't miss" diagnosis, and, as soon as possible, treated with thiamine. Therefore, the burden of permanent brain damage is usually minimal.

Epilepsia

TL 1105088

TUBEROUS SCLEROSIS OR BOURNEVILLE-PRINGLE DISEASE: CASE SERIES AND LITERATURE REVIEW

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Case Presentation: CASE 1: A 58-y-old male with intellectual disability, epilepsy, renal angiomyolipomas, Dupuytren contracture in hands, facial angiofibromas and ungual fibroma in the nail matrix. CT scan showed cortical atrophy and periventricular calcifications (calcified tubers). EEG with epileptiform activity in anterior regions bilaterally. CASE 2: A 33-y-old female with epilepsy, renal angiomyolipomas and facial angiofibromas. Her grandfather, father and cousin had the same signs and symptoms. Brain MRI showed subcortical tubers in both cerebral hemispheres and a small subependymal nodule. The genetic testing showed alteration in the TSC2 gene. CASE 3: A 35-y-old male with epilepsy, intellectual disability, facial angiofibromas and hypopigmented macules on the back. CT scan showed two cortical hamartomas and subependymal nodules. CASE 4: A 36-y-old male with epilepsy, intellectual disability, facial angiofibromas, shagreen patches and hypopigmented macules on the back. **Discussion:** Tuberous Sclerosis (TS), also known as Bourneville-Pringle disease, a rare autosomal dominant phacomatosis or cutaneous syndromes, was first describe in 1862 by von Recklinghausen. Posteriorly, Bourneville and Pringle used the term Tuberous Sclerosis. In 1905, Vogt characterized the clinical triad of mental retardation, seizures and characteristics skin lesions. TS is originates from mutations in the TSC1 and TSC2 tumor suppressor genes located on chromosomes 9q34 and 16p13.3, respectively. The TSC1 gene is responsible for encoding a protein called hamartin and TSC2 for tuberin. TS affects both genders equally with a great diversity of manifestations (cutaneous, renal, pulmonary, cardiac, and neurological). The diagnosis is basically clinical. All patients described have epilepsy, the main neurological manifestation of TS, occurring in up to 90% of the cases, with seizures usually starting in the first three years of life. All patients have renal angiomyolipomas which usually affects about 80% of patients. All of them presented facial angiofibromas and also cortical tubers. Although genetic testing was performed in only one patient, the diagnosis can be made in the other cases based on the clinical and radiological presentation. Complementary exams, like CT and MRI, are important to detect cortical tuberousities. **Final Comments:** TS is a rare disease. Its early recognition is of great importance for a better treatment and follow-up, with multidisciplinary team.

Epilepsia

TL 1105090

LEVETIRACETAM-INDUCED RENAL FAILURE: CASE PRESENTATION AND LITERATURE REVIEW

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Case Presentation: A 24-y-old male patient with a history of epilepsy since the age of 16, in regular use of antiepileptic drugs, was brought to our ER due to a new seizure. He had been using oxcarbazepine since the beginning of treatment. The EEG showed frequent slow wave paroxysms and slow wave peaks of 3-6 Hz. Levetiracetam had been recently added with good seizure control. Normal neurological exam. Brain MRI showed no changes. Laboratory tests showed high levels of creatinine and he progressed to acute renal failure. During Hospitalization, a renal biopsy was performed, which showed acute tubular nephritis. Levetiracetam was then discontinued and valproic acid started, with progressive improvement in renal function. He was discharged from Hospital after a few days and without new epileptic seizures. **Discussion:** Levetiracetam is a broad-spectrum anticonvulsant, being one of the most used drugs in its class. This drug has minimal pharmacokinetic interactions due to the fact that it does not bind to plasma proteins and is independent of the cytochrome p450 system. Thus, its main side effects, already reported in the literature, are dizziness, headache, asthenia, drowsiness and rarely adverse behavioral effects. However, this anticonvulsant, due to its water-soluble properties and low molecular weight, is largely eliminated by the kidneys and may, although rarely, cause repercussions in this system. There are still few cases relating the use of levetiracetam and acute renal failure, with the first case described being that of a 17-year-old girl who developed acute renal failure and interstitial nephritis ten days after starting treatment with the drug. Thus, the case described in this work is one of the few described in the literature, and only one of the three existing cases found involving a male patient. As observed by previous descriptions, the age range is variable, having affected patients from 17 to 45 years old. **Final Comments:** The association of renal failure and levetiracetam is rare, being of fundamental importance to recognize it for a better approach. In addition, it is worth mentioning that the use of this medication, like any other, must be well evaluated, taking into account all possible risks, benefits and adverse effects.

Epilepsia

TL 1105321

DUPUYTREN'S CONTRACTURE SECONDARY TO CHRONIC USE OF PHENOBARBITAL: CASE PRESENTATION AND LITERATURE REVIEW

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Case Presentation: Male, Caucasian, 65 years old, retired (rural area resident), attended the neurology outpatient clinic in October 2021 and showing pain and difficulty moving the fingers of both hands. He observed nodules and hardened tendons, with progressive installation in the last 3 years, with worsening in the last 6 months. He had epilepsy for 26 years: focal motor seizures with bilateralization, and focal impaired seizures, with epilepsy secondary to neurocysticercosis treated with praziquantel in the year 1996, with subsequent ischemic stroke in the year 2000, and status epilepticus, secondary to middle cerebral artery vasculitis due to active perivascular cysticercosis, treated with Prednisone. The patient evolved with control of epileptic seizures through the use of phenobarbital 200 mg/day and carbamazepine 600/day in the last 22 years. Examination of hands: palmar contracture, passive extension deficits of the 3 fingers (2°, 3° e 4°) with retraction and fibrosis of the flexor tendons and nodules. Abdominal ultrasound examinations were performed, in addition broad laboratory evaluation, and they were all normal, excluding secondary causes such as collagen diseases, and additional family's history of Dupuytren's (CD) contracture, being defined a diagnosis of CD secondary to the use of phenobarbital. **Discussion:** Dupuytren's contracture (CD) is a fibroproliferative condition involving the superficial palmar fascia, leading to a progressive and irreversible flexion of the fingers. Dupuytren's (CD) contracture might have a higher incidence in patients with epilepsy. In literature, there are different opinions regarding the phenobarbital, a common antiepileptic drug, and its effective role in the genesis and development of CD. We believe that phenobarbital causes a dose and time-dependent profibrotic effect. The CD's aetiology of some aspects is still unclear. Many factors are already known to be implicated, and there are different opinions regarding the association with the phenobarbital used for the antiepileptic treatment, and its effective role in the genesis and development of Dupuytren's contracture. **Final Comments:** Our case presentation demonstrate that phenobarbital cause a dose and time-dependent profibrotic effect and must to be involved in the genesis of Dupuytren's contracture. Progressive withdrawal of phenobarbital and introduction of Valproate was performed, and he was referred for surgical treatment by an orthopedist.

Epilepsia

TL 1105555

FEBRILE INFECTION-RELATED EPILEPSY SYNDROME (FIRES) AS A CAUSE OF SUPER-REFRACTORY STATUS EPILEPTICUS: A CASE PRESENTATION

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Case Presentation: A previously healthy 14-year-old female was brought to the emergency department with focal to bilateral tonic-clonic seizures that started abruptly at the day of admission. There was a report of odynophagia, headache and fever over the past five days. She persisted with recurrent seizures despite first-line treatment, as well as continuous infusion of sedatives. After therapeutic optimization, including the administration of six classes of antiepileptic drugs, the patient maintained recurrence of seizures. The electroencephalogram (EEG) showed a severe disturbance of the background activity, with a burst-suppression pattern. After a thorough clinical and laboratory evaluation, the hypothesis of an autoimmune encephalitis was raised, and the patient was started on immunosuppressive steroid therapy (methylprednisolone). The clinical response was satisfactory, however, after the third day of therapy, the infusion was interrupted due to a severe respiratory infection. Following the interruption of therapy, a deterioration of the EEG pattern was observed, as well as clinical recurrence of focal seizures, requiring an increase of the intravenous sedatives. Given the respiratory infection, intravenous immunoglobulin was administered (total dose of 2g/Kg). The patient remained without recurrence of clinical seizures, but EEG worsening was maintained. Therefore, barbiturate coma with thiopental was started, with no satisfactory response for 48 hours. After adequate treatment of the infection, a second immunosuppressive steroid therapy was started (methylprednisolone 1g/day for five days). The patient, then, presented clinical control of seizures and improvement of the electroencephalographic pattern, with gradual reduction of sedatives. **Discussion:** Status epilepticus in previously healthy individuals is known as new-onset refractory status epilepticus (NORSE). When it affects young patients with a history of febrile infection preceding the seizure by two weeks to twenty-four hours, the condition is known as febrile infection-related epilepsy syndrome (FIRES). It is a rare condition, with an estimated incidence of one per million. **Final Comments:** The case presented demonstrates the difficulty of clinical management of FIRES, due to the high refractoriness to standardized treatments for status epilepticus.

Epilepsia

TL 1105767

CORTICAL DYSPLASIA AND ANTI-GAD ENCEPHALITIS IN A PATIENT WITH REFRACTORY MESIAL TEMPORAL LOBE EPILEPSY: CASE PRESENTATION

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Case Presentation: Female patient, with type 1 DM and hypothyroidism, with non-perceptive seizures since 14 years old. Dejá Vu aura, oro-masticatory and motor automatism were described. She used to have 3-6 episodes/day. An investigation was performed with brain MRI, evidencing right mesial temporal cortical dysplasia, and video-EEG with right temporal ictal semiology but left temporal electrographic onset. Seizures were refractory to treatment with 1200mg/day carbamazepine. PET compatible with bilateral temporal hypometabolism, worse on the right, neuropsychological evaluation with visuospatial difficulty. Given the suspicion of Cortical Dysplasia, a surgical approach was chosen, with a right temporal lobectomy performed. Pathology compatible with type IB cortical dysplasia. After surgery, she was seizure-free and drug-free for 2 years, when she had a recurrence of seizures with the same ictal pattern. Even after adjusting the antiepileptic medications, there was no satisfactory control of the seizures. In view of the endocrine disease, the possibility of associated anti-GAD encephalitis was raised, with documented antibody positivity in the CSF, in addition to the raising of serum titers in the period seizure recurrence. Immunotherapy with IVIG was started for 5 days in 2021 with marked seizure frequency reduction. **Discussion:** Active neuroinflammation and cell injury seems to play a common pathogenic role in epilepsies. The increased prevalence of autoantibodies in epilepsy reinforces that immune dysregulation participates in this process. As in temporal lobe epilepsy and cortical dysplasias, in immune-mediated encephalitis, there is microglial activation, providing evidence of an immune response, in addition to astroglial activation, which accompanies the progression of cortical damage and probably participates in the inflammatory process. Among patients with epilepsy, 2.58% of them had a positive anti-GAD antibody. The correlation between focal dysplasia and anti-GAD encephalitis seems to raise the idea that an autoimmune response caused by dysmorphic neurons contributes to the pathogenesis of the intense inflammatory response. Outcome: Epilepsies that have autoimmunity as an etiology go beyond this spectrum and it is increasingly important to recognize them as potentially treatable with immune modulation. Even with scarce evidence, patients with focal epilepsies and autoantibodies may benefit from immunotherapy even after surgical intervention.

Epilepsia

TL 1106064

THE ELECTROENCEPHALOGRAM IN THE RESEARCH OF PROGRESSIVE MYOCLONIC EPILEPSY

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Case Presentation: A previously healthy 8-year-old patient, with adequate neuropsychomotor development, started a generalized tonic-clonic crisis (TCGC), afebrile, during sleep in October 2021, associated with the presence of sudden movements contraction of limbs, described as "shocks". He underwent a cranial tomography without alterations, remained asymptomatic for two weeks. He evolved with weekly frequency TCG crisis, with sudden movements (myoclonus) daily and started to present behavioral changes. Two months after the onset of the condition, the patient showed gait and speech disturbances and had daily epileptic seizures. Neurological examination: fluctuating alertness, expression aphasia, slow gaze tracking. Myoclonus on the face, incomplete trunk support. Limb coordination: dyskinesia. Balance/gait: gait with a support, discreetly dyskinetic and on tiptoe. Exams: CSF: clear; 0 leukocytes; 2 red blood cells; proteins 26; Cranial MRI: multifocal signal alteration affecting the subcortical white matter of both cerebral hemispheres, without cytotoxic edema or intramyelinic edema. EEG: abnormal, due to diffuse disorganization of sleep activity, with asymmetry, worse in the left hemisphere. Generalized, polyphasic, high-amplitude periodic activity at 3- to 6-second intervals. After the EEG result, the hypothesis of subacute sclerosing panencephalitis due to measles (characteristic clinical picture, typical EEG) was made, and a new CSF puncture showed negative immunology for measles IgM and IgG 1: 32 and protein electrophoresis with the presence of multiple bands in the gamma region (81.3%). **Discussion:** Subacute sclerosing panencephalitis (SSPE) is a progressive and fatal neurodegenerative encephalitis caused by the persistence of measles virus in the central nervous system (CNS). In the state of Ceará, there were 916 confirmed cases of measles between December 2013 and September 2015. The only concrete strategy against measles and its complications is prevention through vaccination. The COVID-19 pandemic has left a dangerous immunity gap due to the suspension of measles awareness campaigns and delay in immunization, leading to medical concern about the risk of further measles outbreaks in the coming years. **Final Comments:** In this case, we highlight the electroencephalogram as a fundamental exam for the diagnosis, showing the periodic complexes with a long interval, thus directing the investigation in the CSF to confirm the pathology.

Epilepsia

TL 1106106

FOCAL AWARE STATUS EPILEPTICUS IN PATIENT WITH STROKE-LIKE MIGRAINE ATTACKS AFTER RADIATION THERAPY (SMART) SYNDROME: A CASE PRESENTATION

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Case Presentation: A 56-year-old woman was admitted with a three-day history of complex visual hallucinations. She had previous history of right temporo-occipital grade III ependymoma, diagnosed 10 years before, and treated with surgery and radiotherapy. Repeated neuroimaging exams showed no signs of tumor recurrence. She reported neither epileptic seizures nor use of antiseizure medications (ASM). On admission, she reported recurrent episodes of complex visual hallucinations, described as altered depth perception and image distortion, associated with left lateral gaze deviation, without impairment of consciousness. Episodes occurred several times a day and lasted up to an hour. On admission, neurological examination revealed left homonymous hemianopsia, with no other abnormalities. Magnetic resonance imaging (MRI) of the head demonstrated cortical gyriform enhancement surrounding the surgical site in right temporo-parieto-occipital region, virtually pathognomonic of stroke-like migraine attacks after radiation therapy (SMART) syndrome. Continuous electroencephalogram (cEEG) monitoring showed repeated focal epileptic seizures, with ictal onset over posterior regions of the right hemisphere. Clinically, the patient referred visual symptoms; in some of the seizures, she had eye and head deviation to the left. Seizures lasted up to 77 minutes, leading to diagnosis of focal status epilepticus (SE) without impairment of consciousness. She was treated with intravenous levetiracetam plus oral clobazam, which led to seizure cessation. MRI was repeated one month later and showed complete resolution of previous findings. A new EEG demonstrated absence of epileptiform abnormalities. **Discussion:** SMART syndrome is a rare reversible syndrome that occurs as a late effect of brain radiotherapy. Classical features include migraine-like headaches, focal neurologic signs and/or seizures, associated with typical imaging findings. Episodes last days to weeks and usually occur years or decades after radiotherapy. We report a case of a patient with SMART syndrome who presented with focal aware SE with visual symptoms, which is an atypical presentation of this syndrome. After treatment with ASM symptoms regressed, as well as MRI abnormalities, demonstrating the reversible character of the syndrome. **Final Comments:** Albeit rare, SMART syndrome should be considered in patients with history of brain radiotherapy and typical clinical and neuroimaging findings.

Epilepsia

TL 1106111

ATYPICAL ROLANDIC EPILEPSY AND LANGUAGE DELAY ASSOCIATED WITH NR4A2 AND GRIN2A

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Case Presentation: M.A.B, female, 4 years old, normal pregnancy and delivery, non-consanguineous parents. In the first year of life, she had motor development delay, and at 2 years old, her language was incomprehensible, and presented inappropriate social behaviors; At age 3, she started with focal seizures during sleep. Dysmorphic features were absent; Good social interaction, but impaired attention; Low expressive linguistic with predominantly gestural/verbal speech. Initiates a conversation, as it presents difficulties in understanding. The orofacial structures presented low posture, tone, mobility of the lips and cheeks; First EEG: independent projection epileptiform activity in the centrottemporal regions activated by sleep; The use of oxcarbazepine, but the attacks increased in frequency; Control EEG: frequent epileptiform activity and associated with slowing of baseline activity in the left cerebral hemisphere. MRI normal; Clobazam was associated with oxcarbazepine, with no improvement in the seizures and later levetiracetam, but the seizures continued to occur sporadically. The last seizure was 4 months ago. Genetic analysis by next-generation sequencing identified: In heterozygosity, in the NR4A2 gene, a variant promoting the substitution of the amino acid isoleucine. The NR4A2 gene encodes a transcription factor that is highly expressed in critical regions for language development and the normal formation of the dopaminergic pathway. Autosomal dominant inheritance. In heterozygosity in the GRIN2A gene, a variant that promotes the substitution of the amino acid valine for leucine; The treatment was modified, for a gradual withdrawal of oxcarbazepine and maintenance of levetiracetam and clobazam, without new crises.

Epilepsia

TL 1106471

PHENYTOIN-INDUCED RHABDOMYOLYSIS

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Case Presentation: A 41-year-old male patient, previously hypertensive and diabetic, presented a sudden onset of two reentrant generalized tonic-clonic seizures, without regaining consciousness between them, with a total duration of about 10 minutes. In the initial evaluation in the emergency room, was in a typical post-ictal period, and initial assistance was provided and a loading dose of phenytoin with 20 mg/kg, was performed, and later maintained medication at a dose of 100 mg every eight hours. In laboratory tests on admission, the patient had Creatine phosphokinase (CK) of 311 U/L, creatinine 1.3 mg/dL. During Hospitalization, an acute curve of CpK elevation was observed over the days, with peak observed on the fourth day of Hospitalization, in association with Acute Kidney Injury (AKI), with creatinine 5.7 mg/dL. In view of the expressive increase, in large proportion, in the context of a stable patient, without new crises, and without complaints, a hypothesis of association was raised the causal link between the rhabdomyolysis condition and the administration of phenytoin, with the drug being changed to Levetiracetam on fourth day of Hospitalization (CK 20199 U/L). After this event, an immediate progressive drop in serum creatine phosphokinase levels was observed, so that and 24h after phenytoin suspension went to 14000. **Discussion:** The causal association between phenytoin and rhabdomyolysis is considered rare, its first report being in 1976. In some cases, the manifestation and in a context of hypersensitivity syndrome, with other clinical and laboratory findings such as eosinophilia and skin rash, however, there are reports as being the only clinical manifestation, still considered with uncertain pathophysiology. In these cases, the diagnosis is a challenge, because rhabdomyolysis is a frequent finding in seizures. However, in view of being a complication often associated with acute kidney injury, it is important to evaluate differential diagnoses that require rapid management. **Final Comments:** In this context, attention should be paid to this little-known adverse effect of the drug. Thus, acute elevations to unusual CK values, progressive worsening of AKI, and refractoriness to clinical measures to treat the condition should be considered warning signs for phenytoin-induced rhabdomyolysis. Since the response in these cases was observed soon after drug withdrawal, as seen in the case in question.

Epilepsia

TL 1106589

TRANSVERSE MYELITIS ASSOCIATED WITH NEUROLISTERIOSIS: A CASE-BASED UPDATE

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Case Presentation: A 68-year-old man was admitted with a case of suspected chronic lymphocytic leukemia, due to leukocytosis with a lymphocytic predominance. He started investigation for further treatment in 2020, losing follow-up in the pandemic. 2 years after, the patient reported two episodes of falls. In the first, he reported a fall from a "water truck", with pain and swelling in the left knee, but without trauma or excoriation. A week after this event, he fell off the hammock reporting severe pain in the lumbar region, being medicated, but always with the pain returning after the medication ceased its effects. From the second fall, on the following day, the condition evolved with paresis and hypoesthesia in the left lower limb. On this same day, evolving with involvement in the right lower limb. He was referred to the reference Hospital in the region, already with a condition of grade 1 paraplegia and loss of sensitivity at the T10-T11 level. For investigation of the condition, a lumbar puncture was performed, the analysis of cerebrospinal fluid showed a yellow and turbid aspect. Then, antimicrobial treatment was performed with subsequent confirmation of infection by *Listeria monocytogenes*. After imaging tests, myelitis was suspected, which was associated with the bacterial infection. **Discussion:** *Listeria monocytogenes* is a gram-positive bacteria commonly transmitted by ingestion of poorly washed or undercooked food, via the fecal-oral route. This type of bacteria has a certain affinity with the central nervous system, affecting mostly neonates, pregnant women, elderly, and immunocompromised patients. Neurolisteriosis can occur in two ways, through a retrograde neural (associated with rhombencephalitis) or a hematogenous route (resulting in meningoencephalitis). Neurolisteriosis is an uncommon occurrence, but its association with myelitis resulting in a spinal cord syndrome is rare, even in cases with risk factors, such as possible immunosuppression caused by leukemia in the present case, with meningeal involvement being more common. **Final Comments:** Neurolisteriosis is a rare event which may be associated with myelitis, and such an association should be suspected, especially in patients at risk for infection. Its treatment should be initiated quickly, due to its high mortality rate.

Líquido cefalorraquiano

TL 1104966

NEUROLOGICAL ALTERATIONS RELATED TO VITAMIN B12 DEFICIENCY: A CASE PRESENTATION

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Case Presentation: A 56-year-old male patient, in February 2012, presented a neurological condition showing irritability, paranoid delusions, evocation amnesia and apraxia when dressing and performing personal hygiene. Simultaneously, paresthesia and dysbasia in the lower limbs began due to imbalances of insidious onset. On examination, cognitive screening tests were initially unrealisable, and there was diffuse areflexia, hypoesthesia in "boot" and ataxic gait. Laboratory evaluation demonstrated normal blood count, electrolytes, transaminases, nitrogen scores, folic acid, ESR and blood glucose levels, TSH 12.99 mUI/L, free T4 0.87 ng/dL, VDRL and anti-HIV 1 and 2 not reactive, serum vitamin B12 50.0 pg/mL. Magnetic resonance imaging of the skull showed a small cystic lesion in the right basal ganglia and mild leukoariosis. The patient started parenteral use of a compound of B complex vitamins – 5000 U, intramuscularly, once a week, for three months, associated with quetiapine, 50mg/day, and evolved with memory, behavioral disorders and balance improvement as well as functional autonomy, however remaining with a slight sensation of numbness in the feet. **Discussion:** Vitamin B12 deficiency usually causes megaloblastic anemia, which may be absent in one third of cases, associated with neurological symptoms, due to vascular alterations associated with the elevation of homocysteine, or alterations in methylation reactions, affecting myelin. Such alterations compromise axons of the ascending tracts of the posterior funiculus, descending pyramidal tracts and peripheral nerves. The treatment of the disease is carried out through parenteral replacement of the vitamin, with a maximum improvement generally occurring in the first six months, and its early initiation is of great importance to avoid possible neurological sequelae. **Final Comments:** This scenario, consisting of an amnesic and behavioral, apraxia, ataxia and sensory cognitive syndrome, representing subacute dementia, sensory ataxia and sensory polyneuropathy, has, in many cases, a secondary relation to vitamin B12 deficiency, based on its serum dosage, with the possibility of associated problems, such as the existence of a subclinical hypothyroidism, which was also verified in the patient.

Miscelânea

TL 1104970

INADVERTENT INTRATHECAL TRANEXAMIC ACID ADMINISTRATION: A MISTAKE WITH POTENTIAL CATASTROPHIC OUTCOME

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Case Presentation: A 20-years-old pregnant woman was submitted to spinal anesthesia for an elective cesarean procedure at another Hospital. She referred no anesthetic effect after administration of 15 mg bupivacaine, so an additional dose of 15 mg was used. The procedure was finished as usual, however, about 30 minutes later, the patient presented seizures refractory to the first approach with diazepam, and progressed to clinical status epilepticus (SE). There was no history of headache or hypertension during the pregnancy. Her prenatal was unremarkable. After reviewing all the infused drugs, the anesthetist noted that tranexamic acid was wrongly administered during the anesthetic procedure due to the first ampoule being wrongly relabelled. She was sedated, submitted to orotracheal intubation, and transferred to our Intensive Care Unit. At the admission in our Hospital, a new generalized tonic-clonic seizure was observed, so it was treated according to SE protocol. The electroencephalogram demonstrated a burst-suppression pattern due to sedatives. Sedation was progressively reduced until the patient woke up. She was discharged from the Hospital some weeks later without cognitive sequelae, using levetiracetam, which withdrawal was done at our outpatient clinic. **Discussion:** Tranexamic acid is an antifibrinolytic agent indicated in an obstetric scenario to control postpartum hemorrhage. Frequently anesthesiologists have this medication in their medicine box. Its adverse effects may vary from severe back pain and muscle spasm to hemodynamic instability, ventricular tachycardia, seizures and SE. There is no antidote to this drug, so the recommended management is to treat complications. Cerebrospinal Fluid (CSF) lavage, a technique that consists of 10-20mL CSF drainage and infusing the same amount of preservative-free normal saline, is indicated in some cases. Although there are only a few studies considering this procedure, none of them were performed in patients with inadvertent intrathecal administration of tranexamic acid. **Final Comments:** Despite doubtless advances in patient health security, mistakes with catastrophic disclosures still occur. Even with many preventive strategies, the wrongly relabelled drug at the Hospital pharmacy in association with the similar aspects of bupivacaine and tranexamic acid ampoules contributed to that. Thus, the relabel practice should be reviewed.

Miscelânea

TL 1104978

EXTENSIVE LEUKOENCEPHALOPATHY SECONDARY TO VITAMIN B12 DEFICIENCY: CASE PRESENTATION

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Case Presentation: A 43-year-old male patient with a history of sudden cervical pain irradiating to the lower limbs followed by paraplegia 11 years before, with a slight improvement of motor deficit, remained restricted to bed for 2 years, when he presented spontaneous recovery of functionality, without complaints for the following 2 years when he acutely presented symmetrical paraparesis with episodes of daily kinesigenic painful dystonia in the lower limbs with clonus, associated to paresthesia with a sensory level not clearly defined in the T5 topography. The condition has remained stable with no improvement in weakness up to the present day. Personal history: SAH. On examination: alert, oriented, mild intellectual impairment, isochoric and photoreagent pupils, paraparesis (grade 1), spastic tone, normotrophic, superficial hypoesthesia at T5, distal hypopalesthesia with patellar hyperreflexia, Babinski sign on the right and bilateral Hoffman sign. Mini mental with score below the value for the degree of education (27/30). Serum vitamin B12 dosage <148, with macrocytic and hyperchromic red blood cells, without anemia; MRI of the neuroaxis showing confluent punctiform foci in subcortical and deep white matter. U-shaped cortical fibers and diffuse medullary atrophy, with hypersignal on T2 and STIR in the posterior cord at the level of D2 to D5, suggestive of vitamin B12 deficiency. **Discussion:** The combined clinical signs of pyramidal syndrome, with clear predominance of lower limb involvement, sensory changes with spinal cord sensory level suggestive of myelopathy, and quantified cognitive decline, necessitated the need for CNS imaging study that showed extensive leukoencephalopathy pattern and signal enhancement of the posterior spinal cord, which, together with the finding of serum hypovitaminosis B12 and gastric atrophy on AGE, strengthened the hypothesis of specific metabolic myelopathy. **Final Comments:** Vitamin B12 is directly linked to neuropsychiatric functions due to its participation in DNA synthesis and neuronal myelination, and, thus, its deficiency can cause alterations in the central and peripheral nervous system. The above patient presented an atypical clinical picture, with spastic paraparesis and fluctuating painful dystonias, with a surge pattern, extensive radiological involvement, suppressed serum dosage of this substrate, and a definite digestive disabsorptive etiology, consistent with the diagnosis of cyanocobalamin deficiency encephalomyelopathy.

Miscelânea

TL 1104995

FROM OTOLARYNGOLOGY TO HAEMATOLOGY: A BOND CHAINED BY NEUROLOGY

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Case Presentation: A 49-year-old man presenting a growing left neck mass was under etiopathological investigation by otorhinolaryngology for two months before he became bedridden. Upon neurological evaluation, he had upper limbs paralysis, lower limbs paresis, urinary retention and sensitive level at T10 dermatome. Diagnostic work up disclosed chronic anemia and a hypodense cervical mass associated to bilateral pleural thickening and left pleural effusion on chest CT. The Queckenstedt's manoeuvre was positive for spinal block and the CSF analysis revealed significant hyperproteinorachia. Spine MRI unveiled incursion of the cervical mass into the spinal canal through the neural foramen from C2 to C7, compressing the cervical and dorsal cord. Surgical biopsy of the mass was conclusive for nodular sclerosing Hodgkin lymphoma. Steroids had been previously prescribed and had led to some motor improvement. Chemotherapy was initiated and further progress was achieved, especially for upper limbs segments and for left lower limb. After four months of follow-up, however, trunk balance is still unsatisfying. **Discussion:** Hodgkin's Lymphoma (HL) is a hematological malignancy that represents 0.5 to 1% of adult cancers. Spinal cord compression (SCC) is usually a late complication by lymphoma. Moreover, it is three times more frequent in non-Hodgkin's disease. In HL, it occurs only in 5% of cases. In the present report, there is probably a direct spread from cervical lymph nodes, although the cervical spine tends to be the least involved segment. The epidural mass infiltrated the spinal canal via the left neural foramen and spared the vertebral bodies. A retrospective study carried out between 1987-2002 gathered only 12 patients (11 males) with SCC, of which half with vertebral body destruction and only two with cervical compression. Cervical involvement is advantageous in terms of time for diagnosis, since the mass may be promptly noticed and the diagnosis procedures are favored. Differential diagnosis includes: non-Hodgkin's lymphoma, squamous cell carcinoma, tuberculosis and eosinophilic granuloma. **Final Comments:** We present a case of unusual manifestation of Hodgkin lymphoma, that exemplified the need of multidisciplinary integration in order to establish the diagnosis. The neurological propaedeutic was able to predict the large extension of SCC and supported the celerity in the investigation and intervention.

Miscelânea

TL 1105016

DIFFUSE LARGE B CELL LYMPHOMA WITH SECONDARY CENTRAL NERVOUS SYSTEM WITH ATYPICAL PRESENTATION

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Case Presentation: A 67-year-old man presented with 20 days of mental confusion, emotional apathy, dysphagia, followed by decreased level of consciousness. Previously, he had a weight loss of 10 kg in 3 months. He had not fever, night sweats or headache, and had no relevant comorbidities. At examination, he had multiple cervical lymphadenopathy, spontaneous eye opening, localized painful stimulus, no verbal response. MRC grade 3 in all limbs, global hyperreflexia and frontal release signs were present. Brain MRI demonstrated multiple contrasting juxtacortical, hemispherical and bilateral nodular images, sometimes with diffusion restriction, as well as in the hypothalamic-chiasmatic region, medial thalamus, pontomesencephalic transition and right cerebellar hemisphere. There are also areas of discontinuous contracting delineating the ependymal surface of the lateral and III ventricles. It is associated with hypersignal on T2/FLAIR of the adjacent brain parenchyma, determining compression of regional cerebrospinal fluid (CSF). CSF examination showed cell count of 7,6/mm, protein 178 mg/dl, lactate 32 mg/dl, glucose 63 mg/dl and was negative infectious screening and oncotic cytology. He had negative serologies. Incisional biopsy was performed in cervical lymph node, compatible with Diffuse Large B-cell Lymphomas (DLBCLs). **Discussion:** Isolated central nervous system (CNS) relapse involving the brain parenchyma is a rare complication of non-Hodgkin lymphoma. Secondary CNS involvement (SCNSL) affects 5% of patients with DLBCL. Cranial nerve signs, headache, mental status and personality changes were the most frequent manifestations. The neuroimaging characteristics may mimic other neurologic disorders. The immunocompetent patients with primary CNS lymphoma (PCNSL) presents uniformly enhancing mass lesions without necrosis. Intraparenchymal mass and perilesional edema are the most common neuroradiological findings suggesting a SCNSL. SCNSL predominantly presented with multiple lesions, in contrast to PCNSL, while infratentorial and brainstem involvement were rare in SCNSL. In our case, multiple nodular lesions can mimic neurotuberculosis, nocardiosis and fungal infections. **Final Comments:** The suspicion of CSF lymphoma is a challenge in patients without previous diagnosis of aggressive lymphoma. Atypical MRI and CSF features make it difficult to confirm diagnosis. In this scenario, invasive strategies and investigating the primary site could be necessary.

Miscelânea

TL 1105081

ASEPTIC MENINGOENCEPHALITIS AND PERIPHERAL NEUROPATHY – A RARE ASSOCIATION OF NEUROSARCOIDOSIS

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Case Presentation: A 67-year-old female, began symptoms of muscular weakness on both legs in June 2021 that evolved to an ataxia of gait on a two months period. At Hospital admission, she had sensitive ataxia, hyporeflexia and mild weakness on lower limbs. There were neither cranial nerve palsies nor other abnormalities. After five days of Hospitalization, she presented generalized seizures and stupor, requiring mechanical ventilatory support. The liquor was clear with an open pressure of 15 cmH₂O, a protein level of 150 mg/dl, 74 mg/dl Glucose, and 14/mm³ white cell count (78% lymphocytes, 22% monocytes); VDRL, test for M. tuberculosis and PCR for multiple viruses were all negative and no bacteria or fungus were isolated on cultures. Brain and spinal cord MRI were normal. Chest Computed Tomography showed bilateral hilar and paratracheal adenomegaly. There was non-necrotizing granulomatous lymphadenitis on lymph node biopsy. These elements allowed us to diagnose neurosarcoidosis (NS). She treated with methylprednisolone 1g/day i.v. for 5 days followed by oral prednisone 5 mg/day until discharge and two antiepileptic drugs for epilepsy control. Neurological improvement was achieved along the first week with the patient going out the ventilator and becoming ambulatory with almost normal gait after thirty days. However, she developed osteomyelitis and died after clinical complications. **Discussion:** Sarcoidosis is a multisystem inflammatory disorder characterized by noncaseating granulomas in various organ systems, mainly the lungs and the lymphatic system. NS occurs in about 5–10% of patients with sarcoidosis and 50% of patients suspected of NS presents neurologic symptoms as first manifestation. Cranial neuropathy is the most common manifestation (mainly facial nerves) and is seen in 50–70% of cases. Peripheral nerve involvement occurs in 10% and brain and spinal cord lesions are less prevalent. Here we report an even rarer presentation of NS: aseptic meningitis of chronic evolution associated with peripheral neuropathy. Glucocorticoids are considered to be first-line therapy. It is associated with good efficacy and rapid clinical response. Severe presentations may require pulse-dose IV methylprednisolone, followed by a prolonged oral glucocorticoid tapering. **Final Comments:** This case illustrates a rare presentation of neurosarcoidosis that deserves adequate recognition and prompt treatment to avoid neurological sequelae or even the patient's death.

Miscelânea

TL 1105118

WERNICKE'S ENCEPHALOPATHY AND POLYNEUROPATHY FOLLOWING INTRAGASTRIC BALLOON PLACEMENT: A CASE PRESENTATION

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Case Presentation: A 42-year-old woman, with a previous medical history of hypertension, was brought to the emergency department with an acute confusional state that started five days prior to admission. No other symptoms were reported. The patient was submitted to an intragastric balloon (IGB) placement four months before the beginning of symptoms, with significant emesis following the procedure and retrieval of the IGB a week prior to admission. During the physical examination, upbeat nystagmus and clinical signs of polyneuropathy in the lower extremities were noticed. Concomitant to a thorough clinical and imaging evaluation, including the investigation, initial treatment and exclusion of potential causes, such as infectious and autoimmune encephalitis, the patient was treated with intravenous thiamine, which was followed by oral replacement. The diagnosis of Wernicke's encephalopathy was made clinically, although a specific imaging finding (mammillary body hemorrhage) was observed. She presented significant improvement regarding the cognitive impairment, as well as partial improvement of the ocular and motor symptoms. The patient is under physical therapy for rehabilitation and oral replacement of thiamine. **Discussion:** Wernicke's encephalopathy is a potentially severe acute neurological condition caused by thiamine (B1) deficiency. Although typically described as a consequence of chronic alcohol abuse, malabsorptive weight loss surgeries are emerging causes. Restrictive procedures, such as intragastric balloon positioning, are not commonly associated with the condition and scarce reports are present in the literature. Here, we describe the case of a patient that was admitted with an acute confusional state and was diagnosed with Wernicke's encephalopathy after IGB placement. Given the potential severity of the condition and necessity of early treatment to prevent major neurological deficits, clinicians should be aware of this condition. **Final Comments:** The case presented demonstrates a possible, although rare, cause of Wernicke's encephalopathy and polyneuropathy.

Miscelânea

TL 1105121

POST-TRANSPLANT LYMPHOPROLIFERATIVE DISORDER MYELORADICULOPATHY IN A KIDNEY TRANSPLANT RECIPIENT

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Case Presentation: 56-year-old Woman presented with weakness in the right leg and urinary retention. She had undergone living-donor kidney transplantation 21 years before for end-stage renal disease, use prednisone and mycophenolate mofetil (MMF). Started right lower back pain and evolved with muscle weakness of the right leg, worse in distal regions, associated with numbness and tingling. In two weeks she was unable to walk and desolved urinary retention. Physical examination revealed right lower extremity paresis and reduced sensation to pain and touch below L1 level. Magnetic resonance imaging (MRI) revealed cauda equina enhancement and a nodular formation on left nucleocapsular location. Cerebrospinal fluid (CSF) analysis presented with lymphomonocytic pleocytosis (50 cells), high protein (539 mg/dL) and lactate (82 mg/dL), reduced glucose (9 mg/dL), PCR Epstein-Barr virus (EBV) positive and immunophenotyping with mature clonal B lymphocytes. At this point, we made a diagnosis of Post-transplant lymphoproliferative disorder (PTLD) myeloradiculopathy. MMF was suspended and started chemotherapy (MADIT), Dexamethasone and Rituximab. After four months was observed an important reduction of de nodular lesion and normal CSF. Patient had partial improvement on muscle weakness. **Discussion:** PTLD is a rare disorder associated to solid organ, stem cell, or bone marrow transplantation. Pathogenesis is presumed to be associated with impairment in the cellular immunity, leading to proliferation of the lymphoid system. EBV is a relevant factor that plays causative and prognostic roles. The disease have a bimodal pattern of incidence, with a peak in the first and a second occurring after 7-10 years. Central nervous system (CNS) PTLD is considered a rare form of the disease, affects mostly supratentorial regions of CNS, specially the basal ganglia, and involvement of the spinal cord is rare. Diagnostic confirmation is performed with biochemical and cytological analysis of blood, CSF and tissue from biopsy (gold standard). Patterns of alterations in MRI may support suspicion. The treatment is focused in reduction of immunosuppression as the first step, radiotherapy and Rituximab with high response and chemotherapy as the standard option. **Final Comments:** CNS-PTLD represents a potential disabling cause, with specific investigation and treatment. Therefore, it shows the relevance of this discussion in the era of transplantation and immunosuppression.

Miscelânea

TL 1105142

LATE DIAGNOSIS OF SPINAL CORD LIPOMA: A CASE-BASED UPDATE

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Case Presentation: A 20-year-old woman is admitted with progressive paraparesis started two months ago. Magnetic Resonance Imaging (MRI) showed an expansive intradural lesion, with thoracic spinal compression (T7-T8), hyperintense in T1/T2 and hypointense in T2 with fat saturation (T2 FAT). In an axial view, the spinal canal was affected in >75% of its area. Biopsy revealed sections of mesenchymal neoplasia composed of mature adipose cells, confirming the diagnosis of myelolipoma. In the surgical procedure, partial resection was performed due to the wide adherence of nervous structures, with significant spinal decompression. There was complete neurological improvement, without post-surgical complications. **Discussion:** Non-dysraphic spinal cord lipomas compound less than 1% of spinal cord lesions. The diagnosis is usually made in children, without sex predilection. In this work, is presented a late diagnosis case of spinal cord lipoma. Clinical, imaging and surgical discussions are also done. Myelolipoma is a rare non-dysraphic spinal cord tumor, infrequently diagnosed in adults. The most accepted hypothesis of its origin determines an association with defects in neurulation during embryogenesis. Intramedullary lipoma patients usually present neurological deficits secondary to mass effect, having a subtle clinical presentation, slowly progressive myelopathy and a rapid neurological dysfunction after a long period of indolent tumor growth. The clinical manifestations include pain, paraparesis and paraesthesias, varying according to the lesion location and size, commonly the cervical-thoracic region. MRI is the most sensitive exam for diagnosis, with hyperintense lesions in T1 being characteristics of lipomas, confirmed by sequences with suppression of the fat signal. Treatment ranges from conservative methods, such as diet therapy, to aggressive surgical remove of the tumor. The main objective of surgery is to decompress adjacent structures, since lipomas cannot be completely resected. The prognosis of surgically treated patients is satisfactory, including pain reduction and neurological improvement. Tumor remissions have not been reported. **Final Comments:** Although rare, especially in adults, spinal cord lipomas must be known, as these tumors are well detected on MRI, their surgical approach is effective and, if left untreated, they evolve with sudden neurological worsening.

Miscelânea

TL 1105146

CENTRAL NERVOUS SYSTEM INFILTRATION ON MULTIPLE MYELOMA: TWO CASE PRESENTATIONS OF A RARE CLINICAL PRESENTATION

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Case Presentation: A - A 64 years old patient presented to the emergency department complaining of holocranial headache, unsteady gait and drowsiness. She had a history of multiple myeloma inactive for four years after an autologous stem cell transplantation presented to the emergency department. Her neurological examination showed proximal muscle weakness associated with exacerbation of all tendinous reflexes, demanding us to a complementary investigation with cranial neuroimaging. The magnetic resonance imaging showed leptomeningeal, ependymal, supra and infratentorial lesions with involvement of cranial nerves (subclinical at the moment) and parenchymal micronodular lesions. There were also lesions in the cervical level of the spinal cord associated with diffuse leptomeningeal impregnation, affecting the cervical, thoracic, lumbar regions and the cauda equina roots. The lumbar puncture showed monoclonal plasmacytes. Rescue chemotherapy was attempted, without therapeutic response. She died 5 months after the recurrence. B - A 58-year-old female patient diagnosed with multiple myeloma for 4 years since admission, with previous autologous bone marrow transplant presented to the Hospital with vertigo and headache. The magnetic resonance imaging showed lytic lesions on parietal bones with intense gadolinium uptake associated with significant temporal leptomeningeal thickening, inferring invasion of the central nervous system by the underlying disease. The initial cerebrospinal fluid puncture had 27 cells, 100% with neoplastic features. Rescue chemotherapy was performed, however the patient died 5 months after diagnosis. **Discussion:** Multiple myeloma is a hematological disorder caused by abnormal plasmacyte clones with production of monoclonal antibodies. The disease can cause some neurological symptoms secondary to peripheral nervous system disease, amyloidosis and spinal cord accometment. However, while some oncological diseases more commonly presents with brain metastasis or leptomeningeal invasion, these are rare on myeloma multiple patients. Invasion of the central nervous system by multiple myeloma denotes a poor prognosis, with some studies demonstrating an average survival of 3 months, similar to the patients presented in this case. **Final Comments:** These case are rare and severe presentations of multiple myeloma. Knowledge of such manifestations and detailed investigation allows an accurate management.

Miscelânea

TL 1105150

POST-RADIOTHERAPY UNILATERAL HYPOGLOSSAL NEUROPATHY: A CASE PRESENTATION

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Case Presentation: A 54-year-old man had development of involuntary movements of his tongue started one month before the first neurologic evaluation. Those movements were intermittent occurring around ten times a day. One year prior, he presented an airway obstruction that led him to tracheostomy and the diagnosis of invasive squamous cell carcinoma at the right amygdala. He underwent 35 sessions of radiotherapy and was considered cured. His medical history was also relevant for systemic hypertension, chronic kidney disease (due to systemic hypertension) and hepatitis C. He was a former smoker, quitting the habit about 30 years prior. At neurologic examination, tongue fasciculation was observed along with right sided deviation at protrusion. A CT brain scan excluded cancer recurrence. An electroneuromyography was performed and disclosed chronic axonal involvement of the hypoglossal nerve, with the presence of fasciculation, probably related to actinic neuropathy. Low-dose carbamazepine treatment was started and by the dose of 200mg twice a day, the patient had a complete improvement of the fasciculations. Radiation-induced lower cranial nerve neuropathy is a rare complication of radiotherapy. It usually starts around five years after radiation and bilateral involvement has been described. The most affected nerve is the hypoglossal, due to its long and tortuous course from the brainstem to the tongue. The most important risk factor for lower cranial nerve neuropathy is a large total radiation dose (more than 70Gy). No treatment has already been proved as effective, but there are some case presentation on positive outcomes with high-dose steroids. There is also some data about the use of carbamazepine as a membrane-stabilizing drug that may reduce nerve hyperexcitability. Our case presentation an earlier lower cranial nerve neuropathy due to radiotherapy with a very good outcome with low dose of carbamazepine. Cancer patients are surviving much longer nowadays, and it is extremely important to diagnose the treatment-related complications, firstly because of their impact on the patients' quality of life and secondly they may shorten patients life span.

Miscelânea

TL 1105207

OSMOTIC MYELINOLYSIS ASSOCIATED WITH PSYCHOGENIC POLYDIPSIA: CASE PRESENTATION DOUGLAS MARQUES ZARATINI, FRANKLIN TERTULINO FREITAS, ALEX SOARES SOUZA SOUZA

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Case Presentation: Patient with psychogenic polydipsia evolving with hyponatremia, correction of serum sodium levels, and consequent episode of altered level of consciousness. Years later, magnetic resonance imaging showed features of extrapontine myelinolysis. Such lesions were distributed exclusively in the corticosubcortical regions, mainly in the temporal region of the right hemisphere, but also affecting the right parietal region and the left temporal region. **Discussion:** Osmotic myelinolysis (OM) is classified into 3 groups: central pontine myelinolysis (CPM) without extrapontine involvement, extrapontine myelinolysis (EPM) without the involvement of pontine sites, and a form of central pontine myelinolysis with extrapontine involvement (CPM+EPM). The extrapontine form affects regions such as the putamen, thalamus, caudate nucleus, external capsule, and, more rarely, corticosubcortical regions. Isolated cases of corticosubcortical involvement are extremely rare in the literature. The clinical presentation of patients affected by this disease is quite variable, and the use of magnetic resonance imaging is extremely important to confirm the diagnosis. This pattern of exclusive corticosubcortical involvement was identified and well analyzed in an autopsy study, but it lacks reports on its radiological features. **Final Comments:** As the clinical presentation of OM is nonspecific and the radiological aspect can be characteristic, knowing the different imaging patterns is of paramount importance to narrow the differential diagnosis for the various professionals who may face this type of disease, such as neurologists, psychiatrists, emergency physicians, intensivists, among others. It is hoped that this case presentation will provide an expansion of knowledge about this rare condition, generating an eventual diagnostic and therapeutic optimization for future cases.

Miscelânea

TL 1105237

PROLONGED UNILATERAL MYDRIASIS SECONDARY TO CHRONIC IPRATROPIUM BROMIDE NEBULIZATION IN THE OUTPATIENT SETTING: CASE PRESENTATION

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Case Presentation: A 42-year-old female was evaluated at our Neurology service due to blurred vision and dilated pupil in the right eye for three years. There were no pain, diplopia, nor other associated symptoms. Visual complaints and pupil diameter both fluctuated throughout the months, without correlation to any factors. She had hypertension, pre-diabetes, and asthma. Her current medications were losartan and inhalatory combination of formoterol and budesonide, the latter started last month. She had been using salbutamol and ipratropium bromide nebulization since childhood; five years ago, frequency increased to 8-hourly courses every day. She had suspended its use two weeks before consultation, as asthmatic symptoms were improving, and subsequently perceived a discrete pupillary reduction. At examination, there was right mydriasis with absent direct and consensual photomotor reflex (Figure 1). Visual acuity, visual field, direct ophthalmoscopy, and extrinsic ocular motricity were unremarkable, as well as the remaining neurological examination. She had been evaluated by several physicians, and submitted to ophthalmological tests, laboratory work-up, and neuroimaging studies (i.e., computed tomography and magnetic resonance imaging with angiography), without any findings. Ipratropium suspension was reinforced. **Discussion:** Some reports have established the relationship between unilateral mydriasis and nebulized ipratropium bromide in intensive care patients. It acts topically in the pupillary sphincter muscle by blocking cholinergic stimulation. Ill-fitting face masks, especially in patients with facial asymmetries, are attributed as the major cause for unilateral presentation. Inadequate self-medication and chronic misuse of face masks could explain this finding in the outpatient setting. Physical examination, complementary work-up, and follow-up are essential for differential diagnosis, especially with Adie's pupil. Pilocarpine test is also helpful. In most reports, exposure is short and recovery fast after suspension. As the patient had five years of continuous exposure, a longer time to resolution could be expected. **Final Comments:** The present report stresses the importance of an extensive review of patient history, especially in an already over investigated patient. As far as we know, this is the first case in which chronic exposure to ipratropium bromide in the outpatient setting caused a prolonged unilateral mydriasis.

Miscelânea

TL 1105272

UNRESECTABLE HIGH-GRADE PONTINE GLIOMA IN AN ADOLESCENT: A CASE-BASED UPDATE

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Case Presentation: A 14-years-old female has presented a history of sporadic, frontal headache, triggered by movements, associated with vomiting, photophobia and osmophobia for 3 years. There are no reports of similar illnesses in the family and her parents are not consanguineous. The patient developed a visual clouding 7 months ago, which evolved to persistent diplopia and macular edema 2 months after. The symptoms had important intensification 4 months ago, requiring Hospitalization. On admission, she had an episode of convulsive crisis and sensorium lowering, so orotracheal intubation was performed. Skull Magnetic Resonance Imaging (MRI) showed an expansive, infiltrative lesion with imprecise limits, with an epicenter in the pons and extension to the midbrain, medulla and cerebellar peduncles, with a compressive effect on the fourth ventricle and involvement of the basilar artery. In the evolution, convulsive episodes were installed again for a week, and the neurosurgical evaluation did not indicate the possibility of surgical resection or biopsy, thus being transferred to the palliative radiotherapy unit. **Discussion:** Gliomas represent about 24% of primary tumors of the central nervous system, 80% of which are malignant. In general, they are histologically classified into low grade (I and II) and high grade (III – anaplastic astrocytomas/anaplastic oligodendrogliomas and IV – glioblastoma), in addition to molecular criteria. Clinical aspects depend on tumor location or compressive effect and may include focal neurological deficit, headache, nausea, vomiting, seizures, intracranial hypertension and visual changes. Contrast-enhanced MRI is the imaging exam of choice, showing heterogeneous gadolinium enhancement, with non-enhanced signal changes in the T2 images (infiltrative tumor), in addition to edema. Evidence of hemorrhage or restricted diffusion may also be seen. Confirmation comes from the pathological examination of the resection or biopsy. Treatment depends on the degree of cancer, ranging from chemotherapy, radiotherapy and surgical resection. Cases where the brainstem is largely involved are rare and unfeasible for resection, remaining palliative care. **Final Comments:** Early diagnosis and treatment of high-grade gliomas remains a challenge, especially in centers with limited financial resources. Being aware of the presentations of these tumors is necessary for proper and rapid management.

Miscelânea

TL 1105299

HEMATOMYELIA MIMICKING LONGITUDINALLY EXTENSIVE MYELITIS: A CASE PRESENTATION

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Case Presentation: A 54-year-old woman, admitted in 2016 due to sudden severe neck pain with no history of trauma, that radiated to the left side of the body, associated with decreased ipsilateral strength and sensitivity. On admission, she had hemiparesis, hyperreflexia with Babinski's sign and hypoesthesia of left hemibody. Spinal MRI showed T2/FLAIR C1-T2 hyperintense lesion, with extensive vasogenic edema, mild hyposignal on T1-weighted gradient-echo (GRE) inside the spinal cord and gadolinium-enhancement near the spinal canal. CSF analysis with 15 leukocytes (lymphomononuclear cells predominance), 135 proteins, 79 glucose and 11275 red blood cells (puncture accident). Anti-AQP4 antibody was negative, as well as other infectious and inflammatory screening tests. The condition was attributed to a seronegative longitudinally extensive myelitis with probable inflammatory etiology and started 5 days of high-dose steroid treatment, followed by 5 sessions of plasma exchange, with no improvement in symptoms. She was maintained with prednisone and azathioprine, suspended 3 years later due to myelotoxicity. In 2019, new MRI scans showed hyposignal on GRE sequence in the subcortical region of the left inferior parietal lobe, thalamus, bulbo medullary transition and in the C1-T1 spinal cord, suggesting chronic hemorrhage. Idiopathic hematomyelia was the hypothesis made and the patient was kept without immunosuppressive, remaining stable until today. **Discussion:** Hematomyelia is a hematoma in the spinal cord. There are few published case presentation about vascular myelopathy caused by this entity. The etiology can be traumatic (most common) or non-traumatic. Idiopathic hematomyelia is the term used when spontaneous intraspinal bleeding occurs without a clear etiology. Myelopathy occurs due to the mass effect caused by the hematoma, that acutely obstruct the cerebrospinal fluid and blood flow within the spinal cord. The symptoms presented are intense and sudden pain in the neck and back, evolving rapidly (in hours to days) to myelopathic deficits. Recent studies have shown that patients initially diagnosed with idiopathic transverse myelopathy often have alternative diagnoses, with vascular etiology being one of the most common. **Final Comments:** Recognize that hematomyelia can cause extensive spinal cord injury and can mimic an autoimmune longitudinally extensive myelitis is of fundamental importance, allowing for prompt recognition and management for better outcomes.

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TL 1105301

DESMOPLASTIC MEDULLOBLASTOMA IN AN ADULT PATIENT: CASE PRESENTATION

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Case Presentation: A 28-year-old female patient underwent a biopsy of a spinal cord (SP) and cerebellar tumor. Grossly, the fragments from both sites were brownish, firm and irregular. The histopathological examination of the two tumors showed an undifferentiated neoplasm with cells with hyperchromatic nuclei and high biphasic cellularity, ranging from compact to loose. An immunohistochemical panel was performed with synaptophysin, chromogranin, enolase, CD45, TTF1, PAX5, CD99, GFAP and Ki67 cell proliferation index (Ki67-CPI). The results showed positivity for enolase and focal positivity for synaptophysin and chromogranin. The Ki67-CPI was 70%, being lower in nodules. **Discussion:** Medulloblastoma (MB) is a primitive neuroectodermal tumor. Although MB is the most common brain neoplasm in children, it is very rare in adults, with an estimated incidence of 0,6 cases per million per year. Normally, it emerges in posterior fossa structures, specially the cerebellum, and tends to disseminate through the cerebrospinal fluid pathways, being especially prone to leptomeningeal spread. Rarely, other malignant tumors with small cell morphology can be found, including small cell glioblastoma, which can typically be excluded by immunohistochemistry, using lineage markers or entity specific stains. The desmoplastic variant is a subtype of MB with medium to high risk. Our case presents the rare manifestation of a desmoplastic MB in cerebellum and SP in the same patient. Intramedullary spinal dissemination is rare, with very few cases in the available literature. We find no previous scientific report of a desmoplastic MB in SP, but there are seven documented cases of small cells MB in SP, with two of them appearing simultaneously to an extramedullary component, as in our report. **Final Comments:** In adulthood, MB has a higher proportion of desmoplastic histological features, different proliferation rates and a tendency for late relapses. The therapeutic role of adjuvant chemotherapy in adults is still not entirely known due to a shortage of prospective studies. Surgical resection plays a central role in treatment and should be regarded individually, according to the status of the patient's disease. Thus, further studies on the occurrence of MB in adults and on SP involvement are needed, in order to better understand prognostic factors and the best treatment options.

Miscelânea

TL 1105318

CEREBELLAR SUBEPENDYMOMA IN AN ADULT PATIENT: CASE PRESENTATION

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Case Presentation: A 29-year-old male patient underwent a biopsy of a cerebellar tumor. Grossly, the fragments obtained were irregular, brown and soft. Microscopically, circumscribed glial neoplasm was observed, formed by clustered nuclei, without atypia in the midst of occasional microcysts with fibrinoid content. No mitotic or necrotic figures were identified. The association between clinical aspects, macroscopic features and microscopic findings defined the diagnosis of subependymoma, a grade I tumor according to the 2021 World Health Organization (WHO) classification. **Discussion:** Subependymomas are rare, well-circumscribed, slow-growing and often asymptomatic masses with an ependymal origin that are most commonly found in the ventricular system, usually in the fourth or lateral ventricle. Its rarity is reflected in the scarce number of reports in the scientific literature. They represent 0.2 to 0.7% of all intracerebral tumors and can manifest with cerebrospinal fluid obstruction, causing increased intracranial pressure. The intraparenchymal site is very rare and other tumor locations include the septum pellucidum, foramen of Monro and, less commonly, the spinal cord. Most tumors are found incidentally at autopsy. Symptomatic subependymomas most often affect middle-aged patients and may rarely manifest with intratumoral hemorrhage or seizures. Surgical exploration and biopsy are necessary to distinguish the appropriate treatment. Usually, tumor excision and radiotherapy are strongly recommended and often corrective, with tumor recurrence being quite rare. **Final Comments:** Our case intends to register this rare case of subependymoma, recommend more studies to understand the possible origin of this tumor and highlight this possibility for the diagnostic range in clinical picture of dizziness, emphasizing the importance of histopathology in defining the best course of action.

Miscelânea

TL 1105340

FOIX-CHAVANY-MARIE SYNDROME: A CASE PRESENTATION

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Case Presentation: J.J.S, male, 61 years old. This patient was referred with an insidious condition of vocal alteration, difficulty in swallowing and sporadic choking since childhood. From the age of 40, he noticed an intensification of symptoms, presenting lower and slurred speech associated with significant dysphagia for solids and liquids. Neurological examination showed involvement of multiple cranial nerves with hypoesthesia and diparesis facial, reduction of the nausea reflex, dysphagia, dysarthrophonia and paresis of the tongue musculature. Despite the paralysis of the orofacial voluntary muscles, they are preserved when evoked by emotion, such as the act of smiling. He also had mild cognitive impairment, as evidenced by cognitive tests. On MRI, bilateral atrophy was observed in the opercular regions. **Discussion:** Foix-Chavany-Marie syndrome, also known as opercular syndrome, occurs due to bilateral cortical injury in the anterior opercular region. It is a rare suprabulbar palsy condition with dissociation in voluntary and spontaneous (emotional) movements. The clinical presentation reflects bilateral paralysis of voluntary movements of muscles innervated by NNCC V, VII, IX, X and XII, as in this case. However, the initial involvement of impairment in chewing, tongue movement and speech apraxia, but without language impairment, stands out. The main cause is vascular, but cases caused by neuroinfections, traumatic brain injury, neuroinflammatory, demyelinating and neurodegenerative diseases have already been described. On MRI, asymmetric cortical atrophy is seen, predominantly in the lower frontal region, especially in the opercular region, as in this case. **Final Comments:** This case shows that progressive loss of orofacial voluntary motor skills and apraxia of speech with preservation of emotionally evoked movements are the hallmarks of this rare syndrome. The exact diagnosis of the cause is not always possible and the treatment involves a multidisciplinary team. Due to the significant involvement of the facial muscles, malnutrition is the main consequence and alternative feeding methods are necessary.

Miscelânea

TL 1105347

INTRADURAL DISC HERNIA: RARE CONDITION

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Case Presentation: This is a case presentation with an investigative and descriptive methodology about clinical case of intradural disc hernia, a rare condition that brings a lot of information that can enrich the scientific community – A 65-year-old male patient with a history of low back pain without clinical improvement. Lumbar arthrodesis was performed (L3-4-5-S1). Over the months, he evolved with disease of the adjacent level. Neuroimaging exams such as Magnetic Resonance Imaging (MRI) suggested disease of the adjacent level with extruded hernia. Extensive arthrodesis was then performed. The disease evolved with a degenerative behavior, with disease of the adjacent level and later with junctional kyphosis and paraparesis – requiring a new surgery to understand the arthrodesis again, in which a transoperative diagnosis of intradural hernia was made. Microsurgical treatment by arthrodesis was then performed, spinal decompression with wide laminectomy and intradural hernia resection was performed. Postoperatively, he showed significant improvement, returning to ambulation. He later evolved with T8-9 spondylodiscitis that culminated in myelopathy. Intradural disc herniation is a rare complication: 0.04%. In this way, presenting and discussing the diagnostic (neuroimaging exam) and therapeutic (neurosurgical) procedures associated with the unusual epidemiological picture enriches the literature and medical discussion. There is a higher prevalence in males, mainly affecting the lumbar region. The diagnosis of the disease is difficult, with several differentials in the neuroimaging, and immediate surgery is essential to prevent the progression of the pathology and minimize the patient's discomfort, as well as post-surgical neurofunctional follow-up for rehabilitation. The present report discusses a lumbar degenerative disease that, after clinical and intraoperative investigation, was diagnosed as an intradural disc herniation, a rare and difficult to diagnose condition. Therefore, the data presented tend to contribute to the collection of clinical history of the disease, enabling the possibility of early and effective diagnoses.

Miscelânea

TL 1105349

NEURO CUTANEOUS MELANOSIS WITH CENTRAL NERVOUS SYSTEM INVASION

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Case Presentation: An 18-year-old female patient with comorbid cutaneous melanosis was admitted to the emergency room reporting headaches for more than two months. After neuroimaging findings, acute hydrocephalus was observed and an emergency external ventricular shunt was performed. Therefore, endoscopic third ventriculostomy approach was performed for arachnoid biopsy of the pre-pontine cistern, followed by septostomy. In the biopsied pathological analyses, the presence of melanocytic cells was then confirmed. In the immediate postoperative period, there was clinical improvement, however, six months later there was decompensation of condition, with reduced visual acuity, with failure of the third ventriculostomy and the need for a new bypass – however, due to the presence of disseminated skin lesions in the abdominal region, it was then ventriculoatrial shunt was performed by puncture, resulting in clinical improvement. One year after this last event, the patient developed tetraparesis with plural predominance. The entire central nervous system was then reinvestigated, where several syringomyelia and spinal cord injuries were identified. After that, the patient entered palliative and exclusive care, however, thirty days later, she died. Neurocutaneous melanosis is a rare syndrome that is associated with a cutaneous manifestation and even more rarely invades the central nervous system. This condition can evolve in a benign or malignant form, therefore, when presented early in life, it is associated with a high mortality rate. Although skin lesions pose a risk of melanoma, the most likely source of morbidity and mortality comes from “benign” melanocytic proliferation to central nervous system, as presented here. Common neurologic manifestations usually arise in the first few years of life, however, continued melanocytic proliferation can lead to other findings, as discussed here. This way, the present report discussed the clinical history of a young woman with the rare form of neurocutaneous melanosis with evolution to the brain and spinal cord central nervous system. The description of the clinical history of rare conditions enriches knowledge and makes it possible to guide other diagnostic and therapeutic approaches.

Miscelânea

TL 1105377

CERVICAL SPONDYLOTIC MYELOPATHY MIMICKING SUBACUTE COMBINED DEGENERATION: A CASE PRESENTATION

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Case Presentation: A 60-year-old woman with rheumatoid arthritis presented with a 1-year history of difficulty holding objects due to numbness and weakness of the upper limbs. Three months before admission, the symptoms progressed to the lower limbs, and she required support to walk. On physical examination, there was a spastic tetraparesis with extensor plantars and decreased vibratory and position sense in all limbs. The patient also experienced a prominent increase in limb weakness with neck flexion (McArdle sign). The cervical MRI revealed spinal canal stenosis at the level of C2-C8 and symmetric bilateral high signal within the dorsal columns (inverted “V” sign). Serum vitamin B12 was 243 pg/ml (normal 180–500 pg/ml) and serum methylmalonic acid was 0.13 umol/L (normal < 0.40 umol/mL). Flexion–extension radiographs revealed aggravation of cervical misalignment with neck flexion. A standard anterior cervical discectomy was performed and there was a marked improvement in symptoms at 1-month follow-up. The typical MRI finding of subacute combined degeneration (SCD) is a symmetric bilateral high signal of the dorsal columns that begin in the upper thoracic region, with ascending or descending progression. The lateral corticospinal tracts and lateral spinothalamic tract may also be involved. On the other hand, MRI of spondylotic myelopathy reveals a circumferential enhancement sparing spinal cord gray matter on axial images, usually located beneath the site of maximum stenosis (pancake-like sign). We describe a case of compressive myelopathy mimicking clinic and radiologic characteristics of SCD. In addition, our patient also presented with McArdle sign, which is considered highly specific for the diagnosis of multiple sclerosis (MS). In our patient, the normal level of methylmalonic acid and the rapid improvement after surgery ruled out SCD. The clinical features were not compatible with MS. This is a case presentation shows that spondylotic myelopathy can have a great spectrum of clinical and radiographic features and can mimic conditions such as SCD.

Miscelânea

TL 1105378

ISOLATED THIRD CRANIAL NERVE PALSY WITH PUPIL SPARING IN PITUITARY APOPLEXY – CASE PRESENTATION

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Case Presentation: We present the case of 54-year-old man with a 5-day-history of sudden onset, severe, pulsatile headache that was worse in the left retroocular region and associated with nausea and vomiting. Two days after the onset, he developed binocular diplopia and left eyelid ptosis. Physical examination showed complete isolated left third cranial nerve (3CN) palsy but the pupil was spared. Head MRI showed a 2.1x1.4x2.1cm pituitary adenoma, without significant cavernous sinus compression. He also had normal TSH with low free-T4 level; low prolactin and testosterone levels; and normal ACTH and cortisol levels. Cerebrospinal fluid had 13 white blood cells/mm³ and 116mg/dL protein. Based on the clinical history and complementary exams we made a diagnosis of pituitary apoplexy (PA). He was transferred to neurosurgery. **Discussion:** PA is a rare medical emergency caused by abrupt hemorrhaging and/or infarction of the pituitary gland, generally due to a pituitary adenoma. Sudden and severe headache is the main symptom and can be associated with ocular palsy in 52% of the cases. The third 3CN is often the first and the most affected cranial nerve, and is impaired in 50% of the cases, but isolated 3CN palsy is a rare manifestation. Complete nerve palsy characterized by ptosis, impaired pupillary constriction and ophthalmoparesis is the most common clinical presentation, but pupil sparing may occur in 11% of cases of isolated 3CN palsy. Although complete 3CN palsy with pupil sparing is typical of ischemic lesions, compressive lesions may spare the pupil when the pressure of the lesion is evenly distributed and allows the relatively pressure-resistant, smaller-caliber pupillomotor fibers to escape injury; alternatively, the lesion may compress only the inferior portion of the nerve, sparing the dorsal pupillomotor fibers. In addition, several mechanisms have been proposed to explain the occurrence of 3CN palsy in PA, such as direct invasion of the cavernous sinus; compression without invasion, since the nerve is located horizontally in the same plane as the pituitary gland in the lateral wall of the cavernous sinus; and compression of the vasa nervorum of the nerve due to increased pressure. In our opinion, the last mechanism may also explain the sparing of superficial pupillomotor fibers. **Final comments:** PA should be considered an important differential diagnosis in patients presenting with headache and 3CN palsy, even if isolated and with pupil sparing.

Miscelânea

TL 1105379

SPHENOCALVAL INTRAOSSEOUS LESION MIMICKING ACUTE OPTIC NEURITIS: A CASE PRESENTATION

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Case Presentation: This 17 year old patient was referred to the Emergency Room for Neurological evaluation due to acute painful right eye vision loss, involuntary ipsilateral eyelid movements and pulsatile occipital pain. Pain was also triggered by eye movement. It was obtained pain relief with good response to simple analgesics, despite her past medical history of Migraine. Additionally, physical examination revealed decreased sensitivity at right V1 trigeminal branch and ipsilateral reduced visual acuity. Brain and Orbits MRI showed T1 and T2 hyperintensity in the medium clivus, with signs of bone destruction and without gadolinium enhancement or DWI restriction. A contrast Brain CT was also performed which showed no signs of vascular stenosis or venous thrombosis, even though some bone destruction could be noticed. Given the pain improvement, our patient was referred for follow-up at the clinic and possibly elective biopsy. **Discussion:** Depending on the tumor growth direction and speed, infiltrative sphenocaval lesions can mimic acute optic neuritis, including intraosseous lipoma as a distinctive diagnosis to be considered. Although biopsy was not yet performed, MRI seems to be very useful for evaluating this condition. **Final Comments:** Intraosseous lipoma is a hamartoma rarely manifested in the sphenocaval topography and often asymptomatic. MRI can be very suggestive, however biopsy is essential to properly diagnose and manage this condition.

Miscelânea

TL 1105387

METHOTREXATE NEUROTOXICITY WITH ATYPICAL RADIOLOGIC FINDINGS

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Case Presentation: A 18 year old man with infiltration of the central nervous system (CNS) by T-cell acute lymphoid leukemia was treated with intrathecal chemotherapy (ITC) with dexamethasone, cytarabine and methotrexate (MTX). Two weeks later, severe lumbar arachnoiditis was diagnosed, so ITC was interrupted and an Ommaya reservoir was implanted. After 10 days, he developed aphasia and apathy. Brain magnetic resonance imaging (MRI) revealed white matter hyperintensities in T2/FLAIR (fluid attenuated inversion recovery)-weighted images in both frontal lobes, in corpus callosum and around the Ommaya reservoir, with peripheral and irregular gadolinium enhancement and areas of diffusion-water restriction. Leukemia infiltration and reservoir-associated infection were considered as possible diagnoses. CSF analysis had no neoplastic cells and cultures were negative. Despite the achievement of hematologic recovery at the end of that chemotherapy phase, he persisted with neurologic deficits. A few days later he was treated for clinical status epilepticus and a new brain MRI showed progression of the lesions. Spectroscopy had a pattern suggestive of neoplastic infiltration, but considering the normal CSF analysis and the systemic remission of leukemia it was improbable. So, a brain biopsy was performed. Fragments of deep frontal lesions showed edema, necrosis and histiocytic inflammatory reactional areas, without lymphoid atypical infiltrates. Histokymic fungi methods tests and JC virus search were negative. The morphology observed was compatible with MTX neurotoxicity. Given the temporal relationship of the lesions with the drug administration, although there were some atypical MRI findings for this condition, MTX was withheld for 2 months. Gradually, he had neurological improvement with complete resolution of aphasia. Follow up MRI showed a decrease of lesions. **Discussion:** MTX leukoencephalopathy is rare and occurs in about 2 to 3,8% with MTX high dose treatments. It is commonly transient and improves with cessation of the drug. A subsequent MTX dose after the resolution of the acute phase is habitually well tolerated. **Final Comments:** MTX neurotoxicity typically presents as peripheral neuropathy, but it is a main differential diagnosis in CNS lesions in leukemia patients receiving ITC, particularly if neuroinfection and secondary neoplastic invasion were excluded.

Miscelânea

TL 1105429

BRAIN METASTASIS DUE TO RETROPERITONEAL LEIOMYOSARCOMA: A CASE PRESENTATION

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Case Presentation: We present the case of a 53-year-old man with biopsy-proven retroperitoneal leiomyosarcoma (LMS) and pulmonary metastasis. Chemotherapy was started, with stability of the abdominal lesion and regression of the lung metastasis. Nine months after the diagnosis, he presented with progressive headache and an episode of epileptic seizure. Head MRI showed metastatic brain lesion in the right parietal lobe. He underwent surgical resection followed by stereotactic radiosurgery (total dose of 2000 cGy) and pathological examination confirmed LMS metastasis. Control head MRI was performed 1 month after radiosurgery and showed a new right frontal lesion, which was also managed with stereotactic radiosurgery (total dose 3000 cGy). After a period of 6 months of clinical and radiological stability, a new lesion close to the 3rd ventricle appeared, the previous brain lesions increased, and the systemic disease progressed. He evolved with global functional decline, and palliative care was then instituted. **Discussion:** Sarcoma refers to a cancer that arises from cells of mesenchymal origin while LMS is a rare histological subtype that originates from smooth muscle. It is the second most common soft-tissue sarcoma to affect the retroperitoneum. Sarcoma metastasis to the central nervous system is uncommon, occurring in 1-8% of the cases, with LMS being the most common subtype. Among LMS of all sites, the retroperitoneal LMS has the worst prognosis and about 80% - 87% of these patients die within 5 years. LMS responds poorly to chemotherapy and radiation, being considered a resistant cancer; thus the best results come from early surgery, when feasible. Retroperitoneal LMS is often fatal owing to local recurrence and distant metastases. As our knowledge of retroperitoneal LMS brain metastasis is based on a few cases reports, the clinical management and adjuvant therapy of these tumors remain based on individualized decision-making. **Final Comments:** This is one of the few cases described in the literature of brain metastasis from retroperitoneal LMS treated with surgical resection and stereotactic radiosurgery and, to the best of our knowledge, the only case described with documentation of the treatment of the disease progression. Reports like this aim to contribute to the development of a body of evidence that will guide the management of similar cases.

Miscelânea

TL 1105472

PROGRESSIVE MULTIFOCAL LEUKOENCEPHALOPATHY: A CASE PRESENTATION IN A IMMUNOCOMPETENT PATIENT

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Case Presentation: A 23-years-old men, consanguineous parents, present vertigo and left visual cloudiness in November 2020 evolving in 07 days with paraesthesia and weakness in the right hand and dizziness. Pulse therapy was done for 05 days without improvement. He evolved with incoordination, severe dysarthria, right hemiplegia. CSF and serology were performed without changes; MRI of the skull that showed defects of semiovarian centers, bilateral peritrial, posterior horns of lateral skulls, splenium of the corpus callosum and cerebellar with hypointensity in T1 and hypersignal in T2/FLAIR in the white substance of the cerebral tissue. Immunoglobulin was performed without improvement of the condition, with disease progression. Investigation of the immune system without alterations, positive JC virus, stereotactic biopsy and anatomopathological examination characterizing Progressive Multifocal Leukoencephalopathy (PML). Despite therapy, the patient passed away within 7 months. **Discussion:** PML is a central demyelinating disease, related to JC virus reactivation, usually occurring in immunodepressed patients and less usual in immunocompetent patients. The physiopathology is based on the oligodendrocytes and astrocytes lesions of the subcortical hemispheric white matter and cerebellar peduncles, and may also affect a gray matter of the basal ganglia. The prognosis depends on variables such as the degree of contrast enhancement on neuroimaging, CD4 T lymphocyte count and neurological recovery. **Final Comments:** In the case under analysis, there was a peculiarity of the pathology affecting a previously healthy individual, with no history of Acquired Immunodeficiency Syndrome or use of medications that could adjust his immunological competence, making the diagnosis even more challenging. Restoration of the adaptive immune system is the main objective given the fact that the disease still has no specific treatment.

Miscelânea

TL 1105503

PRIMARY SARCOMA OF THE CENTRAL NERVOUS SYSTEM – CASE PRESENTATION

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Case Presentation: A 28-years-old woman, presented with headache in the right frontotemporal region starting in July 2019, evolving with visual acuity changes, dizziness and paresthesia in the left upper limb (LLE). Neurological examination showed only bilateral papilledema. Magnetic Resonance Imaging (MRI) of the skull showed a right frontoparietal dural lesion measuring 5.1x4.4x3.8cm, heterogeneous with T1 hypointensity, intense contrast enhancement and increased rCBV inside. She underwent surgical resection in September 2019 with no residual lesion. Histopathological examination identified features suggestive of high-grade fusocellular sarcoma, with immunohistochemistry demonstrating a Ki-67: Positive 50%. During the planning of radiotherapy, the patient progressed with recurrence of complaints, neuroimaging showed disease progression, and a new surgical resection was performed in January 2020. The patient underwent adjuvant three-dimensional conformal radiotherapy (dose 60Gy). It evolves with recurrence of headache and paresis in LLE, in addition to focal motor perceptual onset crisis. Cranial MRI showed progression of the lesion in the same topography. She underwent a third procedure in June 2021, with wide resection, but with a residual lesion. Stereotactic radiosurgery was performed in a residual lesion (dose 25Gy) in September 2021. During clinical follow-up, a protuberance in the surgical wound was identified and MRI of the skull confirmed the progression of the disease with exteriorization through the frontal bone. A new surgical approach was performed in January 2022 with lesion resection, in addition to subdural empyema drainage and surgical wound cleaning. Antibiotic therapy was started, but the patient evolved unfavorably and died in March 2022. **Final Comments:** Primary sarcoma of the central nervous system (CNS) is a rare neoplasm. It has been hypothesized that these neoplasms originate from a pluripotent mesenchymal lineage within the dura or leptomeninges, extending into the CNS parenchyma. Surgical resection along with chemotherapy and radiation have historically been the cornerstone treatment strategy for CNS sarcoma. Future studies should be focused to understand the histopathological subtypes of primary CNS sarcoma to use different chemotherapeutic agents which be helpful to give patients long survival and best choice of treatment.

Miscelânea

TL 1105525

NEUROPLASTICITY ON DYKE DAVIDOFF MASSON SYNDROME: A CASE PRESENTATION

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Case Presentation: A 57-year-old woman diagnosed with Dyke Davidoff Masson Syndrome presented with cognitive impairment and secondary epilepsy since her childhood. Orphan of both parents as a young infant, her prenatal history is therefore unknown. The patient lives in a public psychiatry institution, where therapy, medical assistance, artistic and ludic activities are offered. The radiological findings evidenced an important unilateral atrophy of the left cerebral hemisphere and an ipsilateral skull hypertrophy. Despite significant brain damage, the patient maintained only cognitive impairment and controlled seizures by the regular use of antiepileptic drugs, being independent for daily living activities. Her neurological examination showed Wartenberg's sign and hipotrophy in the right upper limb and reduction of motion in the same limb during gait. This favorable clinical outcome may be explained by the innate capacity of neuroplasticity. **Discussion:** The Dyke-Davidoff-Masson Syndrome is defined as an atrophy or hypoplasia of one cerebral hemisphere following a prior fetal or childhood insult (1). The etiopathogenesis is not clear, but it could be either a vascular insult during intrauterine life or acquired causes like trauma, infection, vascular abnormalities and intracranial hemorrhage in the perinatal period or shortly thereafter (2). Clinical features depend on the extent of brain injury and include hemiparesis or hemiplegia, seizures, mental retardation or learning disability, speech or language disorders and facial asymmetry (2). Diagnosis is settled by clinical history, neurological examination and radiologic findings. Treatment is symptomatic and aimed at controlling seizures and improving quality of life (3). Some patients evolve with minimum neurologic impairment considering the extension of the brain damage. It could be explained by the nervous system's ability to change its activity in response to stimuli by reorganizing its structure, functions, or connections after injuries (4). The basis of neural plasticity derives from synaptic plasticity that refers to changes in the strength of neurotransmission induced by activity experienced by the synapse in the past (5). **Final Comments:** This report illustrates a rare syndrome that has a wide spectrum of cognitive disabilities affecting multiple domains. We defend that neuroplasticity has a key role on its prognosis, making it possible for one part of the brain to take up the functions of the disabled part.

Miscelânea

TL 1105542

BELL'S PALSY AFTER CORONAVAC VACCINATION: A CASE PRESENTATION

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Case Presentation: Patient is a 66 year old woman, diagnosed with systemic arterial hypertension and type II diabetes, with blood glucose under control. She received the first dose of the CoronaVac COVID-19 vaccine on April 1st, 2021, and the second dose on April 22nd of the same year. Four days after receiving the second dose, the patient reports waking up with left side labial deviation, along with a painful sensation in her right eye. In addition, she exhibited muscle weakness on the right side of her face, causing difficulties in chewing food and swallowing liquids, which poured out of her mouth. Furthermore, the patient states hardship in articulating words. She sought a neurological evaluation on the same day, where peripheral facial palsy House-Brackmann V was noted. No further abnormalities were found in the evaluation, and no signs of Herpes infection were noted in her ear. A brain MRI was performed, with no acute alterations being revealed. She received a prescription of 60mg of Prednisone daily during ten days, together with physiotherapy. In a few months of treatment, the patient had a complete recovery of facial sensibility and motor function. **Discussion:** Peripheral facial palsy or Bell's palsy is an acute mononeuropathy that leads to paresis or paralysis of the upper and lower face. Several etiologies for Bell's Palsy have been reported, such as thermal shock, ischemia, infection and post-vaccination immune trigger reactions. However, there are still few studies on the relationship between peripheral facial paralysis and vaccination for COVID-19, especially with the CoronaVac vaccine. A Chinese study compared the risk of developing peripheral facial palsy between two different COVID-19 vaccines: Pfizer and CoronaVac. The study found an Odds Ratio of 2.385 for CoronaVac and 1.755 for Pfizer – the latter, however, was not statistically significant (CI=0.886–3.77 and p=0.11), not having a significantly higher relative risk than the control group. Therefore, the study concluded that there is an increased risk of developing Bell's Palsy with Coronavac. **Final Comments:** Peripheral facial paralysis can be one of the adverse events of vaccination for COVID-19. However, the risk of developing these effects is minimal and does not outweigh the benefit of getting vaccinated.

Miscelânea

TL 1105597

POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME (PRES): A CASE PRESENTATION IN A MALE PATIENT FROM THE NORTH'S INNER

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Case Presentation: E. B. D. S., a 50-year-old, brown male patient, born and resident in the interior of the Amazonas, was admitted in July 2021 with a history of generalized tonic-clonic seizure preceded by occipital pulsating headache events and sudden loss of visual acuity without pain on eye mobilization. He was referred to the capital emergency department with psychomotor agitation and mental confusion after more than 48 hours of the ictus. Brain Magnetic Resonance Imaging (MRI) was performed and showed signal alteration of the sub-cortical white matter bilaterally in the occipital lobes, which indicated the possibility of reversible posterior encephalopathy (PRES). Previous commemoratives of note included Systemic Arterial Hypertension (SAH) that was difficult to control, diagnosed during Hospitalization, and diabetic nephropathy. After discharge in August, the patient was oriented to drug treatment combined with outpatient neurological and nephrological follow-up; however, geographic and socioeconomic obstacles interrupted follow-up's consistency. **Discussion:** PRES is a clinical-radiological entity associated with a wide array of clinical presentations including visual disturbances, headache, seizures and altered mental status. There is no single mechanism that explains the development of PRES in all cases, and multiple nonexclusive mechanisms likely contribute. Hypertension is the most common precipitating factor, with endothelial dysfunction playing an important role. The cerebral imaging abnormalities are often symmetric and predominate in the posterior white matter. The combination of suggestive clinical manifestations and radiological criteria establishes the diagnosis of PRES. **Final Comments:** The national incidence for Pres is uncertain. Searching main databases, there is an evident discrepancy in the described cases' scarcity in the north compared to other regions. The interior is a neediness' portrait enhanced by obstacles from lack of integration between services which provide proper diagnosis, treatment, and especially follow-up care for this neurological disorder, as in this case presentation.

Miscelânea

TL 1105604

PEDUNCULAR HALLUCINOSIS SECONDARY TO CYCLOSPORINE PONTINE MYELINOLYSIS

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Case Presentation: A 54 years- old female patient, Hospitalized in our service with a diagnosis of acute myeloid leukemia, presented with vivid and complex visual hallucinations consisting mostly of her family members, but with size distortions. She had recently undergone a stem cell transplantation and was under immunosuppressive treatment with cyclosporine. She had a normal neurological exam and denied previous similar episodes of hallucinations of any kind. Her MRI showed diffuse T2 and Flair hyperintensities in pons, suggestive of osmotic demyelination syndrome. She had normal laboratory exams, including serum glucose and electrolytes levels. Due to her immunosuppressive status, CSF was also obtained for investigation of cellularity, biochemistry, cultures, VDRL, Xpert MTB/RIF and viral PCRs (HSV 1 and 2, EBV, HIV). The results were unremarkable. There was no history of ethillism or other metabolic disorder that could result in the MRI findings. Three days before the onset of symptoms her serum cyclosporine level was supratherapeutic. Therefore, the hypothesis of an osmotic demyelination syndrome induced by cyclosporine intoxication was made. Our patient had an atypical presentation, due to the lack of motor, cerebellar or cranial nerve involvement, but presenting only with complex visual hallucinations, compatible with peduncular hallucinosis. After a change in her immunosuppressive therapy and cyclosporine discontinuation, the patient evolved with progressive improvement of symptoms. **Discussion:** Peduncular hallucinosis (PH) is a rare clinical syndrome with dream-like visual hallucinations intruding normal consciousness, most commonly occurring after a midbrain or diencephalic lesion. The pathogenesis is explained by a disconnection between the ascending reticular activating system and retinogeniculocalcarine pathways, particularly at the level of the thalamus. PH occurs mostly after ischaemic infarction and tumoral lesions. Its occurrence after a immunosuppressor induced osmotic demyelination syndrome is rare, with very few cases reported until now. **Final Comments:** Complex visual hallucinations in an immunocompromised patient have a broad differential diagnosis and may require multiple complementary exams. We describe a case of PH secondary to cyclosporine-related pontine myelinolysis, which is potentially reversible with rapid identification and discontinuation of the drug.

Miscelânea

TL 1105606

FAHR SYNDROME SECONDARY TO HYPOPARATHYROIDISM: CASE PRESENTATION

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Case Presentation: A. J.M., 68 years old, history of falls associated with vertigo and syncope for a year. Also, report of behavioral change, visual hallucinations and aggressiveness for eight months, receiving a diagnosis of schizophrenia. No other comorbidities. At physical examination, he was oriented in time and space, but unattentive, with difficulty in understanding commands and bradykinesia. Globally preserved strength and sensitivity. Presence of bilateral dysmetria with ataxic gait. In Computed Tomography (CT) and Magnetic Resonance (MR) of the skull the presence of gross calcifications in base nuclei, supra and infratentorial white substance and bridge. Normal CSF and laboratory tests with low total calcium (CA) corrected by albumin, reduced parathyroid (PTH) (value: 4), 25-hydroxyvitamin D and normal urinary CA. Normal serological and rheumatoid exams, abdominal and chest CT without special features. Cervical ultrasound with reduced thyroid size. In the face of the clinic and examinations, the patient presented criteria for diagnosis of hypoparathyroidism and Fahr Syndrome. Calcium replacement and symptomatic medications were performed, maintaining a stable neurological and neuropsychiatric condition. **Discussion:** Fahr Syndrome is a rare and intractable neurodegenerative condition. It is cured with neuropsychiatric, cognitive and other manifestations such as motor symptoms, ataxia and aphasia. Among its causes, we included hypoparathyroidism, pseudoparathyroidism, vasculitis and hereditary diseases. Hypoparathyroidism results from deficient production of PTH, resulting in inefficient mobilization of bone calcium and inadequate renal resorption, leading to ectopic calcifications in various tissues, which, when intracerebral, characterizes the syndrome, in addition to serum hypocalcemia. The calcium deposit in the central nervous system starts in the blood vessel wall and perivascular space and extends slowly to involve the whole neurone. Progressive calcification generates tissue damage and deposits of minerals. There is a predilection for basal ganglia, also reaching other areas. There is no specific treatment for the Syndrome, and it is only symptomatic. **Final Comments:** The Fahr Syndrome is rare, but an important differential diagnosis in cases of intracranial calcifications, considering the need to discover the primary etiology for adequate treatment and orientation on the neurodegenerative character of the disease.

Miscelânea

TL 1105618

MYELOPATHY INDUCED BY VITAMIN B12 DEFICIENCY IN THE PRESENCE OF PERNICIOUS ANEMIA: CASE PRESENTATION

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Case Presentation: I.F.W, female, 65 years old, with diabetes mellitus and hypothyroidism, with no history of alcoholism or gastrointestinal complaint. She started a progressive picture of paraparesis, hypoesthesia below T10, urinary and fecal retention more than 30 days ago. On physical examination, she was lethargic, a force preserved in the upper limbs and reduced in the lower limbs. Hypoesthesia at T10 level, paresthesia and dysesthesia in feet and hands bilaterally, in addition to hypopalestinian and proprioception abolished in lower limbs. Normal CSF, serological tests for syphilis, HIV, HTLV and normal liver function, blood count with pancytopenia, normal iron and folic acid profile, and vitamin B12 < 50 mg/dL. High digestive endoscopy with biopsy confirming pernicious anemia. Normal magnetic resonance imaging (MR) of the skull and cervical spine, and in the dorsal column, the presence of hypersignal in the posterior portion of the spinal cord at T10 and T11 levels, considering subacute degeneration due to vitamin B12 deficiency. In view of the predominant clinical picture of a posterior cord, the diagnosis of myelopathy due to vitamin B12 deficiency was established. Parenteral replacement was initiated with a slight improvement up to discharge, considering the time of evolution and involvement of nervous fibers. **Discussion:** Vitamin B12 deficiency is caused by bad gastrointestinal absorption, insufficient diet or genetic defects, and anemia is pernicious due to the absence of intrinsic factors. There are several neurological manifestations, including myelopathy, neuropathy, cognitive alterations and others. The motor deficit results from involvement of pyramidal pathways associating the hypopalestinian and batisthesia due to posterior two involvement, and the paresthesias due to polyneuropathy, usually axonal pattern. Such a vitamin is a coenzyme in metabolic functions, and neuronal damage occurs by accumulation of methylmalonyl coenzyme A and increase of methylmalonic acid (MMA) and homocysteine, compromising normal myelin synthesis. The diagnosis is made by the clinic, imaging examination and vitamin B12 and/or homocysteine or urinary MMA dosage. **Final Comments:** Neurological clinical presentation with definition of sensitive level of B12 deficiency is rare and of worse prognosis, and it is essential to suspect myelopathy cases, since early identification with immediate treatment may result in better outcomes for the patient.

Miscelânea

TL 1105624

CASE PRESENTATION PATIENT WITH INTRACRANIAL HYPERTENSION SECONDARY TO THE USE OF USTEKINUMAB

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Case Presentation: Patient, 36 years old, male, with a previous diagnosis of Psoriasis and HIV infection. On 05/18/2022 he woke up with a central scotoma in his right eye and a temporal one on the left. He had episodes of blurred vision in the morning during the week prior to the reported symptoms, in addition to episodes of headache in recent months and tinnitus of mild intensity and spontaneous resolution. Last dose of ustekinumab for psoriasis treatment on 05/06/2022. Subjected to retinal mapping that showed papilledema, worse on the right. In brain resonance, no changes were seen. Laboratory tests showed reagent serum FTA-ABS, VDRL 1:2048 and undetectable Viral Load. A diagnosis of syphilis was made and treatment with Intravenous Crystalline Penicillin was initiated. He was subjected to cerebrospinal fluid puncture with opening pressure of 36 cmH₂O and result of non-reactive VDRL, FTA ABS IgM and IgG in the cerebrospinal fluid. Treatment with acetazolamide was started. The patient evolved with significant improvement in the described visual symptoms. **Discussion:** The signs and symptoms of intracranial hypertension include headache, transient visual blurring, amaurosis, papilledema, and diplopia secondary to sixth cranial nerve palsy. Through the analysis of the clinical condition presented, associating it with the results of the complementary exams, as well as the exclusion of other causal factors for intracranial hypertension, a strong correlation between the situation presented by the patient with the use of ustekinumab is suggested. This is a monoclonal antibody, which binds to the p40 subunit of IL-12 and IL-23 so that they are unable to bind to their receptors, reducing T cell-mediated inflammation in psoriasis. The most common adverse reactions are nasopharyngitis, headache and upper respiratory tract infection. Limited data is available on its potential neurological adverse effects. **Final Comments:** Intracranial hypertension associated with the use of ustekinumab is a rare condition. We emphasize the relevance of knowing the medications in use by the patient in order to make a differential diagnosis in patients with clinical manifestations of intracranial hypertension without anatomical alterations. Therefore, performing a complete anamnesis and careful ophthalmological examination is essential to obtain an accurate diagnosis and the adoption of appropriate therapy.

Miscelânea

TL 1105626

CASE PRESENTATION AN ISOLATED PROGRESSIVE PARANEOPLASTIC MYELOPATHY ASSOCIATED WITH PRIMARY KIDNEY NEOPLASMS

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Case Presentation: Patient R.R.S, 70 years old, male, started in August 2021 with clinical symptoms of monoparesis in the distal region of the right lower limb. The initial condition of distal paraparesis evolved, in a period of 3 months, ascending to proximal involvement of the lower limbs – being unable to walk in November 2021. In January 2022 it evolved with urinary and fecal incontinence, being indicated by the Urology team indwelling urinary catheter. No history of spinal cord trauma was reported. In his past pathological history, he has a history of vaccination for COVID19 (Astrazeneca) in May 2021, Benign Prostatic Hyperplasia, Pulmonary Tuberculosis at age 20. During the reported period was made diagnostic investigation with third parties, not being defined the etiology. Neurological examination revealed a spinal cord syndrome, due to spastic paraplegia associated with hypoaesthesia and crural apalesthesia up to T8 level. Admitted for diagnostic investigation, Neuroaxis Magnetic Resonance (MRI) was performed – an area of signal alteration in the central aspect of the spinal cord of D5-11 was evidenced, apparently without associated expansive effect, compatible with longitudinally extensive myelitis. In addition, Cerebrospinal Fluid was collected and analysis performed (including TRM-TB) – with no changes. Request complementary imaging exams, being evidenced in abdominal magnetic resonance imaging solid heterogeneous lesions in the right kidney, with a suspicious appearance for neoplastic involvement. On contrast-enhanced MRI of the skull, nodular lesions suggestive of secondary implants were visualized. **Discussion:** Due to the clinical picture reported and the imaging tests analyzed, the diagnostic hypothesis was of isolated progressive paraneoplastic myelopathy – with kidney neoplasia being the third most common etiology. As a change in this pathology, they were present in the reported patient T2-weighted signal hyperintensity, with involvement that extends to more than three vertebral segments – associated with a picture of progressive myopathy and evidence of primary neoplasm, Graus criteria can be used during diagnostic investigation. **Final Comments:** The isolated progressive paraneoplastic myelopathy still present as a very rare and poorly described, requiring further studies on the disease.

Miscelânea

TL 1105628

HERPES SIMPLEX ENCEPHALITIS IN PATIENTS WITH MALIGNANT GLIOMA

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Case Presentation: APS, male, 30 years old, began with strabismus and horizontal diplopia, headache, nausea, vomiting, dizziness, weakness and hypoaesthesia in the left side, evolving with difficulty walking, dysphagia, dysarthria three months ago. Neurological exam showed alternating syndrome characterized by a lesion in VI cranial nerve (NC) on the right, complete hemiparesis provided on the left and hemihypoaesthesia on the left. MRI evidenced heterogeneous expansive/infiltrative lesion, with hypersignal on T2/FLAIR, focus of contrast uptake in the central portion, diffusely affecting the pons, extending inferiorly to the medulla oblongata and right superior and middle cerebellar peduncles, compression of the fourth ventricle. Considering the diagnosis of Diffuse Midline Glioma, was prescribed Stupp Protocol. One month later, the patient developed fever, bradypsichism and tonic-clonic seizures. MRI with punctate cerebellar hemorrhages, thrombocytopenia (92,000), cerebrospinal fluid (CSF) with glucose 48, protein 106, leukocytes 53 (predominance of lymphocytes and monocytes), negative cultures and DNA of Herpes Virus 1 was detected in CSF. After 10 days of Acyclovir intravenously, the patient improved the level of consciousness. Maintained follow-up, used adjuvant with TMZ, however there was tumor growth during the fourth cycle, being currently in palliative care. **Discussion:** Herpes simplex encephalitis (HSE) is a very severe infection of the central nervous system, affecting immunocompetent in most cases. Perhaps, nowadays, it has been more commonly HSV encephalitis in immune-suppressed patients than is currently recognized. Some studies suggest an increased risk in patients with cancer and indicate that chemotherapy and steroid administration could even induce HSE without brain irradiation by disrupting host immunity², promoting HSV reactivation. Other studies imply some specific vulnerability in use of TMZ upregulates a protein that results in promoting viral replication, cell death and tissue injury¹. One of the difficulties to identify HSE condition is moderate symptoms in these patients and a cluster of alternative etiologies, like systemic infection, cancer progression, metabolic diseases. **Final Comments:** Otherwise the co-occurrence of HSE and glioma is rare and the benefits of TMZ chemoradiation overtake the risks in malignant glioma, HSE can happen during the standard treatment. So, in case of suspicion, the treatment should be promptly started to avoid more damage.

Miscelânea

TL 1105633

CLINICAL FEATURES OF A LARGE BRAZILIAN COHORT OF PATIENTS WITH MELKERSSON-ROSENTHAL SYNDROME

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Background: Melkersson-Rosenthal syndrome (MRS) is a rare and underdiagnosed neurological condition, characterized by recurrent facial palsy, orofacial edema, and fissured tongue, starting during the second or third decade of life. Its pathophysiological basis is still widely unknown. **Objective:** Our objective was to characterize clinical and epidemiological features of a Brazilian cohort of patients with MRS. **Methods:** We performed a retrospective observational study reviewing medical records from patients with MRS diagnosis from an initial sample of 1550 patients followed-up in a Brazilian Neuromuscular Reference Center. **Results:** We report 16 patients with MRS (7 women, 9 men), with age at diagnosis ranging from 20 to 84 years. All patients presented with peripheral facial nerve palsy as their first sign, starting at age 12 to 50 years, and each episode lasting from 10 to 50 days. Fifteen patients presented with recurrent facial palsy, including one case with 28 episodes during 55 years of disease course. Two cases presented with synchronous bilateral peripheral facial nerve palsy. Six patients presented with the three typical features (orofacial edema, fissured tongue, and facial palsy), while six presented with fissured tongue, twelve with labial edema, and eleven with facial and orbital edema. Mischer's granulomatous cheilitis was identified in four cases. We also identified: episodic dysgeusia (n=7), hypoacusis or hyperacusis (n=3), peripheral neuropathy (n=1), and other cranial nerve involvement (n=4). Facial spasm or myokymia in almost 33% of cases and Marin Amat synkinesis in 12,5% of cases. First clinical suspicion included recurrent Bell palsy (n=11), multiple cranial neuropathy (n=1), neurosarcoidosis (n=1), Guillain-Barré syndrome (n=1), multiple sclerosis (n=1), and ischemic stroke (n=1). Three patients had a familial history of facial palsy. Medical history disclosed: varicella infection during childhood (n=11), chronic rhinitis (n=6), autoimmune thyroiditis (n=3). Diagnostic approach disclosed: 75% of cases with normal neuroimaging studies; 100% with normal cerebrospinal fluid analysis; a large NGS gene panel for neuropathies was performed in seven cases and was unremarkable. **Final Comments:** MRS may be associated with previous autoimmune and autoinflammatory conditions. It is unlikely that a monogenic basis exists for MRS, despite a predisposing genetic risk basis being highly suggestive.

Miscelânea

TL 1105635

MULTIPLE CRANIAL NERVE PALSIES SECONDARY TO SPHENOID SINUS MUCOCELE: A CASE PRESENTATION

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Case Presentation: An 83-year-old male, with previous medical history of hypertension, reported frontal headache with nausea and vomiting of sudden onset, lasting 48 hours, that evolved with bilateral eyelid ptosis, restriction of eye movement and worsening of visual acuity. The ocular motricity deficit progressed with complete bilateral ophthalmoplegia. On physical examination, the following findings were observed: non-photoreactive mydriatic pupils, complete palsy of III, IV and VI cranial nerves bilaterally, with absent oculocephalic reflex. There were no significant visual fields changes and fundoscopic examination showed no abnormalities. Facial sensitivity and motricity, as well as corneal reflex, were preserved. Cranial MRI showed an expansive lesion, with marked spontaneous hyperintensity on T1-weighted images and a fine peripheral contrast-enhancement, centered on the topography of the sphenoid sinuses, measuring approximately $4.2 \times 3.4 \times 2.8$ cm. The lesion determined compression and cranial displacement of the pituitary gland and stalk and was in close proximity with the intracranial internal carotid arteries, without signs of vascular invasion. Laboratory evaluation demonstrated leukocytosis, with no significant change in pituitary function. The patient was, then, diagnosed with sphenoid mucocoele. He underwent endoscopic endonasal surgery, with evidence of a purulent collection in the sphenoid sinus. After surgical drainage, the patient evolved with gradual improvement of the neurological deficits, maintaining only bilateral ocular abduction deficit and right eye adduction restriction. **Discussion:** Sphenoid sinus mucocoeles are uncommon lesions, representing only 1-2% of all paranasal sinus mucocoeles. The condition may cause severe neurological sequelae, including amaurosis and intracranial spread of the infection, if there is a delay in diagnosis or treatment. Symptoms are nonspecific and result from direct mechanical pressure on adjacent structures or nerve involvement in the inflammatory process. The most common signs and symptoms are headache, visual loss, and III and VI cranial nerves palsies. **Final Comments:** The case presented demonstrates a rare condition that may evolve with multiple cranial nerves palsies, and should be considered as a differential diagnosis when identifying expansive lesions in the topography of the sphenoid sinuses.

Miscelânea

TL 1105699

ATYPICAL CLINICAL PRESENTATION OF A PRIMARY NERVOUS SYSTEM GERMINOMA IN A 32 YEARS OLD MAN – A CASE PRESENTATION

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Case Presentation: F.L.G, 32 years old, male, admitted reporting amaurosis in the left eye, of acute onset, three months ago. He presented new visual loss in the right eye, with progressive worsening in 1 month. In addition, the patient reported 6 kilograms weight loss in three months and dyschromatopsia since childhood. In the neurological exam there was dysopia to the right side, inferolateral quadrantanopsia in the right eye and amaurosis in the left eye; besides that, it was identified atrophic papilla on the left eye and afferent pupillary defect and papilledema in the right eye. Magnetic Resonance Imaging showed a cystic lesion in the pellucid septum and right nucleocapsular lesion. This patient was submitted to an endoscopic surgery and biopsy showed a germinoma OCT-4 (MRQ-10), PLAP (8A9), CD 117 (c-kit), Ki-67 (MIB-1) positive (60%). **Discussion:** Primary germinomas are rare malignant brain tumors with usual location in the pineal and suprasellar regions. Its prevalence is approximately 0,2-1,7% of all primary intracranial tumors, most common among male. The reason why germinomas occur is poorly understood but it is secondary to changes during embryogenic development. These tumors mainly are diagnosed until the second decade of life. The treatment involves radiation therapy or combination with craniospinal chemotherapy if the tumor is not metastatic. The case presented above has an unexpected clinical presentation, once the most common are headache, vomiting, ataxia, somnolence and eye movement abnormalities. **Final Comments:** This case is important to be discussed between neurologists because its atypical signs and symptoms could mimic other exclusively neurological conditions, like inflammatory central nervous system diseases. Besides that, the age of the disease, in the reported case, is different than expected, what could delay the diagnosis. From that it is important to neurologists recognize this condition, despite of unlikely epidemiology and clinical presentation, to make early diagnosis and improve survival rate once initiated the treatment.

Miscelânea

TL 1105748

DURAL ARTERIOVENOUS FISTULA AS A CAUSE OF MYELOPATHY

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Case Presentation: A 52-year-old male patient, without comorbidities, presented crural motor deficits with insidious onset about 2 years ago. He had had a car accident one month before the onset of the alterations. Upon admission, the patient presented crural paraparesis, grade III to the left and grade IV to the right, associated with elastic hyperreflexia and exalted patellar and Achilles reflexes. Spinal resonance imaging showed increased volume with diffuse intramedullary hypersignal at T2/STIR, associated with serpiginous congestive dilatation of the intradural pial veins surrounding the thoracic medullary cord, predominantly from the level of D6 to D12, including a lesser degree of edema of the medullary cone, suggestive of venous congestive myelopathy secondary to dural spinal arteriovenous fistula (DASF). Subsequently, a spinal cord angiography was performed, which confirmed the dorsal DASF at the level of the left D8 type 1 intervertebral foramen, and its embolization with adhesive embolic fluid. Anatomical occlusion of the dural arteriovenous malformation was obtained. **Discussion:** Spinal vascular malformations may be arterial or venous, corresponding to rare lesions, with an incidence of approximately 4% of spinal cord masses. They constitute a quite heterogeneous group, the main lesions being arteriovenous malformations (AVMs), DASF, spinal hemangiomas, cavernous angiomas and aneurysms. DASF is the most common (70%), being a direct communication between a radiculomedullary artery directly feeding a radicular vein; it causes venous congestion due to increased pressure in the venous system and constitutes a low-flow lesion. They are much more common in men than in women and occur mainly between the fifth and eighth decade of life, with the thoracolumbar region being the most affected. **Final Comments:** DASF is a rare cause of myelopathy but potentially treatable if diagnosed early. A case was shown that due to the prolonged time between the onset of symptoms and adequate treatment the patient had little clinical response.

Miscelânea

TL 1105765

ACUTE HEMORRHAGIC STROKE AS A TRIGGER FOR MYXEDEMA COMA: A CASE PRESENTATION

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Case Presentation: A 66-year-old woman, with a previous medical history of diabetes, hypertension and hypothyroidism, was brought to the emergency department due to loss of consciousness nine hours prior to admission. At the initial evaluation, a left central facial palsy and left hemiplegia were observed, as well as severe hypertension. The initial non-contrast head CT showed an acute bleeding in the right nucleocapsular and frontal regions, with areas of subarachnoid hemorrhage. After initial evaluation and emergency management, including endotracheal intubation and infusion of intravenous vasopressors, the patient was transferred to the intensive care unit. She was maintained under continuous venous infusion of sedatives, with the following need of neuromuscular blocking agents, due to frequent patient-ventilator asynchrony despite optimization of ventilatory parameters. Over the following days, after empiric treatment of ventilator-associated pneumonia was started, she developed bradycardia and hypertension, but the non-contrast head CT showed stability of the initial findings, without worsening of the cerebral edema. The patient, then, developed anasarca, worsening of the ventilation parameters and hypoxemia (P/F ratio of 98). At the occasion, a CT pulmonary angiogram was performed, but showed no signals of pulmonary thromboembolism. The laboratory workup revealed a TSH of 55,90 ?UI/mL, FT4 < 0,10 ng/dL and total triiodothyronine (T3) < 26 ng/dL. The patient was, then, started on steroids (hydrocortisone) and levothyroxine, with significant improvement of the ventilatory parameters over the following days, in addition to resolution of the bradycardia. **Discussion:** Myxedema coma is a severe and life-threatening endocrine emergency that occurs when physiological adaptations to untreated hypothyroidism are overwhelmed by an acute precipitant. The most common triggers to the condition are hypothermia and infection, while other described precipitants are medications, surgery, GI bleeding and myocardial infarction. Although uncommon, acute hemorrhagic stroke is a possible precipitating event and the condition should be early diagnosed for prompt initial treatment. **Final Comments:** The case presented demonstrates a rare association of acute hemorrhagic stroke and myxedema coma. Given the severity of the condition, early suspicion should be made for prompt initiation of the treatment.

Miscelânea

TL 1105779

SUBACUTE COMBINED DEGENERATION IN A VEGAN YOUNG WOMAN: THE IMPORTANCE OF A CORRECT TREATMENT

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Case Presentation: A 31 year-old woman presented with numbness and complete loss of vibratory sensitivity and proprioception in her legs and arms, significantly decreased temperature and pain sensitivity at T12 level, as well as an ataxic gait and postural instability, which had been present for over a year. When the symptoms started, she sought medical care. Exams then revealed low levels of vitamin B12 and she received three doses of 5.000UI parenteral cyanocobalamin. However, the symptoms worsened over time and she was referred to our service. Electroneuromyography performed at admission showed pure sensitive axonal polyneuropathy; magnetic resonance imaging of her spine suggested subacute combined degeneration from C2 to T12; laboratory findings were normal, including vitamin B12 levels. Since the patient was vegan and had never taken vitamin supplements, the main hypothesis remained incorrectly treated vitamin B12 deficiency. Therefore, she was prescribed 5.000UI of intramuscular cyanocobalamin every week for four weeks and then one dose per month. The patient returned for a new evaluation two months after Hospital discharge. Her gait had improved, along with proprioception and vibratory sensitivity in the lower limbs. Her neuropathic pain was well controlled with 150mg pregabalin. Results of her gastric endoscopy were normal and there was no evidence of autoimmune disease that could hamper vitamin B12 absorption. **Discussion:** Vitamin B12 deficiency prevalence vary among different populations. Some conditions may be associated with it, like reduced intake of animal products and strict vegan diet. The clinical manifestations can range from hematological findings (anemia) to neuropsychiatric changes (mood fluctuations, insomnia, cognitive slowing, ataxia, sensitivity disturbances and weakness). The neurological findings can exist even without hematological alterations. Patients with neurological abnormalities may benefit from more aggressive cyanocobalamin replacement, starting with weekly parenteral replacement for at least four weeks and then once per month. It is important as well to investigate the etiology. **Final Comments:** Despite being a common etiology for neurological abnormalities, this case presentation demonstrates how harmful vitamin B12 deficiency can be if not properly addressed.

Miscelânea

TL 1105798

CEREBRAL BILATERAL MONOCULAR POLYOPIA: AN UNUSUAL PRESENTATION OF AN ISCHAEMIC STROKE

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Case Presentation: A 58-year-old man presented to our emergency unit with a history of a sudden low visual acuity 2 days prior to the admission followed by a polyopia up to four images in both eyes, in all 9 positions of gaze which did not improve with monocular vision. His medical history included type 2 diabetes and ischemic stroke with a mild left hemiparesis. The patient had an otherwise normal ophthalmological exam, except for visual acuity of 20/40 in the left eye and 20/30 in the right eye, besides two hard exudates in the right eye at the direct funduscopy. Visual field testing was unreliable due to the fluctuating diplopia. On examination, he had a mild sequelae left hemiparesis and simultanagnosia. The patient elaborated better that his double vision involved persisting images in his visual field, indicating palinopsia. The remainder of the neurological exam was normal. A CT scan showed chronic watershed right frontoparietooccipital hypodensities and subacute watershed left parietooccipital hypodensities, later confirmed the same infarct areas by magnetic resonance imaging. The patient was diagnosed with a new ischemic stroke and started dual antiplatelet therapy. His clinical presentation was attributed to cerebral polyopia with palinopsia. **Discussion:** Diplopia is a frequent complaint in the neurology clinic. Unilateral monocular diplopia is secondary to intraocular pathology in most cases. Bilateral monocular diplopia can occur due to an intraocular process, but that's unusual to happen simultaneously in both eyes. Polyopia is the visualization of multiple images, often due to an intraocular or a functional disorder. Regardless, it can be a rare manifestation of cortical and subcortical ischemic stroke at the non-dominant or bilateral occipital lobe and even in the dominant parietal lobe. Palinopsia is the persistent visualization of visual images after the stimulus ceased and occur due to cortical non-dominant occipital lesions. **Final Comments:** Bilateral monocular polyopia is a rare manifestation of ischemic stroke. In the presence of this symptom, a complete evaluation of higher order visual function is mandatory, such as visual agnosia, simultanagnosia and palinopsia. We report an illustrative case of watershed ischemic stroke manifesting as polyopia and palinopsia associated with a simultanagnosia.

Miscelânea

TL 1105839

WERNICKE ENCEPHALOPATHY: A LIFE-THREATENING CONDITION, PASSIVE TO REVERSIBILITY IN THE FACE OF EARLY CLINICAL RECOGNITION

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Case Presentation: Patient, male, 43 years old, chronic alcoholic, presented a fall from his own height, evolving with ataxic gait and visual clouding. After the 4th day, he presented worsening of symptoms with acute mental confusion, vertigo syndrome, headache and vomiting. On neurological examination: Vigil with spatial disorientation, Mini Exam of the Mental State : 18 points, Clock test: hit the circle, missed orders of numbers; Vertical Nystagmus at rest and horizontal at the lateral view on both sides; Skew Deviation; tremor of the extremities, ataxic gait, dysbasis with fall to the Romberg maneuver without latency, with preferential side to the right, and vibratory hypoesthesia in the left foot. Among the tests, LDH 594; GGT 902; TGP 696; TGO 642; RT PCR SARS-CoV2 positive. Skull MRI showed hypersignal in nipple bodies and in the bilateral medial thalamic region, in FLAIR. Treatment performed with intramuscular (IM) thiamine for 7 days, progressing with significant improvement in ataxia and partial improvement of nystagmus, but maintained the mental confusion. He was discharged from Hospital, with oral thiamine prescription, for 24 months. **Discussion:** Wernicke's encephalopathy (WE), thiamine deficiency, should be part of the differential diagnoses of acute/subgued ataxic syndromes, especially in the context of chronic alcoholism. The patient in the case described has 3 of the 4 CAINE criteria for clinical diagnosis (acute encephalopathy; oculomotor dysfunction and ataxic gait). Laboratory tests show changes in the markers of liver and canalicular injury, secondary to the chronicity of alcohol consumption. Although neuroimaging is not essential for the diagnosis, its important to the exclusion of other diagnoses, and corroborates the hypothesis of WE: acute diencephalic and periventricular lesions in the III and IV ventricles, and atrophy of nipple bodies are specifics findings. In the suspicion of the diagnosis, patients should receive immediate administration of thiamine, preferably intravenously. Given the unavailability in the service, it had been performed IM. **Final Comments:** The acute evolution of mental confusion or ataxia should have WE as a differential diagnosis, considering the risk factors for vitamin B1 deficiency. Despite being a clinical diagnosis, it continues to be underdiagnosed, even though it is a treatable and reversible neurological emergency, early non-recognition can result in unfavorable outcomes: coma and death.

Miscelânea

TL 1105864

CASE PRESENTATION SEROTONIN SYNDROME ASSOCIATED WITH MEDICATIONS WITH A SECONDARY SEROTONERGIC EFFECT

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Case Presentation: A 31-year-old male patient with a diagnosis of obsessive compulsive disorder, undergoing knee pain investigation for two months, was referred to the emergency room due to myalgia and tremors of the extremities. He was on daily use of fluvoxamine 100mg, quetiapine 100mg and clomipramine 112.5mg increased for 3 weeks in addition to pregabalin and tramadol on demand. During evaluation, tachycardia, hyperthermia, diaphoresis, global hypertonicity and hyperreflexia were also present. Anxiety and restlessness quickly progressed to lethargy. Laboratory tests showed an increase in serum levels of creatine phosphokinase (CPK), characterizing rhabdomyolysis. Computed tomography of brain and cerebrospinal fluid examination showed no changes. Treatment involved discontinuation of offending agents, supportive therapy and use of benzodiazepine, in addition to monitoring for declining CPK levels. **Discussion:** Patients with serotonin syndrome can deteriorate rapidly and require intensive care. This syndrome occurs due to a progression of serotonergic toxicity based on an increase in its concentration levels. Conditions that can alter serotonin regulation include therapeutic doses, drug interactions, intentional or unintentional overdoses, and overlapping transitions between medications. Therefore, it can be avoided with careful titration of drugs and also observing the possibility of interaction with drug classes that do not indicate their serotonergic action in their nomenclature (e. g. opioids or antiepileptic drugs), especially in polypharmacy. **Final Comments:** Serotonin syndrome is characterized by a triad of autonomic dysfunction, neuromuscular excitation and altered mental status caused by high levels of this neurotransmitter in the central and peripheral nervous systems. With the substantial increase in the use of antidepressants worldwide, serotonin syndrome has become common in recent decades. Due to its severity and high correlation with elevated levels of the serotonin, it can and should be avoided.

Miscelânea

TL 1105865

FIRST CASE OF OCULAR CONTRAPULSION FOLLOWED BY IPSIPULSION IN WALLENBERG SYNDROME

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Case Presentation: A 51-year-old man presented with a sudden rotatory dizziness, unsteady gait, and numbness in the left hemibody. He showed left palate paresis, incoordination, horizontal jerk nystagmus with left fast phase, and ocular contrapulsion evoked by eye closure. Magnetic Resonance Imagingresonance imaging (MRI) Flairflair showed left cerebellar hyperintensity. Diffusion-Weighted Imaging (DWI)/Apparent Diffusion Coefficient (ADC) showed infarction in the left lateral medulla. After 7 days of symptoms, he reported intermittent hiccups treated with chlorpromazine. The ocular exam at this time revealed ocular lateropulsion ipsilateral to the lesion (ipsipulsion) **Discussion:** Wallenberg syndrome, also called lateral medullar syndrome, is caused by the infarct in the territory of posterior inferior cerebellar artery. Its clinical signs and symptoms include vertigo, nystagmus, diplopia, ipsilateral Horner syndrome, facial ruddiness and dry skin, dysphonia, dysphagia, dysarthria, ipsilateral loss of gag reflex, ipsilateral ataxia, ipsilateral impaired taste, ipsilateral facial pain and paresthesia, decreased ipsilateral blink reflex, contralateral hypoalgesia and thermoanaesthesia in the trunk and limbs; and ipsilateral facial hypoalgesia and thermoanaesthesia. **Final Comments:** To our knowledge, this semiologic feature of alternating lateropulsion has not been previously described in the literature until now. The scientific study of atypical clinical manifestations can help medical care to identify the syndrome early and to better manage patients.

Miscelânea

TL 1105906

A RARE CASE OF EXPANDING VIRCHOW ROBIN SPACES MIMICKING CYSTIC TUMOR AND CAUSING HYDROCEPHALUS

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Case Presentation: A 57 year women presented with progressive imbalance and right hemiparesis progressing in three months. Chikungunya infection 3 years before was reported. Magnetic resonance imaging (MRI) showed cystic lesions in the midbrain and obstructive hydrocephalus. The features suggested abnormal dilatation of perivascular spaces. The lesion was partially resected, with good evolution of symptoms with three-year postoperative follow-up. **Discussion:** Perivascular enlargements, or Virchow-Robin spaces (VRS), are seen on MR images as small foci that are isointense to the CSF, like small cystic cavities that usually surround the small arteries and arterioles at the level of basal ganglia, the anterior perforated substance and the thalamic-mesencephalic junction. Small Virchow-Robin spaces appear in all age groups. Age, arterial hypertension, dementia, and the incidental white matter, lesions were found to be significantly associated with large Virchow-Robin spaces. In a retrospective study of MR imaging. Typically, they have no mass effect and are asymptomatic, but in our patient and in other studies showed that the perivascular spaces can expand more than usual, acting as space-occupying lesions. Poirier et al reported the first case of expanding lacunae in a 54-year-old woman with dementia. Surgical treatment for the hydrocephalus has produced different results; Ono et al reported a case wherein cystectomy combined with a third ventricle-to-peritoneum shunt improved the symptoms and decreased the size of both ventricles and cysts. **Final Comments:** Virchow-Robin spaces are typically considered incidental but can be symptomatic in rare cases. The typical scan characteristic and location of Virchow Robin spaces can help to differentiate from cystic mass lesions. However, despite advanced MRI techniques, sometimes it is still difficult to differentiate VRS from some cystic brain tumors.

Miscelânea

TL 1105907

PINEOCYTOMA PRESENTING AS A PITUITARY ADENOMA IN A YOUNG WOMAN: A CASE-BASED UPDATE

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Case Presentation: A 18-year-old female patient had a history of progressive frontal headache for about 1 year. The patient evolved with bilateral visual loss 1 month ago, accompanied by vertigo, galactorrhea, amenorrhea and paraparesis, with greater intensity in the left limb. Neurological exam revealed bilateral absence of direct and consensual photomotor reflexes. Brain Magnetic Resonance Imaging (MRI) showed the presence of an expansive lesion around the pineal gland, manifested with T1 hypointense, T1 contrast-enhanced hypersignal, and low signal foci on T2 GRE. This lesion extends to the quadrigeminal plate, left fornix, interior of the fourth ventricle, left lateral ventricle and quadrigeminal cistern, causing tonsillar hernia and obstructive hydrocephalus of the sides of the ventricles and the third ventricle. After neurosurgical evaluation, a right ventriculoperitoneal shunt was performed to resolve intracranial hypertension and collection of cerebrospinal fluid for histopathological study, which confirmed the diagnosis of pineocytoma. **Discussion:** Tumors of the pineal region are relatively rare, accounting for about 0.4-1% of intracranial tumors. These tumors are generally divided into pineocytoma (grade I), intermediately differentiating pineal parenchyma tumors (grade II or III), papillary tumor of the pineal region (grade II or III), and pineoblastoma (grade IV). Pineocytomas on MRI are usually smaller than <3 cm, well defined, iso or hypointense on T1 and hyperintense on T2. They usually do not seed cerebrospinal fluid and have slow growth associated with a good prognosis. The signs and symptoms depend on the tumor grade and its location, being mainly due to cranial hypertension due to obstructive hydrocephalus secondary to compression of the midbrain roof. In this sense, vomiting, behavioral changes and visual disturbances are also observed, which can characterize Parinaud's syndrome. Galactorrhea and amenorrhea are not usually associated with pineocytomas. However, they may be present rarely when there is coexistence with pituitary adenomas in bifocal tumors.

Miscelânea

TL 1106020

WERNICKE'S ENCEPHALOPATHY DUE TO ACUTE PANCREATITIS

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Case Presentation: N.L.S, 19 years old, is reported to the ER, in Salvador – BA, on 04/23/2021, complaining of epigastric pain and vomiting for 30 days, with worsening in the last 24h of admission. She carried an outpatient ultrasound suggestive of acute cholecystitis. Therefore, the possibility of acute biliary pancreatitis was considered. Diet restriction and partial parenteral nutrition were started, with maintenance of nausea and vomiting, being then escalated to total parenteral nutrition on 04/27. On 05/04, she developed fever, tachycardia with leukocytosis. The assistant team prescribed Meropenem and Teicoplanina. Abdominal CT shows slightly hyperattenuating content inside the gallbladder. On 09/05, she evolved with a lowered level of consciousness, GCE 8, being subjected to orotracheal intubation for airway protection. The neurology team is called in to assess the condition. Findings of ataxia, multidirectional ophthalmoparesis, nystagmus, tetraparesis and global hyperreflexia were identified. Two cranial CT were performed without acute changes. Cranial MRI was performed, which showed hypersignal on T2/FLAIR and in the diffusion sequence, affecting the medial and posterior portion of the thalami bilaterally and symmetrically. Such findings are superimposable to those observed in Wernicke's encephalopathy. The patient received IV thiamine replacement in 05/14, presenting an almost complete reversal of the clinical presentation. **Discussion:** Wernicke's encephalopathy (WE) is a rare condition associated with thiamine deficiency, characterized by neurological symptoms such as ataxia, nystagmus, tremors, paresis and cognitive impairment. Although very associated with chronic alcoholism, it is present in 12.5% of the autopsies of addicts, the condition is also associated with situations of malabsorption, poor dietary intake, increased metabolic requirement or in situations of accentuated loss of thiamine (hemodialysis). However, other less common etiologies are described, such as prolonged fasting associated with other pathologies, as acute pancreatitis. The diagnosis of WE is difficult to confirm, and if untreated, patients progress to coma and death. Fortunately, IV administration of thiamine is safe, simple, inexpensive, and effective. Diagnostic tests, therefore, should not delay treatment. **Final Comments:** It is essential to prevent thiamine deficiency in cases of acute pancreatitis, as well as early recognition of the clinical picture.

Miscelânea

TL 1106042

CLIPPERS: CASE PRESENTATION

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Case Presentation: M.S.M., male, 33 years old, was admitted in January 2022 with a report of gait impairment for 4 years, worsening in the last year, associated with imbalance. He also presented, in the last month, blurred vision, diplopia, and slurred speech. On physical examination, he presented with a global strength grade of 4+, all reflexes exalted, and ataxic gait. At the time of admission, a new brain MRI was made with pons lesions, with a salt and pepper aspect, suspected of inflammatory disease, being opted for pulse therapy with corticosteroids and plasmapheresis, with clinical and radiological improvement. **Discussion:** Chronic lymphocytic inflammation with steroid-responsive pontine perivascular enhancement is an inflammatory disease of the central nervous system defined in 2010 by Pittock, who described the case of 8 patients with similar clinical, radiological, and pathological features who were responsive to corticosteroids. For the diagnosis of CLIPPERS is necessary that the patient presents clinical alterations like progressive subacute ataxia and diplopia and symptoms related to brainstem involvement. Associated with MRI alteration with multiple salt and pepper sign in pons, with or without propagation to cerebellar peduncles and cerebellum. Histopathologically presenting a perivascular lymphohistiocytic infiltrate of the white matter, with a predominance of CD3 and CD4 lymphocytes. It's possible to make a definitive diagnosis only with clinical and image characteristics, if other etiologies are excluded. **Final Comments:** CLIPPERS is a rare disease with a high morbidity rate, with a high risk of complications and disability when not diagnosed and treated at the right time, so it is important to know the pathology and make a diagnosis to offer the best treatment to the patient.

Miscelânea

TL 1106053

METASTATIC CERVICAL CARCINOMA WITH EVOLUTION TO ENCEPHALIC MENINGEAL CARCINOMATOSIS

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Case Presentation: Female patient, 38 years old, smoker, diagnosed with stage IIIB squamous cell carcinoma of the cervix in 2019, underwent radiotherapy until February 2020 and after started brachytherapy. During the follow-up, the patient presented progression of the disease to the peritoneum, lung, ribs and dorsal spine, and underwent to palliative chemotherapy with carboplatin and platinetaxel. In September 2020, had a Hospital admission due to severe holocranial headache, fatigue, dysuria, diarrhea, pain in the lumbar region and episodes of fever. Cranioencephalic MRI revealed the presence of multiple contrast-enhanced nodular lesions in the skullcap, compatible with metastatic implants, some causing erosion of the inner bone plate, with thickening and diffuse pachymeningeal enhancement, more evident in the right frontal region, which could be related to pachymeningeal carcinomatosis. Also, the presence of minute nodular images with hypersignal in FLAIR and weak contrast enhancement, located along the leaves of the middle third of the right cerebellar hemisphere, may be related to leptomeningeal or intraparenchymal cortico-subcortical implants. In addition, it was detected small hyperintense T2/FLAIR images in the supratentorial white matter, with no expansive effect, diffusion restriction or contrast enhancement. Other imaging exams revealed the extent of metastatic involvement to the liver, adrenals and mandible. Treatment was ceased due to terminality of the patient, and was kept in exclusive comfort. The patient died a few days later. **Discussion:** There are less than 22 articles published in the literature that describe meningeal carcinomatosis related to gynecological cancers. It is estimated that the incidence of meningeal carcinomatosis in patients with cervical cancer is 0.03%. Patients with meningeal carcinomatosis have a poor prognosis due to the advanced stage of the primary tumor. Literature data indicate that the median survival after diagnosis ranges from 10 days to 26 weeks. **Final Comments:** Meningeal carcinomatosis related to cervical carcinoma is rare. However, this pathology should be considered in patients with primary gynecological tumors who present a clinical picture suggestive of CNS involvement, since palliative therapies can be instituted to improve the patient's quality of life and delay neurological deterioration.

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TL 1106161

MEDULAR COMPRESSION SYNDROME OF METASTATIC ORIGIN

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Case Presentation: CES, male, 62 years old, painter, smoker, alcoholic, previously functional and independent. He reported low back pain for a month. Paresthesia in the lower limbs that evolved a day after to symmetrical paraplegia associated with significant change in sensitivity and urinary retention. Admitted to neurology already with deficit. Neurological examination showed paraplegia, urinary and fecal retention, bilateral biceps, patellar and achilles hyperreflexia. Tromner and Hoffman present bilaterally. Abdominal skin reflex absent. Plantar cutaneous reflex in extension on the left limb. Apalesthesia, thermal, tactile and painful anesthesia with a T6 level. Magnetic resonance imaging of the brain, cervical and thoracic spine were performed, which showed foci of signal alteration in the facets and right pedicle of C3, vertebral bodies of C6 and C7, with a secondary neoplastic aspect. Multiple lesions diffusely distributed in the thoracic vertebrae, especially long expansive and infiltrative lesions involving the vertebral bodies and posterior elements from T4 to T8, with an important soft tissue component at the T5-T6 level. T5-T6 decompressive laminectomy was performed with biopsy of the surgical resection and malignant epithelial neoplasia with clear cell components was visualized. An abdominal tomography was also performed, which showed a suspicious-looking lesion for primary urothelial neoplasm in the right ureter. An investigation was also performed for prostate and lung cancer, both negative. Informed consent was obtained from the patient for a case presentation Spinal cord compression syndrome is a condition that can be caused by different etiologies that cause spinal canal stenosis. It may be the initial manifestation of 20% of metastatic neoplasms. The characteristic manifestations of this syndrome are relatively symmetrical motor deficits, urinary retention or incontinence, loss of sensation, and includes signs of hyperreflexia. Some tumors tend to metastasize to specific regions of the spine. Renal, gastrointestinal or prostate tumors generate metastases to the lower regions of the thoracic spine and lumbosacral spine. The patient in question had a spinal cord compression syndrome, mainly in the thoracic region, with a primary site in the urinary system.

Miscelânea

TL 1106237

NEUROSARCOIDOSIS PRESENTING AS "ACUTE" NORMAL PRESSURE HYDROCEPHALUS: A CASE PRESENTATION

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Case Presentation: A 68-year-old woman, with a previous medical history of diabetes, hypertension and hypothyroidism, was brought to the emergency department with complaints of bilateral leg weakness, cognitive impairment and urinary incontinence that started twenty days prior to admission. The patient also reported unmeasured daily fever over the preceding two weeks, a significant unintentional weight loss and anosmia over the past two years. At the initial evaluation, a mild loss of strength in the lower limbs and significant gait apraxia were observed. Initial head CT showed dilatation of supratentorial ventricular system and transependymal effusion of cerebrospinal fluid (CSF). CSF analysis demonstrated a cellularity of 12 leukocytes/mm³ (100% lymphocytes), increased total protein (63mg/dL) and a low glucose level (CSF/plasma ratio of 52%). The MRI showed signs of leptomeningeal and choroid plexus inflammation, with obstructive hydrocephalus and intracranial hypertension. Following the investigation, a chest CT demonstrated paratracheal and subcarinal lymphadenopathy and a biopsy was performed. The fungal, bacterial and mycobacterial cultures of CSF and lymph node were negative and pathologic examination of the lymph node revealed noncaseating granulomas without evidence of malignancy. A diagnosis of neurosarcoidosis with obstructive hydrocephalus was made and the patient was started on steroids (1mg/Kg/day of prednisone). After the beginning of therapy, the patient presented significant improvement of the symptoms, returned to walk independently and there was no need of neurosurgical procedures to drain the CSF. **Discussion:** Sarcoidosis is a multisystemic granulomatous disorder characterized by the formation of noncaseating granuloma in various organ systems. Neurological involvement may result in peripheral or central nervous system (CNS) manifestations. Gait disorders and cognitive decline are less common forms of presentation. Here, we report an unusual presentation of the disease. A patient with a previously preserved functionality presented a rapidly progressive cognitive impairment, gait apraxia and urinary incontinence. Given the challenging diagnosis of the case and association with obstructive hydrocephalus, clinicians should be aware of this condition. **Final Comments:** The case presented demonstrates an unusual association of neurosarcoidosis and a rapidly progressive presentation of cognitive impairment, gait apraxia and urinary incontinence.

Miscelânea

TL 1106283

IDIOPATHIC POLYARTERITIS NODOSA AND NEUROLOGICAL MANIFESTATIONS: CASE PRESENTATION

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Case Presentation: RBR, 53 years old, with intense and pulsating headache, vertigo, tinnitus, hearing loss on the left, low bilateral visual acuity and behavioral change for 7 months. Past history of an episode of joint pain, fever and nonspecific hepatitis for 4 years, without diagnosis and using corticosteroids since then, in addition to arterial hypertension and dyslipidemia. Neurological examination was lethargic, strength and sensitivity were preserved, grade 3 osteotendinous reflexes globally, low bilateral visual acuity, without other changes. On general physical examination, the presence of hyperemic and scaly lesions on the lower limbs. Cranial MRI showed lesions with hypersignal on T2/FLAIR in the right superior temporal gyrus, ipsilateral superior parietal lobe, with a granulomatous aspect, suggesting an inflammatory nature. In cerebrospinal fluid, lymphocytic predominant pleocytosis, normal glucose and protein, and negative culture. Also, HIV, Syphilis, Hepatitis and other viral infections were excluded. In rheumatological and vascular research, an increase in ESR, with no other changes. Finally, in a biopsy of the lower limb lesion, evidence of mixed inflammatory infiltrate, lymphocytic predominance with foci of aggression in the vascular wall, with thickened central medium-sized vessels, with fibrosis, surrounded by the inflammatory infiltrate, diagnosing polyarteritis nodosa (PAN). **Discussion:** PAN is a vasculitis of medium-sized arteries and rarely of small vessels. It may be idiopathic or secondary to hepatitis and malignancies. Of multisystem involvement, it can cause renal, coronary and gastrointestinal artery disease, in addition to neurological disease, coursing with mononeuritis, peripheral neuropathy and involvement of the central nervous system, more rarely. In the disease, the occlusion or rupture of the inflamed arteries generates ischemia or hemorrhage. Its diagnosis is clinical associated with rheumatological exams of exclusion, serological exams to identify possible causes, and skin biopsy demonstrating infiltrate of inflammatory cells in vessels and fibrinoid necrosis, in addition to angiography that may show saccular aneurysms or stenosis in blood vessels. **Final Comments:** The association of systemic and neurological vascular diseases is common. Thus, it is important to correlate neurological manifestations with those of other organs, as in the present case, being able to direct and reach decisive diagnoses.

Miscelânea

TL 1106326

A RARE CASE OF PARANEOPLASTIC LONGITUDINALLY EXTENSIVE MYELOPATHY, SECONDARY TO MALIGNANT PHEOCHROMOCYTOMA

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Case Presentation: Male patient, 60 years old, with hemophilia type A diagnosed at the age of 13 years old and HCV diagnosed in 1983, with cure control in 2012. 50 days ago from admission, he noticed painless inguinal lymph nodes. After 15 days, he began dysesthesia in the cervical region and upper limbs. He evolved with severe neck pain, dorsal irradiation, worsening of movement and motor symptoms, associated with nausea, vomiting and hiccups. 10 days before admission, he started sensory-motor symptoms in the lower limbs predominant asymmetric to the left and, after 4 days, presented constipation and urinary incontinence. After 5 days, he presented restriction on ambulation and loss of muscle strength in the lower limbs and was Hospitalized. On admission neurological examination showed grade V strength in the upper limbs, grade III in right lower limb, grade II in left lower limb (preserved toe dorsiflexion, grade IV plantar flexion of the toes). Babinski sign present bilaterally. Laboratory tests showed: LDH 795; Anti-HVC 166. Normal rheumatological tests. CSF: colorless, cell 1, proteins 119 mg/dl; missing oligoclonal bands, anti-Aquaporin 4 negative. Total abdominal CT: Retroperitoneal and pelvic lymph node enlargement. Nodule in the body of the left adrenal gland. Neuroaxis MRI: Capture annular lesion, encapsulated with perilesional edema in the left frontotemporal region. Cervical spinal cord injury with irregular contrast uptake, bulb-sacral enhancement. Biopsy left inguinal lymph node tissue with immunohistochemistry: malignant, metastatic pheochromocytoma. During Hospitalization, he evolved with unprecedented quadriparesis and seizures, being submitted to orotracheal intubation and transferred to the ICU. It was decided to perform pulse therapy with methylprednisolone 1g/5 days, with no clinical response. **Discussion:** Paraneoplastic longitudinally extensive myelopathy is a rare event, more associated with neoplasms of bones, liver, lymph nodes and lung. Literary evidence of malignant pheochromocytoma with intramedullary involvement is scarce and constitutes a diagnostic and therapeutic challenge. In this case, in addition to extensive spinal cord involvement, brain involvement is evident. **Final Comments:** The case in question is notable for the rarity of the clinical presentation. The unfavorable evolution is due to the extent of the lesion and concomitant brain involvement, with no previous reports of association with malignant pheochromocytoma in the literature.

Miscelânea

TL 1106357

SMART SYNDROME: A CASE PRESENTATION

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Case Presentation: A 34-year-old man had a progressively developed headache and focal seizures. CT brain scan disclosed a left frontal mass, with gadolinium enhancement. The patient underwent a successful subtotal lesion resection, followed by whole-brain radiotherapy for 3 months (60Gy). Pathological analysis revealed an anaplastic oligastrocytoma, according to the 3rd WHO classification of central nervous system tumors. Due to great post-radiotherapy brain MRI status, the patient was put under conservative treatment, with control images every six months. The four following brain MRIs revealed important gadolinium enhancement in addition to cortical gyriform enhancement, although the patient remained asymptomatic. Two years after the initial diagnosis, the patient presented sudden transient right-sided weakness. A new brain MRI showed diffuse gadolinium enhancement. The patient underwent a second resection and tissue histopathology revealed radiation necrosis, with no signs of neoplastic disease. He also started a daily headache of pulsating quality, in association with nausea. Migraine prophylaxis was started, along with steroids therapy, with great symptom control. Steroid therapy was necessary for the two following years, and the patient presented new clinical recurrences and worsening brain image in every withdrawal attempt. According to Black's criteria, the patient was diagnosed with stroke-like migraine attacks after radiation therapy (SMART) syndrome. **Discussion:** SMART syndrome is a rare late complication of radiotherapy and usually presents 12 years after the treatment. Classic symptoms are migraine attacks and focal deficits, such as aphasia and hemiplegia, although some patients may experience seizures. Some patients also may recur from clinical deficits. Risk factors are not clearly defined but higher radiation doses may increase the chances of developing SMART syndrome. Pathophysiology is still poorly understood, but some authors believe that radiation leads to vascular damage, which causes endothelial dysfunction. Brain MRI is not always specific, and distinction from tumor progression may be difficult. No treatment has been established, but some cases report the use of steroids and antiplatelet agents. **Final Comments:** Our case presentations an early development of SMART syndrome. It is a rare complication of growing importance, especially because cancer patients are surviving much longer, and the symptoms can impact on quality of life.

Miscelânea

TL 1106370

EARLY INFLAMMATORY CHANGES IN THE BRAINSTEM AFTER RADIOSURGERY: A CASE PRESENTATION

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Case Presentation: A 36-year-old woman, without previous comorbidities, was diagnosed with a right petroclival meningioma, who underwent radiosurgery. 30 days after the procedure, she was admitted with initial complaints of vertigo, followed by sensory changes on left hemibody and diplopia. On neurological examination, there were right horizontal gaze-evoked nystagmus, with binocular diplopia, right sensorineural hypoacusia, paresis of the palate elevation on the left, and asymmetry on shoulder elevation, worse on the left. Motor strength was preserved on the four limbs. There was tactile and painful hypoaesthesia on the left hemibody, with deep senses preserved. There was no axial or appendicular ataxia, but a shift to the left on Fukuda gait. CSF disclosed a slight increase in cellularity and protein. MRI showed a hypersignal on T2-weighted images of the brainstem and cerebellar peduncle. Pulse therapy was performed with 1g of methylprednisolone for 05 days, with significant improvement in symptoms. **Discussion:** Inflammatory changes in the brainstem admit a wide differential diagnosis and may be challenging. Imaging changes on MRI are common after CNS radiation and can be seen as a hypersignal on T2-weighted images. However, only about 10% of patients are symptomatic and presented with cranial neuropathies, headache, hemorrhages, edema, epileptic seizures, radionecrosis, among others. Radiation complications can be acute (radiation encephalopathy), subacute (transient demyelination) or chronic. Radionecrosis, the most common chronic complication, especially in the brainstem, is rare and atypical presentations are even less frequent. Usually, radionecrosis occurs about 6 months to 3 years after treatment. First-line therapy is with corticosteroids. **Final Comments:** Acute complications related to radiotherapy are uncommon. Later complications, such as radionecrosis, occur in about 23.1% of cases, in the first year up to decades after treatment. Attention is required for this presentation reported here, in view of its wide differential diagnosis and favorable prognosis after treatment with corticosteroids.

Miscelânea

TL 1106407

MELKERSSON-ROSENTHAL SYNDROME: A CASE PRESENTATION

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Case Presentation: A 44-years-old woman presented to our service due repeated episodes of right Bell's palsy. The begin of symptoms was five years ago, with right facial nerve palsy diagnosed as Bell's palsy and treated with prednisone and aciclovir, showing gradual improvement. One year later, she developed facial disesthesia in right face, associated with right facial edema. In the following 3 years, she had two more episodes of facial nerve palsy, associated with ipsilateral facial edema and tongue fissures, referred to our service. Brain MRI were normal, and laboratory tests showed reagent antinuclear antibody (1/320), with no other biomarkers present and serological tests for the infectious disease negative. CSF showed 16 cells, 65 proteins, with negative infectious tests. Salivary gland biopsy unchanged. The hypothesis of recurrent facial paralysis due to Melkersson-Rosenthal Syndrome was raised and treatment with corticosteroids (prednisone 60mg per day) for two weeks was performed, without recurrence of symptoms. **Discussion:** Melkersson-Rosenthal syndrome (MRS) is a rare syndrome with unclear etiology, comprised of the triad of recurrent facial nerve paralysis, nonpitting orofacial edema and fissured tongue (lingua plicata). All the three features are present in 20–75% of cases. Other neurological manifestations may be seen in MRS, like migraine, facial paresthesias, tinnitus, dizziness, hypogeusia and pharyngeal neuralgias. Non-neurological symptoms include uveitis, diverticulitis, and ulcerative colitis. The investigation must be exclude systemic pathologies, mainly sarcoidosis, Crohn's disease, tuberculosis, herpes and other infectious diseases. Biopsy of a region with orofacial edema demonstrating noncaseating granulomas supports the diagnosis of MRS. There is no specific treatment for MRS. Therapy with corticosteroids leads to improvement in 50–80% of patients and reduced relapse frequency by 60–75%. However, randomized trials are lacking and frequency, dosing or duration are not established. Other options described are NSAIDs, antibiotics (like doxycycline and dapsone) and immunosuppressants (methotrexate, infliximab). Intralesional triamcinolone acetate injection also are used in local edema. **Final Comments:** MRS is a cause of recurrent facial nerve palsy, and etiologies such as multiple sclerosis and infections should be investigated. In idiopathic cases, steroids are often used. More studies are needed to understand and manage these cases.

Miscelânea

TL 1106433

ADULT DIFFUSE HIGH-GRADE BRAINSTEM GLIOMA INDUCED BY BRAIN RADIOTHERAPY IN CHILDHOOD, A CASE PRESENTATION

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Case Presentation: 33-year-old female, in 2002, at age 13, was diagnosed with pineal dysgerminoma. The patient underwent surgical resection of the lesion, followed by chemotherapy and radiotherapy. In 2003, this patient had tumor recurrence and was submitted to a new surgical resection followed by chemotherapy and reirradiation. After achieving complete response, the patient remained in clinical follow-up for 18 years. In May 2020, she evolved with altered mental status associated with diplopia and oculomotor nerve paresis. Cranial MRI was performed and showed an expansive lesion in the right thalamo-mesencephalic region with contrast enhancement. In July 2020, a biopsy of the midbrain lesion was performed and confirmed the diagnosis of high-grade glioma. Immunohistochemical analysis demonstrated positivity for expression of GFAP, ATRX and Ki67 (40% of the tumor cells), while the expression of IDH1, CD34, neurofilament, synaptophysin, p53 and H3K27M were negative. Diffuse high-grade glioma, H3-wildtype and IDH-wildtype was the definitive diagnosis. Radiotherapy was performed initially and later this patient had disease progression and received temozolamide. Currently, this patient is in palliative care. **Discussion:** Aside from genetic factors, the main risk factor already established for the development of glioma is previous exposure to ionizing radiation of the central nervous system (CNS), as occurs from therapeutic radiation therapy for childhood brain tumors or leukemia. The interval between irradiation and glioma development varies from five years to several decades. In this clinical case, the etiology of the glioma is probably related to the brain radiotherapy that this patient underwent for the treatment of pineal dysgerminoma during childhood. **Final Comments:** Radiation induced glioma is relatively rare but well described in the literature. Unfortunately, radiation-induced gliomas promote high morbidity and mortality in patients. In this way, the rational use of brain radiotherapy especially in children and the development of new technologies that reduce the risk of second brain neoplasms induced by radiotherapy is extremely important.

Miscelânea

TL 1106441

REVIEWING AND RECALLING THE KEY ROLE OF NEUROIMAGING IN WERNICKE ENCEPHALOPATHY

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Case Presentation: AMC, 21 years old, 8 weeks pregnant, previously healthy. Admitted to the gynecological emergency care due to repeated vomiting, being diagnosed with hyperemesis gravidarum. On the third day of Hospitalization, the neurology team was called due to mental status change. The neurological examination revealed the presence of confusion, multidirectional ophthalmoparesis and ataxia with significant gait alteration. It was decided to measure serum vitamin B1 (thiamine) and request magnetic resonance imaging (MRI) and analysis of the cerebrospinal fluid. The diagnosis of Wernicke encephalopathy was confirmed due to the finding of vitamin B1 deficiency and, mainly, the presence of hypersignal of the mammillary bodies and lesions of the medial thalamus and of the walls of the third ventricle on MRI. The patient showed significant clinical improvement after thiamine replacement. **Discussion:** Wernicke encephalopathy can occur as a rare complication of hyperemesis gravidarum. MRI plays an important role in elucidating the diagnosis as it presents classic alterations, with the mammillary bodies, medial thalamus and periventricular region of the third ventricle being the typically affected regions. Because it is an acute neurological disease that results from thiamine deficiency, the prognosis of patients depends on the time from the onset of symptoms and the start of supplementation of this vitamin. When the diagnosis and treatment are delayed, Korsakoff psychosis or death may occur. **Final Comments:** Neuroimaging plays a central role in the early diagnosis of this prevalent pathology, which can be reported in up to 36% of patients with acute mental confusion and lead to significant neurological morbidity if not properly managed.

Miscelânea

TL 1106459

VITAMIN B12 DEFICIENCY ASSOCIATED WITH ATYPICAL BRAIN MRI FINDINGS

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Case Presentation: A 68-year-old woman was referred to our Hospital complaining of progressive and symmetrical paraparesis, paresthesia, and gait imbalance over the last five months, currently confined to a wheelchair. She had no previous medical history or noteworthy dietary restrictions. On neurological examination, she had severe lower limb weakness, spasticity, and generalized brisk deep tendon reflexes with a bilateral Babinsky sign. The patient also had reduced light touch and vibration sense in both legs and upper limbs. Laboratory testing revealed reduced levels of B12 vitamin and on brain MRI it was noticed FLAIR and T2 hyperintensities in periventricular and centrum semiovale white matter, bilateral globus pallidus, cerebral and middle cerebellar peduncles, and in the brainstem. The described lesions had no diffusion restriction or contrast enhancement. These findings were therefore compatible with vitamin B12 deficiency and the patient was referred for further investigation in our outpatient clinic. **Discussion:** Vitamin B12 deficiency can be responsible for hematological, gastrointestinal, psychiatric, and neurological symptoms. The causes of depletion are numerous and include malabsorption due to GI tract abnormalities, autoimmune diseases, and dietary restrictions. The most common neurological manifestations are paresthesia, progressive symmetrical weakness and gait disturbances. The disease may progress to spastic paraplegia with cognitive and mood alterations. Spinal cord MRI often shows the altered signal in its lateral and posterior columns. Less commonly there are brain white matter and optic nerve involvement. Bilateral globus pallidus lesions are an even rarer presentation, described in the presented case. Treatment is based on vitamin B12 supplementation and neuropsychiatric improvement can be seen after months, however, some neurologic findings may be irreversible. **Final Comments:** In this case there are classical neurological signs compatible with vitamin B12 deficiency such as combined subacute degeneration, hyperreflexia and peripheral neuropathy. Nevertheless, we present uncommon imaging finding poorly reported in the literature, more specifically bilateral hyperintensities in the globus pallidus.

Miscelânea

TL 1106461

ISCHEMIC MYELOPATHY AND HEMATOMYELIA: A CASE PRESENTATION

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Case Presentation: A 28-year-old woman presented upper thoracic spine pain after supposed trauma in a soccer game, evolving after 03 days with paresthesia of hands and tongue with evolution in 40 minutes to tetraparesis, lowered level of consciousness and acute respiratory failure requiring airway support. The patient underwent a study of the cerebrospinal fluid (CSF) which showed pleocytosis with lymphocytic and neutrophilic predominance associated with normal levels of protein and glucose. The diagnostic investigation was complemented with magnetic resonance imaging (MRI) of the skull and cervical spine, which presented longitudinally extensive myelopathy in addition to internal hemorrhagic foci and presence of hematomyelia in the pons and in the anterior portion of the cervical spine. A hypothesis of acute longitudinally extensive transverse rhombencephalomyelitis of probable immune-mediated etiology was raised and pulse therapy was started with 1g methylprednisolone for 05 days, and, due to an unsatisfactory response, complemented with 05 sessions of plasmapheresis, with imaging improvement afterwards, but without clinical response. She underwent cervical and thoracic spinal cord angiography that showed dissection of the left vertebral artery in the V2 cervical segment and failure of opacification of the anterior spinal artery in the projection of the odontoid process suggestive of occlusion. We reached the conclusion of an ischemic lesion with hemorrhagic transformation, secondary to dissection of the left vertebral artery and probable occlusion of the anterior spinal artery. **Discussion:** Hematomyelia is a very rare intraparenchymal spinal cord hemorrhage. The most common cause is trauma, followed by vascular malformation, coagulopathy, neoplasm, and iatrogenic procedures. A hemorrhagic transformation after spinal cord infarction can sometimes occur, caused by mechanisms such as arterial dissection, atherosclerosis and fibrocartilaginous embolism. Patients may present with acute severe neck or back pain at onset followed by rapid severe myelopathy deficits. The hematomyelia treatment consists of cervical or thoracic laminectomy followed by durotomy and myelotomy, after which the hematoma cavity is evacuated and irrigated. **Final Comments:** Vascular myelopathies are important clinical entities due to its acute and progressive course. A quick diagnosis can potentially result in a life-saving therapy and prevent lasting neurologic damage and death.

Miscelânea

TL 1106500

A RARE CASE OF ALCOHOL-INDUCED OSMOTIC DEMYELINATION SYNDROME

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Case Presentation: Male, 53 years old, is admitted to the emergency room with a subacute picture of dysarthria, central facial paralysis on the left and hemiparesis on the left with evolution of one week. The clinical condition began a day after an episode of alcoholic libation. Previously hypertensive, he made continuous use of telmisartan + amlodipine and chlorthalidone. According to family members, he consumed alcohol excessively on weekends, and after family deconjunction this consumption was exacerbated. In the neurological examination, he was vigilant, oriented in time and space, with severe dysarthria, facial paralysis of central pattern to the left, deviation of the uvula to the left, deviation of the ipsilateral soft palate. He evolved with an asymmetric tetraparesis, with degree muscle strength (DMS) 2 in the left dimidium and DMS 4 in the right upper limb and DMS 2 in the right lower limb. Hoffman is present bilaterally. Slight tactile and painful hypoesthesia on the left. Right dysmetria in the index-nose test. The cerebrospinal fluid was clear and colorless, with an opening pressure of 22cmH₂O, with normal cellularity, normal protein and glucose value. Contrast-enhanced nuclear magnetic resonance imaging (MRI) of the skull revealed the presence of a lesion in the brainstem, especially in a pons characteristic of pontine myelinolysis, with hyposignal on T1-weighted sequences, hypersignal on T2-weighted sequences and Flair, measuring 3.1x3x2cm, bilaterally without circum edema. Intense contrast enhancement. Other similar lesions in splenium of the corpus callosum on the right and right occipital lobe. **Discussion:** The most common cause of Osmotic Demyelination Syndrome (ODS) is the very rapid correction of chronic hyponatremia. Cases of alcohol-related non-hyponatraemic osmotic demyelination result from glycogen depletion and thiamine deficiency that limit the ability to adapt brain to any osmotic stress. The diagnosis is initially suspected from clinical symptoms and is confirmed by magnetic resonance imaging of the brain. **Final Comments:** The authors describe a rare case of pontine and extra-pontine ODS not associated with hyponatremia. The causal factor in question was alcohol libation and the clinical diagnosis was possible from the clinical condition and the exclusion of other differential diagnoses, as well as the performance of the neuroimaging examination, which allowed the identification of the lesions.

Miscelânea

TL 1106512

REPORT OF FORAMEN MAGNUM MENINGIOMA

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Case Presentation: Patient, female, 43 years old, evaluated with a complaint of neck pain and headache that started seven months before. Her neck pain got worse after 3 months, leading her to use a lot of analgesics, without improvement. She was evaluated in other service and a brain CT was performed, without alterations. The condition evolved progressively with pain and weakness in the Right Upper Limb (RUL) and Left Upper Limb (LUL), in addition to low back pain, pain like shocks in the Left Upper Limb (LLL) and difficulty in evacuating. On physical examination, cranial nerve pair testing was obtained without abnormalities. The strength examination (using MRC Scale graduation) showed grade 4 in RUL and grade 5 in LUL, grade 4 proximal in Right Lower Limb (RLL) and grade 5 distal in RLL and LLL. She had hypoesthesia on the right side of the body and her deep osteotendin reflexes were exalted with bilateral Babinski sign. Brain and cervical MRI were requested, revealing an expansive lesion in the craniocervical transition, compressing the spinal cord and the medulla, suggestive of a foramen magnum meningioma prompting neurosurgery evaluation. A complete tumor resection was performed, leading to a great clinical improvement. **Discussion:** Meningiomas originating from the foramen magnum (FMM) are uncommon, being diagnosed in only 1.8% to 3.2% of intracranial meningiomas, with a higher prevalence in females. They present a slow growth pattern with insidious clinical symptoms and late diagnosis. They are classified as ventral, ventrolateral and dorsal, which is essential for the conduct in an integrated manner. The clinic of patients with foramen magnum tumors is diverse, encompassing headache, neck pain, upper limb pain, weakness and low back pain. Magnetic resonance imaging (MRI) is the gold standard test for diagnosis. Surgical treatment is indicated for growing and symptomatic tumors. **Final Comments:** The FMM has a variable clinical presentation, making its diagnosis challenging. The physical examination can be an important clue when bilateral signs of upper motor neuron lesion are present, as it is an important differential diagnosis of spastic paraparesis. Clinicians must be aware that asymmetric sensory abnormalities, as seen in this case, can happen as a result of non-uniform tumor growth and partial compression of spinothalamic tract.

Miscelânea

TL 1106526

INCOMPLETE CAUDA EQUINA SYNDROME SECONDARY TO TARLOV CYST HEMORRHAGE – CASE PRESENTATION

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Case Presentation: A 37-year-old male patient presented sudden onset of severe sacralgia (VAS 9), with paresthesia and allodynia in the right buttock, which radiated to the posterior aspect of the thigh, making it impossible for him to sit down. Five days later, he progressed to urinary and fecal retention and erectile dysfunction. There was no decrease in strength, coordination, or other changes in the neurological examination. MRI of the lumbosacral spine and pelvis showed multiple tubular images in sacral projections with hypersignal on T2 and hyposignal on T1, suggesting multiple Tarlov cysts (Perineural Cyst – PC), in addition to pelvic and perianal varicose veins, edema adjacent to the coccyx and fluid in the pelvis. An area of 50x25 mm was detected in pre-sacral projection at the S4 level, with hyposignal on T2, isosignal on T1 with peripheral areas of hypersignal, without restriction to diffusion, indicating hemorrhage in a PC with compression of the cauda equina. Before hemorrhage, the patient was being treated for hemorrhoidal disease with the drug combination diosmin and hesperidin for two months, possibly contributing to the bleeding of the PC. The patient underwent conservative treatment, with neuropathic pain and anxiety control with duloxetine, pregabalin, chlorpromazine and clonazepam. After 30 days there was an important spontaneous reduction of the cyst to 20x10 mm, with improvement in excretory functions, erectile dysfunction and pain. **Discussion:** Tarlov cyst consists of the formation of saccular dilatations filled with cerebrospinal fluid, located in the nerve sheaths of the posterior roots of the spinal nerves, mainly the sacral roots. It has a prevalence of 1-4%, with asymptomatic clinical course, being symptomatic in less than 1% of cases, due to pressure from the cyst on the nerve roots, caused by trauma, ischemia, inflammation, or hemorrhage, the latter being rare, with few cases reported in the literature. Symptoms can be back pain, sensory and motor radiculopathy, changes in excretory functions, dyspareunia and sexual dysfunction, indicating the need for clinical or surgical treatment. **Final Comments:** This case highlights a rare complication of incomplete cauda equina syndrome secondary to Tarlov cyst hemorrhage, after treatment of hemorrhoidal disease, demonstrating the need to include PC in lumbosacral compressive etiologies, for rapid diagnosis, appropriate therapeutic intervention, and reversal of neurological symptoms.

Miscelânea

TL 1106566

DENTATE NUCLEUS HYPERINTENSITY: AN IMPORTANT DIAGNOSTIC CLUE IN A WOMAN WITH ENCEPHALOPATHY

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Case Presentation: A 56-year-old woman with invasive ductal carcinoma was admitted to the gynaecology department with a 2-week history of somnolence, weakness, mental confusion and reduced fluency of speech. Two months before onset of current symptoms, she had been hospitalized due to a febrile neutropenia, herpes-zoster and a local infected large ulcer on her right hemithorax. Because of this skin lesion, topic metronidazole was prescribed. About two weeks later, she started with the referred symptoms. Metabolic causes were discharged and the only alteration was a pleural effusion with unproven malignancy involvement. Neurologic evaluation was requested and a severe encephalopathy without focal deficits was diagnosed. Brain magnetic resonance imaging (MRI) revealed T2/FLAIR hyperintensities in dentate nuclei and white matter nonspecific lesions. Cerebrospinal fluid was normal and electroencephalogram showed nonspecific findings. The diagnosis of metronidazole induced encephalopathy was made because of history taking and MRI patterns. Hence, the antibiotic was immediately suspended. **Discussion:** Encephalopathy may be caused by toxic, metabolic, infectious and autoimmune mechanisms. Metronidazole-induced brain toxicity can cause altered mental status, seizures and cerebellar ataxia with a wide variation of doses and time of use. In up to 93% of cases of this intoxication, MRI shows bilateral T2/FLAIR hyperintensities in dentate nuclei, which is a very specific pattern. The radiologic differential must be taken with inborn errors of metabolism, totally discharged in the presented case because of the patient's past medical history. Hence, we assume that the extent of the ulcer being treated with topic metronidazole had great cutaneous absorption and within some weeks the antibiotic intoxication led to the current symptoms. In such cases, the antibiotic must be promptly suspended and most patients show mental status improvement. **Final Comments:** The aim of this case presentation is to present an uncommon diagnosis in which MRI pattern recognition and a well-taken anamnesis are essential to the final diagnosis. The faster the metronidazole suspension, the better the patient's prognosis. Neurologists must be familiarized with this cause of encephalopathy.

Miscelânea

TL 1106576

CASE PRESENTATION PATIENT, 24 YEAR, WITH VOGT KOYANAGI HARADA (VKH) INCOMPLETE SYNDROME AND RADIOLOGICALLY ISOLATED SYNDROME (RIS)

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Case Presentation: Female, 24 years old, previously healthy, presented in december 2021 an acute posterior uveitis associated with serous retinal detachment, tinnitus and vertigo. Ophthalmological evaluation was performed with suspected Vogt Koyanagi Harada (VKH) syndrome, treatment with intravenous methylprednisolone was initially with partial response. It remained without maintenance treatment. Two month later she was with worse ocular complaints, binocular involvement, that didn't get better after oral corticoid administration. Magnetic resonance imaging (MRI) orbits showed thickening and enhancement of the posterior contour of the right eyeball (suggestive of VKH) and brain MRI exhibit hypersignal areas in white matter mainly periventricular as well as infratentorial in FLAIR and T2 sequences and post-gadolinium enhancement of two lesion (compliant with criteria for Radiologically isolated syndrome (RIS)). No oligoclonal bands were detected. The patient had no cutaneous lesions. **Discussion:** VKH syndrome is a chronic disorder characterized by inflammation of the uvea, meninges, auditory system, and integumentary system. It may present clinical and/or radiological that can mimic autoimmune demyelinating diseases of the central nervous system (CNS) such as Multiple Sclerosis (MS). Briefly the revised diagnostic criteria for VKH include: 1-absence of penetrating ocular trauma; 2-absence of evidence of concomitant eye disease; 3-bilateral uveitis; 4- neurological and auditory manifestation; 5- cutaneous alteration. We described a case of incomplete VKH disease (based on diagnostic criteria) in which the brain MRI demonstrated demyelinating white matter abnormalities compatible with the revised 2017 McDonald Criteria for dissemination in space. In the absence of clinical demyelinating episodes it was classified as a RIS. Besides the treatment and prognosis for both conditions are different, the early and accurate diagnosis is very important to avoid complications of delayed diagnosis or unnecessary medications. However, differential diagnosis of the combination of uveitis and CNS demyelination may include a systemic or CNS vasculitis, systemic lupus erythematosus, Behcet's disease, sarcoidosis, and paraneoplastic disorders. More studies in the area need to be carried out. **Final Comments:** A rare case presentation of incomplete VKH disease in which the brain MRI demonstrated demyelinating white matter abnormalities compatible with RIS.

Miscelânea

TL 1106583

NEURO-ONCOLOGICAL PERSPECTIVE IN A PATIENT WITH BONE METASTASIS FROM FOLLICULAR THYROID CARCINOMA

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Case Presentation: A 31-year-old female patient underwent a total thyroidectomy 5 years ago for thyroid tumor resection. Recently, she complained of back pain, having a palpable mass at the site. Hypoesthesia was also noted in the right T10 dermatome. 4 months before the neurological consultation, she performed a CT scan in which an osteolytic expansive lesion was observed on T10 in the proximal portion to the right costal arch. Then an MRI was requested, in which a large tumor was found, reaching the vertebral body of T10 and extending to the right thoracic cavity, so a giant cell tumor was suspected. Thus a biopsy of that tumor was requested and the result was compatible with follicular thyroid carcinoma metastasis. Then, tumor resection surgery was performed, including T10 corpectomy and arthodesis with pedicle screws from T7 to L1. Besides, the patient is being followed up with oncology and is awaiting screening tests, but the scintigraphy did not show any other bone lesions. **Discussion:** Thyroid cancer can evolve to its distant metastatic state, such as bone metastases. In a hematogenous route, a carcinoma can lead to bone metastasis, with this malignancy occurring in 2-13% of cases of DTC (Differentiated Thyroid Cancer). This specific type of metastasis is more prevalent in follicular cancer (7-28%) than in papillary cancer (1.4-7%). Bone metastases represent a frequent complication, specially in follicular thyroid carcinoma, and cause pain, fractures and spinal cord compression, also is observed hypoesthesia in some dermatomes, so the patient's quality of life is severely reduced. The diagnosis is established by correlating clinical suspicion with neuroimaging, such as whole-body and skeletal MRI, which are essential to locate and assess the extent of lesions. As an adjuvant treatment, radioiodine therapy is indicated to destroy malignant thyroid tumor cells left over from total thyroidectomy. Thus, it eliminates locoregional micrometastases and distant metastases. **Final Comments:** Since scintigraphy excludes the possibility of another bone lesion, the patient has a good prognosis. According to the neuro-oncological literature, complete remission reaches 20% of clinical cases of bone metastases resulting from follicular thyroid carcinoma, reiterating the matter of radioiodine therapy as an adjuvant treatment. Finally, a clinical improvement in the patient under analysis could already be observed, as the hypoesthesia of the T10 dermatome was reversed.

Miscelânea

TL 1106602

CASE PRESENTATION WILSON'S DISEASE ON A 15-YEAR-OLD BOY WITH SEVERE NEUROLOGICAL IMPAIRMENT AND RARE CORPUS CALLOSUM INVOLVEMENT ON BRAIN MRI

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Case Presentation: A 15-year-old boy presented with a 6-month history of progressive loss of dexterity in his right hand, manifested by difficulty writing and also dysarthria, dysphagia, generalized feeling of weakness and difficulty walking, dragging his right leg in the past months, slowness and shaking of hands. He was under thrombocytopenia on investigation. His parents are non-consanguineous and he has four older siblings and five other half siblings, all of them with no medical problems. He was disoriented in time and place. He also had hypomimia, micrographia, dysarthria, rigidity and bradykinesia on four limbs and also dystonia on both hands and cervical muscles. Lab investigations showed leukopenia with lymphopenia, thrombocytopenia, mild elevated GGT and elevated AST levels. His liver function was impaired, with prolonged RNI, normal albumin and bilirubin levels, but abnormal abdominal ultrasound, demonstrating chronic parenchymal liver disease and splenomegaly. Endoscopy showed esophageal varices without red spots. Serology such HIV, syphilis, HBsAg and HCV Ab were negative or normal. Ophthalmologic examinations were suggestive of Kayser-Fleischer rings on the initial stage. On laboratory low concentrations of serum ceruloplasmin (2.9 mg/dL, normal 20-63), low serum copper (<0.25g/dL, normal 80-140) and increased daily urinary copper excretion (157 µg/24 h, normal 0-60). Gene sequencing later identified ATP7B c.3061-12T>A (intronic) / ATP7B deletion (exon 7), confirming Wilson's Disease. MRI of the brain showed bilateral and symmetric T2 hyperintensity in basal ganglia (especially putamen), mid-pons, cerebellar peduncles, thalamus, which are typical regions of involvement for WD, but also showed and evident hyperintensity in the splenium of corpus callosum, an uncommon finding in the disease. The patient was started on biperiden 2mg/8h for symptomatic control and we prescribed D-Penicillamine as a treatment plan. Parkinson-like syndrome is the most common neurologic presentation of Wilson Disease, especially in adolescents. On Brain MRI the most common finding is T2 hyperintensity in putamen and involvement of other basal ganglia, pons, thalamus and the midbrain. Midbrain involvement led to the classic "face of the giant panda sign". Is it uncommon the involvement of frontal, parietal, temporal lobes, cerebellum and corpus callosum, the last being the rarest site of lesion. Corpus callosum involvement was associated with more severe neurological and psychiatric dysfunction, more extensive brain lesions and longer course of disease.

Miscelânea

TL 1106613

OPTICAL NEUROTOXICITY BY TOLUENE

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Case Presentation: A 32-year-old male, illicit drug user, was evaluated for bilateral visual loss after inhaling large amounts of thinner and accidentally ingesting the product, in addition to touch his eyes with the product's dirty hands. A day after the event, he developed bilateral intense retrocular pain that worsened with eye movement, associated with photophobia and diffuse myalgia. He evolved with significant loss of visual acuity, remaining with only light perception. He was assessed by an ophthalmologist who did not find ocular or retinal changes. Eight days later the accident, he was then referred to this neurology service with non-reactive mydriatic pupils, bilateral amaurosis, with only light perception. Cranial CT was normal and CSF showed high protein levels. He was subsequently diagnosed with a bilateral optic nerve damage due to optic neurotoxicity caused by toluene present in thinner. **Discussion:** Thinner is a solvent for paints and varnishes, used in automobiles paint and body shops. Its main compound is toluene, a fat-soluble which is rapidly absorbed by the lipid-rich CNS, its primary target. Its distribution in the body and excretion are quite rapid. All the absorbed compound is almost completely eliminated from the body 18 to 20 hours after exposure. Acute manifestations of toluene toxicity include euphoria, headache, weakness, vision disturbances, tremors, confusion, ataxia, paralysis, epileptic crisis, narcosis, and death. In addition, toluene optic neuropathy may occur, which leads to a slow and progressive decline in visual acuity. The initial appearance of the optic disc is usually normal, but there is gradual optic atrophy. **Final Comments:** Hippuric acid is a biological indicator of exposure to toluene, excreted in the urine. However, some authors claim that there is no diagnostic value in its dosage, as it is a normal constituent of urine. Thus, after discussing the condition with the toxicology team of a reference Hospital, it was decided not to measure this marker, since the main suspicion would remain, even with a negative test. A fat-suppressed MRI of the skull and orbits were performed, showing discreet tortuosity of optic nerves and peri sheath fluid at the insertion of the eyeball, suggestive of neuritis. He underwent pulse therapy for 5 days with methylprednisolone, without clinical improvement.

Miscelânea

TL 1106633

LEPTOMENINGEAL NEOPLASTIC SYNDROME: A CASE PRESENTATION

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Case Presentation: E.D.S; female, 65yo, white, smoker (50 pack-years) with consumptive syndrome and report of cognitive decline, behavioral and gait disorders, with a slowly progressive course plus a recent episode of exacerbation and possible diagnosis of Normal pressure hydrocephalus (NPH). The patient presented, in the last 5 months, worsening of the symptoms associated with headache, evolving with dependence for activities of daily living. In addition, he presented hearing deficit with a unilateral sensorineural pattern on the right, in addition to central vestibular dysfunction and left third cranial nerve paresis. The patient sought care when she was Hospitalized and tonic muscle spasms were observed with associated loss of consciousness, in addition to an increase in serum inflammatory and infectious markers and in the cerebrospinal fluid (Hyperproteinorrachia with hypoglycorrhachia and slightly increased cellularity; BAAR and GeneXpert negative). Magnetic resonance imaging showed leptomeningeal enhancement, mild dilatation of the supra and infratentorial ventricular system, associated with signs suggestive of transependymal CSF transudation and ventriculitis. Computed tomography of the abdomen, pelvis and neck were performed in order to promote neoplastic screening, however the results were normal. Cervical ultrasound showed lymph nodes increased in number in left cervical chains. A leptomeningeal biopsy was performed and the result consisted of infiltration by atypical cells with the presence of KI67 marker in 100% of the cells of interest. The patient had an unfavorable outcome and died within 1 month. **Discussion:** Despite a clinical course suggestive of NPH, the history of smoking and the consumptive syndrome, associated with diffuse meningeal manifestation and characteristic cerebrospinal fluid profile, showed a probable neoplastic syndrome in the central nervous system, associated with the KI67 marker, which is more frequently expressed in upper respiratory tract epidermoid carcinomas. **Final Comments:** The presence of hypoglycorrhachia in the diagnostic context of meningoencephalitis should expand the possibilities of differential diagnosis, with neoplastic diseases among the hypotheses. In the case in question, the histochemical finding allowed the correct diagnosis of meningeal carcinomatosis, with the central nervous system being an uncommon site for metastatic implantation of squamous cell carcinomas of the upper respiratory tract.

Miscelânea

TL 1106562

EPIDEMIOLOGICAL ANALYSIS OF PATIENTS WITH DIFFUSE GLIOMAS TREATED IN PARAÍBA

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Background: Diffuse glioma (DG) is the most frequent and aggressive type of malignant tumor of the Central Nervous System with high morbidity and mortality. Epidemiological data on patients with diffuse gliomas in Brazil are scarce. **Objective:** To characterize the epidemiological profile of patients with gliomas in Paraíba. **Method:** This is a cross-sectional, retrospective observational study that included participants of both genders, over 18 years of age, with DG diagnosed and treated in João Pessoa. **Results:** The sample consisted of 28 patients, with a mean age of 59 years. The most relevant symptom at diagnosis was seizure (46.15%), followed by headache (23%), progressive forgetfulness (23%), aphasia (12%), progressive visual loss (8%), and hemiparesis (8%). Astrocytomas were the most prevalent type of tumor (16 patients), with grade 4 being the most prevalent of them. There were 5 glioblastomas multiform (GBM) diagnosed, all in males, while in astrocytomas, only 37.5% were males. The mean KPS scale was 81.15 with only 19.2% (5 patients) having KPS below 70, and the mean overall survival was 41 months. Patients with KPS greater than 85 had better overall survival (57.8 months) than patients with KPS less than 85 (36.4 months). When we associated age and overall survival, we noticed that patients younger than 45 years had a mean overall survival of 105 months, while patients aged 45 years or older had a mean overall survival of 30.9 months. None of the five patients with GBM survived after 5 years (mean overall survival of 10 months). As for Astrocytomas, 37.5% of patients were alive after 5 years (6 patients, with a mean overall survival of 46.9 months). As for the other gliomas, 2 of the 5 patients were alive after 5 years (median overall survival 74.7 months). **Discussion:** Current literature highlights that patients with older ages (>45 years) have a worse prognosis than younger patients. In addition, KPS <85 is also an important predictor of unfavorable outcomes, and may increase mortality. In contrast, low-grade gliomas (1 and 2) reduce mortality by up to 22%. In GBMs, the 5-year survival after diagnosis is less than 5%. **Final Comments:** The data brought by this study corroborate the current literature, in addition to reinforcing the influence of age, KPS at diagnosis and histological type as prognostic factors, assisting in decision making by health professionals.

Neuroepidemiologia

TL 1104697

CASE PRESENTATION LEFT FRONTAL EXTRA AXIAL TUMOR CAUSED BY EPITHELIAL NEOPLASM OF THYROID FOLLICULAR ORIGIN

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Case Presentation: From antiquity to the present day, tumors have always aroused curiosity in the field of medicine and instigated science in an obsession to understand its mechanism of action and seek its cure. Neoplasms were initially described around 2600 years before Christ in an Egyptian papyrus containing reports of 48 cases of the disease carried out through the studies of the Egyptian physician Imhotep. It was only centuries later, with the Renaissance movement and the beginning of the modern age in the 18th century, that the first anatomical and histopathological descriptions were made thanks to the development of microscopy and the work of the German physician Virchow and the Belgian anatomist Andreas Versalius. In addition, with the advance of the scientific revolution in the 19th century, factors related to the heredity of tumors as well as their compression of genetic aspects were elucidated through analytical observations and categorical writings developed by the Austrian monk Gregor Mendel and later applied to the study of the genetic basis. Modern. Since then, with the advent of technology, hundreds of scientists have embarked on a mad dash to improve new techniques and innovative treatments that allow us to understand and treat the mechanisms of the disease. In this context, epithelial neoplasms are known to be tumors that can be classified as benign or malignant, originating from a disordered proliferation of follicular cells in tissues influenced by thyroid hormones. Benign follicular nodules are the most common types presented, which contain in their cytological analysis variable proportions of colloid and benign-looking follicular cells arranged as macrofollicles or macrofollicle fragments in some tissues of the body. Therefore, in the **Case Presentation:** in question, a 50-year-old female patient was identified with the presence of a left frontal extraaxial tumor caused by an epithelial neoplasm of thyroid follicular origin.

Neurofisiologia Clínica

TL 1104975

PARRY-ROMBERG SYNDROME – CASE PRESENTATION

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Case Presentation: A 25-year-old female referred to the vascular neurology outpatient clinic after hemorrhagic stroke in July 2021. She complained of daily pulsatile headache associated with nausea and left hemicranial location. Besides progressive deformity in the right hemiface. Physical examination: good general condition, conscious and oriented, afebrile, acyanotic, anicteric and eupneic. Besides left hemiparesis and deformities in the skull, this one being hypotrophy in the right hemiface. Being requested Magnetic Resonance Imaging of skull and subsequently having the confirmation of the diagnosis of Parry-Romberg Syndrome. **Discussion:** The Parry-Romberg Syndrome (PPR) or progressive facial hemiatrophy is a disease of unknown origin, usually develops in the first or second decade of life, prevailing female gender, most often affects only one hemiface, as in the patient's report. It is manifested by a craniofacial atrophy that can affect part or all facial structures from skin, muscle, fat, bone tissue, trigeminal nerve and even the brain parenchyma (PRADO et al., 2011). The SPR can affect the central nervous system, among the neurological manifestations, headache and seizure disorders are the most common. Neuropathies involving several cranial nerves (the third, fifth, sixth and seventh), vascular inflammation and destruction of the bone surrounding the trigeminal nerve causes secondary trigeminal neuralgia in these patients, manifesting with chronic facial pain that is usually resistant to treatment (ARIF et al., 2020) Computed tomography and MRI are methods for diagnosing CNS changes, with the characteristic findings being: white matter hypersignal on T2 and FLAIR, leptomeningeal enhancement, and brain atrophy, being found on the patient's skull MRI. The differential diagnosis includes diseases that course with cerebral hemiatrophy, such as Rasmussen's encephalitis and Sturge-Weber syndrome, but in these we did not observe the typical hemifacial changes found in Parry-Romberg syndrome (PAULA et al., 2014). **Final Comments:** The reported case and exposed publications bring a rather rare diagnosis. As the patient's facial involvement has not yet significantly affected the quality of life of the patient, the professionals responsible for the case did not prescribe a surgical approach.

Neurofisiologia Clínica

TL 1105119

RABIES ENCEPHALITIS: FOLLOW-UP WITH ELECTROENCEPHALOGRAM

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Case Presentation: A 2-years-old boy from the interior of Maranhão was referred for suspected encephalitis. He had a history of fox scratch on the left leg without rabies prophylaxis 2 months ago of symptoms onset (fever, irritability, spasms and incoordination). The diagnosis of rabies was made through splitte and PCR test. CSF (9th day) and brain MRI (15th day) were initially normal. The CSF (16th day) showed WBC 106; PMN: 76% Prot: 117; Glucose: 35. Serial evaluations were performed with electroencephalogram (EEG), which showed: The EEG was performed under continuous sedation on the 15th, 18th and 22nd day.; 15th day: Diffuse slowing with the presence of frontocentral spindles; 18th day: Burst-suppression pattern, consisting of delta and theta activity with frontal accentuation and maximum interburst interval of up to 5 seconds; 22nd day: Activity suppression. Patient evolved with the absence of brain stem reflexes; 32nd day: Burst-suppression. Its bursts were constituted of rhythmic theta activity (5 to 7 Hz) in the frontocentral region, with a lack of alpha and beta rhythms. The maximum interburst interval was 10 seconds. 38th day: Activity suppression and occasional bursts of rhythmic theta activity in the left frontocentral region. The last brain MRI showed: areas of diffuse hyperintensity in the cortex and basal ganglia on T2 and Flair without expression in the diffusion sequence. The patient died on the 51st day of symptoms onset. **Discussion:** Rabies encephalitis is caused by a Lyssavirus. It is transmitted through the bite or scratch of mammals carrying the disease. Despite reports of survivors, the mortality of the disease is close to 100%. There are descriptions of multiple EEG patterns of this encephalitis, such as: slowing, periodic discharges, isoelectric pattern mimicking brain death, among others. None of them are specific to the disease. In the present case, it was possible to follow, through the electroencephalogram, the catastrophic evolution of the disease, being observed patterns of progressive hypometabolism, as respectively: diffuse slowing, burst-suppression and activity suppression. The EEG is not indicated for the diagnosis of brain death in this population. **Final Comments:** In rabies encephalitis, there are no specific findings on EEG. It is used as a non-invasive method at the bedside to identify subclinical seizures, monitor the level of sedation and follow-up the patient's clinical course.

Neurofisiologia Clínica

TL 1105148

AUTONOMIC NEUROPATHY FOLLOWING COVID-19 VACCINATION

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Case Presentation: Patient, male, 24 years, presented in emergency service following a 4-day history of intense abdominal pain with associated nausea, after the first dose of a mRNA COVID-19 vaccine. Upon interrogation, he stated that he had not defecated for 2 days and reported having difficulties urinating. On physical examination, he was alert, with normoactive reflexes, without appendicular and facial motor deficits, and apparent hypoesthesia in his left lower limb, with normal deep sensitivity. In a later evaluation, he reported sporadic urinary retention, requiring effort to initiate diuresis. He denied nausea. Mentioned intestinal colic with diarrhea and hypotension, reporting episodes of dizziness when standing in orthostasis. Laboratory tests showed no changes. Electroneuromyography showed absence of plantar cutaneous reflex in the right foot region, which is compatible with suspicion of autonomic neuropathy. After 10 days from the onset of symptoms, the patient evolved with clinical improvement and normal, pain-free urination. **Discussion:** Autonomic neuropathy is a condition where the nervous connections of the autonomic nervous system are affected, causing denervation that has repercussions on various body systems, such as the gastrointestinal, urinary and cardiovascular. There are several associated etiologies, as well as different pathophysiological. The current literature already has published case studies associating the COVID-19 mRNA vaccine with autonomic dysfunction. However, most cases mention Guillain-Barré Syndrome, an autoimmune disease, in which autonomic neuropathy is a secondary complication, not the main symptom. Primary autonomic dysfunctions can occur as an autoimmune complication of viral infections, including being associated with the post-COVID-19 syndrome itself. As an adverse post-vaccination effect, it has already been reported in studies after immunization with the quadrivalent HPV vaccine, but with regard to COVID-19 vaccines, so far only two cases have been reported worldwide, one with a recombinant vaccine and another with a mRNA vaccine. **Final Comments:** The patient's clinical history together with the complementary exams performed confirmed the diagnosis of autonomic neuropathy, which by the chronology of the facts can be attributed to a rare complication of the vaccination. Furthermore, the authors believe that further studies on the comorbidities of vaccination for COVID-19 will may explain its complete pathophysiology.

Neurofisiologia Clínica

TL 1105833

MYOPATHY MISS DIAGNOSED AS AMYOTROPHIC LATERAL SCLEROSIS FOR SEVEN YEARS – DIAGNOSTIC CHALLENGES OF NEUROMUSCULAR DISEASE

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Case Presentation: Male, 56 years old, was referred to EMG lab by a rheumatologist for evaluation of his Amyotrophic Lateral Sclerosis (ALS). He arrived with complaints of weakness and sadness because he received a diagnostic seven years ago. Since the diagnostic, he was expecting his physical decline, as said by his attendant. It slowly evolves. That moment was the first time some doctor asked him to repeat the electromyography (EMG) test after the first two diagnostic tests. Physical exam: Medical research council (MRC) strength in the triceps and the quadriceps was IV, He had prominent atrophy of facial, arms and legs muscles. Sensory examination was markedly normal. The two previous EMG reports demonstrated prominent fibrillations (Fibs) and positive sharp waves (PSWs) with increased motor unit potentials (MUPs) and with a final statement of motor neuron disease. We performed a new EMG that showed Fibs and PSWs. Interesting that was no high amplitude MUP, instead, we are able, with quantitative EMG, to show a great number of small amplitude and short duration (SASD) MUPs, consistent with active myopathy. Muscle biopsy show inflammatory infiltrate. Corticosteroids were started and the patient slowly evolves with significant recovery of symptoms. **Discussion:** The ALS diagnostic is a dramatic report to be given to any patient. The findings of widespread active denervation and chronic reinnervation in three different corporal segments were easily obtained in this case. The normal sensory examination also corroborates for the establishment of the diagnosis of ALS. High amplitude and long duration MUPs also were there. The patient stayed waiting for his slow decline and it was a surprise when he shows up with seven years of ALS history walking with some difficulty but without any assistive device and speaking normally. To us, it was an easy diagnostic as the patient still has widespread positive sharp waves with fibrillation potentials, small amplitude short duration MUPs and no HALD MUPs. Many reasons can throw us in the wrong diagnostic direction in this case. In Brazil, in our lab, we see too many HALD without reasonable explanation in a way that we don't have seen in the USA. As in this case, the true cause of weakness was hidden under HALD MUPs. **Final Comments:** We encourage the routine use of quantitative EMG techniques to discover and quantify the SASD that otherwise can pass masked as seen in this case.

Neurofisiologia Clínica

TL 1106280

ABDOMINAL PAIN AND FLACCID TETRAPARESIS IN A YOUNG ALCOHOLIC MAN: AN ATYPICAL CASE OF ACUTE INTERMITTENT PORPHYRIA (AIP)

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Case Presentation: A 38-year-old man is admitted in January-2022 with worsening abdominal pain that started 4 months ago. The pain was triggered by alcohol ingestion, associated with abdominal distension, nausea and vomiting. He was Hospitalized. He realized a computed tomography (CT) scan of the abdomen, which showed distension of the loops, without obstruction. He was previously healthy, had been drinking for 10 years, denied smoking and illicit drugs, hadn't had a similar episode nor family history. No urinary changes. He had seizures, psychomotor agitation, dysphagia, dysphonia, diparesis, signs of dysautonomia and progressive proximal paraparesis, progressing to areflexic flaccid tetraparesis. Normal brain CT, cerebrospinal fluid (CSF) study with 3 erythrocytes, 2 cells, glucose 85, proteins 95, LDH 25 and electroneuromyography (ENMG): a predominantly sub-acute motor axonal polyneuropathy of the four limbs with F waves and reflex normal H. He evolved with respiratory failure, was intubated, and plasmapheresis was initiated, as it was not possible to exclude an acute polyradiculoneuropathy. The hypothesis of AIP was also raised, and the urinary dosage of porphobilinogen (UDPBG) and genetic tests were positive. **Discussion:** Acute hepatic porphyrias (AHP) are those in which enzyme deficiency occurs in the liver, with AIP being the most common. AHP mainly affects the central and peripheral nervous system by increasing the production of neurotoxic precursors (delta-aminolevulinic acid and porphobilinogen). It is an acute predominantly motor axonal neuropathy similar to Guillain-Barré syndrome (GBS), which usually occurs after muscle pain and weakness commonly preceded by abdominal pain and psychiatric disorders that can progress to quadriplegia and death. Alcoholism, sex, negative family history, and protein-cytologic dissociation in the patient's CSF made the diagnosis challenging. **Final Comments:** ENMG may be helpful in differentiating GBS from porphyric neuropathy with GBS showing absent H reflex and F waves or with prolonged latency, in contrast to the relative preservation of the H reflex and F wave in porphyric neuropathy, as well as prolonged distal latency and block. conduction block on GBS with normal latency and without conduction block on AHP. The definitive diagnosis of AIP is made with DUPBG and genetic testing, both positive in our case.

Neurofisiologia Clínica

TL 1106294

THE IMPORTANCE OF ELECTRONEUROMYOGRAPHY IN THE DIFFERENTIAL DIAGNOSIS OF ACUTE FLACCID PARALYSIS

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Case Presentation: A 53-year-old hypertensive pharmacist reports that 2 months ago he began insidiously with bilateral paresthesia associated with mild brachial diparesis. In the last 2 weeks before admission, patient reports worsening of the condition with paresis in the 4 limbs, predominantly brachial and proximal. In the evaluation, the patient was alert, without alterations in the cranial nerves, reduced strength in the upper limbs (Grade I) and in the proximal segment of the lower limbs (Grade III) and with areflexia. In the laboratory analysis, the cerebrospinal fluid showed cellularity of 38 (90% lymphomononuclear), proteins of 36 and glucose 70; in serum tests, the only change was VDRL reagent in the titer of 1/128, when FTA-Abs was requested (which later turned out to be negative). In the imaging evaluation, cranial and cervical spine resonance was performed without significant changes. Therefore, Electroneuromyography (ENMG) was requested and the condition was initially managed as neurosyphilis, until there was no therapeutic response and ENMG proved to be a sensory-motor syndrome with a demyelinating pattern. Based on the hypothesis of Guillain-Barré Syndrome, he underwent treatment with Human Immunoglobulin and showed significant improvement in the condition, with improvement in sensitivity and strength in all segments. **Discussion:** The use of Electroneuromyography as an auxiliary tool in the diagnosis of neurological diseases is already a reality. However, ENMG has been shown to be particularly useful in the diagnostic distinction between acute flaccid paralysis, being a fundamental element for more adequate therapeutic guidance. **Final Comments:** In the same way that complementary exams can, on the one hand, help the diagnosis, on the other hand, they can delay it due to numerous diagnostic possibilities that remain open. However, the ENMG, when in correlation with the clinic, has been shown to be a valuable element in the therapeutic decision

Neurofisiologia Clínica

TL 1106413

HEREDITARY AXONAL MOTOR NEUROPATHY DUE TO DYNC1H1 GENE MUTATION, NEW FORM OF MARIE TOOTH CHARCOT DISEASE

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Case Presentation: C.A.A, male, 53 years old, hypertensive and asthmatic. He reports adequate neuropsychomotor development and at 6 years of age he began to have difficulty walking, due to distal weakness in the lower limbs and the presence of left tendon retraction. At age 18 he walked with unilateral support. Referred for evaluation at age 49 years after performing electroneuromyography with the presence of neurogenic motor unit potentials in the soleus and tibialis anterior muscles, with anterior tip potentials and absence of current denervatory activity. A genetic test was performed for progressive spinal amyotrophy and showed no deletions in the SMN1 gene. Thus, the investigation was continued with hereditary neuropathy panel and evidenced a heterozygous mutation in the DYNC1H1 gene (OMIM* 600112). This variant is associated in this case with axonal type 20 Charcot-Marie-Tooth disease. Referred to motor rehabilitation, with improvement in ambulation afterwards. Thus, the investigation was continued with the hereditary neuropathy panel and evidenced a heterozygous mutation in the DYNC1H1 gene (OMIM* 600112). This variant is associated in this case with axonal type 20 Charcot-Marie-Tooth disease. Referred to motor rehabilitation, with improvement in ambulation. **Discussion:** Hereditary neuropathy encompasses a group of genetic disorders with a phenotypic spectrum that ranges from oligosymptomatic to severe disability. Charcot-Marie-Tooth disease (CMT) is the most common form of hereditary neuropathy, and it may exclusively affect peripheral nerves and produce symptoms of their dysfunctions. **Final Comments:** The advancement of genetic testing allowed new mutations to be discovered or those that were previously not considered pathogenic to become later. In the case described, it was discovered that the patient's variant could manifest with the Charcot-Marie phenotype or spinal muscular atrophy predominantly in the lower limbs type 1. Thus, given the clinical picture and the neurological examination, it was associated with Charcot-Marie, this is a new form of the disease.

Neurofisiologia Clínica

TL 1106453

SEIZURE CONTROL INDUCED BY TACTILE STIMULI, NOT ALWAYS A CONVERSION DISORDER

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Case Presentation: A 50 years old male, with previous medical history of systemic arterial hypertension and diabetes mellitus, presented a 20 days onset of paroxysmal abnormal movements at left side limbs, with 5 minutes duration, and ataxia in between episodes. At neurological evaluation, a complex movement disorder was observed, with predominantly hemiballism, but clonic-tonic paroxysmal seizures, with 2 minutes duration and EEG electric ictal activity, were observed. These seizures ceased clinically and electrographically with tactile stimulus. Excluding structural and other metabolic causes after normal brain MRI, CSF analysis and blood tests, chemical control of blood glucose with insulin showed improvement in clinical symptoms. Patient was discharged and in follow-up was asymptomatic. **Discussion:** Separately, seizures and hemiballism are an uncommon complication of non cetotic hyperglycemic states. In this case presentation, we showed a very rare complex movement disorder, with components of both clinical conditions. More importantly, although pathophysiologically explained in few studies, with cases in which seizure cessation was obtained by auditory stimulus, both clinically and eletrographically, with tactile stimulus have no case presentation, being the presented above unique. **Final Comments:** Although rare, hyperglycemia induced seizures and movement disorders must be in differential diagnosis, especially because they can be easily treated. Furthermore, more data about seizures inhibitor effects of sensorial stimulus are needed and possibly supports the development of new therapies.

Neurofisiologia Clínica

TL 1104807

LABRUNE SYNDROME: CASE PRESENTATION OF AN RARE GENETIC ENCEPHALOPATHY

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Case Presentation: Our patient is a young man with a normal development and previously healthy until 30 years old, when he presented a clinical picture of a subacute intracranial hypertension characterized by progressive headaches, vomiting, gait difficulties and papilledema. At the time, he was seen by a neurosurgery team at another Hospital, and after going through a CT scan and a brain MRI, was suspected to have an intracranial tumor of the right frontal lobe. He underwent surgery for drainage of the lesion and a biopsy was performed, that later came inconclusive. He responded well to the procedure and was relieved of his symptoms. 2 months later, he came to our Hospital after having two episodes of a focal to generalized tonic clonic seizure. While doing the workup for the seizure disorder, our team reviewed the images and noted an aspect of diffuse leukoencephalopathy with cysts, and asked our pathology team to look at the biopsy, noting and chronic gliosis with Rosenthal fibers. The patient underwent and genetic testing that excluded COATS syndrome. Overall, the findings were consistent with Labrune Syndrome, an extremely rare form of genetic encephalopathy. The patient remained asymptomatic with seizures controlled by low doses of antiepileptic drugs for 7 years, when he again presented with a clinical picture of intracranial hypertension and imaging exams revealed formation of new cysts over the cerebral hemispheres, cerebellum and brainstem. The neurosurgery team at our Hospital decided to place a ventriculoperitoneal shunt this time, for which the patient responded well. At the same time, the family agreed to do genetic testing specific for Labrune Syndrome, which confirmed the diagnosis. Labrune Syndrome is a rare genetic disorder that is characterized by the triad of encephalopathy, brain cysts and calcifications. It was described first by Labrune et al. in 1996, and was for a while thought to be related to the syndrome of cerebroretinal microangiopathy with calcifications and brain cysts, but later the genetic mutation specific for Labrune was discovered to be the SNORD 118 mutation, and these patients, as our own, do not have retinal telangiectasias as would be expected in the COATS syndrome. Due to its rarity, there are no large studies that give information about natural history, life expectancy and prognosis of the syndrome.

Neurogenética

TL 1104874

LATE-ONSET ATYPICAL PARKINSONISM AND MOTOR NEURONOPATHY RELATED TO POLG GENE VARIANT

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Case Presentation: A 60-year-old patient presented with a 4-year-history of stiffness and slowing of the left upper limb, and shortly afterwards in the left leg. He also developed neck stiffness, urinary retention, and weight loss of 10kgs. The patient had no sleep complaints, REM sleep behavior disorder, cognitive complaints, or anosmia. Divergent strabismus in the right eye was present since childhood. Neurological examination showed dysarthria, exotropia of the right eye, and facial hypomimia. Asymmetric bradykinesia more pronounced on the left side. Festinating gait with significant postural instability and camptocormia without ataxia. Treatment was initiated with levodopa 1000mg and pramipexole 1.5mg daily with partial clinical response and wearing off after 1h30min. Cranial CT disclosed punctiform calcification of the basal ganglia. Brain and spinal cord MRI showed marked calcification in the basal ganglia. DaT-SPECT showed asymmetric reception of the radiotracer right less than the left side. Electrodiagnostic studies showed chronic preganglionic involvement in C6-T1 bilaterally and L4-S1 bilaterally. NGS-based gene panel testing was performed, showing the heterozygous variant c.678G>C (p.Gln226His) in the POLG gene. **Discussion:** The human Polymerase Gamma (POLG) gene encodes the mitochondrial DNA polymerase responsible for replication and repair of the mitochondrial DNA. In adults, the most common movement disorder is parkinsonism, occurring in up to 43% of the patients. POLG-related parkinsonism has an earlier age of onset, at about 40 years and is initially asymmetric clinically and in imaging. Tang et al. described Q226H POLG variant as potentially pathogenic. Different from what is seen in the literature our patient had a late-onset parkinsonism and poor response to levodopa. Nevertheless, our patient had early postural instability and camptocormia, that can be the presenting sign in patients with POLG variant, sphincter dysfunction, asymmetric DAT-SPECT findings, and neuronopathy on electrodiagnostic studies. These findings led to the differential diagnosis of a genetic metabolic cause for parkinsonism, and the diagnosis of POLG-related disorder was made. **Final Comments:** Our case further expands our knowledge regarding POLG gene variants and phenotypes, as a highly heterogeneous spectrum, and shows the importance considering it in patients with parkinsonism.

Neurogenética

TL 1104940

ADCY5 RELATED DYSKINESIA PRESENTING AS DYSTONIA-PLUS RESPONSIVE TO CAFFEINE

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Case Presentation: We present a case of a 40 years old male with early childhood onset dystonia, paroxysmal worsening of varying frequency triggered by fasting and improved by coffee consumption, important orofacial dyskinesia and normal cognitive function, neuroimaging and laboratory screenings. This suggestive story brought up the suspicion of a ADCY5 related dyskinesia, confirmed by genetic testing and with excellent response to caffeine with clonazepam. **Discussion:** ADCY5 related dyskinesia is a newly recognised heterogeneous entity with various phenotypes caused by many different pathogenic variants in the ADCY5 gene in chromosome 3q21-3p21. Clinically it is characterized by hyperkinetic movement disorder of early childhood onset including choreoathetosis, myoclonus and dystonia with paroxysmal worsening related to sleep, waking up, fatigue, physiological or psychological stresses, little to no progression and important facial dyskinesia. The majority of cases have normal neuroimaging, normal cognitive function and unrevealing laboratory finds. Some cases are associated with axial hypotonia, painful dystonias, hyperreflexia and proeminent nocturnal paroxysms. This phenotype suggests the R418W mutation as the cause. The two most frequent variants (first the R418W and second the A726T) both occur at conserved residues in the catalytic pocket of the protein, probably resulting in gain of function. Many therapies have been tried in those patients, with conflicting data on response. Treatment with caffeine has been proposed by many authors. The rationale behind this relies on the antagonism of the A2A adenosine receptor by caffeine counterbalancing the theoretical gain of function of ADCY5. There are reports in literature of autosomal dominant heritage, autosomal recessive cases with a more severe phenotype, de novo mutations, mosaicisms and even possible dual diagnosis. The most prevalent seems to be the de novo mutation in R418W. **Final Comments:** This report helps to consolidate the phenotype of ADCY5-related-dyskinesia, to describe a new case of R418W as a causative variant and adds evidence to the safety and efficacy of caffeine in association use with benzodiazepines.

Neurogenética

TL 1104996

A CASE OF ADULT-ONSET LEUKODYSTROPHY OF RARE UNDERLYING AETIOLOGY: THE IMPORTANCE OF EXPANDING TOOLS FOR EARLIER DIAGNOSIS

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Case Presentation: A 43-year-old male developed dementia in the previous three years. He presented with dysarthria, dysphasia, speech and ideomotor apraxia and, mainly, gait apraxia, which was followed by knee walking. On examination there was paratonia, global hyperreflexia despite preserved strength. MRI disclosed diffuse cortical atrophy with unspecific bilateral white matter hypersignal, suggestive of advanced leukodystrophy (LKD). Electroneuromyography was unremarkable, as well as, infectious and metabolic screening. CSF revealed hyperproteinorrachia. Genetic investigation from salivary sample identified one likely pathogenic mutation in ETFDH gene, associated to autosomal recessive multiple-acyl-CoA dehydrogenase deficiency (MADD). Dietary orientation and supplementation with riboflavin were initiated. **Discussion:** MADD impairs the riboflavin-dependent electron transfer system in mitochondria. This genetic mutation compromises the fatty acid oxidation, particularly branched chain ones, and the amino acids metabolism. Therefore, treatment recommendations include: protein and fat-restricted diet every 4 h; conjugation of toxic metabolites with L-carnitine and glycine reposition; supplementation with riboflavin; besides the promising replacement of endogenous ketone body with sodium D,L-3-hydroxybutyrate. Clinical spectrum varies widely depending on the degree of hypoketotic hypoglycaemia, metabolic acidosis and defective protein metabolism. It ranges from lethal malformation to adult onset, throughout a myriad of complications: hepatic dysfunction, cardio/myopathy, neurodevelopmental delay and LKD. In late-onset MADD, it is more prominent the neuromuscular phenotypes, with demyelination directed to peripheral nervous system. On the other hand, LKD in adults shelters others inherited and acquired disorders (e.g. vascular, toxic, degenerative or inflammatory) without precise genotype-phenotype correlation. Symmetrical pyramidal findings and varying degrees of cognitive impairment are common. **Final Comments:** Adult-onset LKD requires a puzzling investigation, considering the differential diagnosis of the underlying pathology. The prevalence is lower than in childhood and specific radiological/clinical markers may be missing. Our case illustrates MADD leading to adult-onset LKD, while it would be expected milder findings at this age. Expanding the access to ancillary exams might widen the etiological panel for young-onset dementia and guide therapeutic interventions.

Neurogenética

TL 1105073

NEUROFIBROMATOSIS TYPE 1: CASE PRESENTATION

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Case Presentation: Neurofibromatosis is a neurocutaneous syndrome characterized by tumors in the central or peripheral nervous system and in the skin, and can be of three types: 1 (NF1), 2 (NF2), and Schwannomatosis. NF1 is the most common type, also called Von Recklinghausen's disease, and is a rare genetic disorder with vertical autosomal dominant transmission and an estimated incidence of 1: 2500-3000 live births. It is caused by a de novo or inherited loss-of-function mutation in the neurofibromin 1 gene, located in band 17q11.2, and encodes the tumor suppressor neurofibromin. The diagnosis is clinical, and there are seven criteria for recognition, two of which are sufficient for that purpose: at least six café-au-lait spots, larger than 5 mm in pubertal period or larger than 15 mm post-pubertal; at least two neurofibromas or one or more plexiform neurofibromas; axillary or groin freckles; optic glioma; two or more Lisch nodules; sphenoid dysplasia, dysplasia, or enlargement of the cortex of long bones; first-degree relative with NF1. The objective of this paper is to describe the case of a patient with neurofibromatosis type 1 who died due to the appearance of neurofibromas in internal organs. Female, 8 years old, diagnosed with neurofibromatosis at birth because her father was a carrier of the disease. She developed nodules only internally, but in vital regions, so she underwent seven surgeries until she was 8 years old. She was diagnosed with a nodule in the mediastinum at that age, and 3 to 4 months later in the throat, which was impossible to remove. She began to lose speech and have recurrent respiratory infections, and was hospitalized at least eight times with pneumonia. She was placed in the ICU, placed on non-invasive mechanical ventilation, and required respiratory physiotherapy to try to maintain airway patency. She was on drug therapy with traditional corticoids, antibiotics, and analgesics. She had café au lait spots and Lisch nodules. Eight months after the appearance of these nodules, a brain nodule was discovered, also impossible to remove, all benign but located in vital organs. Afterwards, he began to present frequent headaches, important visual alterations and convulsive crises, dying about a year and a half after the discovery of the nodules. It is important to highlight that, even though the disease was inherited from the father, the patient had a greater evolution due to the appearance of the nodules being in internal organs.

Neurogenética

TL 1105082

MULTIPLE CONGENITAL ANOMALIES HYPOTONIA SEIZURES SYNDROME (MCAHS) – MULTIPLE GENES, ONE PHENOTYPE? A CASE SERIES

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Case Presentation: We report 3 patients from two unrelated families with Multiple Congenital-Anomalies-Hypotonia Seizures Syndrome (MCAHS) due to pathogenic variants in different genes and discuss phenotype-genotype associations. Case 1: A 26-year-old woman with a history of myoclonus in both hands during breastfeeding, generalized seizures, delayed milestones, hypotonia and dysmorphisms. She also had recurrent flu and pneumonia because of an immunodeficiency. Parents were not related but both were of Lebanese descent. Case 2: Female patient, sibling of Case 1, died 6 years ago, was diagnosed with tetralogy of Fallot and had a similar history of generalized seizures, myoclonus, global delayed milestones, dysmorphisms, hypotonia, urinary infections and recurrent flu and pneumonia also because of an immunodeficiency. Whole exome sequencing in both sisters disclosed two pathogenic variants in heterozygosity (Chr20: 44.047.933 A>G and Chr20: 44.047.955 C>T) in PIGT (OMIM* 610272). Case 3: 9-year-old male, presented with a history of epilepsy, delayed milestones, gastroesophageal reflux disease and myoclonus since birth. He also exhibited obstructive sleep apnea syndrome. Immunodeficiency was not present. Brain Magnetic Resonance Imaging showed cortical and cerebellar atrophy. Whole exome sequencing revealed a missense variant in homozygosity (Chr18: 62.157.747 G>A) in PIGN (OMIM* 606097). **Discussion:** MCAHS is a heterogeneous autosomal recessive disorder characterized by global developmental delay, hypotonia and early-onset seizures, as well as variable congenital anomalies in urinary, cardiac and gastrointestinal systems. MCAHS has also been classified under the more encompassing term of "glycosylphosphatidylinositol biosynthesis defects" (GPIBD). It can result from variants in different genes (PIGN, PIGA, PIGT, PIQ), and MCAHS1 and MCAHS3 are related to PIGN and PIGT mutations respectively. Despite some information in the literature about the clinical spectrum, it is still unclear if the phenotypes are really similar between the different gene mutations. Immunodeficiency was the atypical finding in our two cases of mutation in PIGT, which can suggest some particularities related to the different genes. **Final Comments:** Immunodeficiency is a symptom not yet described in patients with PIGT mutation and, despite the visible similarities between our 3 cases, further studies may demonstrate phenotypic specificities in different patients with MCAHS/GPIBD.

Neurogenética

TL 1105171

MELAS SYNDROME &NDASH; MOTHER AND DAUGHTER WITH RARE MUTATION (3291) IN MITOCHONDRIAL DNA

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Case Presentation: BVSM, 11 years old, in early childhood difficulty in gaining weight and median school performance due to inattention. At 9 years of age, he started to have epilepsy associated with severe metabolic acidosis. After 1 year, the patient evolved with visual scotomas, headache, vomiting, daily seizures, right hemiparesis, fatigue and palpitations. At the time, she underwent tests that showed an increase in lactate, magnetic resonance imaging (MRI) of the skull with ischemic stroke (IS) and echocardiogram with the presence of dilated cardiomyopathy. Genetic tests were requested to investigate the underlying pathology, and a mutation was found in the mitochondrial gene MT-TL1, variant c.3291T>C in heteroplasmy (71%), closing the diagnosis for MELAS. The patient died in a few months from congestive heart failure. Due to the mitochondriopathy, a genetic investigation of the mother was guided, and the same mutation was found (heteroplasmy in 32%). She was asymptomatic and recently had an unprecedented seizure. **Discussion:** In MELAS syndrome (mitochondrial encephalopathy, lactic acidosis and stroke) there is a genetic mutation that compromises the mitochondrial DNA, being exclusively maternally transmitted. Mitochondria generate ATP, and their dysfunctions lead to energy insufficiency for various organs. The clinical diagnosis is based on the presence of IS (<40 years), encephalopathy (convulsions/dementia), mitochondrial myopathy (lactic acidosis and/or irregular red fibers), criteria found in the patient in the case in question. The disease can lead to other symptoms such as headache, vomiting, dementia, deafness, diabetes, short stature and cardiomyopathy. About 80% of cases result from leucine tRNA mutation, with m.3243A>G being the most common. The c.3291T>C variant, present in the patient, is found in <1% of cases. MRI lesions are not classic, restricted to the cortex and subcortical white matter. In spectroscopy there is a reduction of n-acetylaspate and accumulation of lactate. Treatment is symptomatic and with a multidisciplinary team. Antioxidants and cofactors can be administered with the aim of improving quality of life and reducing the risk of complications. **Final Comments:** MELAS presents different phenotypes in the same mutation, making it a diagnostic challenge. Still intractable and with devastating consequences as in the case described, it is expected that in future research new medications can bring better quality of life to patients.

Neurogenética

TL 1105190

CPEO DUE TO TK2 MUTATION, A RARE AND ORPHAN DISEASE – CASE PRESENTATION

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Case Presentation: female patient, 32 years old, previously healthy, with proximal limb weakness since childhood, described as incapacity to jump, difficulty to run and frequent falls. In adulthood, she had progression of the weakness associated with pain in proximal limbs. There were no diplopia, dysphagia, cervical weakness or systemic complaints. No fluctuation of symptoms was noted. Patient denied similar cases in family and consanguinity. Neurological examination showed bilateral, symmetrical and proximally appendicular weakness with myopathic gait associated with facial diparesis and paresis of the bilaterally superior rectus muscles. Complementary investigation showed a slight increase in hepatic transaminases associated with significant increase in CK (1.341 UI/L) and lactate levels. Electroneuromyography evidenced polyphasic motor unit potentials with reduced amplitude and short duration in the right deltoid muscle. Muscle biopsy showed marked variation in the diameter of fibers, single or multiples internal nuclei, some degenerating fibers and evidence of post-necrosis regeneration, derangement of the intermyofibrillar cytoarchitecture without inflammatory infiltrates and also substantial number of “ragged-red” and “ragged-blue” fibers with COX-negative and fiber hyperintensity in the NADH-TR and SDH reactions. Genetic panel revealed a homozygous pathogenic variant in the TK2 gene (c.323C>T, p.Thr108Met). **Discussion:** TK2 is a nuclear gene that encodes the mitochondrial thymidine kinase (TK), an enzyme involved in the maintenance of the deoxynucleotides of the mitochondria and crucial in the biosynthesis of the mitochondrial genome. Bi-allelic mutations in the TK2 gene relates with diverse clinical presentations, mainly with a severe myopathy, which is rapidly progressive, or with chronic progressive external ophthalmoplegia (CPEO) associated with limb muscle weakness and dysphagia, both with premature death. Although not approved by FDA yet, deoxynucleosides therapy for TK2 deficiency are under research with positive results, mainly in early-onset cases. **Final Comments:** In clinical practice, from molecular point of view, CPEO is a diagnostic challenging. This case presentation illustrates that testing CPEO in genetic panels is worthwhile. Considering the tragic natural history of the disease, it is essential to identify the TK2 patients, not only for diagnosis, but also to offer these promising therapies.

Neurogenética

TL 1105225

A CASE PRESENTATION ON THE DIAGNOSIS OF NEURONAL CEROID LIPOFUSCINOSIS TYPE 2 (CLN2) AND ITS FOLLOW-UP IN A FEMALE PATIENT IN THE STATE OF CEARÁ

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Case Presentation: Female patient, 15 years old. With normal neuropsychomotor development up to 5 years old, when she began to have seizures. A computer tomography scan of the skull was requested, which was normal. The patient progressed with limb tremors, ataxia, pyramidal syndrome, and speech difficulties. At 9 years of age, she had difficulty walking and loss of gait; the cranial resonance showed alterations in the white matter and signs of cerebral cortico-subcortical and pontine-mesencephalic atrophy. In 2016, after full exome sequencing, the diagnosis of Neuronal Ceroid Lipofuscinosis Type 2 (NCL 2) was confirmed. In 2018, the patient started using the drug Brineura. At age 12, in 2019, she scored 1 on The Hamburg Motor and Language and CLN2 Clinical Rating Scale (motor=0 + language=1), in 2020, she scored 3 (motor=1 + language=2). **Discussion:** NCL is a group of neurodegenerative diseases with 4 types of variation and clinical and pathological classification according to age at onset of neurological symptoms, neuroradiological and pathological findings. However, the final distinction is genetic. Among them, the late infantile variant or Jansky-Bielschowsky disease starts between 2 and 4 years of age, with rapid progression. NCL2 occurs as a result of the deficiency of the lysosomal enzyme tripeptidyl-peptidase 1 (TPP1) when the gene encoding the protein expresses an error. The usual findings are myoclonus, seizures, ataxia, difficulty walking, or progressive loss of pronunciation of words. Encephalic volumetric reduction causes a decrease in the levels of intelligence, phonation, and motor function. Speech and motor function disorders are related to the reduction in pons and bulb diameters, with cerebellar atrophy being the most prominent factor. In the patient, manifestations ranging from seizures to changes in white matter and signs of cerebral cortico-subcortical and pontomesencephalic atrophy were found. **Final Comments:** We present a case of CLN2, a very rare disease, occurring in 6: 100,000 inhabitants. Effective management and treatment of this condition require an early diagnosis. Therefore, we bring this case to encourage the investigation of CLN2 disease in children presenting seizures, loss of motor and cognitive function, or speech delay without a diagnosis.

Neurogenética

TL 1105241

GENETIC LEUKOENCEPHALOPATHY: A DIAGNOSTIC CHALLENGE

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Case Presentation: C. A. S., male, 35 years old, presented in 2014 with depressive symptoms, developing visuospatial and executive impairment, as well as behavioral disorder, after a few months. He presented worsening of cognition over the years. In 2019 he developed gait ataxia with early falls, becoming wheelchair bound within few months. He became totally dependent for daily life activities since then. In 2021 he developed dysarthria and dysphagia. There was no consanguinity nor similar cases in the family. Physical exam: awake, bulbar dysarthria, disconnected words, spastic tetraparesis, bilateral Babinski sign, dysmetria in four limbs. Brain MRI showed diffuse confluent white matter T2/FLAIR hyperintensity and cortical atrophy. The possibility of leukodystrophy was raised. Genetic testing showed heterozygous mutations in the mitochondrial gene alanyl-tRNA synthetase 2 (AARS2). The patient passed away in April 2022. **Discussion:** The diagnostic evaluation of genetic leukoencephalopathy includes screening test that should be performed to rule out the main causes: biochemical for inborn errors of metabolism, spinal fluid analysis, magnetic resonance of the brain and spinal cord, and genetic testing, if possible. Diseases associated with mitochondrial aminoacyl-tRNA synthetase proteins (mt-aARSs) enzymes are a group of genetic leukoencephalopathies associated mainly with three specific clinical diseases: aspartyl-tRNA synthetase (DARS2), mitochondrial glutamate tRNA synthetase (EARS2) and AARS2. [1]. AARS2 is associated with a rare autosomal recessive leukodystrophy. In adults the most prevalent phenotype, first described in 2014, is characterized by premature ovarian failure in women and progressive leukodystrophy [2]. The age of onset is usually around 25 years, with cognitive decline, neuropsychiatric changes, pyramidal and extrapyramidal signs, ataxia and neuropathy [2,3]. Some cases have been described with developmental delay or motor problems in childhood [2]. **Final Comments:** This case presentation illustrates the diagnostic challenge presented by the primary leukodystrophies, which is only possible with a genetic study, unfortunately difficult to obtain in our country. The possibility of identifying a specific mutation related to leukodystrophy is the first step that will allow the development of targeted therapeutic treatment for this group of diseases, that are currently associated with high morbidity and mortality.

Neurogenética

TL 1105247

HICKAM'S DICTUM ON A CASE OF PROGRESSIVE WEAKNESS AND HYPERMOBILITY: A DOUBLE GENETIC HIT DISORDER

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Case Presentation: Son of consanguineous parents, a 14-year-old male patient presented with slowly progressive, symmetrical, distal predominant neurogenic weakness since he was 1-year-old. He also experienced striking joint hypermobility and spinal deformities. His whole exome sequencing (WES) first revealed a homozygous class 4 variant at the IGHMBP2 gene (c.2796delC, p.Cys932TrpfsTer46), related to distal hereditary motor neuropathy (dHMN) type VI and CMT2S. This variant, however, failed to explain the hypermobility. Therefore, a thorough search at the WES was conducted, revealing a second class 4 homozygous variant at the ALDH18A1 gene (c.121 C>T, p.Arg41Cys) associated with cutis laxa, and thus elucidating the clinical picture. **Discussion:** Occam's razor is a heuristic tool much applied in medicine. It values simplicity, asserting that multiple symptoms should have a common explanation. Hickam's dictum, on the other hand, states that concomitant symptoms can be due to different reasons. In this case, we demonstrate the antitheses. When in face of a pathogenic variant that only partially explained the clinical scenario, we decided to further investigate by expanding molecular analysis. This led to the discovery of a second pathogenic variant, in a double-hit genetic disorder. The clinical evaluation remains sovereign to the point of directing the reasoning and defining whether the eventual finding is sufficient for the diagnosis or if we need to look for a second explanation, invoking Hickam's dictum. **Final Comments:** The implementation of the next-generation sequencing (NGS) techniques in clinical practice changed the molecular assessment and improved patient and family care. However, the massive information obtained with these tests is still not fully manageable. We emphasize the importance of interpreting genetic findings in light of clinical manifestations. We also stress that Occam's razor is not always superior to Hickam's dictum. In genetic conditions, when a strong pathogenic variant does not explain the full clinical picture, the possibility of a double-hit genetic disorder should be considered. In such cases, a judicious clinical analysis is essential to shape the genetic data search.

Neurogenética

TL 1105344

ARGINASE 1 DEFICIENCY: AN IMPORTANT MIMICKING OF EARLY-ONSET HEREDITARY SPASTIC PARAPLEGIA

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Case Presentation: A 19 years-old male patient, born from consanguineous parents, presented with an insidious complaint of progressive weakness and frequent falls since the age of nine. At fourteen he needed bilateral assistance to walk, and at eighteen he already needed the use of a wheelchair. His mother highlighted that the patient never liked meat and that he had a lack of appetite and frequent vomiting since childhood. She has also noticed that he has greater learning difficulties, is more childlike than people of the same age, and is more irritable and anxious. Neurological examination shows MRC grade II weakness and severe spasticity in the lower limbs, in addition to being unable to stand in an orthostatic posture. Brain MRI showed diffuse cortical and cerebellar atrophy. Whole-exome sequencing revealed a homozygous pathogenic variant mutation p.Arg308Gln in the ARG1 gene. **Discussion:** Arginase-1 deficiency (OMIM #207800) is a rare autosomal recessive genetic disorder caused by mutations in the ARG1 gene, resulting in partial or complete loss of enzyme function that affects the liver-based urea cycle. ARG1-deficient patients mainly exhibit hyperargininemia with spastic paraparesis, progressive neurological and intellectual impairment, and persistent growth retardation. Early symptoms include irritability, global developmental delay, failure to thrive, recurrent vomiting, feeding/protein aversion, and anorexia. Spastic paraparesis with onset in early childhood is the most obvious sign of the disease. Brain imaging may reveal cerebral atrophy and cerebellar atrophy may also occur. The ARG1 gene sits on chromosome 6 (6q23) composed of eight exons and there are at least 43 potentially disease-causing variants in ARG1 with the majority being missense/nonsense mutations and small deletions. In this case, a rare homozygous missense mutation was identified. Approximately half of the reported subjects are homozygous with a moderate number of cases arising from consanguineous relation, as in this case. **Final Comments:** This case shows that progressive loss of mental and motor skills, increasingly more severe spasticity, and pyramidal tract signs are the hallmarks of the disease caused by ARG1 deficiency, and this is an important differential diagnosis of childhood-onset hereditary spastic paraplegia.

Neurogenética

TL 1105382

CANTU SYNDROME AS A DIFFERENTIAL DIAGNOSIS OF DEPOSIT DISEASES: A CASE PRESENTATION

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Case Presentation: The patient is a female child, who sought consultation with a neurologist at 1 year and 8 months of age due to delay in neuropsychomotor development and excessive irritability. She showed features of thick hair up to the forehead and increased body hair, prominent mouth with full lips, macroglossia, coarse face, wide nasal bridge; also had a history of surgically corrected congenital heart disease (patent ductus arteriosus) at 9 months old and a hypertrophic cardiomyopathy of the left heart ventricle. The hypothesis of lysosomal storage disorder was raised and a genetic testing was requested. The result was an alteration in the ABCC9 gene, indicative of Cantu Syndrome. Patient was referred to orthopedist for the investigation of osteochondrodysplasia, speech therapy and physiotherapy, and advised to keep follow-up with a cardiologist. **Discussion:** Cantu Syndrome is a rare disease, with little more than 100 cases described in the literature, caused by alteration in two genes, ABCC9 and KCNJ8. It is inherited in an autosomal dominant manner and is proposed as a potassium channelopathy. Clinical features include face coarse with bushy eyebrows, prominent supraorbital ridges, anteverted nostrils, prominent mouth with full lips, macroglossia, wide nasal bridge, long and wide philtrum, hypertrichosis with generalized increase in body hair, cardiomegaly, patent ductus arteriosus requiring correction, mild speech delay, broad ribs, skull thick, scoliosis, flaring of the metaphyses. The patient in the clinical case has the main features of the disease. Differential diagnoses of the disease includes Beckwith-Wiedemann syndrome, Zimmermann-Laband, dilated and hypertrophic cardiomyopathy, mucopolipidosis III and mucopolysaccharidosis (types I, II and IV), the latter one being the patient's initial diagnostic hypothesis. **Final Comments:** So far, there is no treatment for Cantu Syndrome, however the manifestations can be managed individually, such as follow-up with a cardiologist and correction of correctable changes; dermatological treatment for hypertrichosis; surgical correction of possible bone changes; monitoring of DNPM in children with delay, among others. There is great potential in therapeutic options with future clinical studies.

Neurogenética

TL 1105401

DEVELOPMENT DELAY, IMPAIRED GROWTH, DYSMORPHIC FACIES, AND AXONAL NEUROPATHY (DIGFAN) WITH EPILEPSY: CASE PRESENTATION

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Case Presentation: LSG, 31 years-old man, presented development delay (motor and cognitive), short stature, dysmorphic facial features, deafness and epilepsy since childhood. The brother had motor and intellectual impairment and his father progressive ataxia related to SCA 2. Neurologic exam revealed: short stature, facial dysmorphism (figure 1), hearing loss, incomprehensible sounds, comprehends some basic commands with gesture, no pyramidal nor cerebellar signs were documented. Brain MRI revealed white matter hyperintensities on T2 and FLAIR, brain and cerebellar atrophy and thinning of the brainstem. Auditory evoked potential showed bilateral involvement with peripheral topography. The genetic test showed a pathogenic variant c.79G>A in the MORC2 gene. **Discussion:** MORC2 is a gene that through its ATPase activity is essential for epigenetic silencing. Mutation in this gene seems to cause a hyperactivation of silencing in neural cells, impairing neurological development [1]. These mutations have been reported most in Charcot-Marie-Tooth type 2Z and DIGFAN syndrome [2,3]. DIGFAN syndrome is characterized by development delay, short stature, hypotonia, cognitive impairment, dysmorphic facies, and axonal neuropathy. This syndrome can also present hearing loss and brain atrophy [4,5]. In the literature, patients with the c.79G>A variant are associated with developmental delay, growth retardation and abnormal brain MRI, as in our case. Epilepsy is not usually associated with DIGFAN syndrome in MORC2 mutation as in our case, being a rare manifestation of this phenotype. **Final Comments:** Epilepsy should be considered as a manifestation of MORC2 spectrum (DIGFAN syndrome), depending on the variant. Some variant exerts a more intense action on neural silencing, resulting in more severe phenotypes related to central nervous system that could be involved in epileptic manifestation as in our case. [4].

Neurogenética

TL 1105428

AN ATYPICAL PRESENTATION OF LESCH-NYHAN SYNDROME MIMICKING INHERITED NEUROTRANSMITTER METABOLISM DEFECT WITH A DOPA-RESPONSIVE PHENOTYPE

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Case Presentation: A 10-month-old Brazilian male infant presented with irritability, generalized hypotonia and global developmental delay since the neonatal period. Since age 4 months, he started with paroxysmal events, tonic hyperextension of upper limbs several times a day. Afterwards, he started having apneic spells, vomiting, and diaphoresis. Examination at age 1 year and 3 months disclosed axial hypotonia, no ability to sit without support, dystonic posture of neck, arms, and legs, along with choreoathetoid movements in hands and feet. CSF analysis and lab tests were unremarkable, including normal serum uric acid levels. As an inherited disorder of monoamine metabolism was suspected, a levodopa trial was started with marked improvement of dystonic and autonomic disturbances. NGS-based multigene panel for movement disorders disclosed the pathogenic missense variant c.233T>C (p.Leu78Pro) in hemizygoty in the HPRT1 gene (Xq26.2-q26.3), establishing the diagnosis of Lesch-Nyhan syndrome (LNS). Five months after the genetic diagnosis, the patient presented with the typical so-called self-injurious behavior. **Discussion:** Although classically considered in the differential diagnosis of early-onset presentation of complex movement disorders, LNS is rarely considered for further diagnostic work-up after normal serum and urinary screening for uric acid levels. Even in the absence of abnormal serum biomarkers and the typical automutilation features, LNS diagnosis must be kept in the diagnostic work-up of highly suggestive neurological phenotypes. This case highlights the importance of performing large NGS-based multigene panels to screen complex phenotypes of suspected inherited neurometabolic disorders, including LNS and inherited disorders of neurotransmitters metabolism. Marked very early-onset autonomic disturbances were present in this patient and presented with a levodopa-responsive pattern, as well as the dystonic involvement. **Final Comments:** Infancy or neonatal-onset cases of global developmental delay, generalized hypotonia, early-onset paroxysmal events resembling dystonic posturing, and dysautonomia must include LNS in the diagnostic work-up. Levodopa-responsive cases should alert clinicians about LNS as a differential diagnosis.

Neurogenética

TL 1105483

CLINICAL RESPONSE TO SELUMETINIB: CASE PRESENTATION

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Case Presentation: Neurofibromatosis type 1 (NF1), neurofibromatosis type 2 (NF2) and schwannomatosis are tumor suppressor syndromes characterized by multiple nervous system tumors, which manifest in different locations depending on their genetic etiology. NF1 affects approximately 1:3000 individuals and is a tumor caused by mutations of the NF1 gene on chromosome 17q11.2. The treatment of NF1 consists of surgical excision of cutaneous neurofibromas (CNFs) when symptomatic. Surgical removal of plexiform neurofibromas (PNFs) are not always possible due their diffuse and multiple nerve roots involvement. The recent advance is the FDA approval of the MEK inhibitor selumetinib (Koselugo™) for the treatment of children with symptomatic, inoperable PNF based on a trial that demonstrated 72% response rate in tumor volume shrinkage by ? 20% in NF1 children with PNFs. However, this is not curative as no subjects had complete tumor disappearance and tumors regrew particularly after dose reduction or cessation due to toxic effects, such as diarrhea, weight gain, paronychia, skin ulceration, and elevated creatinine level. This case presentation is about a patient with NF1 diagnosed at 14 years old in 2008 who started compassionate use of Selumetinib 80 mg/day (divided into two doses a day) in august 2020 due to clinical and functional worsening at this time. He had difficulty swallowing due to neurofibromas in cervical region, also had inability to walk, was able to move only on a wheelchair, and difficulty in evacuating and urinating, using a urinary catheter and laxative medications continuously. Neuroaxis resonance showed countless nodular images in the subcutaneous tissue of the scalp, compatible with neurofibroma, sometimes confluent, predominance in the occipital regions. Other similar lesions can be seen in the deep cervical spaces. Countless PNFs involving all intervertebral foramina evaluated, confluent in the paravertebral regions, sometimes forming masses. Intracranial and extracranial lesions within the vertebral canal stand out, the largest at the level of C1 and C2, compressing the spinal cord, without altering its signal. After starting Selumetinib the patient had a significant reduction in neurofibromas, and significant clinical improvement. At the moment, the patient is able to walk with the help of a walker, has sphincter control and improved dysphagia. The case is maintaining follow-up in the neuro-oncology sector at UNIFESP, with stable clinical status.

Neurogenética

TL 1105540

DOUBLE TROUBLE: COEXISTENCE OF FULLY PENETRANT CAG REPEAT EXPANSIONS IN THE ATXN3 AND ATXN7 GENES

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Case Presentation: A 16-year-old young man presented with a 5-year history of slowly progressive imbalance and visual impairment. On neurological examination there was cerebellar ataxia, loss of visual acuity, macular degeneration, slow saccades, ophthalmoparesis, upper motor neuron signs and mild cognitive impairment. Interestingly, there was a positive medical history of an ataxic disease with male-to-male transmission on both paternal and maternal sides in up to six consecutive generations. Brain MRI demonstrated mild cerebellar atrophy. Target analysis for CAG expansions through PCR with fragment analysis detected 59 repeats in the SCA3 gene and 57 repeats in the SCA7 gene. **Discussion:** The Spinocerebellar Ataxias are heterogeneous group of autosomal dominant neurodegenerative diseases characterized by progressive cerebellar ataxia, different genetic mutations and frequently associated a further signs and symptoms. SCA3 is caused by a monoallelic CAG trinucleotide repeat expansion in the gene coding for the cytoplasmic protein ataxin-3 (ATXN3) located on chromosome 14. Normal individuals have up to 44 glutamine repeats and complete penetrance of the disease is commonly seen in individuals with more than 52 repeats. Age of onset usually occurs in the 4th decade of life, and correlated with the CAG repeat size. SCA7 is caused by a heterozygous CAG repeat expansion in the gene encoding ataxin-7 (ATXN7) located on chromosome 3. Pathogenic fully penetrant expansions have 37-460 repeats. Disease spectrum may vary from an aggressive early-childhood ataxia with failure to thrive, rapidly deterioration and early death or as a progressive adolescent/adult form. Both forms have visual impairment. As observed in SCA3 there is a correlation between CAG repeat size and disease severity and age of onset. **Final Comments:** The presence of more than one SCA mutation in a single individual is rare. The age of onset, the presence of macular degeneration with visual impairment suggests SCA7 is contributing to his clinical picture. This patient and his family also may provide unique opportunities to study the interactions of two genetic mutations and their effect on clinical expression of disease. Furthermore, finding two genetic mutations in a patient may have important implications regarding counseling on genetic risk, genetic testing, and disease prognosis.

Neurogenética

TL 1105541

SPINAL MUSCULAR ATROPHY TYPE 2 – USE OF NUSINERSENE IN ADULT PATIENT

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Case Presentation: VLO, male, 23 years old, at 6 months of age, started to develop muscle weakness, evidenced by the delay in developmental milestones. At 2 years of age, he was able to walk with support, when the hypothesis of Spinal Muscular Atrophy (SMA) type 2 was raised, which showed chronic denervation and a DNA study with mutation of the SMN1 chromosome 5q gene (absence of exons 7 and 8). Since then, there has been a progressive worsening of symptoms. In recent years, he developed dysphagia and dyspnea, especially during sleep, requiring CPAP. In 2021, he started treatment with intrathecal Nusinersena at a dose of 2 mg, with the first three doses administered at 14-day intervals, the fourth dose after 30 days of the third, following treatment with maintenance doses every 4 months. During therapy, an improvement in the strength of limbs and patient autonomy was observed, demonstrated by the increase in the SCORE of the HFMSE scale (Hammersmith functional motor scale expanded), from 10 to 37 points. **Discussion:** SMA is an autosomal recessive genetic disease with mutation in the SMN1 (motoneuron survival 1- the gene product is a key protein for RNA division that is reduced in SMA), inhibiting functional SMN protein production and leading to degeneration of anterior horn cells in the spinal cord, which results in progressive proximal weakness with varying degrees of muscle atrophy and paralysis of the respiratory muscles. It presents a wide spectrum of clinical severity, and can be classified according to the age of onset of symptoms and the developmental milestones reached. Recently, disease-modifying therapies have emerged, such as Nusinersene, a nonsense oligonucleotide that increases the amount of complete SMN protein, thereby reducing mortality and improving the patient's respiratory and motor function. Its use is beneficial at any stage of the disease. In children, mainly, with little application in adults. **Final Comments:** The advent of new treatments for AME has highlighted the importance of early diagnosis, allowing pre-symptomatic treatment and resulting in better quality of life for patients. More studies are needed, especially those that include the adult population with the disease, where a good response is also observed.

Neurogenética

TL 1105547

MORC2 RELATED NEUROPATHY ASSOCIATED WITH AUTONOMIC DYSFUNCTION

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Case Presentation: A 27-year-old female, borned from non-consanguineous and healthy parents. She presented in her first decade of life with distal and proximal asymmetrical weakness, sensory loss, generalized areflexia. She also presented cognitive impairment. Nerve conduction studies revealed sensory and motor axonal neuropathy. WES revealed a class 5 variant in the MORC2 gene (NM_001303257.2: c.754C>T ; p.Arg252Trp) confirming the diagnosis of CMT2Z. Sanger sequencing of her parents confirmed it was a de novo variant. At 26 years old, she reported episodes of recurrent abdominal pain, early satiation, bloating, nausea, vomiting, constipation and progressive weight loss. Blood pressure standing and lying, as well as heart rate due valsalva maneuver were normal. Gastric emptying scintigraphy showed that gastric emptying time T1/2 was greater than 30 minutes, with gastric retention of about 70% of the content at the end of the study, confirming gastrointestinal specific autonomic dysfunction. **Discussion:** Charcot-Marie-Tooth disease type 2Z is a rare form of autosomal dominant axonal neuropathy. Clinical findings include sensory impairment, distal and proximal weakness, distal atrophy and muscle cramps. Also, there are reports of global hypotonia, including MORC2-related disorder in the differential of non-51 SMA. Additionally, intellectual disability, hearing loss, microcephaly, pyramidal signs and brain atrophy have been reported. MORC2 protein regulates DNA transcription and is important for cytoplasmatic function. In a literature review, dysautonomia is not reported in patients with CMT2Z, but it is well established the secondary involvement of MORC2 in mitochondrial function and maintenance. Additionally, there is a Leigh-Like case previously reported sin association to MORC2. **Final Comments:** We report a case of CMT2Z caused by a de novo MORC2 gene mutation in a patient presenting with gastrointestinal isolated dysautonomia. We hypothesized that this mutation may affect mitochondrial machinery. Further data studies are necessary to elucidate the relationship between mutation in MORC2 gene and dysautonomia.

Neurogenética

TL 1105781

WHEN A HINT TURNS INTO A HINT – A RARE CASE OF AN AXONAL SENSORY-MOTOR NEUROPATHY WITH NEUROMYOTONIA

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Case Presentation: A 17-year-old woman born from consanguineous parents presented to our neurogenetic outpatient clinic with a history of progressive unsteady gait since early childhood. She had normal milestones achievement and no significant medical history until the age of 6-year-old when she began to have frequent falls. For the subsequent years, she had worsening of symptoms with slowly progressive muscle weakness and sensory loss, especially at distal parts of the limbs. The patient was actively asked about difficulties in releasing grip after a strong voluntary hand contraction and reported distress doing this task since the beginning of the symptoms. In the neurological examination she had winged scapula, absent Achilles tendon reflexes, pinprick and vibration sense impairment with a length-dependent pattern and a slap gait with distal lower limbs weakness combined with fasciculations and action myotonia. Nerve conduction studies revealed an axonal motor-greater-than-sensory polyneuropathy and needle monopolar examination revealed increased insertional activity with myotonia and neuromyotonia (discharges of 90 to 120 Hz). The patient's parents also performed nerve conduction and needle studies with normal results. On complementary investigation, no cardiologic or pulmonary involvement was detected. Initial molecular analysis ruled out MPZ, PMP22, GJB1, GDAP1 mutations. An American College of Medical Genetics and Genomics (ACMG) class 5 homozygous missense variant on histidine triad nucleotide binding protein 1 (HINT1) gene (c.110G>C, p.Arg37Pro) was found leading to the diagnosis of autosomal recessive neuromyotonia and axonal neuropathy (NMN). **Discussion:** An early-onset (mostly around the age of 10-years-old) axonal form of motor-predominant peripheral neuropathy associated with neuromyotonia has been linked to loss of functional HINT1 gene. About 70-80% of patients present with neuromyotonia, which is a clinical hallmark for the diagnosis. Our case brings to light a classical presentation of a rare autosomal recessive mutation gene that produces a neuropathy within the Charcot-Marie-Tooth (CMT) spectrum. **Final Comments:** Despite the increasing diagnosis of HINT1 patients world-wide, it remains a diagnostic challenge to assess the pathogenicity of novel variants. This reinforces the importance to recognize the clinical syndrome allowing for request exome sequencing, identify molecular etiology and define treatment strategies of this CMT subtype.

Neurogenética

TL 1105844

STURGE WEBER SYNDROME IN ADULT: A CASE PRESENTATION

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Case Presentation: Patient FB, 41 years after entering the service complaining of left hemiparesis (strength grade 4-). The same refers that since childhood (5 months - 5 years) he presents recurrent episodes of reported deficit, which improve with the use of phenytoin, causing delay in his motor development. He denies other comorbidities or neurological pathologies in the family. In addition to the deficit, diagnosed as a focal crisis, a cutaneous vascular malformation on the right hemiface, compatible with a flaming nevus, called attention on physical examination. Due to this clinical finding and previous history, a brain resonance imaging (MRI) was performed, which showed volumetric reduction in the right parietal hemisphere and cortical leptomeningeal uptake compatible with anomaly of the cortical venous system, prominence of varicose leptomeningeal veins and prominence of the brain-related choroid plexus. trigeminal angiomatous. The seizures were controlled with phenytoin, a drug that the patient received when he was discharged, without any focal deficits. Scheduled outpatient follow-up for seizure control and follow-up. **Discussion:** Sturge Weber syndrome is a rare condition with an incidence of 1/50,000 to 1/230,000 live births. It is explained by the abnormal persistence of the embryonic vascular system which is located around the cephalic region of the neural tube. Its most common clinical presentation is the vascular dermato alteration manifested in the face, in addition to seizures. Less presented per patient, in addition to classic change seen on MRI of the skull. **Final Comments:** The management of patients with this syndrome shows challenges due to its rarity and general medical ignorance about it. Its treatment aims to control epilepsy and decrease visual deficits. In view of the benefit of the correct treatment, it is important to disseminate knowledge about this pathology in the medical and academic environment, prioritizing investigation with an image exam which demonstrates lesions characteristic of the disease.

Neurogenética

TL 1105867

EXPANDING THE PHENOTYPIC SPECTRUM IN OF HEREDITARY SPASTIC PARAPLEGIA TYPE 6 (SPG6): A CASE PRESENTATION OF SPASTICITY, EPILEPSY, MOTOR AXONAL NEUROPATHY, AND DISARTROPHONIA

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Case Presentation: A 70-years-old man presented with a long standing history of complex hereditary disorder characterized by spastic paraplegia, epilepsy and axonal motor peripheral neuropathy. In the last decade, he observed that his voice became laborious, leading to dysarthria and hypophonia. Otorhinolaryngological evaluation revealed vocal fold paresis, which was attributed to the neuromuscular disease after excluding acquired conditions. Brain and spinal cord MRI showed T2 hyper-signal along the corticospinal tract. The SPG6 diagnosis was confirmed by an exome sequencing that identified a missense variant, (NM_144599.5: c.316G>A, p.Gly106Arg), within the microtubule binding site domain of the NIPA1 gene. This variant was classified as class 5 according to ACMG criteria, which implies it has known functional consequences. Sanger sequencing showed this variant was also present in his affected son. **Discussion:** Mutations in the NIPA1 gene have been associated with autosomal dominant SPG6, which is a progressive neurodegenerative disorder. Although most cases present with a pure HSP phenotype, there has been some descriptions of complex cases, with variable clinical pictures. About 23% of all published cases report a complex phenotype, with epilepsy found in about 10% and a sensory and motor length-dependent peripheral neuropathy in only 5,5% of them. Dysarthria, hypophonia and a distal motor neuropathy have not been described, according to the best of our knowledge. **Final Comments:** We presented an unusual case of SPG6 with a complex phenotype including epilepsy, axonal motor peripheral neuropathy, dysarthria and hypophonia in a patient harboring a pathogenic variant in NIPA1 gene, which expands the phenotypic spectrum of NIPA1-related disorders.

Neurogenética

TL 1105880

ADULT-ONSET MELAS: WHAT IS THE BEST WAY TO INVESTIGATE?

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Case Presentation: Female patient, 41 years old, with a previous history of non-migraine headache and cramps during exercise, presented, at the age of 36, a sudden change in consciousness associated with reduced visual acuity on the right, auditory and visual hallucinations, with complete recovery in two days. Here, she was diagnosed with a psychiatric illness. After four years, she had a new episode, but now with associated right hemiparesis. Within two months, she recovered, but remained with some cognitive sequelae (mainly memory and executive dysfunction). After months, she had a new crisis, now with left hemiparesis, which was initially treated as herpetic encephalitis. His exams revealed an increase in CPK, an increase in CSF lactate (despite normal cellularity and proteinorrhea) and cranial resonance with extensive DWI / FLAIR lesion in the occipital, temporal and frontal regions on the right, not respecting vascular territory and without marked hyposignal in the ADC map, in addition to lactate peak spectroscopy. Panel of autoantibodies, electroneuromyography, echocardiogram, CT of the abdomen, thorax and pelvis without alterations. Due to the clinical suspicion of mitochondrial disease, a molecular study was carried out, collected by oral swab, aimed at the variants most associated with MELAS. This showed the presence, in heteroplasmy (in 36% of the readings performed), of the pathogenic variant m.3243A>G in the MT-TL1 gene, the most associated with this syndrome, confirming the patient's diagnosis. **Discussion:** Adult-onset MELAS, especially after the age of 20 years, is considered very uncommon, which leads to a delay in diagnosis by being confused with isolated psychiatric conditions or other causes of encephalitis. In cases where clinical suspicion is form, it is initially recommended to evaluate the variants in the MT-TL1 gene. The m.3243A>G variant is the one most associated with MELAS syndrome, and the degree of heteroplasmy in oral swab collection is in agreement with what has been previously reported. If it were low, due to the high clinical suspicion, one possibility would be to perform the analysis on epithelial cells by urine collection, since it is the sample that shows the highest percentages of heteroplasmy, among those of easy access. **Final Comments:** In adult-onset forms, diagnostic delay of MELAS is common. In addition, collection by blood or oral swab is the most performed, although urinary analysis has a higher diagnostic yield.

Neurogenética

TL 1105882

SYNDROME 47, XYY: A CASE PRESENTATION FROM A NEUROPSYCHIATRIC PERSPECTIVE

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Case Presentation: Male, 15 years old, had his first consultation approximately 6 years ago due to complaints from his parents regarding psychomotor agitation, heteroaggressiveness, hypoprosia, and a possible tonic-clonic and generalized ictus pattern at 5 years of age without tongue biting and apparent triggers. He arrives on chlorpromazine, risperidone, biperidene, and valproic acid. Drug therapy is corrected to risperidone, carbamazepine, and clonazepam. Physical examination revealed macrotia, gait with right lateral-everted foot, plagiocephaly due to early fusion of the right lambdoid suture, and absence seizures. The psychomotor symptoms improved significantly with continued use of the prescribed therapy, but hetero-aggressiveness persisted. In complementary exams, cranial computed tomography (CT) scan showed encephalic hypotrophy with malformed gyrus dysmorphisms; electroencephalogram (EEG) showed negative spikes in temporal lobes and negative sharp wave asymmetry and the karyogram confirmed the diagnosis by resulting in a 47, XYY genotype. **Discussion:** Jacobs' syndrome or 47, XYY consists of an aneuploidy of the male sex chromosome. Typically, this nosological entity usually presents in association with cognitive, behavioral, and motor alterations. Van Rijn, in a recent systematic review, highlighted the prevalence of ASD and ADHD symptoms in XYY patients in most cases. Executive functions, such as working memory, cognitive flexibility, and inhibitory control, also suffer greater impairment compared to the general population. The cognitive alterations caused by syndrome 47, XYY are also linked to neuroanatomical alterations caused by the overexpression of homologous and holandric regions of the Y chromosome. The NLGN4Y gene, which is related to autistic spectrum symptoms and social responsiveness is an example of these overexpressed genes. **Final Comments:** The understanding of anatomical, physiological and cognitive alterations allows a better elucidation of the relationship between the Y chromosome and neuropsychomotor development. A striking feature of Jacobs syndrome are the behavioral and cognitive changes present in carriers; such changes are closely connected to neuroanatomical variations triggered by genetic reasons. This disorder, therefore, manifests itself as a peculiar intersection of genetic, neurological, and psychiatric factors, thus being a fertile field for the development of research that integrates several areas of medical knowledge.

Neurogenética

TL 1105900

MILLER-DIEKER SYNDROM: A CASE-BASED UPDATE

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Case Presentation: A 5-year-old female presents with significant delay in neuropsychomotor development, first observed at 3 months of age, and refractory epileptic seizures, despite multiple regimens with anti-seizure drugs. Previous physiological history revealed gestational bleeding until the 4th month of pregnancy. Neurological exam revealed adequate visual contact and social smile, but no language, and significant axial and appendicular hypotonia, with absence of cervical tone. Morphological examination revealed the presence of microcephaly, divergent strabismus to the right, epicanthus, ocular hypertelorism, flat nasal base, small and upturned nose, palpebral fissures turned down and prominent forehead. Computed Tomography and Magnetic Resonance Imaging of the skull were performed, which showed lissencephaly and agria. A molecular study to search for microdeletion syndromes was performed, which detected deletions in the METTL16 and PAFAH1B1 genes of chromosome 17p13.3. **Discussion:** Miller-Dieker syndrome (MDS) is a rare genetic disease that occurs due to deletions in chromosome 17p13.3. It is characterized by lissencephaly, craniofacial alterations, growth retardation, delay in neuropsychomotor development and it is common to present refractory seizures. Lissencephaly associated with MDS tends to be more severe when compared to other diseases of chromosome 17p13.3, being classified as grade 1 lissencephaly (agria), also distinguished by its craniofacial alterations. The genetic deletion of MDS usually occurs between the PAFAH1B1 and YWHAE genes, with the PAFAH1B1 isolated deletion resulting in classic lissencephaly. In this case, there was a deletion of two genes between these chromosomes, which configures a MDS case. There was also a deletion of METTL16 gene that succeeds the PAFAH1B1, and, despite being associated with the MDS, the exact effects on the progression or severity of the syndrome are not elucidated. It is known, however, that it may be associated with hepatocellular carcinoma. **Final Comments:** MDS is rare and has a poor prognosis, with a reduced life expectancy in most cases. Its suspicion is very important when there is a typical phenotype associated with refractory seizures, requiring evidence of lissencephaly by imaging.

Neurogenética

TL 1105904

SUSCEPTIBILITY TO INFECTION-INDUCED ACUTE ENCEPHALOPATHY (IAE1/ANE1) TRIGGERED BY COVID-19

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Case Presentation: A 3-year-old male patient had a first episode of epileptic seizure at the age of 1 year, 2 days after the onset of fever. After this episode, he had several Hospitalizations for epileptic seizures, always preceded by fever, and in the last one he had status epilepticus and required sedation. His cerebrospinal fluid showed increased proteinorrachia (122), 6 red blood cells and 10 cells (69% macrophages) and MRI of the Skull showed hypersignal on T2 FLAIR temporal poles, external capsule, insula and posterior trigone, bilaterally. Normal interictal electroencephalogram. In 1 year, when he had COVID, he started with severe epileptic encephalopathy, requiring ICU admission. New MRI showed extensive white matter lesions on T2 FLAIR. After treatment with pulse therapy with corticosteroids and tocilizumab, he had a response. When he started with fever again, he made early use of steroids, preventing further deterioration. His father was diagnosed with encephalitis at age 4. After Exome sequencing, found a probably pathogenic variant in heterozygosis in the RANBP2 gene c.1754C>T: p. (Thr585Met), suggesting the diagnosis of Susceptibility to Infection-Induced Acute Encephalopathy (IAE1/ANE1), an autosomal dominant disease. Segregation analysis showed that her father is also a carrier of the variant. **Discussion:** The most frequent clinical presentation of IAE1/ANE1 is characterized by three stages: prodromal (febrile viral infection), acute (encephalopathy, on average 3 days after the beginning of the prodromal phase, which may be accompanied by epileptic seizures and focal neurological signs), and recovery. Episodes can be recurring. MRI findings are characterized by T2/FLAIR hypersignal in the thalamus and brainstem. The outcome ranges from complete recovery to persistent deficits and death. Several infectious triggers have already been reported, but here we report one of the first triggered by COVID-19. Early high doses of corticosteroids seem to be the best treatment. IL6 (tocilizumab) and TNF alpha inhibitors appear to play a role, but there are very few studies to date. **Final Comments:** Although rare, the investigation of genes associated with susceptibility to Acute Encephalopathy should be investigated in recurrent cases triggered by infectious triggers, as early treatment with corticosteroids or immunomodulators seems to change the outcome of patients.

Neurogenética

TL 1106067

RECESSIVE TAY-SACHS DISEASE: A CLINICAL CASE STUDY

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Case Presentation: Male patient, 3 years and 8 months old, born in Brazil, son of parents with no history of consanguinity, pregnancy discovered at 5 months, with prenatal care and delivery without complications. The patient has a family history of a first cousin with the disease. History of developmental regression from baseline to 2 years, with progression to global ataxia. Cranial magnetic resonance imaging showed signal alteration with atrophic effect affecting the white matter adjacent to the trigones and posterior horns of the lateral ventricles, hypersignal and volumetric reduction of the inner and posterior surface of the thalamus. Fundoscopy showed cherry red spots in the macular area bilaterally. Genetic panel showing homozygous pathogenic mutation of the HEXA gene and deficiency of oxidative phosphorylation by mutation in the PNPT1 gene compatible with the diagnosis of Tay-Sachs disease, with Sodium Valproate being prescribed for the onset of crises. At 2 years and 11 months, the patient had preserved transitive praxis, global ataxia, axis instability without preferential fall, broad-based gait and little movement of the upper limbs, exalted global osteotendinous reflexes with reduced axial tone, and preserved trophism. At 3 years and 5 months, he developed seizures that had not been contained with prescribed medication since the time of diagnosis. The electroencephalogram showed changes with generalized epileptiform activity and carbamazepine was prescribed, with a 90% regression in the number of seizures after two days of medication. **Discussion:** Tay-Sachs disease is a rare genetic disease transmitted from parents to children. It is caused by the absence of an enzyme that helps break down fatty substances called gangliosides, which reach toxic levels in the brain and spinal cord and affect nerve cell function. Signs and symptoms of the disease usually begin at 3 to 6 months of age. Having a reduction in muscle development and atrophy, evolving, over time, to seizures, loss of vision and hearing, paralysis and other major problems. Tay-sachs disease results in mortality within a few years of life. **Final Comments:** This case study reveals the importance of clinical and imaging follow-up of patients with diseases whose prognosis is reserved, as this, associated with adequate therapy, brings results that in the short term provide a better quality of life for the patient and family.

Neurogenética

TL 1106107

BRAINSTEM ISCHEMIC SYNDROME IN JUVENILE NEUROFIBROMATOSIS TYPE 2: CASE PRESENTATION

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Case Presentation: An 18-year-old male patient admitted to our Hospital complaining of intense headache, with spontaneous relief after three days, evolving then with left hemiparesis that worsened during the first week and with posterior gradual improvement. Physical examination demonstrates left spastic hemiparesis with hyperreflexia and right third cranial nerve palsy (congenital). Ectoscopy showed multiple fibroelastic skin nodules and left crystalline lens opacity. Patient brought a magnetic resonance (MRI) (Jul-15, 2021) that showed a focus of T2/FLAIR and DWI hyperintense signal with reduced ADC values, contrast ring enhancement on T1 and SWI slight hypointense signal localized in right midbrain and pons measuring 2x2,5 cm indicating hemorrhagic-ischemic injury or neoplastic nature lesion (glioma). Subsequent MRI (Ago-06, 2021) showed extension of the lesion to right internal capsule persisting the same patterns of signal. Stroke mechanism was extensively investigated with no relevant findings. Because of the possibility of neoplastic nature of the lesion, patient underwent MR spectroscopy and perfusion (Oct-10, 2021) in which it was noticeable volume reduction of the initial lesion, persistent restricted diffusion in addition of multiples contrast enhanced nodules of cranial nerves and cervical roots. The spectroscopy study showed discreet increase of choline and lactate indicating ischemia. CBV perfusion MR values were normal. This set of findings may suggest the diagnosis of brainstem ischemic syndrome in juvenile neurofibromatosis type 2 (NF2). **Discussion:** NF2 is an autosomal-dominant inherited tumor-prone disorder, predisposing the occurrence of meningiomas, ependymomas and specially schwannomas. Patients are also characterized by skin abnormalities, juvenile cataract, retinal hamartoma and scoliosis. Case series studies identified progressive brainstem ischemia as a rare presentation of NF2 occurring in young patients. These cases had no obvious reason for stroke like occlusion of a visible larger vessel, especially not in the vertebral or basilar arteries. Thus, a microvascular affection is the most likely cause and the midbrain/brainstem area seems to be the predilection site for these ischemic events in NF2. **Final Comments:** We presented a case of progressive brainstem ischemia with persistent restricted diffusion on MRI after 3 months, representing a challenge diagnosis due to its rarity and differential diagnosis with neoplastic lesions.

Neurogenética

TL 1106177

POMPE DISEASE (PD) IN A PATIENT WITH ACROMEGALY PREVIOUSLY DIAGNOSED AS POLYMYOSITIS – A CASE PRESENTATION

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Case Presentation: A 46-year-old male patient, sixth son of non-consanguineous parents and with no similar family history, was previously diagnosed with acromegaly (enlargement of nose, hands and ears, mandibular prognathism and deep voice) due to a pituitary adenoma. Six years ago he started to have proximal muscle weakness. Muscle biopsy revealed mild muscle atrophy with a lymphocyte infiltrate, suggestive of an inflammatory myopathy. He was diagnosed with polymyositis and treated with prednisone, with little response. One year later he was forwarded to a neurology outpatient clinic with progressive fatigue, difficulty standing up and low back pain. Symptoms have become worse over time. The last examination showed grade 4 muscle weakness globally and marked hypotrophy of proximal muscles, including pectorals, associated with a myopathic gait. Deep tendon reflexes were hypoactive globally. Plantar responses were indifferent. Cranial nerves, sphincter function and bulbar muscles were preserved. He had elevated creatine kinase (327 IU/L). A next-generation sequencing panel for neuromuscular disorders was performed and showed two GAA pathogenic variants (c.-32-13T>G – intronic; and Exon 18 deletion). Enzymatic testing for acid alpha-glucosidase was 0,64 (reference range – 4.57 ± 1.37 umol/L/h), which confirmed Pompe disease (PD). **Discussion:** PD is still underdiagnosed in Brazil and may affect people of all ages with varying degrees of severity, usually presenting as a limb-girdle myopathy, predominantly in lower limbs. Symptoms such as dysarthria, dysphagia, scoliosis, pain, fatigue and sphincter involvement can also be found. Mild PD is common in late-onset patients. Because of its similar presentation, PD can be mistaken for polymyositis, which also exhibits a progressive proximal weakness pattern. This patient had a biopsy reportedly compatible with polymyositis, but search for serum antibodies was negative and corticosteroid therapy didn't show any improvement. In those situations, testing for genetic myopathies might reveal alternative diagnoses of potentially treatable conditions, such as PD. **Final Comments:** To the best of our knowledge, this is the first case of PD in a patient with a typical presentation of acromegaly. Here we also highlight the recognition of Pompe disease as an important differential diagnosis of polymyositis. This knowledge can reduce the delay in etiologic determination and might reveal potentially treatable conditions.

Neurogenética

TL 1106256

AUTOSOMAL DOMINANT HEREDITARY SPASTIC PARAPLEGIA WITH PURE PHENOTYPE AND EARLY ONSET CAUSED BY VARIANT IN THE KIF1A GENE

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Case Presentation: A 14-year-old male patient started walking on tiptoe at one and a half years of age. These symptoms evolved progressively. On current physical examination, he has grade IV strength in dorsiflexion of the feet and interosseous in the hands, patellar and achilles hyperreflexia, bilateral Achilles clonus, bilateral Babinski sign and reduced vibratory sensitivity. Mild hyperreflexia in upper limbs. He also has low-amplitude, high-frequency postural and kinetic tremor. His parents are not consanguineous and there are no similar cases in the family. He performed investigation of sporadic and metabolic causes that showed no changes, as well as MRI of the skull and spinal cord. We opted for genetic investigation with a diagnosis of pure Hereditary Spastic Paraplegia (HSP), as an isolated case, through a panel of genes. In this, the probably pathogenic variant was found in the KIF1A gene c.1021A>G (p.Thr341Ala), in heterozygosity. This was also evaluated in their parents, who did not have the mutation. Thus, the diagnosis of SPG30, pure form, autosomal dominant (AD), with early onset, was concluded. **Discussion:** Hereditary Spastic Paraparesis is a heterogeneous group of rare neurogenetic disorders caused by variants in more than 80 genes, with all forms of inheritance already described. They are divided into pure or complicated forms, according to the clinical presentation. SPG30, caused by variants in the KIF1A gene, which encodes a motor protein expressed exclusively in brain tissue and transports cargoes from the neuronal body to the periphery of neurites. In addition to the two forms of inheritance, AD conditions have already been described as pure in some families and as complicated in others, and also as a cause of polyneuropathy, sometimes making it difficult to confirm the pathogenicity of the variant. Few cases with this phenotype have been described in the Brazilian population. **Final Comments:** A better understanding of the variants found in the Brazilian population and, consequently, which forms of HSP we find here, helps in the interpretation of molecular results and even in the creation of more directive gene panels, depending on the origin of the patient. In addition, better understanding the changes caused by mutations in the KIF1A gene further broadens the spectrum of diseases associated with it.

Neurogenética

TL 1106349

ADULT-ONSET LEUKOENCEPHALOPATHIES: A DIAGNOSTIC CHALLENGE

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Case Presentation: Male patient, 58 years old, hypertensive, with a history of colon cancer in 2011 (treated with resection and chemotherapy) and hypogonadotropic hypogonadism. He began with attention difficulties in September/2021, which worsened over the next months with cognitive-behavioral changes like aggressiveness, irritability and apathy, sleep disturbances, memory dysfunction and difficulties to perform daily tasks (paying bills on the phone, financial controlling). On neurologic examination, the patient only presented cognitive changes, with pronounced executive dysfunction. He was then Hospitalized for investigation. Cerebrospinal fluid examination showed slightly elevated protein level, and brain MRI showed symmetric frontal white matter hyperintensity with spared subcortical U fibers on FLAIR images, besides no contrast enhancement and no restricted diffusion foci. Metabolic, infectious, and serologic tests were normal. The patient underwent further investigation, including genetic testing to Adult Leukodystrophies, which revealed heterozygosity for the GBE1 gene. **Discussion:** Leukodystrophies are hereditary disorders that present with progressive white matter dysmyelination or defects of myelin biosynthesis and metabolism. The pattern of white matter involvement is essential to narrowing the diagnosis process. Although rare, it is an important differential diagnosis to be considered in patients with cognitive decline. Even with advances in imaging and genetic testing, about 40% of patients remain undiagnosed. Mutations in the GBE1 gene is associated with Adult Polyglucosan Body Disease, an autosomal recessive disease that presents as neurodegenerative condition with involvement of the central and peripheral nervous system. **Final Comments:** Leukoencephalopathy disorders represent a diagnostic challenge for the neurologist, requiring an extensive investigation, including infectious, immune-mediated, degenerative, vascular, metabolic, and neoplastic process, in addition to genetic causes. This last group constitutes the major diagnostic challenge, mainly due to the availability of diagnostic resources in health services. For this reason, a systematic approach to the investigation of these patients is mandatory.

Neurogenética

TL 1106376

EFFECTS OF LONG-TERM TREATMENT IN A PATIENT WITH ISOVALERIC ACIDEMIA

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Case Presentation: An 11-year-old female patient, daughter of consanguineous parents, was born with no complications during pregnancy, childbirth or puerperium. At the age of 3 years, she required Hospital admission with a history of recurrent vomiting, fever, metabolic acidosis, lethargy, and diarrhea. It was also reported neuropsychomotor development delay. The patient was suspected to have encephalitis, which led to treatment with ceftriaxone, aciclovir and vancomycin, but remained a concern for underlying metabolic disorder. During Hospitalization, the patient presented one convulsive seizure, beginning phenobarbital therapy. Plasma amino acids and urine for organic acids were performed to investigate metabolic disorders. The metabolic studies revealed a marked excretion of isovalerylglutamate, along with a moderate excretion of 3-hydroxy-isovaleric acid. The diagnosis of Isovaleric Acidemia (IVA) was made in the same year. The genetic examination revealed, in homozygous, the variant p.Arg414Trp C>T in the IVD gene (OMIM 607036). The patient was placed on a protein-restricted diet and use of L-carnitine 100 mg/kg/day therapy. Eight years later, she made significant progress on speech and motor development, with improvement on strength, gait, and academic performance. **Discussion:** IVA is a rare inborn error of leucine metabolism caused by biallelic pathogenic variant in the isovaleryl CoA dehydrogenase (IVD) gene. The accumulation of isovaleric acid, toxic to the central nervous system, is related to the phenotypic abnormalities in this disorder. Patients may present an acute neonatal form of the disease, leading to massive metabolic acidosis and rapid death in the neonatal period, or a chronic intermittent form, with retarded psychomotor development, aversion to dietary protein, hyperammonemia, and pernicious vomiting. If a patient is not treated on time, the patient may proceed to severe acidosis, coma, and death. Therapy consisting of L-carnitine supplementation and protein restriction may be sufficient for effective management of IVA, starting as soon as possible after birth. **Final Comments:** Our results demonstrate the neurocognitive improvement of long-term treatment with L-carnitine supplementation and protein restriction in our patient with IVA. We encourage the analysis for this disorder when investigating patients with recurrent vomiting and lethargy, born from consanguineous marriage.

Neurogenética

TL 1106381

A RARE CAUSE OF NEURODEVELOPMENTAL DISORDER WITH INVOLUNTARY MOVEMENTS: GNAO1 ENCEPHALOPATHY

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Case Presentation: A 6 years old girl born from vaginal delivery, at term, by the age of 8 months started with involuntary hyperkinetic jerk movements most notably in the hands, and jaw, especially when anxious, with progression of symptoms. Crawling and sitting at the age of 2 years old and first words at the age of 5. At the age of 6, the patient was admitted to the current service and parents reported anxious behavior and worsening of the chewing movements and uncoordinated arm movements. Neurological examination showed axial hypotonia, compromising trunk sustentation, sialorrhea, uncoordinated crawling, chewing movements, Babinski sign and achilles tendon shortening. A full exome sequencing was performed, demonstrating, in heterozygous, the variant chr16: 56.351.397 A>T (or alternatively c.737A>T - ENST00000262493), which promotes the substitution of the glutamate amino-acid by a valine on the codon 246 (p.Glu246Val) in GNAO1 (OMIM 139311). This variant has never been described in the medical literature. **Discussion:** GNAO1 encodes the alpha subunit of the Go subclass of heterotrimeric guanine nucleotide-binding proteins (G proteins), which is expressed in the brain and has an important role in the regulation of neuronal excitability and neurotransmission. Heterozygous mutations in GNAO1 are associated with a wide range of manifestations, from Developmental and Epileptic Encephalopathy-17 (DEE17) to significant Neurodevelopmental Disorder with Involuntary Movements (NEDIM). Patients with NEDIM may present with severe choreoathetosis, developmental delay, and hypotonia. The hyperkinetic movements were often exacerbated by specific triggers, such as voluntary movement, intercurrent illnesses, high ambient temperature, and specific emotions, such as anxiety, a phenotype similar to that of the patient described herein. **Final Comments:** Our study reports a Brazilian patient with NEDIM, confirming the association between the GNAO1 p.Glu246Val pathogenic variant and this phenotype, which has not been described before in the literature. Therefore, we encourage that GNAO1 pathogenic variants should be taken into consideration when investigating patients with unexplained early-onset hyperkinetic movement disorders and neurodevelopmental delay.

Neurogenética

TL 1106396

MORC2 VARIANT PRESENTING WITH DEVELOPMENTAL DELAY, DYSMORPHIC FACIES, POLYDACTYLY, AND MICROCEPHALY

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Case Presentation: 12-year-old male patient, nonconsanguineous parents, born at term, with cesarean delivery, no complications during pregnancy, childbirth or puerperium. At 9 months, neuropsychomotor development (NPDM) delay and dysmorphic facies were observed. He also presented preaxial polydactyly, which was resolved. At 12 months, he had an episode of febrile seizure. Posteriorly, it was also verified intellectual disability and hyperactivity. During etiological investigation, brain Magnetic Resonance Imaging (MRI) revealed thin corpus callosum. Cytogenetic tests were performed, with suspicion of Fragile X Syndrome, and identified no alterations. Besides the NPDM delay, and dysmorphic facies, the neurological examination was notable for generalized hyporeflexia, normal strength, microcephaly, and digitigrade gait. An analysis of the exome sequencing was solicited, which showed, in heterozygous, the variant chr22: 30.958.684 C>T (or alternatively c.79G>A - ENST00000397641) in the MORC2 (OMIM 616661) gene, which promotes the substitution of the glutamate amino acid by lysine at the 27th codon (p.Glu27Lys). This is a rare variant, and was described in the medical literature associated with developmental delay, dysmorphic facies, and microcephaly. **Discussion:** Microorchidia CW-type zinc finger protein 2 (MORC2) encodes a DNA-dependent ATPase involved in epigenetic silencing through chromatin modification, necessary for chromatin remodeling, DNA repair, and transcriptional regulation. Heterozygous variants in the MORC2 are associated with Charcot-Marie-Tooth disease type 2Z; and developmental delay, impaired growth, dysmorphic facies, and axonal neuropathy (DIGFAN syndrome). DIGFAN syndrome is an autosomal dominant neurodevelopmental disorder, associated with de novo mutations. Besides the mentioned characteristics, the patients may also present delayed acquisition of motor skills, gait abnormalities, hyporeflexia, and abnormalities on brain imaging, a phenotype similar to that of the patient described herein. **Final Comments:** Our study presents a 12-year-old male with a rare MORC2 p.Glu27Lys heterozygous pathogenic variant. We highlight the importance of MORC2 variants analysis in front of patients with global developmental delay, short stature, microcephaly, and variable dysmorphic facies.

Neurogenética

TL 1106525

PHENOTYPIC VARIABILITY IN 3 SIBLINGS WITH CLCN1 ASSOCIATED BECKER'S MYOTONIA CONGENITA

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Case Presentation: Three affected siblings whose parents and grandparents were consanguineous. The parents had 3 affected and 7 non-affected children. Case 1: A 40-year-old woman presented with a history of muscle cramps, difficulty standing-up after being seated for a long period, stiffness in legs while walking, dysphonia and sporadic transfer dysphagia. She exhibited pronounced muscular hypertrophy and generalized myotonic phenomenon (GMP) at examination, was able to walk unassisted and did not need medication for myotonic phenomenon. Electroneuromyography (EMG) showed diffuse myotonic discharges. Case 2: A 48-year-old male presented with GMP and complaints of dysphagia for liquids, difficulty climbing stairs and brushing his teeth. He reported weakness since childhood and had frequent falls and needed intermittent assistance when walking. Myotonic phenomenon did not respond well to carbamazepine or phenytoin. EMG demonstrated non-dystrophic myotonic syndrome. Case 3: A 51-year-old male with a history of muscle cramps, dysphagia and dysphonia, muscle stiffness with difficulty running and standing-up after being seated for a long period. Examination showed GMP, hyporeflexia and myoclonus in upper and lower limbs. Myotonic phenomenon was well controlled with phenytoin 200mg/day. None of the siblings reported exacerbation of myotonic phenomenon by cold. The 3 siblings were submitted to a genetic panel for neuromuscular disorders which revealed a CLCN1 c.1886T>C variant in homozygosity. **Discussion:** Myotonia Congenita is an inherited disorder caused by biallelic pathogenic variants in CLCN1, which encodes chloride channel CLC-1 expressed predominantly in skeletal muscles. These variants cause muscle hyperexcitability and delayed muscle relaxation. The disease usually presents in childhood but has a highly variable phenotype even in patients with the same variants, such as in this family, where one patient had muscle hypertrophy and mild myotonic phenomenon without disability while her brother had intense, drug-resistant myotonic phenomenon leading to falls, difficulty walking and quality of life impairment. **Final Comments:** Our study reports three patients with the same pathogenic variant presenting with different phenotypes. There's still a lack of description on the multitude of repercussions related to chloride channel alterations. We therefore encourage more research regarding this topic in order to unveil the potential repercussions of this condition.

Neurogenética

TL 1106539

MULTIPLE ABDOMINAL ANEURYSMS IN AN ASYMPTOMATIC LAMA2 MYOPATHY AND LEUKOENCEPHALOPATHY

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Case Presentation: A 50-year-old female, hypertensive, daughter of non-consanguineous parents, presented with acute pain at the upper left abdominal quadrant. Abdominal ultrasound revealed left non-obstructive renal lithiasis. Computed tomography scan incidentally showed multiple saccular aneurysms close to the renal hilum and to the splenic hilum, in addition to expressive volumetric loss of paravertebral and lumbar dorsal spinal muscle bellies. Her creatine phosphokinase (CPK) was 359 IU/L. A next generation sequencing panel for neuromuscular disorders revealed two pathogenic variants in LAMA2: c.2461A>C (p.Thr211Pro). Pulmonary function tests disclosed only mild obstructive respiratory disorder. Brain MRI revealed extensive involvement of the white matter of the cerebral hemispheres, mainly in the deep regions of the brain, confluent and bilateral, compatible with leukoencephalopathy. **Discussion:** LAMA2-Related Muscular Dystrophy (LAMA2-MD) is part of a group of muscle disorders called Congenital Muscular Dystrophies (CMD) and the clinical manifestations represent a continuous spectrum, varying from a severe congenital muscular dystrophy to milder late-onset LAMA2-MD. Late-onset LAMA2-MD usually presents with proximal muscle weakness (not present in our case) and delayed motor milestones (but independent ambulation can be achieved). A leukoencephalopathy is also present and is an important finding to suggest this diagnosis. Reported atypical findings include seizures, rigid spine syndrome with joint contractures, progressive respiratory insufficiency and cardiomyopathy with or without conduction defect, but aneurysms were not yet mentioned. LAMA2 pathogenic variants can lead to defects in laminin alpha-2 protein, also called merosin. As merosin is a subunit of laminin and laminin isophormes are expressed in a lot of tissues, including endothelial and perivascular basement membranes, we might observe vascular abnormalities as the aneurysms described in this case. **Final Comments:** Here we report a case not yet described, as far as we know, of multiple abdominal aneurysms in a patient diagnosed with muscular dystrophy and leukoencephalopathy related to LAMA2. Although there is a lack of literature regarding an association of LAMA2 with aneurysms, we hypothesize that alterations in laminin could interfere with vessel wall architecture.

Neurogenética

TL 1106550

NOVEL SH3TC2 GENE MUTATION IN A BRAZILIAN INDIVIDUAL WITH CHARCOT-MARIE-TOOTH DISEASE TYPE 4C

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Case Presentation: A 42-year-old Brazilian male with a history of hypertension was admitted to our medical department due to progressive numbness and weakness in the lower extremities. The patient described his motor performance as relatively poor compared to his peers since childhood. At age 39, he noticed decreased sensation in his feet, causing difficulty in wearing shoes and walking. He slowly developed progressive weakness of lower limbs, unsteadiness, and clumsiness at the age of 41. He is the son of a consanguineous couple and has a sister with similar symptoms. Physical exam was remarkable for pes planus deformity, stork legs, and scoliosis. Neurological examination revealed slowing of saccades, hypoacusis, occasional tongue fasciculations, and mild facial diplegia. He had weakness of the proximal (4+/5) and distal lower limbs (4-/5) with relatively preserved muscle strength of the upper limbs. Sensation of vibration, light touch, and pinprick were markedly reduced in both lower extremities. Hyporeflexia, a broad-based gait, and a positive Romberg sign were also noted. Biochemical analysis showed elevated serum creatine kinase (1278 U/L) and neuron-specific enolase (10 µg/L). His complete blood cell count, electrolytes, HbA1C, Vitamin B12, folic acid, liver, thyroid, and kidney function tests were all unremarkable. Autoimmune panel, serum tumor markers, and cerebrospinal fluid analysis disclosed no abnormalities. Electrophysiological studies indicated a symmetrical, length-dependent, demyelinating sensorimotor polyneuropathy with axonal loss and markedly reduced motor conduction velocity. Genetic testing revealed a novel homozygous pathogenic variant, c.1144G>T p.Glu382*, in SH3TC2. **Discussion:** Our patient presented with the typical features of Charcot-Marie-Tooth disease type 4C (CMT4C), including demyelinating neuropathy, foot deformity, scoliosis, and cranial nerve involvement. We identified a new mutation in SH3TC2, predicted to be disease causing since it results in a very premature stop codon at amino acid position 382 and a shortened protein with a potentially abnormal function. The identified variant is absent in 141000 control chromosomes from gnomAD; it has never been reported in the medical literature or in the ClinVar database. **Final Comments:** In conclusion, we present a Brazilian patient with CMT4C associated with a novel mutation of SH3TC2. This case highlights the importance of considering rare diagnoses in the new generation sequencing era.

Neurogenética

TL 1106567

PHENOTYPIC VARIABILITY IN TWO SIBLINGS WITH UNVERRICHT-LUNDBORG DISEASE: JUVENILE MYOCLONIC EPILEPSY PHENOTYPE X CLASSIC PHENOTYPE

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Case Presentation: Here, we present two siblings with consanguineous parents (first-degree cousins) and an uncle with a history of epilepsy. **Case 1:** A 28-year-old female with a history of myoclonic epilepsy since she was 10-years-old that became worse over time. She had normal psychomotor development until 11-years-old. Physical examination demonstrated global ataxia, intense action myoclonus and psychotic symptoms. Genetic testing revealed homozygous variant for the dodecamer expansion in the cystatin B (CTSB) gene, which confirmed the clinical diagnosis of Unverricht-Lundborg Disease (ULD). She scored 21/30 in the Mini-mental state examination (MMSE) and 28 in the Scale for the Assessment and Rating of Ataxia (SARA). Brain MRI disclosed diffuse cerebral and cerebellar atrophy. **Case 2:** Brother of Case 1, 23-year-old, who developed tonic-clonic seizures and occasional myoclonus that started at 11-years-old. Psychomotor development was normal. On examination he didn't exhibit action myoclonus. Coordination tests and gait were preserved. He scored 30/30 in the MMSE and 29/30 in the Montreal Cognitive Assessment (MOCA). Electroencephalography showed sparse paroxysms of generalized spike-wave and polyspike-wave epileptiform activity, with normal baseline activity. Brain Magnetic Resonance Imaging was normal. **Discussion:** Unverricht-Lundborg Disease (ULD) is a rare, disabling and severe genetic condition associated with pathogenic variants in CTSB. ULD is considered the most common form of Progressive Myoclonus Epilepsy (PME). In our cases, the sister expressed a classic ULD phenotype while the brother presented a relatively mild phenotype, which can be mistaken for similar conditions as Juvenile Myoclonic Epilepsy (JME). The relative intensity of the various symptoms and the speed at which the disease progresses can vary, even within the same family. Therefore, it isn't possible to assume that the brother's disease will progress in a similar way to his sister's condition. **Final Comments:** In this report we aimed to describe the first case of a JME-like phenotype and a classic ULD phenotype in siblings. This highlights the possibility of searching for ULD in some cases of families with clinical diagnosis of juvenile myoclonic epilepsy.

Neurogenética

TL 1106598

ANALYSIS AND THE IMPORTANCE OF THE DIFFERENTIAL DIAGNOSIS BETWEEN KENNEDY'S DISEASE AND AMYTROPHIC LATERAL SCLEROSIS

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Case Presentation: Male, 57 years old felt weakness in the right upper limb in rural work, dropping the instrument used five years ago. Three years ago he presented difficulty in flexion of the right index finger and atrophy of the right upper limb, diagnosed with ALS. The symptoms started on the right side and progressed to the left side. He also reports polyuria, urgency, cries easily, imbalance and falling, night sweats and coldness in the upper limbs. History of 10kg loss and use of Riluzol 50mg. On physical exam he presented grade 3 strength deficit in the four limbs, fasciculations in the right upper limb, upper back and tongue that presented atrophied right muscles, interosseous atrophy of hands, hyporeflexia and staggering gait. **Discussion:** Spinal and bulbar muscular atrophy (SBMA) or Kennedy's disease, is a rare, X-linked, inherited lower motor neuron disease. It is characterized by progressive muscle weakness. An expanded trinucleotide repeat (CAG > 37) in the androgen receptor gene, which encodes glutamine, is the responsible mutation. Toxicity of this mutant protein affects both motor neurons and muscles. The main symptoms are weakness in the lower limbs, more proximal than distal, hyporeflexia, fasciculations, and muscle atrophy. Non-motor symptoms include dysphagia, dysarthria, testicular atrophy, Brugada syndrome, urinary disorders, and dementia due to reduced frontal brain volume. Amyotrophic Lateral Sclerosis is the main differential diagnosis and is ruled out by the absence of hyperreflexia and sensory changes. Early diagnosis is important because the prognosis is related to the risk of weakness-related falls and bronchoaspiration, and sudden death from Brugada Syndrome. **Final Comments:** SBMA is not only a lower motor neuron disease, but a complex disorder affecting different systems, including the central nervous system. The differential diagnosis with (ALS) is of utmost importance, since the age of presentation of SBMA is 30-40 years and the patient is already in a very advanced picture at diagnosis.

Neurogenética

TL 1106622

CEREBELLAR ATAXIA ASSOCIATED WITH LANGERHANS-CELL HISTIOCYTOSIS: A CASE PRESENTATION

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Case Presentation: A 36-year-old male patient suffered 2 episodes of skull fracture related to low kinetic energy accidents at the age of 3. When he was 4 years old, considering the imaging alterations and skull bones fragility, a bone biopsy was performed and suggested Langerhans-Cell Histiocytosis, which was confirmed with immunohistochemistry. The patient was submitted to chemotherapy for 1 year and 6 months with complete remission. When he was 12 years old, gait ataxia, urinary volume increase and cognitive delay started. At 14 years, ataxia stabilized. He was restricted to a wheelchair due to gait impairment. Cerebellar atrophy was identified by Magnetic Resonance Imaging (MRI). Diabetes Insipidus (DI) was diagnosed and therapy with desmopressin was introduced. During the following years, the patient presented pseudobulbar affect, lower limbs myoclonus, dysphagia and dysarthria. An MRI of the spine when he was 33 years old revealed T2 hypersignal in the lateral funiculi of the whole spinal cord, more prominent in the thoracic segment. A skull MRI when he was 35 years old showed midbrain and pontocerebellar atrophy with central pontine hypersignal and marked globus pallidus T2 hyposignal, in addition to focuses probably representing hemosiderin or calcifications at the temporo-occipital transitions and a slight increase in the supratentorial cerebrospinal fluid spaces. **Discussion:** Langerhans-cell histiocytosis (LCH) is considered the most common histiocytic disorder and is associated with an abnormal accumulation of immature histiocytes in some parts of the body, usually bone, skin, lungs and pituitary. Diabetes insipidus (DI) is the most common neurological manifestation, but cerebellar ataxia has been reported as an extremely rare presentation and may mimic an autosomal recessive ataxia. The mechanisms leading to cerebellar ataxia in Langerhans-cell histiocytosis are still unclear. In our case, DI and ataxia presented almost simultaneously, and the patient also exhibited pyramidal syndrome and a pontine gliosis that may be related to osmotic demyelination events. **Final Comments:** Ataxia is a rare finding in LCH with a still unclear pathogenesis. It is possible that in some cases neurological complications of this disease are related to osmotic demyelination episodes caused by diabetes insipidus.

Neurogenética

TL 1104712

MARBURG MARIANT OF MULTIPLE SCLEROSIS AFTER COVID-19 SUCESSFULLY TREATED WITH BETAINTERFERON 1A

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Case Presentation: The Marburg variant of MS is a rare variant that leads to an severe clinical course, with a high rate of mortality or severe residual deficits and unclear pathophysiology. A 20-year old female patient, presented at the Hospital emergency with left inferior limb paresis and visual blurring. The neurologic exam showed complete and proportionate left hemiparesis with pyramidal signs and clonus, loss of proprioception and vibration in lower limbs, tactile and painful hypoesthesia on the left side. This report describes a rare case of Marburg variant associated with COVID-19 infection.

Neuroimunologia

TL 1104728

HASHIMOTO ENCEPHALOPATHY: DIAGNOSTIC CHALLENGE

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Case Presentation: Woman, 49 years old, with hypothyroidism and fibromyalgia. The first symptom reported by her husband was crisis of anxiety that in a month was more frequent and associated with hallucinations. In 2 months, he noticed changes in memory with temporo-spatial disorientation and difficulty in recognizing family members. Involuntary tremors in the upper limbs of myoclonus were observed more frequently during sleep. During a 9-month diagnostic investigation, the patient underwent a psychiatrist's Hospital being treated as a psychotic break. She was Hospitalized with the hypothesis of limbic encephalitis after neurologic evaluation. The tests revealed; MRI of the brain with alteration signal in the hippocampus bilaterally, electroencephalogram with record of electrographic crisis and presence of paroxysm in the previous temporal regions bilaterally. Serum tests identified anti-TPO: 940. Markers for autoimmune encephalitis were all negative and the possibility of paraneoplastic syndrome was ruled out. Pulse therapy with methylprednisone was performed and lamotrigine was prescribed. The patient showed significant clinical improvement. **Discussion:** Hashimoto encephalopathy (HE) is a term used to describe a rare encephalopathy, of presumed autoimmune origin, associated with Hashimoto's thyroiditis. It is included in a group of nonvasculitic autoimmune inflammatory meningoencephalopathies. The patient described presented all the characteristic symptoms: behavioral and psychiatric symptoms, movement disorders and convulsions. The way symptoms are on set up is typically acute or subacute which can become chronic. **Final Comments:** Behavioral complaints, hallucinations, accompanied by memory deficits and language disorders, are often found at an early stage of HE and may be confused with psychiatric disorders. Common differential diagnoses should be made with Creutzfeldt-Jakob disease, rapidly progressive dementias and limbic encephalitis.

Neuroimunologia

TL 1104814

INTRACRANIAL HYPERTENSION AS A RARE NEUROLOGICAL PRESENTATION OF SJÖGREN'S SYNDROME: A CASE PRESENTATION

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Case Presentation: A healthy, young female patient presented a sudden short-term loss of consciousness. In the next day she complained of moderate headache that worsened in supine position and emesis. It evolved with bilateral diplopia and convergent strabismus. Brain MRI showed signs of intracranial hypertension (ICH). Her examination revealed good general condition, gait deviation to the right in the Fukuda's Test, star gait with eyes closed and bilateral paresis of the VI cranial nerve. Pupils were isocoric, photoreactive, visual acuity was normal, as it was the fundoscopy. Lumbar puncture revealed proteins 41, glucose 40, negative anti-AQP4, anti-MOG and OCB, and intrathecal synthesis of IgA and IgM antibodies, indicating blood-brain barrier dysfunction, negative IgG, and intrathecal production of kappa free light chain (Kappa index 7.23), and neurofilaments = 4852. Laboratory tests showed VHS 34, anti-RO 240, anti-LA 174. She was referred to the Rheumatology Service for Sjögren's Syndrome (SS) investigation. Because of complaints of dysphagia and xerostomy during this period, salivary gland biopsy was performed, and the findings were compatible with SS. She was treated with corticosteroids, acetazolamide, azathioprine, hydroxychloroquine and lubricating eye drops, and all symptoms resolved. **Discussion:** ICH is a clinical syndrome characterized by increased intracranial pressure without brain or cerebrospinal fluid lesions that justify it, maybe explained by recent discoveries of the glymphatic and lymphatic systems of the brain. It mainly affects overweight or obese women. Although female, the patient in this case was eutrophic. SS is a rheumatologic disease with 8.5 to 70% of the patients presenting with neurological complains. The diagnosis of SS is based on clinical grounds and laboratory findings with positivity of anti-Ro and anti-La in 52% and 34% of the patients respectively. There are few cases described where ICH was the primary symptom of SS. In our case, the CSF analysis revealed a high Kappa index and neurofilaments, important biomarkers related to important neuroaxonal damage. Kappa index and neurofilaments are promising biomarkers for the diagnosis, prognosis and evaluation of the disease progression in immunomediated compromise of the Central Nervous System. **Final Comments:** Although rare, neurological manifestation of SS may be the first symptom of the disease. Among the neurological manifestations described, ICH is even less frequent.

Neuroimunologia

TL 1104943

DIAGNOSTIC CHALLENGES IN ANTI-NMDAR ENCEPHALITIS: A CASE PRESENTATION

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Case Presentation: A 29-year-old woman, lawyer, no comorbidities or psychiatric history, presented with agitation and insomnia. In few days, there was worsening of symptoms, associated with delusional speech and executive dysfunction. Present to psychiatry evaluation, with TAB diagnoses, Olanzapine started, but no improvement. After three weeks, the patient needs help for basic activities. She presented perioral movements and unexplained coma, with Hospitalar admission. The MRI, CSF, laboratory tests for infection and metabolism were normal. Acyclovir started due to suspected herpetic encephalitis. The patient was transferred to a tertiary center, anti-NMDA encephalitis was suggested and a sample of CSF and serum was sent for antibody testing. Screening imaging and markers excluded teratoma or other occult neoplasms. Empirical treatment was started with methylprednisolone 1g daily for five days, followed by five sessions of plasmapheresis and rituximab. The tests has been found positive for NMDAR antibodies, and the patient remains in intensive care for clinical support.

Discussion: Autoimmune encephalitis is caused by the production of antibodies against neuronal autoantigens. It affects mostly young women, presenting with acute or subacute psychiatric symptoms, seizures, cognitive deficits, coma, and autonomic dysfunction. Often psychiatric symptoms are the initial symptoms and these patients are mistakenly treated as primary psychosis or mood disorders, leading to diagnostic delay and worse prognosis. The association of anti-NMDAR encephalitis with neoplasms is large, and investigation should be extensive, focusing on ovarian teratoma. Treatment primarily involves intensive care associated with immunosuppressive therapy. The first line constitutes corticoids, immunoglobulin or plasmapheresis. The second line of treatment is done in selected cases, usually in patients with late diagnosis or in those with no association with tumor, being Rituximab the drug of choice. For refractory cases, Bortezomib is third line. The prognosis is generally satisfactory, however, because it is a long-stay disease in the ICU, it is susceptible to several clinical and neurological complications throughout treatment. **Final Comments:** Anti-NMDA encephalitis should be considered as a diagnostic possibility in cases of subacute psychiatric alterations with neurological symptoms, and empiric treatment having to be considered for better therapeutic results.

Neuroimunologia

TL 1105006

LYMPHOMA ASSOCIATED WITH FINGOLIMOD TREATMENT: FIRST REPORT OF ADULT-ONSET CASE IN BRAZIL

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Case Presentation: A 31-year-old white woman had been suffering from relapsing-remitting multiple sclerosis (RRMS) for more than 8 years. She had been taking fingolimod properly for one and a half years when she developed subacute behavioral changes, dysarthria, right numbness, and hemiparesis. The general laboratory showed 696 lymphocytes. As if she had relapsed, she received intravenous (IV) methylprednisolone with no significant improvement. Four months later, a repeat MRI scan showed new lesions with gadolinium enhancement in the posterior white matter and brainstem, in addition to meningeal enhancement in the frontal lobe. Fingolimod was discontinued because of possible progressive multifocal leukoencephalopathy. After its discontinuation, two months later, a comprehensive malignancy workup failed, including normal CTs, serous immunofixation and beta-2 microglobulin, a cervical lymph node biopsy, and a brain biopsy with typical MS findings. Subsequent CSF examination revealed 16 cells, and immunophenotyping revealed 3% non-neoplastic clonal plasma cells. Given this atypical evolution, our team of experts concluded a diagnosis of B-cell lymphoma during immunosuppression in advanced RRMS due to immune reconstitution. She received IV rituximab and IV methylprednisolone and had significant improvement. **Discussion:** Fingolimod is a sphingosine-1-phosphate (S1P) receptor agonist and the first oral agent approved for the treatment of RRMS. After phosphorylation in vivo, the drug acts as an S1P receptor modulator that binds with high affinity to four of the five known S1P receptors, retaining lymphocytes in lymph nodes and preventing their egress into the peripheral circulation. As a result, the infiltration of autoreactive lymphocytes into the CNS is reduced. It is believed that due to this mechanism, fingolimod may alleviate and reduce the disease burden of lymphoma, increase the efficacy of other treatment options, and improve the function of the immune system in destroying neoplastic cells. Therefore, the drug can be used not only to treat lymphoma, but also to control and prevent disease recurrence in patients in remission. **Final Comments:** To our knowledge, this is the first report of fingolimod-associated lymphoma confined to the CNS in Brazil. It is a reminder that immunosuppressive drugs have a full spectrum of side effects, not just in infectious diseases. Both solid tumors and hematologic cancers should be considered as possible causes of deterioration.

Neuroimunologia

TL 1105066

DIFFERENTIAL DIAGNOSIS IN DEMYELINATING DISEASES: A CASE PRESENTATION

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Case Presentation: A 46 years old female patient came to our Neurology Service in 2003 with decreased visual acuity in her left eye, which evolved to amaurosis after 7 days. Brain MRI revealed two hyperintense signs on T2 and FLAIR in the semioval center, without post-contrast enhancement, and hyper signal in the left optic nerve on the STIR sequence. She was treated with pulse therapy and partially improved. She returned after a few months complaining of low visual acuity in her right eye. A cervical MRI revealed contrast-enhanced lesions in C2-C3, CSF test was positive for oligoclonal bands (OCB) and a serum aquaporin-4 antibody was negative. She was treated with three cycles of pulsetherapy and began Interferon β1-a 44mcg, being episode free for several years, even though routine brain MRI revealed new lesions in 2009 and 2011. In 2020, at the age of 61, she complained of paresthesia in her left inferior limb, which ascended to the dorsal region. Neurologic examination revealed crural paraparesis and posterior cord syndrome. A new cervical and dorsal MRI showed an extensive hypointense lesion on T1, and hyperintense on T2, with peripheral contrast enhancement involving the central and posterior portions of the spinal cord, with an expansive aspect from C5 to D1, surrounded by edema from C2 to D3. A new CSF study demonstrated positive OCB, positive aquaporin-4 antibody in CSF and serum, and non-reactive anti-MOG. A new pulsetherapy and azathioprine were initiated after the denial of rituximab by her healthcare plan, with a good response to treatment. **Discussion:** Neuromyelitis optica (NMO) is a demyelinating disease with severe and more disabling outbreaks than multiple sclerosis (MS). In this reported case, the patient had neuroimaging studies suggestive of MS, with positive OCB in the CSF and negative anti-aquaporin-4 antibody in the serum. After 16 years without any recurrent episodes, or significant gain of lesion load, she presented with a new severe bout with posterior cord ataxia and crural paraparesia, with evidence of extensive myelitis on cervical-dorsal MRI. **Final Comments:** As a highly sensitive test, aquaporin-4 antibody has gained prominence to differentiate MS from NMO, increasing the diagnostic accuracy of NMO to 90% of cases, when associated with clinical criteria. Therefore, in diagnostic doubt aquaporin-4 antibody must be tested more than once. The differential diagnosis between these two entities is essential and requires specific treatments.

Neuroimunologia

TL 1105104

STERIOD-RESPONSIVE PONTINE PERIVASCULAR ENHANCEMENT DUE TO CHRONIC LYMPHOCYTIC INFLAMMATION (CLIPPERS) IN A YOUNG PATIENT AS A DIFFERENTIAL DIAGNOSIS WITH INFLAMMATORY CENTRAL NERVOUS SYSTEM DISEASES: A UNCOMMON REPORT CASE

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Case Presentation: M.L.N.P, female, 22 y, admitted to a neurology ER reporting a holocranial headache associated with 3 weeks of intermittent rotational vertigo, diplopia and strabismus. On physical exam observed sixth cranial nerve ophthalmoplegia leading to right eye convergent strabismus and horizontal nystagmus, rapid phase to the left side, ataxia in left hemibody with dysmetria and ataxic gait. Hypothesis of CNS inflammatory disease was evented and brain MRI with positive signal anomaly in the right nucleocapsular region, extending to midbrain, pons, middle cerebellar peduncles and left cerebellar hemisphere, with punctate enhancement was identified, suggesting CLIPPERS diagnosis. Other labs and supplementary tests were normal results. Performed intravenous therapy with methylprednisolone 1g/d for 5 days and improvement mainly strabismus was identified. Prednisone 20mg daily was prescribed, in accordance with literature findings just until Rituximab as a Corticosteroid-Sparing Therapy (CST) starts. Outpatient care 30 days later, demonstrated no neurological deficits, with remission of previous symptoms and new MRI identified no signs perivascular enhancement. CST was introduced 4 months later, without adverse events or new episodes of inflammatory outbreaks at last 6 month. **Discussion:** CLIPPERS has become a crucial differential diagnosis of CSN immune-mediated diseases. Its pathophysiology, characterized by autoreactive lymphocytes-mediated perivascular inflammation, remains unclear. In Brazil, only few cases of female patients with this syndrome has been reported since it classically affects men, with a wide range of age distribution. This clinical case is relevant to brazilian neurological may the use CST, so far, has been scarcely described. **Final Comments:** Diagnosis of CLIPPERS was established using clinical and complementary criteria, described by Pittock et al, and treatment is successfully correlated by symptoms remission. This case might contributes to the knowledge that CLIPPERS is a differential diagnosis of immune-mediated CNS diseases, understanding follow-up and therapy used with positive results. Cases estimated in the world is uncertain and a literature review founded about 40 cases, predominant in older men, where reports on female patients is uncommon.

Neuroimunologia

TL 1105109

ATYPICAL PRESENTATION OF ACUTE DISSEMINATED ENCEPHALOMYELITIS (ADEM) IN ADULT AFTER SARS-COV-2 INFECTION

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Case Presentation: A 31-years-old man, gardener, previously obese and hypertensive, presented with binocular diplopia associated with SARS-Cov-2 infection. After two weeks, he presented gait abnormalities and nausea with progressive worsening of symptoms, and onset of peripheral facial paralysis. The patient was admitted to Emergency Department for investigation, and the neurological examination showed right dysmetria, cerebellar gait, horizontal nystagmus with fast phase to the right, and right peripheral facial palsy. CT in admission was normal. Extensive laboratory tests were normal. CSF has been showed protein 83mg/dL and 2 cells. MRI showed in FLAIR and T2 a periaqueductal hypersignal at mid-brain-bridge transition, in the posterior region of the midbrain and cerebellar vermis. On T1 contrast, there was gadolinium enhancement. Neoplastic screening revealed no signs of tumor. Then the patient was treated with intravenous methylprednisolone 1000mg/d each for 5 consecutive days. After 5 days, the neurological examination showed gait and cerebellar tests normal, with persistence of mild facial palsy. New brain MRI showed significant improvement in aspect of the lesion, with mild hypersignal in cerebellar vermis. **Discussion:** ADEM is a rare demyelinating disease more commonly seen in children, presenting clinical heterogenic, generally causing encephalopathy and multifocal deficits. Several studies reported an increased incidence of ADEM after SARS-CoV-2 epidemics. In the typical presentation of ADEM, neurological symptoms develop 7–14 days following an infection and may involve headache, emesis, behavior alterations and coma. The case presented showed dysmetria, cerebellar gait, nausea and facial palsy. ADEM CSF examination reveals inflammatory findings, consisting of elevated protein levels and lymphocytic pleocytosis. Isolate increased protein level was present in up 40% of ADEM linked to SARS-Cov-2 in some case series. A few atypical presentations can be seen in some cases, and the differential diagnosis with multiple sclerosis (MS) remains challenging due to the overlap in the clinical presentation. In ADEM, the lesions can be seen in the brainstem and cerebellum, resulting in symptoms that are like those found in the present case. **Final Comments:** The varied clinical manifestations caused by SARS-Cov-2 infection in the nervous system have been constantly updated. In this context, coronavirus infection can be seen as a trigger to ADEM.

Neuroimunologia

TL 1105131

STERIOD-RESPONSIVE ENCEPHALOPATHY ASSOCIATED WITH AUTOIMMUNE THYROIDITIS (SREAT): A SEVERE CASE IN A YOUNG WOMAN

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Case Presentation: A 39-years-old woman, previously healthy, presented in the Emergency department with tonic-clonic seizures, followed by refractory status epilepticus requiring induction of barbituric coma to achieve seizures control using standard antiseizure drugs in high doses. Initial blood test and serologies were normal. Thyroid function tests showed TSH: 7,98, free T4: 0,81, Anti-thyroglobulin: 37,4 and Anti-thyroid-peroxidase: 97,6. Cerebrospinal fluid cell count, protein and glucose were normal. CSF VDRL and Criptococcal antigen were negative. Cultures for bacteria, fungi and BAAR were negative. Brain magnetic resonance imaging revealed in FLAIR/T2 sequences increased signal in hippocampus bilaterally, extending to hypothalamus, with swelling features, exhibiting cortical diffusion restriction. The patient received human immunoglobulin, remaining with consciousness impairment, orofacial and lower limbs mioelonus at the end of immunotherapy. Infusion therapy with methylprednisolone were indicated. After pulse therapy, the patient showed improvement of consciousness level and the mioelonus ceased. However, within ten days, the seizures recurred, associated with drowsiness. Prednisone 40mg daily associated with anti-seizures drugs gradually improved the clinical picture. She was discharged within six days, lucid, with mild-cognitive impairment and normal thyroid function tests. She is currently in physical rehabilitation, with regular follow ups. **Discussion:** Previously named Hashimoto's encephalopathy, Steroid Responsive Encephalopathy associated with autoimmune thyroiditis is a rare disease, associated with antithyroid antibodies. Women tend to be more affected than men and the mean age of onset is 51 years. It may comprise neuropsychiatric signs, such as progressive cognitive impairment, disturbance of consciousness, seizures, psychosis, behavioral changes, hallucinations, myoclonus, involuntary movements, language barrier and ataxia. This wide range of clinical findings leads to a underrecognition of the syndrome. The pathophysiology, as well as the relationship between Hashimoto thyroiditis remains unclear. SREAT has a pronounced response to corticotherapy, the first-line treatment. **Final Comments:** The relevance of this case presentation lies on the fact that SREAT is a rare and severe, but treatable syndrome. In order to avoid and ease complications that could jeopardize the patient's recovery, the early diagnosis and treatment are decisive.

Neuroimunologia

TL 1105169

EXTENSIVE SPINAL WALLERIAN DEGENERATION IN A PARANEOPLASTIC GANGLIONOPATHY CASE

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Case Presentation: A 52-year-old woman presented a three-month history of weight loss and incoordination that turned her bedridden. She was a longstanding smoker without comorbidities. Neurological evaluation unveiled severe appendicular and axial sensitive ataxia, uncomfortable paresthesia, despite preserved strength. Previous electroneuromyography endorsed the hypothesis of ganglionopathy. Etiologic investigation disclosed a small cell lung cancer (SCLC) and the final diagnosis was a paraneoplastic syndrome. Intravenous pulse steroid therapy was prescribed without symptomatic change, and the patient was referred to Oncology. After 2 months of chemotherapy, surveillance exams revealed lung response though the appearance of dorsal spine hyperintensity, not seen in the first MRI. Neurological examination was unchanged. Patient was submitted to radiotherapy and a 3rd MRI still showed the hyperintensity. Nine months after the treatment beginning, the oncologic disease remains under control without neurological gain. **Discussion:** Paraneoplastic ganglionopathy is most associated to SCLC indeed. The main antibodies involved are: anti-Hu, anti-CRMP5 and anti-amphiphysin. MRI dorsal column hyperintensities have been reported in ganglionopathy, a differential diagnosis for tabes dorsalis, vitamin B12 and copper deficiency and nitrous oxide toxicity. In ganglionopathy, it has been reported a precocious emergence of this spine hypersignal, besides a radiological improvement after immunomodulatory therapy. However, in most cases, MRI finding represents irreversible damage due to gliotic injury. This latter scenario reinforces the physiology basis of Wallerian degeneration (WD) started from dorsal root ganglia (DRG), pseudobipolar neurons that ramify into the peripheral branch and the central branch which carries the somatosensory information to the spinal cord. Although the underlying cause has been addressed, the lesion of the DRG, namely the proximal cell body, triggered a cascade to distal axonal degeneration. Therefore, first MRI was unremarkable but the following exams testified a permanent finding. **Final Comments:** Our case highlights WD in central afferent tracts, secondary to disrupted synapse at the dorsal-horn. The persistence of the novel hyperintensity and the absent recovery from sensitive ataxia supports MRI as a tool for chronicity ascertainment. Moreover, MRI was able to illustrate the pathogenesis behind the natural history of ganglionopathy.

Neuroimunologia

TL 1105183

SEVERE GUILLAIN-BARRÉ SYNDROME DURING RECENT DENGUE OUTBREAK IN THE SOUTH OF BRAZIL

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Case Presentation: Fourteen days before admission, a 62-year-old woman was diagnosed with dengue virus infection. She had typical clinical features – high fever, retro-orbital pain, headache, muscle pain and cutaneous rash – and positivity to NS1 antigen. She recovered well and remained asymptomatic for 5 days. However, 2 days before Hospital admission, the patient presented with progressive paraparesis and lower limbs paresthesia. At the first neurological exam at the emergency room, she had distal lower limb weakness (MRC = 4) and walk impairment. In less than 24 hours, she rapidly evolved to global paresis (MRC = 1-2) associated with diminished upper limbs reflexes, lower limbs areflexia, dysphagia and urinary retention. After ICU admission, she required orotracheal intubation and mechanical ventilation due to ventilation distress. Sudden changes in heart rate and blood pressure suggested autonomic dysfunction. Cerebrospinal fluid analysis revealed protein elevation (111 mg/dl) and normal cell counts. Due to the probable Guillain-Barre Syndrome (GBS) diagnosis, the patient underwent 7 plasmapheresis sessions on alternate days. During ICU period, she had other complications such as neuropathic pain, ventilation associated pneumonia and urinary tract infection. She received intense physical and ventilatory rehabilitation. The first sign of clinical improvement was the resolution of dysautonomic symptoms. On the 12th day, she started to recover spontaneous movements from axial muscles. Tracheostomy was removed on day 24. After 46 days, she was discharged with partial recovery, being able to stand and walk with assistance, and partial diuresis control. She was referred to outpatient rehabilitation and neurological follow-up. **Discussion:** Dengue virus infection was very uncommon in the Rio Grande do Sul state, but recent outbreaks and spread of *Aedes aegypti* vector changed the disease epidemiology. Fast evolving neurological manifestations of dengue are rare and may be overlooked by general practitioners. case presentation of GBS after dengue virus infection are scarce. Management of severe GBS requires quick diagnosis with rapid treatment, ICU support and intensive rehabilitation to reduce disease related morbidity and mortality. **Final Comments:** Dengue virus infection should be considered in the diagnostic workup of triggers for GBS, including in regions with previous low prevalence. Plasmapheresis is an alternative treatment when human immunoglobulin is not available.

Neuroimunologia

TL 1105216

HASHIMOTO'S ENCEPHALOPATHY: CASE PRESENTATION AND LITERATURE REVIEW OF A RARE ENTITY

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Case Presentation: A 62-year-old man presenting altered mental status and a history of four seizure episodes in the past 2 months, along with aggravated neuropsychological symptoms, such as anxiety, delirium, and depression. Previous history of steroid-responsive encephalopathy, suspected Hashimoto's encephalopathy (HE). He was treated with prednisone 3 months prior current admission, with a gradual withdrawal. Magnetic resonance imaging (MRI) report was unremarkable, serology tests positive for Anti-TPO antibodies, EEG not available. HE suspicion remained. Pulse corticosteroid therapy was administered, previous treatment reinstated. **Discussion:** HE is a rare and controversial disease encompassing a heterogeneous neurological and psychological presentation. Diagnosis of HE is a clinical challenge since there isn't a well-defined criteria and the pathophysiological mechanisms are still unclear. Literature review presents neither a gold-standard test for diagnosis nor a reference value for serology exams, although elevated anti-TPO seems to be the hallmark of HE. Other case presentation presented highly heterogeneous patient profiles with long-ranging age groups, mostly female, and diverse clinical presentations, varying from acute to insidious onset. Studies showed the relapse and remitting course in 50% of the patients. Most common treatment is still corticosteroid administration, but intravenous immunoglobulin, methotrexate, mycophenolate, and azathioprine have been used with variable efficacies. **Final Comments:** Thus, it is important to raise awareness of HE since it's a neglected differential diagnosis for various neuropsychological symptoms and early intervention can define prognosis. Furthermore, the relapse after corticosteroids treatment emphasizes the need of therapeutic alternatives, which demands a better understanding of its pathophysiology.

Neuroimunologia

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MYELITIS AS THE FIRST NEUROPSYCHIATRIC MANIFESTATION IN A SYSTEMIC LUPUS ERYTHEMATOSUS PATIENT – RARE CASE PRESENTATION

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Case Presentation: M.C.J.S., 30 years old, female, diagnosed with Systemic Lupus Erythematosus (SLE) at the age of 20, presenting, over 10 years, cutaneous, joint, hematological and renal activity. During this period, she received treatment with hydroxychloroquine and prednisone. In June 2021, she started a prodromal condition flu-like associated with paresthesias, distal weakness in the lower limbs and urinary incontinence. After 48 hours, she evolved with worsening of the legs weakness, being unable to stand and walk without assistance, and in less than 24 hours progressed to flaccid tetraparesis. Analysis of the cerebrospinal fluid showed pleocytosis with lymphocytic predominance, hyperproteinorrachia and normal levels of glucose and lactate faced with the suspicion of autoimmune inflammatory myelopathy. In addition, we noticed a high value of anti-double-stranded DNA antibody 400 U/ml, indicating high disease activity. Magnetic resonance evidenced hypersignal lesions, with a diffuse and irregular distribution, longitudinally from the medulla oblongata transition to the level of the conus medullaris, with a slight swelling effect and without contrast enhancement. She was treated with high-dose intravenous methylprednisolone with little initial improvement, showing a better therapeutic response after plasmapheresis. During follow-up, 6 months later, she was able to walk unassisted, maintained temporary treatment with cyclophosphamide, but due to the potential nephrotoxic damage, she started using rituximab with excellent results. As additional data, anti-aquaporin-4 antibody (AQP4) was measured with a negative result. **Discussion:** the occurrence of myelitis in SLE is estimated at 1.2%. In cases where signs of gray matter predominate, high disease activity was observed by high levels of anti-double-stranded DNA antibodies, while when white matter disorders predominate there is an association with anti-AQP4 positivity, indicating, in fact, a great possibility of overlap with neuromyelitis optica spectrum disorder (NMOSD). **Final Comments:** although uncommon, promptly diagnosing and treating SLE myelitis can prevent severe neurological damage. Also, it is important to pay attention to the differential diagnosis with other autoimmune demyelinating diseases such as NMOSD, which can coexist with SLE and be responsible for the involvement of the spinal cord and not the activity of the underlying systemic disease.

Neuroimunologia

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OVERLAP OF MILLER FISHER SYNDROME (MFS) WITH BICKERSTAFF SYNDROME (BS)

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Case Presentation: A 15-year-old male patient presented with subacute weakness of the lower limbs associated with incoordination within 1 week, with a 48-hour progression to impaired gait, axial instability and diplopia. He denied fever and diarrhea but reported having a flu-like syndrome three weeks before the current condition. On examination: hypotonia and exalted osteotendinous reflexes with bilateral Babinski, and bilateral ophthalmoparesis was noted. On the day after admission, the patient developed ebrious dysarthria, dysphagia, ineffective cough and decreased level of consciousness, and orotracheal intubation was chosen. After 1 week of sedation withdrawal, he presented some improvement in the sensorium, proximal grade 0 and distal limbs grade IV strength, sustained bilateral ophthalmoparesis, facial diparesis, absent cough reflex, associated with dysautonomia with pressure variability and of heart rate. Tomography (CT) and Magnetic Resonance of the skull performed: no alterations; cerebrospinal fluid (CSF) with protein of 23mg/dL, glucose: 118mg/dL; leukocytes: 3, Oligoclonal bands absent. Electroencephalogram without changes. Laboratory tests/cultures/serologies without alterations. Chest CT: normal. Based on the above, the suspicion of SMF superimposed with SB was raised. He received treatment with plasmapheresis in 4 sessions on alternate days, being interrupted after motor worsening in the left side and evidence of intraparenchymal hematoma with hemoventricle and midline deviation on a new cranial CT, evolving with complications and death. **Discussion:** MFS is a variant of Guillain-Barre syndrome characterized by a subacute, monophasic onset and the classic triad of ophthalmoplegia, ataxia and areflexia. If there is progressive impairment of the level of consciousness and/or associated hyperreflexia, we recognize brainstem encephalitis, or BS. Sharing similarities in the autoimmune mechanism, both syndromes are associated with anti-GQ1b antibodies, which are highly specific for diagnosis. One of the main limitations of this case was the unavailability of dosing this antibody. Both intravenous immunoglobulin and plasmapheresis have been used in the treatment. **Final Comments:** The overlap of SMF with SB is rare. It is important to remember the need for attention in clinical practice for possible diagnosis because the prognosis is generally good and most patients recover well when the syndrome is quickly recognized and treated.

Neuroimunologia

TL 1105274

PAROXYSMAL DYSTONIA AS ATYPICAL INITIAL MANIFESTATION OF MULTIPLE SCLEROSIS

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Case Presentation: A 14-year-old male with autism spectrum disorder started with gait disturbance and vertigo, evolving with tingling in his hands and feet, in addition to several daily episodes of sustained and painful flexion of the left upper limb, lasting 1-2 minutes. On examination, he had dystonic posture in the left hand and forearm, hyperactive patellar reflex, and bilateral clonus. No changes in cranial nerves, strength, sensitivity and coordination. Therapy with carbamazepine showed progressive improvement of dystonic spasms. Investigation demonstrated CSF with oligoclonal type 2 bands and cranial resonance with T2/FLAIR hypersignal in the juxtacortical, periventricular, callosal and bulbar paramedian interface and gadolinium enhancement in the corpus callosum. He was diagnosed with Multiple Sclerosis (MS) and treated with steroid pulse therapy followed by plasma exchange. At discharge, the patient was asymptomatic, with complete improvement of dystonia. **Discussion:** Paroxysmal symptoms in MS are a heterogeneous group of motor, sensory, cerebellar, or brainstem changes, characterized by repetitive, stereotyped and short episodes that can last from days to months. The incidence is 1.6 to 17% in MS cases, of which 24% are the initial manifestations. The most important are dyskinesia, myokymia, hermitte sign, trigeminal neuralgia, vertigo, diplopia and ataxia. Paroxysmal dystonia is an involuntary, abrupt and also stereotyped movement. It may be painful or not, kinesigenic or not, focal or generalized. Although not completely understood, it is speculated that this phenomenon may occur due to erratic transmission of ectopic nerve impulses by partially demyelinated axons. These may present a dysfunction in ion channels, which generates neural desensitization to stress factors. This is also strengthened by its good therapeutic response to anticonvulsant drugs. Furthermore, its location occurs due to lesions in the motor pathway, such as in the posterior horn of the internal capsule, basal ganglia, brainstem and cervical spinal cord. We report a case of a patient who presented with a first onset of MS associated with dystonic spasm, with dramatic improvement of dystonia after the introduction of carbamazepine. **Final Comments:** Paroxysmal symptoms are uncommon in MS, posing a diagnostic challenge. Recognition as a possible manifestation of MS is of fundamental importance, as it can avoid delay in diagnosis and provide adequate early treatment.

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TL 1105324

VOGT-KOYANAGI-HARADA SYNDROME AFTER SARS-COV 2 VACCINE: CASE PRESENTATION

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Case Presentation: A 29-year-old female patient, who was vaccinated against SARS-COV-2 (Pfizer), sought a neurological emergency service 24 days after vaccination due to a severe, progressive headache, mainly in the right temporal region, with subsequent bilateral involvement associated with phonophobia, photophobia, nausea and dizziness. She also reported visual scotomas, tearing of the eyes and intense bilateral conjunctival hyperemia, with the development of blurred vision and diplopia. She was Hospitalized and in the initial neurological evaluation showed a reduction in bilateral visual acuity, not accompanied by changes in extrinsic ocular muscles or direct pupillary changes. Other focal deficits were also not detected. She underwent imaging studies of the skull and initial orbits, which showed no changes. She also underwent lumbar puncture, which showed lymphomonocytic pleocytosis, with manometry within the normal range. An ophthalmologic evaluation was performed, which showed bilateral conjunctival hyperemia with ciliary injection and posterior synechiae of almost 360 degrees. The funduscopy examination revealed cloudy media, pink papilla with clear edges, sensorineural retinal detachment in the macular region, presence of nodules in the middle periphery. papillitis, with Vogt-Koyanagi-Harada syndrome as the main suspect. **Discussion:** Vogt-Koyanagi-Harada syndrome (VKH) is an inflammatory and autoimmune condition characterized by panuveitis, serous retinal detachment and extraocular manifestations (auditory, integumentary and central nervous system). Its autoimmunity mechanism may be associated with viral infections as well as vaccines. In the current scenario of mass vaccination due to the SARS-CoV2 pandemic, this association has also been evidenced, but with mechanisms still poorly described. The diagnosis of VKH is essentially clinical and can be confirmed with imaging tests. Treatment consists of the use of immunosuppressants, preferably started early to avoid the emergence of complications, mainly neuro-ophthalmological. **Final Comments:** In the context of a pandemic and in the course of a global mass vaccination campaign, it is important to recognize possible complications associated with vaccination against COVID-19. However, to date, these risks are low and the benefits of vaccination outweigh any side effects.

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CASE PRESENTATION ENCEPHALOMYELORRADICULITIS OF UNKNOWN ETIOLOGY IN A YOUNG PATIENT

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Case Presentation: 49 year old male patient with controlled hypertension was Hospitalized with a first-time seizure. Three months before Hospitalization he presented a persistent low back pain which progressed to an intermittent left hemiparesis. Those symptoms were previously assigned to a C4-C5 disc herniation. Magnetic resonance imaging showed an irregular lesion in the right temporal lobe causing mass effect in addition to an extensive cervicothoracic myelopathy with contrast enhancement and a diffuse leptomeningitis affecting even cauda equina roots. The cerebrospinal fluid (CSF) sample had 50 cells/microL, with a lymphocytic predominance, 663 mg/dL proteins and glucose consumption. Oncocytic cells and the antineuronal antibody panel were negative. Adenosine deaminase (ADA) at CSF was 1 U/L, not suggestive of tuberculous meningitis. Oligoclonal bands were present in both CSF and serum suggestive of a systemic inflammatory process. The CSF immunohistochemistry and rheumatologic/infectious workup were negative. Angiotensin-converting enzyme (ACE), anti-myelin oligodendrocyte glycoprotein (MOG) and anti-aquaporin-4 (AQP4) were non-reactive. Chest and abdomen tomographies showed no neoplasms. Biopsy of the temporal lesion resulted in nonspecific meningoencephalitis, with no signs of malignancy. The CSF metagenomic sample is still without a definitive result. Patient was referred for an external ventricular drain implantation for intracranial pressure monitoring. In addition, empirical plasma exchange was started. However, it was not possible to observe the patient's response since he progressed with a septic shock due to a *Staphylococcus haemolyticus* bloodstream infection. By the sixth day after admission brain death was certified. **Discussion:** This case presented itself as a challenging clinical case in a previously functional young patient with multiple active central nervous system lesions. Considering the main hypothesis, inflammatory disease and neoplasm, several procedures were performed, especially MOG, anti-neuronal antibody panel, CSF immunohistochemistry and metagenomic sample. Some of these exams are still not easily available in medical practice and may be extremely important for some etiological diagnosis. **Final Comments:** Neurological cases without etiological definition occur occasionally in medical practice. Therefore, it is important for neurologists to share experiences for the establishment of new approaches in daily practice.

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POST-COVID19 ACUTE TRANSVERSE MYELITIS TREATED WITH PLASMA EXCHANGE AND RITUXIMAB: A CASE PRESENTATION

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Case Presentation: 51-year-old woman with severe obesity and no other medical history, admitted with dyspnea and anosmia for 5 days. SARS-COV-2 pneumonia was confirmed by RT-PCR. The patient developed respiratory distress within 24 hours and required mechanical ventilation for 4 days, without hemodynamic instability. After pulmonary recovery the patient noticed difficulty in standing, received discharge in a wheelchair. At home, progressively worsens motor function, not being able to support her trunk associated with bladder dysfunction and lower limb paresthesia. Readmitted 15 days after, the first neurological examination described severe muscle weakness of lower limbs, feet drop and mild involvement of intrinsic hands muscles; global deep tendon absence and babinski sign bilaterally; large and small fiber anesthesia below T12 level. Spine MRI concluded transverse myelitis from C7 to T12; CSF's routine was normal and RT-PCR for SARS-COV-2 and Herpesviruses went negative. Investigation for neuroimmune and rheumatologic markers, serological status, onconeural antibodies, hypovitaminosis and metabolic causes were negative. Methylprednisolone 1g for seven days was administered with poor motor outcome. Afterwards, seven plasma exchange sessions followed by Rituximab 2g total dose led to substantial improvement. At deinstitutionalization the patient regained bladder and trunk control with poor lower limb mobility. At three-month follow-up the patient could sustain her legs; at sixth month she could walk with crutches. Nine months after discharge she could walk without assistance. **Discussion:** Acute Transverse Myelitis (ATM) is a rare and acquired neuro-immune disorder characterized by perivascular lymphomononuclear infiltration of spinal cord affecting white and gray matter, explaining sensitive, motor and autonomic dysfunctions. Besides it's difficult to assuredly exclude differential diagnosis, post-viral infection immune-mediated TM is a main etiology, classically associated with the human Herpesviruses. Nevertheless, as described in other reports, immune-mediated TM is an emerging complication of SARS-COV-2 infection, which by analogy, has been treated how other viruses justifying an immunosuppressive therapy. **Final Comments:** Albeit evidence is needed to guide decision through post-COVID19 TM, this case corroborates for an combined aggressive immune-therapy after a suitable investigation for severe presentations with high rate of disability and complications.

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THE IMPORTANCE OF DYNAMIC MAGNETIC RESONANCE IMAGING IN THE DIAGNOSIS OF CERVICAL MYELOPATHIES: A CASE PRESENTATION

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Case Presentation: A 62 year old man was admitted to our service due to weakness in lower limbs and unsteadiness. Symptoms began 3 months before, with pain and numbness in his hands, evolving to bilateral and symmetrical weakness in lower limbs. The weakness became worse 4 days before admission, after receiving the 2nd dose of Covid-19 vaccine. There was no history of trauma, constitutional nor sphincters complaints. On neurologic examination, he showed a global MRC strength grade 5, except for 4+ on hip flexion and 4+ on hip abduction. There were signs of bilateral pyramidal release in lower limbs. He had a reduced proprioception and doubtful touch and pain hypoesthesia at T12 and T4 level, respectively, and he was unable to stand without assistance. He had sensory ataxia in all limbs, worse in his legs. Results of laboratory exams and CSF were unremarkable. The cervical spine MRI showed an nonspecific T2 signal heterogeneity at C4-C5. Based on the findings, the hypothesis of a post vaccinal inflammatory myelopathy was made and the patient went under pulse therapy with 1g methylprednisolone for 5 days, without objective improvement. Because of the atypical presentation of an inflammatory myelitis and for the differential diagnosis of compressive spondylotic myelopathy (CSM), a dynamic cervical spine MRI was requested. The exam revealed a canal stenosis at C4-C5. The patient underwent laminectomy, spinal cord decompression and posterolateral cervical arthrodesis from C3 to C6. The procedure was uneventful and he evolved with progressive improvement of symptoms. **Discussion:** Cervical spondylotic myelopathy is the most common spinal cord disorder in the elderly. Diagnosis is based on clinical signs and symptoms associated with a radiological exam. The spinal cord is made under dynamic changes during flexion and extension. These changes lead to biomechanical stress with movement, which may aggravate damage in patients with critical cervical stenosis. Normal MRI is unable to detect those changes. Therefore, the dynamic MRI has a higher sensibility for the detection of CSM. **Final Comments:** This case shows a patient with a typical presentation of subacute cervical of CSM, but no significant changes in MRI. The diagnosis was only confirmed after a dynamic cervical spine MRI. Hence, dynamic MRI is an additional image resource able to provide useful information when there is a high clinical suspicion for CSM and the standard radiological exam is inconclusive.

Neuroimunologia

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GUILLAIN BARRÉ SEVERE SYNDROME (GBS) PROBABLY ASSOCIATED WITH VACCINATION FOR COVID-19

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Case Presentation: Female patient, 60 years old, hypertensive, presented insidious lower limb (LL) weakness associated with tactile-painful hypoesthesia. In five days, there was a progression of weakness with impairment of walking, which motivated the visit to the Hospital. She denied fever, diarrhea, or respiratory infection symptoms preceding her current condition, but a vaccine reference for COVID-19 (Astrazeneca) fifteen days earlier. On examination: hypotonia in the LL, reflex absence globally, grade IV strength in the upper limbs (UL), grade III proximal, grade II distal in the LL, and peripheral facial diparesis. Performed cranial CT with no alterations and cerebrospinal fluid (CSF) with protein of 141mg / dL, glucose: 80mg / dL; WBC: 0. Screening for nasopharyngeal SARS-CoV-2 using rapid SWAB test, negative. Based on the above, the suspicion of SGB was raised. Intravenous immunoglobulin infusion (IVIG) was started, followed by clinical worsening of the degree of strength (LL and UL grade 0) and presence of dysautonomia (sustained tachycardia and arterial hypertension). After the end of IVIG, the patient established dysphagia, dysphonia, and dyspnea. Due to the clinical deterioration, we opted for a new IVIG cycle, with an additional 1g / Kg being a total dose, divided into three days. There was an improvement in swallowing, dyspnea, upper limb strength (proximal grade II, distal grade III), and reversal of dysautonomia. **Discussion:** GBS is an acute inflammatory disease of the peripheral nerves that commonly presents with progressive symmetrical weakness and areflexia, resulting in respiratory failure. Annual report of 1 to 2 cases / 100,000 inhabitants and peak between 20-40 years, being somewhat more common in men. It can result from an immune response that occurs 3-6 weeks after a previous acute illness, infection, or trigger in 50% of cases. Still, it can also be associated with immunization, surgery, and trauma, for example. A relationship between COVID-19 infection and many neuropathies, including GBS, has been reported. However, we still do not have full knowledge of GBS related to immunization against COVID-19. **Final Comments:** It is essential to remember the need to pay attention in clinical practice to GBS causes in a scenario of such a recent and devastating infection as COVID-19 and its newly discovered vaccines. Extensive studies are needed.

Neuroimunologia

TL 1105399

ANTI-LEUCINE-RICH GLIOMA-INACTIVATED 1 ENCEPHALITIS: A CASE SERIE

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Case Presentation: Three elderly patients, mean age of 67 years old, two women and one man, presented to our Hospital clinic due to subacute faciobrachial dystonic seizures (FBDS) as main symptoms. FBDS were bilateral and high frequency (mean sixty episodes per day). One patient also had dystonic seizures in legs. Mild chorea was noticed in one woman concurrently. Executive dysfunction and short-term memory loss were noted in all of them, leading to rapidly progressive dementia phenotype in two. The man also showed visual hallucinations, behavior changes, depressed humor and hyponatremia (120 mmol/L), which prohibited the initial management of FBDS with carbamazepine (CBZ). Electroencephalogram showed disorganized background activity with bifrontal slowing in both women. The Magnetic resonance images evinced white matter hyperintensities presumed of vascular origin in totality, besides hippocampal volume reduction in the woman with worse cognitive status. Search to occult neoplasm was negative to all. There were no cerebrospinal fluid abnormalities besides anti-LGI1 antibody positive, excepting the last patient whose testing is still in progress. Methylprednisolone was the first line therapy to all, added to plasmapheresis (PLEX) in two. CBZ and levetiracetam associated with immunotherapy were essential to achieve FBDS full control in all patients in the mean period of 1 month. Micofenolato and prednisone were the choices for long term immunosuppression. Cognitive functions were improved in all cases. **Discussion:** Anti-LGI1 encephalitis mainly affects elderly (median age 60 years) who present with seizures, cognitive and behavioral disorders. FBDS are very specific for the disease otherwise presenting in a minority of cases. Usually there is no EEG correlation with FBDS. Moreover, about 60-65% of patients develop hyponatremia, which is a clue to the diagnosis. Treatment consists in immunotherapy with corticosteroids, IV immunoglobulins or plasmapheresis. **Final Comments:** This case series adds knowledge of Brazilian and local epidemiology of this rare cause of rapidly progressive dementia, especially since patients presented with the classical clinical features of the disease had a great response to treatment. This reinforces the importance to recognize the clinical syndrome allowing for promptly request antibody testing and initiate the immunotherapy even before antibody results are available.

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MOG ASSOCIATED DISEASE FOLLOWING HERPES 7 INFECTION: A CASE PRESENTATION

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Case Presentation: A 34-year-old woman, on the 4th postoperative day of video surgery due to ruptured ectopic pregnancy, was Hospitalized with a CSF suggestive of viral meningitis with a positive HHV-7 PCR and acyclovir was started. Evolved with a somnolence, associated with ampicillin for 21 days and a brain MRI was performed with a finding of diffuse hyperintensity on T2/flair in the brainstem without contrast enhancement, associated with lesions in the hippocampus and in the internal and posterior capsule/right lentiform nucleus, coexisting with diffuse leptomeningeal enhancement, with a fine and regular pattern. She developed urinary retention. Three days after discharge, she was diagnosed with an optic neuritis. A new CSF analysis revealed just a lymphocytic pleocytosis 53 cells/mm3 and no OCBs. She was readmitted with a classic clinic of an optic neuritis on the right (20/40). All types of blood tests were requested, with no changes. Therefore, she was treated with IV 5 grams methylprednisolone, and a new MRI revealed findings of involvement of the right optic nerve, in addition to signal hyperintensity on the side of the pons on cranial MRI associated with the finding on spinal MRI with anomalous gadolinium enhancement at the level of the conus medullaris, suspicious findings of an autoimmune nature due to MOGAD. Foscarnet was started for 14 days and on the Hospital discharge, she presented resolution of all symptoms. Positive result of serum anti-mog antibodies. Primary infection with HHV-7 can occur in adults which can lead to different manifestations such as encephalomyelitis, however studies show that cases of secondary optic neuritis are uncommon. Onset of demyelinating diseases, such as multiple sclerosis, due to EBV infection have already been scientifically proven and the occurrence of MOGAD spectrum diseases have also been described after reports of infectious mononucleosis by EBV and VZV infection. Case presentation showing an association between MOGAD and HHV-7 infection are rare. However, due to molecular similarity between virus from the herpes family to EBV, it can be suggested that the herpes virus family may be triggering agents of autoimmunity. Therefore, in the rare clinical presentation of MOGAD spectrum disease possibly triggered by an infection caused by HHV-7, the description of this case is very important for this etiology to be considered in the differential diagnosis of an infectious condition that evolves with demyelination.

Neuroimunologia

TL 1105420

DIAGNÓSTICO DE NEUROMIELITE ÓPTICA, EM PACIENTE COM MIASTENIA GRAVIS

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Case Presentation: G.C.D.S. started at 29 years of age, floating weakness in arms and legs, prevailing in the proximal muscles, associated with intermittent diplopia and ptosis. Reported worsening of symptoms with stress and warm weather. Electroneuromyography with repetitive nervous stimulation at 3 Hz showed a 25.4% decrease in amplitude; she was then diagnosed with myasthenia gravis (MG), and started treatment with pyridostigmine, azathioprine and prednisone and thymectomy was performed. Evolved with good response to treatment with control of symptoms; 10 years later, during an interruption of azathioprine use, she noticed recurrent paresthesias in the arms and developed sudden visual acuity loss of the left eye; funduscopy with optic disk with low-defined margins and pale color suggestive of optic neuritis; Treatment with pulsotherapy with methylprednisone 1g for 5 days in Hospital was performed, with partial improvement of visual acuity; Brain MRI: smooth signal change and apparent enhancement by contrast of the left optic nerve. Cervical MRI: focus of signal change in the vertebral cord, most evident at the bulb and at the level of C2, C4 and C5, without significant enhancement by contrast; dosage of anti-aquaporin 4 IgG: reagent 1: 20; receiving a diagnosis of neuromyelitis optic (NMO); **Discussion:** MG and NMO are immune-mediated disorders with low prevalence in the general population. MG is a neuromuscular disorder associated with the presence of anti-acetylcholine receptor antibodies (AChR-Ab) and characterised by fluctuating muscle weakness. NMO is an inflammatory disease of the central nervous system that mainly affects the spinal cord and optic nerve, and is associated with presence of anti-aquaporin-4 antibodies (AQP4-AB) in 60%-80% of patients. Both AQP4-AB-positive NMO and AChR-AB-positive MG are associated with other autoimmune diseases and autoantibodies, both organ-specific and systemic. Despite the rarity of MG and NMO, several cases and small series of patients with both disorders have been reported over the years. Aqp4-NMO is recognized to be associated with other autoimmune manifestations in about 25%-50% of cases, both organ-specific and systemic. **Final Comments:** Clinicians should have a low threshold to screen for comorbid autoimmune disease in patients with MG and NMO, in which non-invasive testing for antibodies is specific and allows for early treatment and better outcomes.

Neuroimunologia

TL 1105430

MULTIPLE SCLEROSIS AFTER ANTI-NMDAR ENCEPHALITIS: CASE PRESENTATION AND REVIEW OF THE LITERATURE

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Case Presentation: A 19-years-old woman presented with seizures in June/2019. Brain MRI and EEG were unremarkable and she received levetiracetam. After 30 days she developed depression, aphasia, anosognosia, apathy, dyscognitive seizures, sleep disorder and recurrent vomiting. Physical examination disclosed temporal disorientation, mild ataxic gait, tremor and incoordination in left limbs. New EEG showed epileptiform discharges in right temporal lobe. CSF showed cells 11 cells/mm3, protein 23 mg/dL, glucose 68 mg/dL and positive BOC. Brain MRI showed T2/FLAIR hyperintense lesions in subcortical WM in left temporo-occipital lobe and left precuneus associated with SWI hypossinal areas that could represent hemosiderin deposition. Anti-NMDAR antibody was detected in serum and CSF. The patient received IVIG and MPIV with improvement. After 16 months she became impulsive, irritated, with difficulties in planning complex activities. Brain MRI showed multiple demyelinating lesions in brainstem, with leptomeningeal enhancement. CSF showed 6 cells, protein 26mg/dl, glucose 56 ml/dL, BOC +. Anti-MOG, anti-AQP4 and anti-NMDAR were negative. EEG showed rare epileptiform discharges in right frontoparietal lobe. According to 2017 MacDonald criteria she was diagnosed with Multiple Sclerosis (MS). HLA-DRB1*15: 01, HLA-DQA1*01: 02/06: 02 were negative. She was treated with MPIV, and received rituximab. In After 22 months she developed dizziness, nausea, mild upper limb ataxia, pallor of optic disc with bilateral visual acuities of 20/50. EDSS was 2,0. Brain MRI showed new infratentorial and supratentorial demyelinating lesions. Alemtuzumab was indicated. **Discussion:** We report a patient that developed MS after anti-NMDAR encephalitis, a rare association. An overlap between anti-NMDAR encephalitis and neuro-inflammatory disorders, mainly with NMOSD and MOGAD was described in a large series. However, association with MS is limited to only a few case-reports. Hypothesis for such association include: medication-induced autoimmunity, exposed epitopes during disease course, or EBV trigger. **Final Comments:** In conclusion, recurrence of neurological symptoms in anti-NMDAR encephalitis, the presence of unspecific brain MRI lesions at first attack and BOC + should trigger additional investigation and raise the possibility of MS. A question is whether the overlap between anti-NMDAR encephalitis and the demyelinating disorders occurs by chance or whether they may be mechanistically linked

Neuroimunologia

TL 1105442

COMBINED CENTRAL AND PERIPHERAL DEMYELINATION: A CASE PRESENTATION

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Case Presentation: Our patient is a forty-six year old woman, with a previous 9 year diagnosis of Multiple Sclerosis (MS) and using intramuscular interferon beta-1a, a disease-modifying therapy. She has been asymptomatic for 3 years, until she started to report numbness and weakness beginning at her feet and knees ascending to upper limbs symmetrically over the course of 48 hours. She denied a previous history of gastrointestinal or respiratory symptoms. At examination, she presented with grade 2 and 3 muscular strength on lower and upper limbs, respectively, associated with hypotonia and areflexia. We also observed a sensory level at T4 and Lhermitte sign. Cerebrospinal fluid showed albumin-cytologic dissociation and nerve conduction studies (NCS) presented the findings of acute sensory-motor demyelinating polyradiculoneuropathy, characteristics of Guillain-Barré Syndrome (GBS). Spinal cord Magnetic Resonance Imaging (MRI) presented a T2 high-intensity lesion from C1 to C5. We opted to treat the patient with intravenous Methylprednisolone for five days. She started to improve significantly and was walking again 5 weeks after the first symptoms initiated. **Discussion:** The co-occurrence of MS and GBS is rare, with less than 20 cases described around the world until now. Demyelinating diseases involving both central nervous system (CNS) and peripheral nervous system (PNS) are usually etiologically heterogeneous. Recently, autoantibodies against nodal and paranodal proteins were discovered (NF186, NF155, CNTN1 and CASPR1) and associated chronic and acute demyelinating diseases. Combined Central and Peripheral Demyelination (CCPD) is defined fulfilling of the following criteria: a presentation of T2 high-signal intensity lesions in the brain, optic nerves, or spinal cord upon MRI, or abnormalities on visual-evoked potentials (VEPs), and conduction delay, conduction block, temporal dispersion or F-wave abnormalities according to NCS. This condition is highly sensitive to intravenous corticosteroids, with an outcome of symptom recovery of 83.3%. Our patient presented both criteria to CNS and PNS involvement simultaneously and a significant response to treatment with intravenous Methylprednisolone. **Final Comments:** We hope to shed light on the rare association between CNS and PNS demyelinating diseases and encourage a better investigation of ascending acute sensitive symptoms in patients with a previous diagnosis of MS.

Neuroimunologia

TL 1105445

AUTOIMUNE ENCEPHALITIS AFTER 1ST DOSE OF PFIZER COVID-19 VACCINE IN A 16-YEAR-OLD PATIENT: CASE PRESENTATION

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Case Presentation: A 16-year-old male patient, from a countryside city in Amazonas, was referred to our service reporting sudden onset of paresthesia and paraparesis in the lower limbs, associated with chest tremors, dyspnea and anterograde amnesia one day after receiving his 1st dose of Pfizer COVID-19 vaccine. When admitted to the Hospital, he had an GCS of 14, spasms in the abdominal and thoracic muscles and plastic hypertonia in the left upper limb. After admission, he presented with generalized myoclonic seizures without loss of consciousness associated with constant high-pitched tinnitus bilaterally and his neurological examination showed grade 3 reflexes in right biceps and patellar, left brachioradialis and triceps bilaterally, in addition to hypoesthesia in the left hemibody and bilateral nystagmus. The patient was treated with phenytoin, which was later replaced by sodium valproate, associated with diazepam and empiric acyclovir. Given the information presented, the hypothesis of autoimmune encephalitis was raised. Due to logistical difficulties caused by the pandemic, the patient underwent only a cranial magnetic resonance imaging 18 days after admission, where no changes were evidenced. After an increase in the frequency of seizures, he was referred to the ICU for treatment with plasmapheresis. One month after his admission, he was discharged using levetiracetam and the case was followed up in our outpatient service. **Discussion:** Autoimmune encephalitis presents as a range of acute neurological symptoms, which can include headaches, altered mental status, seizures and autonomic and movement disorders, and can or can not be associated with a paraneoplastic syndrome. Its treatment is based on immunosuppressive therapies such as steroids, immunoglobulins or plasmapheresis, favoring the patient's prognosis in case of an early diagnosis. **Final Comments:** In this case, we bring a severe and rare presentation of post-vaccination encephalitis. It is essential to shed light on such adverse reactions, not to question the safety of the vaccine campaign, but to show that treating them effectively can enable complete patient rehabilitation, in addition to contributing to the formation of more accurate epidemiological evidence about neurological disorders related to COVID-19 vaccination.

Neuroimunologia

TL 1105524

GANGLIONOPATHY AND LONGITUDINALLY EXTENSIVE MYELOPATHY ASSOCIATED WITH SJÖGREN'S SYNDROME, IN A PATIENT WITH A BACKGROUND OF POST-VACCINATION POLYMYELITIS: A CASE PRESENTATION

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Case Presentation: In March 2020, arthralgias, perioral numbness, difficulty to speech, voice, swallowing, walking and forgetfulness, developed in a 44-years-old woman. November/2020, she had COVID-19, and trunk/limbs weakness, with abnormal hand movements appeared. In childhood, she presented post-vaccination poliomyelitis (atrophy in the right leg). June/2021, Neurological exam: Alertness, tachycardia, tachypnea, and hypotension. Performed simple commands, reduced fluency. Hypomimic face, dysphonia, dysarthria. Equine right foot and fasciculations in the right leg. Global hypotonia. Force graded MRC 2. Reflexes abolished. Indifferent plantar responses. Apalesthesia from C3. Normal superficial sensibility. Choreic and dystonic movements in face and upper limbs. Brain MRI-hypersignal in deep white matter, pons and gliosis in the right temporal pole. Cervical MRI-hypersignal region C3-T1 (posterior). Electroneuromyography- lower motoneuron pathology with a sensory ganglionopathy in four limbs. Polysomnography revealed significant oxygen desaturation. FAN 1: 320 and anti-Ro >240. Schirmer's test was positive to xerophthalmia. Anti-aquaporin-4 negative. Initially, methylprednisolone (1g/5days) was performed, without improvement, followed by immunoglobulin (0.4mg/Kg/5days), without major motor gains. Subsequently, she underwent cyclophosphamide (750mg/m2/day-3months), with interaction, swallowing and abnormal movements improved, but continued with force graded MRC 2. Then, Rituximab was realized (1000mg-15/15days), with recovery of cervical and trunk support. She is in homecare, with speech-physical therapies, using nighttime beeper, and waiting for Rituximab (2nd cycle). Ten months Hospitalized, there were complications, requiring ICU, due to dysautonomia, acquired tuberculosis, respiratory infections, which were circumvented. **Discussion:** Sjögren's syndrome (SS) is a chronic autoimmune inflammatory disease with exocrine gland involvement and extraglandular disease features, affecting peripheral and central nervous systems. Autonomic neuropathy cause difficult control of the symptoms. Myelitis leading a diagnosis of NMO spectrum. The importance of distinguishing NMO and SS is important for different treatments and prognosis. **Final Comments:** Neuro-Sjögren is a defiant illness to immunology, pathophysiology and possible complications inherent to this rare disease. Advances in disease-modifying therapies and early interventions can greatly improve the prognosis.

Neuroimunologia

TL 1105528

PARANEOPLASTIC NEUROMYELITIS OPTICA SPECTRUM DISORDER: A CASE PRESENTATION

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Case Presentation: A 53-year-old female patient was admitted to our Hospital with back pain history, progressive spastic paraparesis, with urinary incontinence and constipation, being diagnosed with acute transverse myelitis. Her magnetic resonance imaging showed longitudinally extensive spinal cord lesions; and her capillary electrophoresis (CE) showed polyclonal gammopathy (PG). She received high-dose intravenous methylprednisolone with satisfactory results. During follow-up, she tested positive for AQP4-IgG, fulfilling criteria for Neuromyelitis optica spectrum disorder (NMOSD), and was discharged with Azathioprine (AZT) plus prednisone. After 4 months, she was hospitalized again for cellulitis and infectious ulcers. Her complete blood count showed pancytopenia that was associated with the use of AZT, therefore, the medication was suspended. The patient progressed with worsening cytopenias. Another CE disclosed monoclonal gammopathy (MG), and a myelogram was performed, which showed 60% of clonal plasma cells in the bone marrow. As she was diagnosed with Multiple Myeloma (MM), with IgA/Kappa monoclonal protein, isolated Kappa light chain and beta-2 microglobulin, AZT was discontinued and myeloma treatment started with Cyclophosphamide, Bortezomib and Dexamethasone (CYBORD protocol). **Discussion:** Studies report a link between autoimmune diseases accompanied by diagnosis of MG. A study described 6 cases of paraneoplastic NMOSD. In his results 5 patients were older than 50 years old, and 4 had positive AQP4 antibodies. The median time between NMOSD and cancer was 12 months. A case series reported a 58-year-old female with visual defects and acute transverse myelitis, with a concomitant finding of MG in CE. Similarly to our case, there were no lytic bone lesions, hypercalcemia and renal impairment. Generally, patients with prolonged autoimmune diseases have PG. Therefore, CE with protein immunofixation and free light chain quantification is adequate for MG detection. The pathogenesis involving inflammatory states and clonal transformation is still unclear. Polyclonal proliferation may progress to monoclonal proliferation of B cells and induce MG. Possibly, there is involvement of dysregulated cytokines. **Final Comments:** We suggest that MG should be ruled out for patients with prolonged autoimmune diseases. There is probably a direct mechanism between cancer cells and development of NMOSD, however, further studies are required.

Neuroimunologia

TL 1105538

NEUROMYELITIS OPTICA & TONIC SPASMS AS ASSOCIATED CLINICAL MANIFESTATION

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Case Presentation: DNG, female, 26 years old, with inappetence, asthenia and paresthesia in the lower limbs, started 40 days after cesarean section and with progressive worsening. She had painful tonic limb spasms of short duration provoked by voluntary movements and emotional factors. Former alcoholic/addict for 6 months, she reported reduced visual acuity for 4 years after first pregnancy, with partial improvement. On physical examination, in addition to spasms, there was dysesthesia and paresis of limbs, hyperreflexia, bilateral reduced visual acuity with diffuse left papilla atrophy. Magnetic resonance imaging (MRI) of the orbits showed atrophy and hypersignal in the left optic nerve, suggesting inflammatory optic neuropathy, cervical spine MRI with hypersignal in T2 bulbomedullary, in C2-C3 and C6-C7, corresponding to demyelinating lesion, increased CSF of proteins and positive anti-aquaporin 4 antibodies (anti-AQP4). Pulse therapy with 1g Methylprednisolone was performed for 5 days and 40 mg of prednisone was maintained with gradual weaning. There was a good response with complete recovery. Currently on Azathioprine 50mg 2 times a day. For the treatment of spasms, we opted for the use of benzodiazepine and carbamazepine, with good control. **Discussion:** Neuromyelitis optica (NMO) is an autoimmune inflammatory disease of the central nervous system (CNS), characterized by simultaneous or consecutive cases of optic neuritis and transverse myelitis. The majority (80%) is caused by IgG autoantibodies anti AQP4 – a protein that forms a water channel in CNS. The clinical manifestations are diverse, being normally present the reduction of visual acuity and paresis/paresis in limbs with sensitive level. Tonic spasms are not common and are characterized by sudden onset of tonic postures, short duration, triggered by voluntary movements, sensory stimulation or hyperventilation, with or without pain and are due to axonal irritability due to the release of inflammatory mediators. On MRI there is hypersignal in the optic nerve and spinal cord, in addition to anti-AQP4 in blood/liquor. Treatment is with pulse therapy, followed by immunosuppressive medication. Tonic spasms respond well to carbamazepine. **Final Comments:** NMO is a disease with multiple clinical manifestations, sometimes presenting atypical symptoms as in the case above. It is up to the physician to make an early diagnosis for appropriate treatment, seeking control of symptoms and a favorable outcome.

Neuroimunologia

TL 1105571

CASE PRESENTATION OF AUTOIMMUNE ANTI-NMDA ENCEPHALITIS IN ADOLESCENTS IN THE STATE OF RIO GRANDE DO SUL

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Case Presentation: Patient GCS, female, 18 years old, in July 2020 started to present behavioral changes, hypersexualization, depression, anxiety, crying crises, emotional lability, refractory to psychiatric treatment. In April 2021, the exacerbation of the symptoms evolved, followed by language alteration, agitation, aggressiveness, visual hallucinations, and seizures, evolving to status epilepticus. On further investigation she presented small area of hypersignal in T2W and FLAIR in bilateral mesial temporal region topography, no pathological enhancement on skull MRI and pleocytosis with 69 cells, exclusive intrathecal oligoclonal bands and presence of anti-NMDA-receptor antibodies in cerebrospinal fluid. She was initially approached with acyclovir, pulse therapy in another service, with no response. We followed treatment with immunoglobulin, again without response, with ongoing epilepsy and orofacial dyskinesias. Therapy with Rituximab was subsequently followed, resulting in severe pharmacodermatitis in the condition of Toxic Epidermal Necrolysis (TEN), combined to secondary complications such as multidrug-resistant infections, that greatly increased morbidity. At the first opportunity of clinical stabilization, cyclophosphamide therapy was proceeded, obtaining good response in neuropsychiatric manifestations. The patient maintained the rehabilitation with a slow and gradual recovery, and today she has returned to her family, walks without help, and eats orally. **Discussion:** In this case, we discuss the importance of autoimmune encephalitis among the differential diagnoses of neuropsychiatric conditions, its diagnosis delay, mainly due to the onset of psychiatric manifestations and consequently delay in treatment. We also present the reality of a committed neurology service in the northern interior of the state of Rio Grande do Sul. The N-methyl-D-aspartate (NMDA) anti-receptor encephalitis first described in 2007 is the most common, and children and young adults should always be considered. The research of these receptors in the cerebrospinal fluid confirms the diagnosis, since brain images may be normal. The therapeutic approach consisting of immunosuppression should be initiated as early as possible to improve the outcome. The possibility of adverse reactions to potent and consolidated medications should always be taken into consideration.

Neuroimunologia

TL 1105592

PARANEOPLASTIC ENCEPHALOMYELITIS DUE TO ANTI-HU ANTIBODIES: A CASE PRESENTATION

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Case Presentation: A 75-year-old man, with multiple cardiovascular risk factors and a previous history of heavy smoking, presented to our clinic due to a forty-five days history of progressive right hemiparesis and hemiparesis. On neurologic examination he had normal cranial nerve evaluation. Motor examination revealed a right hemiparesis with upper motor signs (DTR were pathological and cutaneous response was extensor). Pinprick was reduced distally and vibration was severely impaired with positive Romberg sign. Brain MRI showed previous lacunar infarcts and no abnormalities in diffusion-weighted imaging. The patient had no signs of infection whatsoever. The CSF analysis showed normal cell count with slightly increased protein. Nerve conduction studies were normal and needle examination revealed chronic neurogenic features. EEG revealed diffuse desorganized background with no epileptic discharges. After three weeks of Hospitalization, the patient's level of consciousness got progressively worse and orotracheal intubation was needed. In the setting of sensory ataxia, progressive encephalopathy, without focal injury on neuroimaging or signs of infection in an elderly patient, a paraneoplastic panel was ordered, revealing anti-Hu antibodies in CSF and sera. The patient underwent neoplastic screening and a pulmonary nodule was identified. Unfortunately, before it was possible to biopsy the pulmonary mass, he died from infectious complications. **Discussion:** Anti-Hu antibodies is part of the onconeural antibodies. The anti-Hu paraneoplastic neurologic syndrome may present as sensory neuronopathy, chronic gastrointestinal pseudoobstruction, encephalomyelitis, and limbic encephalitis. When anti-Hu is identifiable in blood tests, cancer is almost always found. being small cell lung cancer the most common. Our patient presented risk factors for oncologic disease to his age, previous smoking history, and clinical course. Paraneoplastic syndromes are a differential diagnosis in these cases and investigation of this cause is required. **Final Comments:** Paraneoplastic neurologic syndromes can affect any part of the nervous system. This group of rare disorder is becoming more prevalent as the population age and with new paraneoplastic panel being affordable. The neurologist must include these group of disorder in the differential diagnosis, specially when the patient has risk factors for neoplasm.

Neuroimunologia

TL 1105609

MYELIN OLIGODENDROCYTE GLYCOPROTEIN ANTIBODY-ASSOCIATED DISEASE (MOGAD) WITH PNS INVOLVEMENT: A CASE PRESENTATION

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Case Presentation: GLF, male, 13 years old. The patient came for consultation in June 2020. At the age of three, he had a generalized tonic-clonic febrile seizure with post-ictal drowsiness. From then on, a difficulty in school learning was noticed. At the age of 7, he evolved with bilateral visual impairment with pain at eye mobilization. Two years later, he started with distal weakness and frequent falls. He had no family history. The patient was oriented, but with childish behavior for age. Mini-mental state exam: 22/30. He also presented flaccid tone and global hypotrophy associated with symmetrical tetraparesis predominantly crural. Deep tendon reflexes and cutaneous-plantar reflex were absent. Cerebellar gait with dysdiadochokinesia and dysmetria was evident. Distal hypopallesthesia was noticed. Visual acuity: 20/300 in the left eye and 20/200 in the right eye. Fundoscopy showed bilateral optic nerve atrophy. Considering the phenotype of presentation, findings in complementary exams and epidemiological profile, we chose to measure the anti-MOG IgG, with a positive result, corroborating a diagnosis of Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) with PNS involvement, manifested in our patient as polyradiculoneuropathy. **Discussion:** MOGAD is an immune-mediated demyelinating inflammatory disorder of the central nervous system predominantly affecting the optic nerves, brain, and spinal cord. The disease has a predilection for children. Oligodendrocyte myelin glycoprotein (MOG) is a protein on the surface of oligodendrocytes that has historically been considered as a potential antibody target in multiple sclerosis, until in 2007 it was described as a different disease phenotype. According to a database review (PUBMED), there are a few studies that corroborate the occurrence of peripheral impairment associated with MOGAD, present in 7% of MOGAD patients in an Australian cohort. There are some hypotheses to justify the concomitant central and peripheral demyelination processes: one of the most accepted claims that MOG is found in an isoform that can be released into the CSF, triggering or "spreading" autoimmunity when drained into the PNS. **Final Comments:** The presence of central demyelination syndrome and SNP involvement may be associated with MOGAD and may respond to immunotherapy, and should not discourage the search for MOGAD in patients with an appropriate clinical setting.

Neuroimunologia

TL 1105625

HYPERMOTILENCE AND ENCEPHALOPATHY AS A MANIFESTATION OF NEUROMYELITIS OPTICA: A CASE PRESENTATION

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Case Presentation: Female, 77 years old, hypertensive. In January 2021, she started presenting behavioral changes, difficulty looking up and binocular horizontal diplopia. The medical team performed brain MRI which presented a slight expansive effect on the mesial portions of the thalamus, hypothalamus and midbrain. After being hospitalized for investigation, the patient evolved to an altered level of consciousness, with severe drowsiness. She was submitted to a lumbar puncture and a non-reactive anti-AQP4 test. Part of the liquor was sent to Vienna. On 02/02, a stereotaxic biopsy was performed due to suspicion of a tumoral lesion. Treatment with IV pulse therapy was started. On 02/08 she had reentrant seizures and one week after started IV immunoglobulin. On 02/25 she was discharged from Hospital with partial improvement in her consciousness level. The next day she returned due to worsening of the drowsiness. In a new examination, a reduction of previously altered areas in the thalamus was seen. A new therapeutic attempt was performed, including Cyclophosphamide 1g IV, evolving with a significant improvement in the level of consciousness, but still presenting periods of drowsiness and cognitive impairment. On 03/26, a result of anti-AQP4 from Vienna was sent back with a reactive result. **Discussion:** Neuromyelitis Optica (NMO) is characterized by the presence of antibodies called Aquain 4 present in the hematoencephalic barrier. It has predominantly female occurrences and the first symptoms occur between the 3rd and 4th decades of life. The most common clinical manifestations include visual impairment and bilateral motor symptoms due to optic neuritis and transverse myelitis. There are other NMO spectrum syndromes, including an encephalopathy characterized by other levels of consciousness syndromes and seizures. The diagnoses are based on the presence of AQP4 antibody associated with at least one outbreak of clinical syndromes. One can still have the diagnosis without the presence of the antibody along with two clinical outbreaks and the presence of extensive lesions in the optic nerve, spinal cord or brain stem. **Final Comments:** In patients with positive AQP4 antibody, it is possible to make the diagnosis of NMO even with atypical clinical presentations. Our patient presented from the beginning the typical symptoms of NMO Encephalopathy in addition to radiological changes, but the diagnosis was concluded after the second AQP4-AB test with a positive result.

Neuroimunologia

TL 1105638

CASE PRESENTATION OF A 73-YEAR-OLD PATIENT WITH INCLUSION BODY MYOSITIS EVOLVING WITH BRADYCARDIA AFTER VACCINE FOR COVID

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Case Presentation: A 73-year-old male patient, from Manaus, Amazonas, previously diagnosed with Inclusion Body Myositis (IBM) confirmed in 2017 by the muscle biopsy method, which presented an inflammatory myopathic histological pattern with a large amount of COX- negative. The patient was taking prednisone 5mg/day, ramipril 10mg/day, hydrochlorothiazide 25mg/day and glicazide 60mg/day at home. On the twenty-sixth day after the third dose of Pfizer vaccine for covid-19, the patient appears for an outpatient follow-up consultation for underlying disease and for investigation of complaining of lipothymia and vertigo, denying any other sign of infection. During the evaluation, dyspnea was verified in MRC Class 3 and, in physical examination, a heart rate of 34 bpm was verified. Due to the condition, the patient was immediately advised by our neurology team to go to the emergency Hospital. In the emergency room, an electrocardiogram (ECG) was performed and a Complete Heart Block (CHB) was identified and the implantation of a temporary transvenous pacemaker was chosen as an emergency therapeutic approach, later, a permanent pacemaker was implanted. Patient was Hospitalized for 21 days in an Intensive Care Unit (ICU) for follow-up and procedures, today he has relief from dyspnea and lipothymia. **Discussion:** Cardiovascular damage is a known comorbidity of myopathies and can evolve with functional and conduction limitations. For example, cases of atrioventricular conduction delay have already been reported in a patient with IBM and, similarly, cases of CHB after vaccination for COVID-19 have already been reported. However, the causes of CHB are vast, making it difficult, in some cases, to identify its etiology. Thereby, the interconnection between vaccination, CHB and IBM can be arduous. **Final Comments:** In this case, we present a possible correlation between the Pfizer vaccine for covid-19 and abnormal electrical conduction in a patient affected by IBM. However, some limitations should be mentioned: despite the temporal correlation between the vaccine dose and the onset of symptoms being suggestive of inference and the existence of subsidies for connection with the baseline diagnosis, tests were not performed to directly prove the correlation between the three factors.

Neuroimunologia

TL 1105656

CASE PRESENTATION OF A 59-YEAR-OLD PATIENT WITH TRANSVERSE MYELITIS, CEREBELLAR SYNDROME AND ADEM AFTER VACCINE FOR COVID IN BRAZILIAN AMAZON

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Case Presentation: A 59-year-old patient from Manaus, Amazonas, seeks emergency care at an emergency Hospital after the fifth day of AstraZeneca vaccine for COVID-19, presenting progressive, symmetrical and ascending loss of strength in the lower limbs with 3 days of evolution, without other associated symptoms, denying diabetes, systemic arterial hypertension, alcohol consumption or smoking. Upon admission, physical examination revealed a Glasgow Coma Scale 15, isochoric and photoreactive pupils, no signs of meningeal irritation, grade 5 muscle strength in the upper limbs and grade 0 in the lower limbs according to MRC, areflexia in the lower limbs and decrease of superficial and deep sensitivity on them. Initially, the patient was admitted to the Intensive Care Unit (ICU), staying for 11 days for investigation for Guillain-Barré Syndrome (GBS) and for pulse therapy with methylprednisolone 1000mg IV for 5 days, but without improvement of symptoms. He was later transferred to our teaching Hospital, where a new neurological assessment was performed, identifying persistence of the previous findings, in addition to the presence of horizontal choreic movements of the head, intention tremor in the upper limbs, significant dysmetria, worse on the left, nystagmus and without alteration of cranial nerves. During Hospitalization, two biochemical evaluations of CSF were performed, but with unspecific results. The patient evolved with complications of cutaneous, urinary and pulmonary infections and died of sepsis after 110 days of Hospitalization. **Discussion:** Acute disseminated encephalomyelitis (ADEM) is a rare inflammatory disease of autoimmune etiology that may have a presentation similar to GBS. In the current context, there are already a few reports that point to cases similar to the one we present, showing a possible correlation between the vaccine for COVID-19 and ADEM. **Final Comments:** In this case, we present a possible and rare correlation between the AstraZeneca vaccine for COVID-19 and ADEM. That said, the present report seeks to encourage more discussions about the possible complications of vaccination without, however, questioning its safety, since adverse effects are extremely rare.

Neuroimunologia

TL 1105691

OPHTHALMOPARESIS SECONDARY TO IGG4-RELATED DISEASE – A CASE PRESENTATION

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Case Presentation: A.L.O.S, female, 61 years old. The patient related binocular diplopia, ophthalmoparesis – there was no horizontal movement and limited vertical movement – and fixed semiptosis in the left eye that started 3 days before the medical care. The patient experienced similar symptoms two years ago, which solved spontaneously. Previous diseases were osteoporosis, dilipidemia and systemic arterial hypertension. The Magnetic Resonance Imaging showed expansive/infiltrative formation compromising the left lateral rectus musculature and a lesion with the same sign in the temporal musculature in the same side. Cerebrospinal fluid analysis was normal. Lateral rectus muscle biopsy confirmed IgG4-related disease. The patient started corticotherapy (prednisone 40 mg/ per day). **Discussion:** IgG4-related disease is an immunomediated disease characterized by enlargement of the organs and lymphoplasmatic infiltration. It is a rare condition that affects 2,2 to 100,000 people. Extraocular muscle involvement in this disease has been related but with a poor description of the eye movement. The literature describes IgG4-related disease eye movement muscles involvement as a condition that is associated to pain, proptosis and ophthalmoplegia. The case presented above had a different clinical presentation because semiptosis and severe ophthalmoparesis that is uncommon in IgG4-related disease; also, there was no eye pain. **Final Comments:** IgG4-related disease usually appears as a condition with normal ocular motility. Once there are lots of other diseases that compromise eye movement muscles this case report is important to neurologists to show an unexpected clinical presentation and call attention to the need to make a correct diagnosis despite of an atypical presentation.

Neuroimunologia

TL 1105694

CASE PRESENTATION ACUTE ISCHEMIC STROKE IN BEHÇET'S DISEASE

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Case Presentation: A 16-year-old male patient, previously healthy, presented a sudden onset of severe headache associated with paresis of the right hemibody. Undergoing Hospitalization in a highly complex local referral Hospital for treatment and investigation. During the Hospital stay, he develops a new condition of left hemiparesis, suddenly. In both events, areas of cerebral ischemia were evidenced, confirmed by magnetic resonance imaging, in different locations. After active investigation, patient and mother confirmed a personal history of recurrent oral ulcers, episodes of ulcers in the penile region, and ulcerated skin lesions. Complementary exams: Skull Resonance: Temporo-parietal signal alteration on the right represented by hypersignal on T2 and FLAIR which may correspond to an ischemic event. Entry labs exams (2/16/2022): ESR 150, Bilirubin 2.4, TGO 38, TGP 51, Reticulocytes 2.5%, DHL 519. Pathergy test was positive. Patient submitted to Hospital pulse therapy treatment with 1g/day of Methylprednisolone for five days, associated with full anticoagulation, evolving with stability of the condition. Patient referred for outpatient follow-up after Hospital discharge, with oral corticosteroid maintenance, therapeutic anticoagulation and immunosuppressive therapy planning. **Discussion:** Behçet's Disease is an autoimmune syndrome, and among the systemic vascular diseases, is notable for its ability to involve blood vessels of all sizes. Affects young adults aged 20 to 40 years and the most common clinical features are the presence of recurrent and painful mucocutaneous ulcers, painful urogenital lesions and various skin lesions. The neurological impairment of the syndrome occurs in less than 10% of patients and more commonly seen in men and the diagnosis is made on the basis of clinical findings. The goal of treatment is to promptly suppress inflammatory exacerbations and recurrences to avoid irreversible organ damage, and immunosuppressive medications are mainly indicated. **Final Comments:** Despite being rare, the neurological impairment in Behçet syndrome has high rates of morbidity and mortality, given the vast clinical involvement of this pathology. Considering that the diagnosis of the disease is clinical, a careful anamnesis and the direct search for complications caused by this disease are essential for the treatment to be established and the systemic damage to be stopped.

Neuroimunologia

TL 1105705

A REMARKABLE RADIOLOGICAL IMPROVEMENT IN A PATIENT WITH NMOSD

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Case Presentation: A 43-year-old woman presented to the neurology department quadriplegic, with hypoaesthesia below the C4 level, bilateral impaired vision, no alterations in other cranial nerves and bilateral Babinski sign. She had a history of similar symptoms 7 months before, but was not examined at that time. The dosage of vitamin B12 was 88 pg/mL, the AQP4-IgG was negative, MOG-IgG was not available, and other laboratory results were non-significant. Lumbar puncture was not performed because the patient was in full anti-coagulation. A magnetic resonance imaging (MRI) of the spine evidenced areas of hyperintensity on T2-weighted sequences from the area postrema to the conus medullaris, compatible with longitudinally extensive transverse myelitis. The patient met the diagnostic criteria for neuromyelitis optica spectrum disorders (NMOSD) without AQP4-IgG, and a course of pulse therapy with methylprednisolone was prescribed, as well as one plasma exchange therapy, and vitamin B12 replacement. She used oral prednisone until azathioprine was available. Five years after this episode, the patient returned to the neurology department for a follow-up consultation. She was in a good general state, no longer in need of a wheelchair, with a spastic parietic gait, no alterations in cranial nerves, muscular strength of 4/5 in the upper limbs and 3/5 in the lower limbs, and hypoaesthesia below T10 level. Her EDSS was 5.5. A new MRI of the spinal cord showed a great radiological improvement, with only small areas of hyperintensity at the cervical spine. The reported case shows a patient diagnosed with NMOSD with an important clinical and radiological improvement after 5 years of evolution. The initial manifestation on MRI, with a centrally located lesion on axial imaging, a large cord expansion and post-contrast enhancement, was compatible with the general presentation of NMOSD. While vitamin B12 deficiency could be a confounding factor, radiological findings do not support the hypothesis, although it could enhance symptoms. Different studies discuss the link between AQP4-IgG, MOG-IgG and the disease prognosis, with better outcomes for patients who are AQP4-IgG-seronegative and MOG-IgG-seropositive. In the present case, MOG-IgG was not tested, which weakened the ability to completely interpret the clinical observations. Nevertheless, a remarkable reduction of the lesions extent on MRI and a less likely chance of relapse are seen in AQP4-seronegative cases, as in this case.

Neuroimunologia

TL 1105712

TITLE: PAI-1 INHIBITOR GENE 4G5G POLYMORPHISM IN THE CONTEXT OF RECURRENT STROKE AS A DIFFERENTIAL DIAGNOSIS OF RELAPSING-REMITTING MULTIPLE SCLEROSIS

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Case Presentation: A – Woman, 40 years old, in 2017 had a visual blurring in the left eye upon waking up, and her brain CT scan suggested stroke. Due to further motor deficit, brain MRI was performed and were observed multiple hyperintense focal lesions on T2/FLAIR in white matter, one of them enhanced contrast. CSF analysis, cervical and dorsal spinal cord MRIs were normal. A screening for microvascular disease, autoimmune diseases, bacterial/viral infections and hereditary and acquired thrombophilia were negative. In 2019, she presented motor aphasia and a new brain MRI showed a left frontal ischemic lesion. In 2020 it evolved transient stroke episodes. An investigation for thrombophilia causes, a mutation in the polymorphism in the PAI-1 gene, 4G allele, was detected. B – A woman, 26-year-old in December 2020 had sequential generalized motor seizures, a brain CT scan showed right cortical and subcortical hypodensity with signs of subarachnoid bleeding. He remained with mild right hemiparesis. A control MRI showed areas of right parietal encephalomalacia, with hemosiderin residues, and multiple small hyperintense focal lesions on T2 and Flair in white matter, some of them with apparent enhancement by gadolinium, suggesting demyelination. The CSF analysis showed positive oligoclonal bands. Cervical and dorsal spinal cord MRI, visual and somatosensory evoked potentials were normal. An investigation for polymorphic alteration of the PAI-1 inhibitor gene, configured as 4G5G, was detected. **Discussion:** In PAI-1, the 4G allele, suppresses fibrinolysis by inhibiting tPA and uPA, plasminogen activating enzymes, thereby suppressing plasmin formation. Normal polymorphism is 5G/5G. The 4G5G and 4G4G polymorphic presentations are associated with cardiovascular thrombotic conditions and cerebral infarctions. The clinical relevance of PAI-1 has been documented in several clinical and laboratory studies (1,2) Cases of Multiple Sclerosis, an inflammatory disease, may include fibrinolysis disorders in its pathogenesis. (3) The increased presence of the PAI-1 inhibitor, through polymorphism with 4G alleles, can destabilize the clinical picture, inducing infarctions and ischemia. **Final Comments:** the participation of these polymorphisms may have contributed to the neurological conditions presented by the patients and may be a confounding or intensifying factor in the development of Multiple Sclerosis and other diseases that course with thrombosis.

Neuroimunologia

TL 1105728

CASE PRESENTATION OF STEROID RESPONSIVE ENCEPHALOPATHY ASSOCIATED WITH AUTOIMMUNE THYROIDITIS (SREAT)

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Case Presentation: Patient S.C.D.S., 51 years old, female, started in May 2020 with gait ataxia with postural instability determining falls. Report of incoordination to handle objects, hand tremor when performing movements. Later, difficulty in balancing oneself in an orthostatic position, even at rest. There was progressive worsening over the subsequent month, requiring support to walk. Neurological examination showed cerebellar syndrome with ataxic gait, dysmetria, dysdiadochokinesia, Romberg sign, Stewart-Holmes test, and moderate frequency and amplitude intention tremor. Complementary tests showed thyroid hormones (TSH and free T4) within the normal range but high Anti - T.P.O. and Anti - Thyroglobulin. Skull M.R.I. without alterations. Given the findings, the diagnosis of steroid-responsive encephalopathy associated with autoimmune thyroiditis (S.R.E.A.T.) was proposed. The patient started pulse therapy with methylprednisolone with excellent therapeutic response. In outpatient follow-up, cerebellar syndrome improved, especially in gait, with the return of the ability to walk without support. SRAT characteristically presents as an encephalopathy in which seizures, movement disorders, and psychosis are common findings. Rarely, the disease is predominantly expressed as cerebellar ataxia, as presented in the case described. The criteria for diagnosis of this condition were established with the elevation of anti-thyroid peroxidase (anti-TPO) and anti-thyroglobulin (anti-Tg) antibodies. Patients usually have an excellent response to immunotherapies, including steroids, as corroborated in the case we describe. Considering the atypical clinical presentation of the predominantly cerebellar S.R.A.T., the relevance of the case is reinforced.

Neuroimunologia

TL 1105742

ACUTE DISSEMINATE ENCEPHALOMYELITIS AFTER PFIZER VACCINE: CASE PRESENTATION

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Case Presentation: A 44-year-old male patient from Manaus, Amazonas was admitted in our Hospital with a history of upper limbs paraparesis starting on April 21, 10 days after Pfizer COVID vaccine (it was his 3rd dose; the 1st and 2nd doses were Coronavac), which progressively affected his lower limbs associated with stiff neck, nausea and vomiting. He evolved with dysphagia, dysarthria, urinary retention and severe tetraparesis, looking for an emergency service on May 15, where he performed a cranial CT which showed right frontal hypodensity and enlargement of the right temporal horn of the lateral ventricle, being later referred to the neurology Intensive Care service for better investigation. CSF collection did not show infectious processes. Neurological examination revealed grade 2 strength tetraparesis in the proximal lower limbs and plegia distally, while the upper limbs had 1-strength proximally and 1+ distally. Spastic tetraparesis associated with clonus in superior and lower limbs was also found. Reduced protopathic touch globally. Also bilateral vertical nystagmus and paresthesia in the left hemiface, without other alterations. In view of the imaging findings in association with clinical manifestations, the diagnosis of Acute Disseminated Encephalomyelitis (ADEM) was confirmed. After the condition worsened, due to dyspnea and hypoxic respiratory insufficiency, the patient was referred to the ICU where he underwent endotracheal intubation and, to date, 7 plasmapheresis sessions. Now, he is in a lethargic state, with sepsis due to urinary tract infection. **Discussion:** ADEM is a rare disease that can be associated with other neurological diseases, such as Guillain-Barré syndrome or isolated encephalitis. It is a diagnosis of exclusion and, therefore, imaging tests and CSF collection are essential. In the clinical aspects, several signs of encephalopathy can be found, such as headache, ataxias, hemiparesis and cranial nerve alterations. Treatment revolves around immunosuppression, typically with human immunoglobulin, steroids, and, in severe cases, plasmapheresis. **Final Comments:** The case presented shows a rare case of ADEM after the 3rd dose of the vaccine against COVID-19. The present report does not aim to question the safety of vaccines, since adverse effects of this severity are rare, but rather to share this specific case and, in this way, foster discussions about the preparation of health professionals to deal with such a situation.

Neuroimunologia

TL 1105758

PARANEOPLASTIC NEUROMYELITIS OPTICA SPECTRUM DISORDER WITH ANTI-AQUAPORIN-4 ASSOCIATED WITH SQUAMOUS CELL CARCINOMA OF THE LUNG

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Case Presentation: A 61-year-old man with a past medical history of liver transplantation in 2001 due to alcoholic cirrhosis was admitted for untreatable hiccups and vomiting, which had started 14 days earlier. Initial imaging showed a left lung apex mass. A biopsy was made and the patient was diagnosed with squamous cell lung carcinoma. One month after the first symptoms, the patient developed paraparesis, upper right limb paresis, complete anesthesia of the lower limbs, urinary and fecal retention. During the following 24 hours, the symptoms worsened and he became paraplegic. Brain and spine MRI showed a longitudinally extensive transverse myelitis, with high T2 signal involving C5 to T11 segments, along with high T2 signal within the area postrema. CSF analysis were as follows: protein 472 mg/dl, 124 cells (81% neutrophils), with no other relevant findings. Anti-aquaporin 4 antibodies were detected on a blood sample. The patient was not tested for onconeural antibodies due to unavailability. The patient was started on high dose EV corticosteroids (methylprednisolone 1g for five days), without any improvement. A second treatment attempt with a five-day course of plasma exchange therapy was also unsuccessful. During Hospital stay, the patient developed sepsis and was intubated. Due to respiratory muscular weakness, ventilator weaning was challenging. After a prolonged ICU stay, the patient died. **Discussion:** Paraneoplastic neuromyelitis optica is a rare condition, with an estimated incidence of less than 4 per cent of all NMOSD cases. It is more often associated with lung and breast adenocarcinoma. However, other cancer types can be involved. The mean age of onset is 55 years. Brainstem lesions and longitudinally extensive transverse myelitis are the most common presentations, and some studies suggest cancer screening in these patients. **Final Comments:** Although the condition is rare, it is important for the neurologist to recognize paraneoplastic NMOSD as a cause of demyelinating disease in cancer patients, but also as a red flag for possible occult tumors in older patients who develop neuromyelitis optica, especially those over the age of 55.

Neuroimunologia

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VOGT-KOYANAGI-HARADA DISEASE, A RARE ETIOLOGY OF STERILE MENINGITIS – A CASE PRESENTATION

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Case Presentation: A 16 year old Venezuelan boy with no previous comorbidities, who was admitted at the emergency room presenting headache associated with bilateral conjunctival hyperemia and reduced visual acuity, limited to light perception. The signs and symptoms presented have been developing during the course of 14 days. There were no other neurological findings. A head computed tomography scan accused a diffuse enlargement of the ventricular system, suggesting hydrocephalus, and there was an altered signal inside both orbits compatible with retinal detachment. In addition, the analysis of the cerebrospinal fluid revealed pleocytosis, with a monocytes predominance. The patient underwent neurosurgery and a ventriculoperitoneal shunt had been installed. The ophthalmologic evaluation pointed out a serous panuveitis. The hypothesis of Vogt-Koyanagi-Harada disease has been raised and the patient was treated with a 5-day course of methylprednisolone followed by oral glucocorticoid and ophthalmological follow-up. **Discussion:** Vogt-Koyanagi-Harada (VKH) disease is an important and rare cause of sterile meningitis. It is a systemic autoimmune disorder that target pigmented tissues of the body, affecting particularly the eyes, but also the skin, meninges and inner ear either, resulting in neurological symptoms such as headache, cranial nerves palsy and tinnitus. The prevalence of VKH varies in different populations in the world, accounting for 2 to 4% of all uveitis in Brazil. The diagnosis is based on clinical findings and must follow the criteria proposed by the American Uveitis Society, including bilateral ocular involvement with no history of penetrating ocular trauma or surgery, along with neurological or integumentary findings. The goal of treatment is to suppress intraocular inflammation and prevent potential visual impairment, therefore early diagnosis and treatment are essential. **Final Comments:** It's important sharing cases like this since it is a relatively rare diagnosis of sterile meningitis in clinical practice, hence often underdiagnosed. It is vital that physicians pay attention on the ophthalmological manifestations associated since they are the most frequent ones and the base of the diagnostic criteria. Once more, it is of utmost importance the fast diagnosis followed by and early and aggressive treatment with systemic corticosteroid to reduce subsequent recurrences and avoid complete and irreversible visual loss.

Neuroimunologia

TL 1105761

DIAGNOSIS OF AUTOIMMUNE CEREBELLITIS DETECTED BY 18 F-FDG-PET/CT AFTER SARS-COV-2 INFECTION

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Case Presentation: A 57-year-old man presented with Sars-Cov-2 infection in March 2020 with severe viral pneumonia, without the need for mechanical ventilation. He evolved after 2 weeks with gait ataxia, dysmetria, dysidiadochokinesia, vertigo with cerebellar features, cerebellar tremor and bilateral vertical nystagmus. Cranial magnetic resonance imaging was performed, which did not show any acute changes. Lumbar puncture was performed to collect cerebrospinal fluid that showed no changes. Brain PET-CT with 18F-FDG was performed, which showed anomalous hyperconcentration of the radioisotope in cerebellar tonsils and cerebellar vermis. Hypothesized post-COVID-19 autoimmune cerebellitis. Pulse therapy with methylprednisolone 1g/day was performed for 5 days, with considerable improvement in symptoms. He is discharged from the Hospital with improvement of gait symptoms and improvement of nystagmus. After 15 days, improvement of dysidiadochokinesia and dysmetria, maintaining mild vertigo. After 2 months of pulse therapy, the patient was asymptomatic from a neurological point of view. **Discussion:** The diagnosis of encephalitis/cerebellitis using the PET-CT technique with 18-F-FDG infusion is effective in cases with normal CSF results described in the literature. Reports of autoimmune cerebellitis after COVID-19 infection are described in the literature with inconclusive results regarding the use of methylprednisolone or immunoglobulin. **Final Comments:** The report on the use of PET with 18-F-FDG for the diagnosis of autoimmune involvement of the central nervous system appears to be a promising alternative with increasing use. Considering the possible relationship between Sars-Cov-2 infection and autoimmune cerebellitis, it is necessary to publicize the method used to the scientific community in order to facilitate future prospective studies.

Neuroimunologia

TL 1105791

MARBURG VARIANT OF MULTIPLE SCLEROSIS AFTER COVID-19 SUCCESSFULLY TREATED WITH BETAINTERFERON 1A – A CASE PRESENTATION

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Case Presentation: A 20-year-old female patient previously healthy had a mild Covid-19 one month before neurological symptoms. Four days before admission, she started with paresthesia in the inferior left limb. The neurological examination showed complete left hemiparesis and painful hypoesthesia on the left side. She was submitted to a brain magnetic resonance imaging (MRI) that evidenced hyperintense subcortical lesions on T2/FLAIR and hypointense on T1. She received a five days course of methylprednisolone 1g with partial improvement of the symptoms and she was discharged to ambulatory following. After one month, the patient went back to emergency with focal seizures in the face and worsening of left hemiparesis. The new MRI showed an increase of hyperintense T2/FLAIR lesions on the right frontotemporal. Due to the fast and aggressive presentation with neurological deterioration and failure of initial therapy, the possibility of Marburg's variant of MS was suggested. It was decided to start beta interferon 1a with 44 mcg three times a week. After one month of the new therapy, the patient presented an improvement of symptoms, reduction of lesions in MRI. **Discussion:** The Marburg variant is a particular form of MS. It is known this results in myelin sheath instability that contributes to the formation of demyelinating plates. Generally, the injuries show a widespread and inflammatory aspect. The neuroimaging generally reveal multiple extensive lesions, involving the white matter. Our case presentation showed an evolution of neurological injuries in a healthy patient, except history of Covid-19, possibly a trigger of this demyelinating disease. The first-line therapy is the use of glucocorticoids in high doses, and the second-line is intravenous immunoglobulin and plasmapheresis. After the progression of deficits, we decided to treat with beta-interferon 1a, which stops that progression, improved neurological symptoms and promoted regression of brain lesions. Although this medication is not commonly indicated for this form of disease, it shows the immune factor in the Marburg variant and its potential to respond to immunomodulatory drugs. **Final Comments:** The Marburg variant of MS requires a high level of clinical suspicion and knowledge about different MS presentations. We present a peculiar case because a correlation with Covid-19 made this a possible trigger and the good response with beta interferon 1a showed a possible drug to consider in management of this disease.

Neuroimunologia

TL 1105795

ADULT AUTOSOMAL LEUKODYSTROPHY: RARE FAMILY CASE, WITH DYSAUTONOMIA PRECEDING ATAXIA

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Case Presentation: A family in which 4 of its members were diagnosed with autosomal dominant adult leukodystrophy with dysautonomia (LADA) was studied in our service: 1. PJ, first child, onset of symptoms with pain in the lower limbs (LL) in 2012 at age 42, evolving with loss of strength in the lower limbs and ataxia in 2016. He currently has hoarseness, urinary retention, pain and ataxia in the lower limbs. On MRI: hypersignal in the supratentorial, periventricular white matter, with preservation of the "U" fibers, internal capsule, trigeminal nerve nucleus and midbrain course, cerebellum, cerebellar peduncles with bulbar pyramid extension. 2. AF, second child, started in 2010 at age 40 with a complaint of generalized loss of strength and ataxia with progressive worsening in ambulation and urinary retention. The current physical examination shows hypoesthesia in the right hemibody, in addition to dysmetria in the lower limbs. On magnetic resonance imaging (MRI): symmetrical hypersignal in white matter of the semiovals and periventricular centers, cerebellar peduncle, critical spinal path and medullary bobus transition. 3. AA, third child, individual already in death secondary to a car accident, siblings reported that he had dysmetria in the lower limbs, was diagnosed through imaging tests, but did not maintain medical follow-up. 4. CA, mother started symptoms at 40 years old, with frequent falls, at 56 years old, need for a wheelchair, intestinal and urinary dysautonomia, dying after 14 years. FA and PJ presented. The two brothers present in cervical MRI, medullary thinning and hypersignal. **Discussion:** Autosomal dominant leukodystrophy in adults with dysautonomia is a rare condition to the point that its prevalence is unknown. It is secondary to the accumulation of lamina b1 proteins, by genetic defect 5q23 Lmnb1, leads to demyelination with astrocyte dysfunction and oligodendroglia preservation. The disease has a slow evolution in about 18 years, with paraparesis, spastic tetraparesis and finally pseudobulbar palsy, leading to death. Clinic in which the mentioned family presents, in addition to MRI. **Final Comments:** In the literature, only one case is presented in which dysautonomia complained of ataxia, which, like the aforementioned family, alternates the classic order of symptoms. Which makes the reported case even more rare

Neuroimunologia

TL 1105822

EIGHT-AND-A-HALF SYNDROME AS A RARE INITIAL PRESENTATION OF MULTIPLE ESCLEROSIS (MS)

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Case Presentation: A 22-year-old male with no previous medical history is admitted with a new onset strabismus and double vision, both installed three days prior to the consultation. He had no history of trauma, head or ocular pain, vision loss or systemic symptoms. Ophthalmology was the first specialty evaluating the patient, concluding no alterations in their field. Neurological examination showed an ocular misalignment at primary gaze, with right eye exotropia, right conjugate gaze palsy, impairment of right eye adduction and complete right facial nerve palsy. Remaining neurological assessment was preserved. Initial blood investigation tests were negative for inflammatory markers, serological status and rheumatological panel. The Cerebrospinal fluid analysis showed 1 leukocyte/mm³ and 24mg/dL proteins. Magnetic resonance imaging identified several T1-weighted hypointense images with no gadolinium enhancement compatible with "black-holes" in periventricular white matter, as long as T1-weighted hyperintense lesions on cervical spinal cord and right facial colliculus site at the pons, also with no gadolinium enhancement. Patient was treated for MS outbreak with Methylprednisolone 1g IV for five days with complete resolution of diplopia and partial recovery of the eye movements and strabismus. As disease modifying therapy, the patient was put under Natalizumab schedule. MS is an autoimmune demyelinating disease whose lesions can potentially cause irreversible disabilities when untreated. Among these, neuronal pathways to ocular extrinsic motility are commonly affected. Nevertheless, the presentation of eight-and-a-half syndrome is a very rare and intriguing presentation where a single lesion compromises three pons structures: medial longitudinal fasciculus, leading to homolateral eye adduction deficit (called internuclear ophthalmoplegia); abducens nuclei, impairing homolateral eye abduction and fascicular facial nerve fibers at the genu site where they arch behind abducens nerve, clinically presented as "periferic" pattern facial palsy besides being a central lesion. In our case we illustrate how the knowledge of anatomical pathways is crucial to clinical neurology, useful for identifying patterns which help correct diagnosis and treatments. Despite continuous improvement, MS remains a challenge to clinicians and neurologists because of its extensive variety of initial presentations, sometimes making the correct diagnosis a true hardship.

Neuroimunologia

TL 1105848

MARBURG VARIANT OF MULTIPLE SCLEROSIS AFTER COVID-19 SUCESSFULLY TREATED WITH BETAINTERFERON 1A – A CASE PRESENTATION

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Case Presentation: A 20-year-old female patient previously healthy had a history of mild covid-19 one month before neurological symptoms. Four days before admission she started with paresthesia in the inferior left limb, evolving with paresis and visual blurring. Neurological examination showed complete left hemiparesis with pyramidal signs and clonus, loss of proprioception and vibration in lower limbs, tactile and painful hypoesthesia on the left side, slightly scanning speech and hemiparetic gait. Differential diagnosis (DD) was with infectious, inflammatory and rheumatologic tests, all negative. Magnetic resonance imaging (MRI) showed diffuse hyperintense subcortical lesions on T2/FLAIR and hypointense on T1, involving periventricular white matter and brainstem. Methylprednisolone was initiated, with partial improvement of the symptoms. After one month, the patient had focal seizures in the face and worsening of left hemiparesis, with MRI revealing increase of hyperintense T2/FLAIR lesions on the right frontotemporal and parietal lobes, pons and cerebellar regions. DD was tumefactive demyelination or tumor. The family didn't agree about the brain biopsy. The possibility of Marburg's variant of MS was suggested. Interferon beta therapy was initiated three times a week. After one month, the patient presented improvement of symptoms and reduction of lesions in MRI, with continuous improvement in the following months. Marburg variant is a form of MS that has a single-phase course and can lead to death in weeks to months after the initial presentation, an uncommon outcome in the general MS patients. The distribution of the lesions can be exactly as MS, though, in this variant, they spread to diverse brain areas, even including the brainstem. Differential diagnoses, such as tumor, infectious, granulomatous, and vasculitic lesions must be excluded. Neuroimaging alterations are not specific, involving the white matter of the hemispheres and brainstem. Our report showed evolution of neurological injuries in a patient with no previous conditions, except history of covid-19, possibly a trigger of the disease. The patient was treated with doses of steroids with partial response. After the progression of deficits, treatment with betainterferon 1-a was proposed, regressing symptoms and brain lesions. We present Marburg variant of MS its correlation with covid-19 as a possible trigger and the good response with beta-interferon.

Neuroimunologia

TL 1105855

MIOCLONUS-OPSOCLONUS-ATAXIA SYNDROME POST COVID-19 INFECTION: CASE PRESENTATION

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Case Presentation: A 65-year-old man attended the outpatient clinic referring tremors for 30 days. The condition progressed with dizziness, malaise, vomiting, aphasia, opsoclonus and myoclonus, which were presented 9 days after confirmed Covid-19 infection. Denies vertigo, seizures and history of cerebral vascular accident (CVA). Neurological examination showed bilateral dysmetria, dysidiadochokinesia in left upper limb and involuntary coarse movement with chorea and athetosis, in addition to muscle contraction. The cerebral magnetic resonance revealed global atrophy and microangiopathy. Requested electroencephalogram (EEG), which showed normal activity with the presence of rapid eye movement artifacts, in addition to laboratory tests for paraneoplasia and other possible etiologies screening (CA 19.9, antigen carcinoembryonic (ACE), ESR, PCR, occult blood test, ammonia, ANA, anti-beta2-glycoprotein, homocysteine, vitamin B12 and vitamin E dosage, folic acid, PII, anticardiolipin, ALT and AST, AP and GGT, HbsAg, anti-HCV, Elisa HIV, VDRL and FTA-ABS). The results did not show paraneoplasms or infections, thus closing the diagnosis of Kinsbourne Syndrome. A lumbar puncture was made to evaluate the cerebrospinal fluid in addition to a high dose methylprednisolone pulse therapy. At the return visit, the patient showed a significant improvement in the clinical status. **Discussion:** The Kinsbourne Syndrome or Opsoclonus Myoclonus Ataxia syndrome is a rare neuroinflammatory disorder with a variable combination of opsoclonus, myoclonus, and ataxia¹. The etiology may be paraneoplastic, such as for neoblastoma, parainfectious (after viral or bacterial infections), toxic/metabolic or idiopathic². The exact pathogenesis is not fully known, however, in recent years, the emergence of neurological disorders, such as Kinsbourne syndrome itself, after COVID-19 infection has been reported in several studies¹. The syndrome is believed to be an inflammatory phenomenon mediated by the autoimmune system, being secondary to the Sars-Cov 2 infection². **Final Comments:** Although it is a rare disease, the diagnosis of Kinsbourne syndrome by the neurologist, in the present case, was verified from the permanence of sequelae, mainly related to motricity and persistence of opsoclonus, as reported in the literature, in addition to the exclusion of occult neoblastoma and other conditions.

Neuroimunologia

TL 1105897

DEPERSONALIZATION AS A SYMPTOM OF A PARIETO-OCCIPITAL LESION IN A PATIENT WITH RELAPSING REMITTING MULTIPLE SCLEROSIS: A CASE PRESENTATION

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Case Presentation: In June 2021, a 52-year-old female with relapsing remitting multiple sclerosis (MS) since the age of 14 with left hemiplegia, attended to an outpatient follow-up with a history of 4 monthly episodes of temporal disorientation and disorganized thinking in the past few years, after one of the MS outbreaks. Each episode begins by waking up; lasts throughout the day; ends at bedtime. While in this situation, the patient describes that she watches her body from above, also, she cannot perform a sequence of tasks properly, also she notices a change in processing speed, characterizing a depersonalization disorder. The patient underwent a new brain MRI that showed a focal T2 hypersignal in the subcortical white matter of the right frontal lobe present in a previous exam and a new lesion in the right parieto-occipital subcortical white matter next to the posterior horn of the lateral ventricle with hypersignal in T2 and FLAIR and no gadolinium enhancement that might temporarily correspond to the onset of the current symptom. **Discussion:** Cognitive impairments and MS are often related, suggesting they are associated with the demyelination process. This finding is more common in patients with progressive MS. Most cases report deficits in episodic memory and processing speed, although symptoms such as verbal fluency, visuospatial analysis, difficulty in multitasking and in word finding may occur. These declines often begin early in disease, being one of the major concerns of the patients once they are diagnosed. Besides cognition, psychiatric symptoms are also reported in some MS phenotypes, including psychosis, depersonalization. Despite the link between the relapse and the depersonalization, it is more frequently seen the association between MS and cognitive disorders. Also, neuropsychiatric disorders appeared in a bigger percentage associated with frontal lobe lesions. **Final Comments:** Cognitive disorders and behavioral symptoms may be associated with multiple sclerosis outbreaks and represent an important factor to the patient's quality of life. In this patient, the parieto-occipital lesion in the new MRI correlates with symptom onset, suggesting a possible correlation between that region and depersonalization. Although not described before, it could be attributed to the spatial recognition function of the parietal lobe, prompting more studies regarding the correlation of depersonalization and its topography.

Neuroimunologia

TL 1105925

CADASIL AND/OR MULTIPLE SCLEROSIS? A CASE OF DIFFICULT DIAGNOSIS

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Case Presentation: A 48-year-old female was referred to our clinic for investigation of possible MS, which had started 3 years earlier with an episode of dysarthria and right lower limb weakness. MRI showed focal and confluent T2-hyperintense lesions in the subcortical and periventricular white matter as well as infratentorial regions and cervical and thoracic spinal cord. Since she lacked evidence of dissemination in time, a diagnosis of high-risk clinically isolated syndrome was made and she was started on glatiramer acetate. Over the next year, there were new symptoms accompanied by gadolinium-enhancing brain lesions. At this point she fulfilled the diagnostic criteria for MS and was treated with IV. methylprednisolone, with improvement. She was switched to natalizumab, which effectively controlled MS activity, and five years later to fingolimod due to increasing titers of serum JCV antibodies, but later on developed insidious worsening of physical and cognitive disability (EDSS 5.5), suggestive of secondary progressive MS, as well as epilepsy. She had no personal or family history of migraine, stroke or psychiatric conditions. At age 62 years, no-cost testing for genetic leukoencephalopathies became available for our site and revealed one likely pathogenic variant in NOTCH3, the gene associated with autosomal dominant cerebral arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL). In retrospect, her MRI scans were assessed as compatible not only with MS but also with CADASIL, due to involvement of the external capsule and temporal lobes bilaterally. Genetic counseling was performed, management of vascular risk factors was intensified, and she was kept on fingolimod. **Discussion:** CADASIL is a monogenic disease associated with recurrent small-vessel ischaemic strokes, migraine, and dementia. As CADASIL often has a relapsing-remitting presentation and widespread cerebral and brainstem hyperintense white matter lesions, misdiagnosis with MS can occur. However, in this case, features such as gadolinium-enhancing and spinal cord lesions and response to steroids were rather suggestive of MS. We found similar cases in the literature, indicating that the association between MS and CADASIL may be more than casual. **Final Comments:** Our case highlights that CADASIL may mimic or coexist with MS, yet the relationship between these conditions is yet to be determined. Genetic testing for leukoencephalopathies may prove useful for selected patients with MS.

Neuroimunologia

TL 1105934

POST COVID-19 VACCINE-ASSOCIATED ACUTE MYELORADICULONEUROPATHY RESPONSIVE TO PLASMAPHERESIS

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Case Presentation: A 31-year-old male patient presented with acute tetraparesis and urinary retention one day after the first dose of the Covid-19 vaccine. Covid-19 PCR (polymerase chain reaction) was negative, and IgM (Immunoglobulin M) Covid-19 was reactive. Electromyography showed asymmetric motor-sensory axonal polyneuropathy. Cervical MRI (Magnetic resonance imaging) revealed a longitudinally extensive cervical transverse myelitis. Immunoglobulin for five days improved arms strength. Methylprednisolone for five days was started without improvement. He also underwent plasmapheresis with improvement. Blood tests and cerebrospinal fluid were performed, excluding autoimmune diseases, other infections, and neuromyelitis optica. He returned walking unassisted after 60 days with mild hypoesthesia in his left foot and mild urinary retention. Previous reports with similar and worse outcomes were reported after the Covid-19 vaccine, after Covid-19, and other viral infections^{1,2}. Our report presents one of the earliest cases described after vaccination; however, it has already been registered at a similar time³. Such cases usually occur with extended latency periods, probably by SARS-CoV-2 antigens in the Covid-19 vaccine or its chimpanzee adenovirus adjuvant. These antigens could cause myelitis by immune mechanisms².

Neuroimunologia

TL 1105950

ANTI-HU SYNDROME WITH SIMULTANEOUS MOTOR, SENSITIVE AND AUTONOMIC NEURONOPATHY: A CASE PRESENTATION

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Case Presentation: A 67 year-old male with a medical history of prostate cancer, with no evidence of active disease in the last year, started numbness in the right leg and left hand in May 2019, which progressed to paresthesias and paresis in the four limbs (asymmetrical, worse in MSE), reporting difficulty picking up objects, writing and walking. In July 2020, he began to present episodes of seizures reported as a cephalic version, automatism and unmotivated laughter. Neurological examination showed diffuse paresthesia and areflexia with diminished vibration and proprioception sensitivity in addition to sensory ataxia. Also, prominent and asymmetric weakness was evidenced on the left hand, with signs of muscular atrophy and grade two strength. Pupils were symmetric with no photomotor response. Nerve conduction study and electromyography showed absence of diffuse sensitive responses and chronic motor commitment of left C8-T1 roots, suggesting motor root or anterior horn lesion. Brain MRI was normal, and spine axial T2-weighted sequences showed hyperintense lesions in the dorsal columns, compatible with sensory ganglionopathy. Further investigation revealed positive serum anti-Hu antibodies. Neoplastic screening with chest and abdomen CT and whole-body PET-CT were negative. The patient was treated with intravenous methylprednisolone 1g/d per five days during three consecutive months, intravenous cyclophosphamide for three months, and five plasmapheresis sessions for each month during two consecutive months, but showed no improvement. In the follow-up, patient presented constipation and PET-CT showed important gastroparesis. New neoplasia was not identified until two years after the first symptoms. **Discussion:** Anti-Hu-syndrome occurs when there is a dysfunction of the peripheral, autonomic and central nervous systems caused by an immune disorder. Small cell lung cancer is found in more than 90% of cancer. Anti-Hu syndrome may present with neuropathy, ganglionopathy, limbic encephalitis, brainstem encephalopathy, opsoclonus-myoclonus, paraneoplastic cerebellar degeneration or myelopathy. We report a case of anti-Hu syndrome presenting simultaneous evidence of motor, sensitive and autonomic neuropathy, with no neoplasia identified. Motor commitment was prominent with extension of the lesion to anterior horn. **Final Comments:** Anti-Hu antibodies should be tested in patients with simultaneous resensation of motor, sensitive and autonomic neuronopathy.

Neuroimunologia

TL 1105951

POST COVID-19 STEROID-RESPONSIVE ENCEPHALOPATHY ASSOCIATED WITH AUTOIMMUNE THYROIDITIS (SREAT) PRESENTING AS A NEW ONSET REFRACTORY STATUS EPILEPTICUS (NORSE) RESPONSIVE TO TREATMENT

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Case Presentation: Neurological symptoms after Covid-19 are common, as seizures.^{1,2} In addition, refractory status epilepticus has also been described after Covid-19.³ This article aims to describe a patient with steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT) after Covid-19, presenting as a new onset refractory status epilepticus (NORSE), with good response therapy. **Discussion:** Our patient presented with NORSE.⁷ The most common etiologies of NORSE were ruled out, such as structural damage, vascular, infectious, toxic and metabolic causes. Markers for autoimmune encephalitis were negative. However, thyroid autoantibodies raised the hypothesis of SREAT. The diagnosis of SREAT is currently a diagnosis of exclusion. The clinical picture usually involves encephalopathy, cognitive impairment, tremor, and other symptoms (such as seizures).⁵ The patient in the present case met criteria for this disease.⁸ The treatment of this condition involves immunotherapy with methylprednisolone and/or immunoglobulin.⁵ The relationship between Covid-19 and the subsequent appearance of SRAT may be due to the autoimmunity triggered by SARS-CoV-2, as already described in other conditions (referencia NMDA covid). The presentation of SREAT with NORSE is rare and, after Covid-19, it is unprecedented.⁹ The treatment of NORSE, in turn, involves the control of seizures, with anticonvulsants and anesthetic drugs.¹⁰ In addition, immunotherapy should always be used, attempted when autoimmune causes are part of the diagnostic hypothesis.⁸ Although other causes were investigated and excluded, the relationship between Covid-19 and SRAT may have been a fortuitous finding. The present case showed improvement after the attempt with the addition of ketamine to propofol and midazolam aiming at suppression burst with inter-surge intervals longer than 5 seconds, for 48 hours and after immunotherapy with IVIG and MPIV. **Final Comments:** Our case illustrates an unprecedented case of NORSE by SREAT after Covid-19 with excellent therapeutic response.

Neuroimunologia

TL 1106025

ANTI-MOG DISEASE A DIFFERENTIAL DIAGNOSIS TO BE CONSIDERED

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Case Presentation: Patient G.A.M.C.R, 13-year-old, black woman. In December 2018 suffered a sudden event of amaurosis in the right eye followed by mild spontaneous improvement. Visual acuity 20-800 in the right eye and 20-20 in the left eye. With papilla pallor at direct funduscopy. Under the hypothesis of acute inflammatory optic neuritis, cerebrospinal fluid collection and magnetic resonance of the brain were performed. Received treatment with methylprednisolone 1g/day for 5 days. A extensive propaedeutics for etiology of optic neuritis revealed MRI without alterations with absence of monoclonal peaks or oligoclonal bands in cerebrospinal fluid. Magnetic resonance imaging without criteria for multiple sclerosis. Negative anti-AQP4. Under the chance of isolated optic neuritis or spectrum of optic neuromyelitis, after treatment with methylprednisolone the visual acuity was for 20-200 in the right eye, remaining 20-20 in the left eye. Another test for anti-mog and anti-AQP4 was tried at the moment. However, on 06-01-22, the patient seeks an emergency complaining of sudden low visual acuity, pain on eye movement with 20-800 in the right eye again and 20-20 in the left eye. But complaining of loss of definition for colors in both eyes. Direct funduscopy with bilateral pallor. Made new therapy with methylprednisolone 1g a day for 5 days. The patient brings the result of the new test for anti-AQP4 with non-reagent value but brings the result of the positive anti-mog test. After pulse therapy there is improvement for color vision and visual acuity returns to the previous level with visual acuity 20-20 in the left eye and 20-200 in the right eye. Star using azathioprine with gradual dose escalation reaching 150mg-day and is maintained prednisone 10 mg day. Since then, the disease is kept under control. **Discussion:** In this case, we discuss the importance of considering differential diagnostics beyond the spectrum of optic neuromyelitis in cases of severe optic neuritis. In this sense, anti-mog disease has proved very relevant, especially in cases where there is severe optic neuritis but with specific examination of negative anti-AQP4. **Final Comments:** Severe optic neuritis can be highly disabling and elucidation aiming at the best treatment of the conditions involves knowing the diagnostic hypotheses, considering them and well testing them. Anti-mog disease should be remembered in this context.

Neuroimunologia

TL 1106080

ACUTE NECROTIZING ENCEPHALOPATHY: CASE PRESENTATION

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Case Presentation: We describe a case of acute necrotizing encephalopathy in a 3-year-old boy. His father has been previously diagnosed with encephalitis at age four. The patient case started with epileptic seizures time-locked to several viral infections. The first seizure happened at 1 year, two days after the onset of fever. He followed up with several Hospitalizations for convulsive crises, always preceded by fever. He was diagnosed with febrile seizures. At the age of 2 years and 6 months, he was affected by COVID 19, presenting a status epilepticus concomitant with the infection, requiring intubation. Treated with methylprednisolone for five days. Brain MRI with ventricular dilatation, signs of cytotoxic edema and cortical necrosis. Ventriculoperitoneal shunt was performed, and immunoglobulin was sequentially administered for 5 days. Sequencing of the exome was performed showing a probably pathogenic variant in the RANBP2, suggesting the diagnosis of Infection-Induced Acute Encephalopathy Susceptibility (IIA1/ANE1). It was then decided to perform treatment with Tocilizumab, subsequently better controlling symptoms, culminating in extubation 4 days after the infusion. The patient is currently 3 years and 9 months old, remains stable, without new infections, relapsing intermittently with drop attacks. There is no motor deficit, only cognitive. **Discussion:** The disease hallmark is the presence of a viral infection related to seizures, evidencing multiple symmetrical lesions due to edema and necrosis in the thalamus, cerebellum, brainstem and white matter. The etiology arises from the uncontrolled release of cytokines during a febrile illness. There are a variety of viroses that has been already identified and related to ANE, Influenza A is the most commonly related disease trigger. Most cases are sporadic, but familial cases are associated with mutation in the gene RANBP2. The clinical presentation is characterized by three stages: prodromal (febrile viral infection), acute (encephalopathy, on average 3 days after the beginning of the prodromal phase) and recovery. After recovery, brain lesions may resolve completely or result in atrophy, or hemosiderin deposition. **Final Comments:** The patient had the classic clinical presentation, confirmed with the exome, despite the absence of the most frequent radiological alteration. The purpose of this report is to draw attention to potentially serious complications in patients with these comorbidities.

Neuroimunologia

TL 1106090

FLAIR-HYPERINTENSE LESIONS IN ANTI-MOG-ASSOCIATED ENCEPHALITIS WITH SEIZURES (FLAMES): A MISUNDERSTOOD ENTITY WITH DIAGNOSTIC LIMITATIONS

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Case Presentation: A 73-year-old male was admitted in a tertiary Hospital due to progressive weakness in the right arm and later in the right leg, over three days, along with dysarthria and temporospatial disorientation. Was initially approached as an acute ischemic stroke with associated dementia, and, after Hospital discharge, about one week from the start of the symptoms, he started to present focal aware clonic seizures in the right side. At admission, he was globally aphasic, with frontal release signs and right spastic hemiplegia. Brain magnetic resonance imaging (MRI) did not reveal ischemic stroke signs, but retrospective analysis of previous MRI showed left parieto-occipital cortical and bilateral insula hyperintensities on fluid attenuated inversion recovery (FLAIR) with cortical post-gadolinium enhancement, with notable progression. CSF analysis did not show inflammatory markers and Multiplex PCR was negative. Due to the symptoms progression and radiologic features, an hypothesis of FLAIR hyperintense cortical lesions in MOG associated Encephalitis with Seizures (FLAMES) was made. The patient was then treated with high dose intravenous steroids and albumin plasmapheresis, with an excellent response, being discharged after two weeks speaking and walking without help. After one month, the patient is taking azathioprine and on corticosteroids tapering, still able to walk independently and speak with only a small latency of speech response. Myelin oligodendrocyte glycoprotein (MOG) antibody associated disease (MOGAD) is an inflammatory affliction of the central nervous system (CNS), and similar cases were described with FLAMES (4). Clinical features include seizures, headache, fever and cortical symptoms referable to the FLAMES location, and 95% reported cases at least two of these four findings (5). Although initially described as a unilateral cortical encephalitis, bilateral cortical involvement and possible meningeal inflammation could indicate a broader disease spectrum. The syndrome is highly steroid-responsive (1). FLAMES is a rare condition that remains underdiagnosed. Although, we still have a lot of limitations to diagnoses, considering the unavailability and high prices of the tests. Recognition of this distinct clinico-radiographic syndrome may facilitate prompt diagnosis and treatment. Therefore, the reported cases suggest excellent response to adequate treatment.

Neuroimunologia

TL 1106104

DYSARTHRIA-CLUMSY HAND SYNDROME: AN ATYPICAL PRESENTATION OF MULTIPLE SCLEROSIS RELAPSE

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Case Presentation: A 39-years-old woman with a history of hypothyroidism presented in August 2020 with progressive slurred speech. The following day she noticed weakness in her right hand and was evaluated at the emergency room. She was three weeks pregnant according to obstetric ultrasound. Neurological examination revealed mild weakness of the right upper limb and discrete dysarthria. The remainder of the examination was normal. Brain magnetic resonance imaging (MRI) revealed a tumefactive lesion on the right corona radiata and rare foci of T2/FLAIR high signal lesions in the white matter, suggesting a demyelinating disease. Cerebrospinal fluid was normal except for the presence of type 2 oligoclonal bands. Due to her pregnancy and improvement of symptoms, she did not receive any treatment initially, and she had an incomplete recovery. Six months after delivery, the MRI was repeated and showed a new mass lesion involving the left internal capsule. She decided to breastfeed, and a DMD was initiated a year later. **Discussion:** Multiple Sclerosis (MS) is the most common demyelinating disease of the central nervous system, and its typical presentations include optic neuritis, internuclear ophthalmoplegia, and myelitis. Stroke-like presentation of MS is uncommon, with a few cases described in the literature. Dysarthria-clumsy hand syndrome (DCHS) is typically a lacunar syndrome characterized by unilateral pyramidal signs and speech disturbance. The most frequently described sites of lesions are the internal capsule, corona radiata and pons. Our patient also had a tumefactive demyelinating lesion (TDL), a rare form of MS relapse. Typical pathological findings of TDLs include abundant foamy macrophages containing myelin debris, reactive astrogliosis, relative axonal preservation, and variable perivascular and parenchymal lymphocytic inflammation. The TDL could be explained by a procoagulant state that represents an increased innate immune response, probably needful to compensate for reduced cell-mediated immunity inherent to the pregnant state. **Final Comments:** There is no description in literature of MS relapse causing DCHS, but the lesion's location seen on MRI and the patient's symptoms are compatible with the syndrome. This atypical case shows that MS can present in various forms. In this context, the systematic approach is fundamental to diagnosing and expanding the knowledge about this disease.

Neuroimunologia

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NEUROLOGICAL COMPLICATIONS IN LYMPHOHEMATOPOIETIC LINEAGE NEOPLASMS: REPORTS ON CRANIAL NERVE INFILTRATION AND ITS DIAGNOSTIC CHALLENGE

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Case Presentation: CASE 1: LPS, male, 46y. Presented with pain in the lumbar region for 6 months with irradiation to the left lower limb and bilateral inguinal lymphadenopathy. Laboratorial investigation showed leukocytosis with left shift. Immunophenotyping (IPT) suggestive of chronic myeloproliferative disease. During Hospitalization, developed Peripheral Facial Palsy (PFP), being managed as Bell's Palsy (BP) while CNS infiltration was investigated. CASE 2: ACDS, male, 70y. Presented with melena, enterorrhagia, hepatomegaly and mobile painless lymphadenopathy. IPT suggestive of chronic lymphoproliferative B cells disease. During Hospitalization, evolved with ptosis of the right eyelid, hypotropia and esophoria. **Discussion:** We report the investigation of two patients diagnosed with neoplasm of the lymphohematopoietic lineage that developed cranial nerve (CN) palsy. Case 1 reports a patient with chronic myeloid leukemia that develops PFP, a rare finding. Thus, BP was raised as a possible diagnosis. Contrast-enhanced MRI of the brain showed thickening of the 7th CN suggestive of leptomeningeal metastasis, a finding also seen in BP. Patient was transferred without being able to perform specific LCR analysis. Case 2 presents a patient with lymphoma that develops an incomplete 3rd CN palsy. Contrast-enhanced MRI of the brain showed no significant changes. CSF analysis did not present with neoplastic cells, despite hypoglycorrhachia and increased protein. Reports suggest that such findings do not rule out the cranial nerve involvement. **Final Comments:** Neurological involvement can occur during the natural history of hematological neoplasms, especially in leukemia and lymphoma. Despite several clinical presentations, cranial nerve impairment is the most common and more easily noticeable of the extra-hematological manifestations. Even so, its diagnosis is not simple, as diagnostic confirmation requires invasive and difficult-to-access complementary investigations, which makes the confirmation a challenge. Therefore, even though CN infiltration by hematological neoplasms is rare, if a CN impairment appears in a patient with positive neoplastic history, it is worth investigating. The investigation should include imaging and cerebrospinal fluid study. Sometimes the diagnosis is presumptive and it is plausible to believe that the challenge of confirming the diagnosis is the probable cause of its low epidemiology.

Neuroimunologia

TL 1106140

ANTI-GABA-AR POSITIVE ANTIBODY ENCEPHALITIS AS A RECURRING ENCEPHALITIS ASSOCIATED WITH THYMOMA

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Case Presentation: A 53-year-old otherwise healthy female presented with a six-month history of cognitive and behavioral symptoms. Six months before, she presented difficulty in planning and performing multiple-step household tasks and progressive apathy. After two months, she manifested persecutory and religious delirious thoughts, sexual disinhibition and irritability. After another two months, she presented recurring focal onset seizures, starting with a speech disturbance and quickly evolving to a generalized tonic-clonic episode. Neurological exam revealed a frontal syndrome characterized by apathy, social disinhibition and executive cognitive impairment. Brain magnetic resonance imaging showed multifocal, non-enhancing, cortico subcortical T2/FLAIR signal abnormalities predominantly on frontal and temporal lobes. Cerebrospinal fluid study demonstrated the presence of oligoclonal bands despite normal cell and protein counts. A chest computed tomography revealed a thymoma that was surgically removed. Autoantibody panel confirmed the diagnosis of anti-GABA-A receptor encephalitis. The patient was treated with pulse therapy using methylprednisolone and plasmapheresis followed by cyclophosphamide, obtaining good seizure control. Six months later the disease relapsed presenting this time also with hemiparesis. Rituximab was administered, seizure control was once more obtained but the patient remains to this day with important behavioral symptoms. **Discussion:** Anti-GABA-Ar encephalitis can be one of the thymoma associated paraneoplastic encephalitis. Seizures, cognitive impairment, decreased level of consciousness, altered behavior and movement disorders are the most frequent symptoms. Distinctive multifocal cortical-subcortical T2/FLAIR MR abnormalities provide an important clue to the diagnosis, since they rarely occur in other autoimmune encephalitis. The disorder is severe but most patients respond to treatment. However, up to a third of patients may relapse and recurrent disease may occur even after surgical removal of the thymoma. **Final Comments:** Anti-GABA-Ar positive encephalitis should be considered in patients with rapidly progressive cognitive decline, particularly in the setting of thymoma. Relapse may occur even after the removal of the associated tumor.

Neuroimunologia

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PRIMARILY PROGRESSIVE MULTIPLE SCLEROSIS WITH LATE PRESENTATION

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Case Presentation: Male patient, 67 years old, without comorbidities, five years ago he started a postural imbalance, weakness of the lower limbs and falls. Four months ago, he presented: mental confusion; apathy; attention alteration; excessive sleepiness; depressed mood, dysphagia; major dysarthria; urinary incontinence and became bedridden. At Hospital admission, on neurological examination, he was disoriented and uncooperative, dysarthric, with emotional lability, tetrasegmental hypotrophy, elastic hypertonia and hyperreflexia with signs of pyramidal release, reduced strength, worse in lower limbs. In the imaging exams, the patient presented characteristic lesions of multiple sclerosis, comparing magnetic resonance imaging of the brain performed in a period of one year, the disease activity is visible, demonstrated by the contrast enhancement in some of the lesion areas. Multiple lesions were defined in the periventricular white matter, subcortical, corona radiata, in the left portion of the pons and in the right cerebral peduncle, most with a periventricular aspect (Dawson's fingers), some with contrast enhancement. In the cerebral spinal fluid: leukocytes: 2, proteins: 100; albumin: 56.2 / gamma: 15.7 / total proteins: 118 / presence of suggestive band in the gamma region; IgG: 17.2 / IgM: 163; Serum protein immunofixation: IgG: 907 / IgM: 159. No monoclonal bands were detected. Serum albumin 3.67g/dL CSF IgG index was 1.24. The investigation of neoplasms was performed, and did not show lesions. The patient and the family were oriented about the prognosis of the disease and was improved the clinical support. Multiple sclerosis (MS) is the most common immune-mediated inflammatory demyelinating disease of the central nervous system, the mean age of MS onset ranges from 28 to 31 years; clinically it becomes apparent between the ages of 15 to 45. Clinical onset rarely occurs in the first years of life or after the seventh decade. In only about 5% of cases, MS is diagnosed after age 50. There is growing concern of a more progressive form of MS in late-onset cases. Unlike adult-onset MS, the first presentation of the late form is usually motor dysfunction. This case presentation exemplifies what we see in the literature, although it is uncommon, late presentation, after 50 years of age, occurs, and has some characteristics such as the onset with a motor condition. It is important to emphasize unusual presentations so that we do not fail to recognize them and manage to treat them earlier.

Neuroimunologia

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AMYLOID-? RELATED ANGIITIS AS A NEUROCOVID MANIFESTATION?

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Case Presentation: A previously independent 75-years-old woman was admitted in April of 2021 to a neurological department in the city of Fortaleza, Brazil. She presented a three week report of progressive speech impairment, difficult interaction and comprehension associated with right hemiparesis and sphincter incontinence. She had an unremarkable medical history. Upon neurological examination, it was additionally observed mutism, hyperreflexia, clonus and babinski sign. On further work up, a magnetic resonance imaging (MRI) exhibited numerous microbleeds on SWI surrounded by vasogenic edema on T2 sequences. RT-PCR test for COVID-19 was revealed to be positive. She had been experiencing mild dyspnea one week prior to first evaluation. Cerebrospinal Fluid Evaluation revealed normal opening pressure, 40 RBC/mm, 2 WBC/mm, Protein level of 126 mg/dL, Glucose 80 mg/dL. It was negative pathogens. After initial approach the diagnosis was considered to be probably ABRA (Amyloid-? Related Angiitis), she was submitted to therapy with a 5 day course of methylprednisolone evolving with progressive clinical improvement. Was discharged in a few days and was followed at the outpatient care. She remained at follow up with motor aphasia and a slight upper right limb weakness with relatively independence requiring little assistance to daily routine. **Discussion:** Neurological symptoms associated with COVID-19 have been described with manifold aetiologies, such as ischaemic stroke, hemorrhagic encephalopathy and others. The largest published autopsy series of COVID-19 neuropathology reported microthrombi and acute hemorrhagic infarction in a significant number of patients, while another more recent study found evidence of lymphocytic encephalitis and meningitis. Endotheliitis of the brain and extraneural organs has been shown in Sars-CoV-2 infected patients. Amyloid-beta related angiitis is a predominantly granulomatous angio-destructive inflammatory mediated disease affecting leptomeningeal and cortical vessels characterized by meningeal lymphocytosis and abundant amyloid-beta deposition within the vessel walls. The pathophysiology of ABRA is not fully understood. One theory suggests an immunologic response toward amyloid-beta, resulting in leptomeningeal and parenchymal inflammation. Reports of rare cases such as this one of amyloid-? related Angiitis associated with COVID infection, may help to clarify roles of immune response, its triggers and incite further questions in the scientific community.

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SUSAC SYNDROME: DO WE NEED THE TRIAD?

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Case Presentation: A 35-year-old male presented with rapid onset headache and visual disturbance in May 2022. The symptoms evolved in two days with worse in the intensity of the headache, associated with vomiting, dizziness and imbalance. Physical examination revealed disorientation, confusion, amnesia and executive dysfunction. The Mini mental state examination (MMSE) score was 14. Cerebrospinal fluid analysis showed lymphocytic pleocytosis, hyperproteinorrachia and normal glucose levels. Brain MRI disclosed supratentorial and infratentorial white matter lesions with high signal on T2-weighted imaging. Snowball lesions in the corpus callosum, leptomeningeal and linear contrast enhancement were also observed. Retinal angiography with fluorescein showed peripheral vasculitis and areas of peripheral arterial occlusion in both eyes. Audiometry was normal. After reasonable exclusion of alternative causes, the diagnosis of Susac Syndrome was made. Intravenous methylprednisolone for 5 days was started followed by Prednisone 1mg/kg/day and Azathioprine for maintenance therapy. After the acute treatment, the patient presented marked cognitive improvement in the MMSE (22), semantic fluency and clock drawing-test. **Discussion:** Susac syndrome is a rare pathology, believed to be caused by autoimmune microvessels vasculitis of the brain, retina, and inner ear. These occlusions lead to the clinical triad: encephalopathy, visual disorders and hearing deficits. The occurrence of two features of the classical triad suggests the diagnosis of incomplete susac. In a review study with 304 patients, only 13% presented the triad at the clinical onset. In the disease course, in up to 21 weeks, 85% presented the complete triad. **Final Comments:** Despite being a rare disease, a better understanding of the syndrome is needed. Neurologist should be aware that basing the diagnosis solely on the presence of complete triad may not be appropriate on the clinical onset of this disease. Once excluded alternative diagnosis and in the presence of suggestive radiological features of Susac, immunosuppression treatment should be immediately considered in patients with incomplete syndrome in order to avoid further disability.

Neuroimunologia

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GUILLAIN-BARRÉ SYNDROME AFTER COVID-19 VACCINE: CASE PRESENTATION AND LITERATURE REVIEW

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Case Presentation: We report the case of a male patient, 50 years old, who started an acute condition of sensory and motor loss of progressive ascending evolution, associated with low back pain, global areflexia, evolving with peripheral facial palsy on the right side, 10 days after the administration of the first dose of the AstraZeneca laboratory vaccine against COVID-19. The patient had no infections or other vaccinations prior to the neurological condition. Clinical examination and complementary investigation findings, including cerebrospinal fluid (CSF) and electroneuromyography (ENMG), were consistent with the diagnosis of Guillain-Barré syndrome (GBS). GBS is an acquired acute inflammatory demyelinating polyradiculoneuropathy that commonly occurs after respiratory or gastrointestinal viral infections, immunizations or surgical interventions, which precede neurological symptoms by 5 days to 4 weeks. Its etiology is not fully understood, but there is evidence that it is mediated by autoimmune mechanisms with self-tolerance suppression by molecular mimicry. The symptoms of this syndrome are wide-ranging, present in a wide variety of ways, and its diagnosis is made based on clinical suspicion, associated with neurophysiological confirmation and CSF examination. Treatment consists of immunotherapy with immunoglobulin or plasma exchange, supportive care and associated clinical complications. The case presented and the literature review show that there is a correlation between vaccines against SARS-Cov-2 and the occurrence of Guillain-Barré Syndrome.

Neuroimunologia

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BICKERSTAFF BRAINSTEM ENCEPHALITIS A CASE PRESENTATION

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Case Presentation: Bickerstaff brainstem encephalitis (BBE) is a rare post-infectious neurological disease defined by the triad of ophthalmoplegia, ataxia and decreased consciousness. It is considered to be a variant of Miller Fisher syndrome. **Discussion:** We present the case of the patient T.A.O., 40 years old, female, admitted to the aforementioned nosocomium on 11-24-21 with flaccid quadriplegia and sensorium lowering. Aphasia. Currently, patient with no history of previous illness. But she had a history of using oral contraceptives and smoking. Initially, under suspicion of a cerebrovascular event, she was submitted to imaging tests and, due to acute respiratory failure, she was intubated and kept under intensive care. After brain and cervical imaging exams did not show signs of vascular disease, blood tests were collected with serology for virus as well as cerebrospinal fluid collection, this one, without testing for viral serology. After a detailed interview with the family members it was reported, 15 days before the beginning of the events, episode of diarrhea with malaise lasting 24h. This fact reinforced the hypothesis of post-infectious encephalitis. In this intercourse the patient was receiving anti-viral following treatment with human immunoglobulin for 5 days. Physical examination revealed. Facial diplegia, complete internal and external ophthalmoplegia. Flaccid tetraplegia. Chart, which according to medical literature, can be confused with brain death. In fact, electroencephalogram was performed that showed wakefulness activity, without pathological graphoelements. In fact, electroencephalogram was performed that showed wakefulness activity, without pathological graphoelements. The results of viral serologies showed positivity for acute reaction to cytomegalovirus. Reinforcing the diagnosis. Slowly, after treatment with immunoglobulin, physiotherapeutic, nutritional and pressure wound prevention care, the patient recovered from diplegia. Improvement of ophthalmoplegia, flaccid tetraplegia. With slow improvement of ventilatory support. Six months after the beginning of the condition and treatment, the patient maintains tetraparesis with improvement of aphasia, without the need for ventilatory support, improvement of diplegia gradually recovering to basic activities such as feeding and verbal communication. **Final Comments:** Bickerstaff brainstem encephalitis may even be confused with brain death. This requires a lot of holistic care.

Neuroimunologia

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MYELIN OLIGODENDROCYTE GLYCOPROTEIN ANTIBODY-ASSOCIATED DISEASE (MOGAD) AFTER RECENT COVID-19 INFECTION AND COVID-19 VACCINATION: A CASE PRESENTATION

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Case Presentation: Female patient, 53 years old, previously diabetic, hypertensive, and dyslipidemic; with a clinical picture of gradual visual loss installation of central predominance in the left eye (LE), pain at ocular mobilization in the LE, and cephalaea. On physical examination, she presented visual acuity (VA) of counting fingers in the LE, presence of a relative afferent pupillary defect in the LE, papilledema in the LE on dilated eye examination, and remaining neurological examination without alterations. Symptoms began 22 days after the first dose of the COVID-19 vaccine (AstraZeneca) and 15 days after COVID-19 infection with bilateral mild-moderate lung impairment. She was admitted to the Neurology Department, with laboratory tests presenting serologies for COVID-19 IgM and IgG Reagents, increased PCR, other general tests without alterations, infectious serologies without acute conditions, negative rheumatological tests, and liquor with a normal result. Magnetic resonance imaging (MRI) of the orbits shows signs of optic neuritis on the left, with paramagnetic contrast enhancement, without extension for optic chiasm. The MRI of the brain, cervical and thoracic spine was performed without any relevant changes. Opted for methylprednisolone 1 gram/day for 5 days, and the patient evolved with partial recovery of VA in the LE (20/200). Due to atypical optic neuritis, serum tests of antiaquaporin 4 antibody with negative result and anti-MOG antibody with positive result were requested. Was performed a visual evoked potential evidencing demyelinating process in the pre-chiasmatic visual pathways of the LE. In view of the findings, a diagnosis of Myelin Oligodendrocyte Glycoprotein Antibody-associated Disease (MOGAD) was made, and subsequently, azathioprine was prescribed associated with corticosteroid therapy, without new neurological events and with progressive improvement of the visual picture. **Final Comments:** The aforementioned patient presented a picture of optic neuritis in the LE with diagnostic possibilities of post-vaccination etiology, post-infectious COVID-19 or first event of demyelinating disease. The visual evoked potential with signs of demyelinating optic neuritis and the positivity of the anti-MOG antibody contributed to the diagnosis of MOGAD and probably the previous infection and/or previous vaccination worked as triggers for the manifestation of the autoimmune disease.

Neuroimunologia

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RELAPSE OF ANTI-NMDAR ENCEPHALITIS IN COVID-19 INFECTION: A CASE PRESENTATION

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Case Presentation: A 19-year-old male presented with visual and auditory hallucinations, insomnia, confusion, and mood oscillations, as well as three episodes of fever in the week prior. A day before Hospital entry, he had four episodes of stereotyped movements on the left side of his body, as well as sialorrhea, sweating and visual hallucinations, lasting 9 minutes. At examination, he was afebrile, agitated, disoriented, without focal deficits or meningeal signs. He had been diagnosed with anti-NMDAR encephalitis in our service (confirmed by serum antibody dosage) in 2014, when he exhibited severe agitation, autonomic signs, chorea, and status epilepticus, having undergone treatment with two courses of methylprednisolone 1g, one course of immunoglobulin 1g, followed by 1g of cyclophosphamide, having remitted completely of the symptoms at discharge. He was on carbamazepine 200mg 8/8h from 2014 to 2020, having suspended the medication himself, with no epileptic events until the day prior to admission. He underwent diagnostic testing with unremarkable head CT, MRI, and lumbar puncture. His SARS-CoV-2 PCR test was positive, and sequential EEGs showed diffuse slowing, without epileptic activity. Considering an anti-NMDAR encephalitis relapse triggered by COVID-19 infection, he was treated with two courses of methylprednisolone 1g for five days, two days of cyclophosphamide (1g/d), and one course of immunoglobulin (0,4mg/kg/d) for five days and kept on 30mg/d of prednisolone after discharge, with progressive tapering. At the time of discharge, the patient had mild executive dysfunction and echolalia, and was oriented to return for 6 monthly courses of cyclophosphamide. **Discussion:** COVID-19 has been described as trigger to autoimmune or autoinflammatory conditions; anti-NMDAR encephalitis is a multistage acute autoimmune disease, progressing from psychiatric symptoms to memory disturbances, dyskinesias, seizures, and catatonia. Several cases of anti-NMDAR encephalitis associated to the infection by the novel coronavirus have been reported, many confirmed by SARS-CoV2 PCR and anti-NMDAR dosage in CSF/Serum. Given our patient response to treatment and unremarkable laboratory and image, we believe this might be the case. **Final Comments:** The coronavirus pandemic gave rise to neurological manifestations, including triggering of Anti-NMDAR encephalitis or their relapses, which should be considered in the differential diagnosis of patients in this context.

Neuroimunologia

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VOGT-KOYANAGI-HARADA SYNDROME, AN IMPORTANT DIFERENCIAL DIAGNOSIS

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Case Presentation: Female, 44 years old attended at Neurology service with 15 days of bilateral, throbbing headache, intensity 8/10, associated with nausea, vomiting, hearing loss (HL), low visual acuity (VA) and pain on eye movement in the left eye (LE). After 7 days, she evolved with low VA in the right eye (RE), with loss of color saturation and presence of central stochoma. She reported a history of previous, mild, perimenstrual headache and no ophthalmologic complaints. Denied other symptoms, trauma, use of medications, allergies or Hospitalizations. On examination: preserved eye movements. VA of the RE: counting fingers (CF) 50 cm. VA of LE: CF 30 cm. Reduced photomotor reflex bilaterally, and biomicroscopy with thin keratic precipitates and posterior uveitis (2+/4). Bilateral optic disc edema, with vascular tortuosity and bilateral serous retinal detachment. Further investigation showed: lumbar puncture with opening pressure 23 cmH₂O (and 1 leukocyte, non-reactive VDRL, total proteins 25); Normal magnetic resonance imaging (MRI); Mixed HL audiometry and orbital magnetic resonance imaging with signs of chorioretinitis. In addition to negative serological tests and negative antiaquaporin 4 research. **Discussion:** Vogt-Koyanagi-Harada Syndrome (VKHS) is a systemic autoimmune disease that mainly affects melanocyte-rich structures such as the retina, inner ear and central nervous system. It contains 4 distinct clinical stages: prodromal, uveitic, chronic and recurrent. The previously healthy patient sought care due to unilateral eye pain, which developed bilaterally after 7 days, associated with other ocular symptoms and HL. MRI rule out tumors or signs of increased intracranial pressure and demyelinating diseases. Additionally, acute worsen of HL, along with serous retinal detachments bilaterally and posterior uveitis. And a negative viral serology and anti-aquaporin 4 narrows criteria for VKHS. Treatment with 5 days of 1g methylprednisolone intravenous was made with significant outcome, showed by improvement of VA and retinal detachments at OCT, along with no more headache episodes. **Final Comments:** As a rare syndrome, the diagnosis of VKHS requires a complete anamnesis and a good physical examination, associated with complementary tests, such as OCT and serological tests, to aid diagnosis.

Neuroimunologia

TL 1106378

PULMONARY EMBOLISM IN A PATIENT WITH MULTIPLE SCLEROSIS DURING THE TREATMENT WITH TERIFLUNOMIDE: A CASE PRESENTATION

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Case Presentation: A 44-year-old woman, former smoker, was previously diagnosed with relapsing-remitting multiple sclerosis (RRMS) and had been receiving interferon beta-1a over the past five years, with no signs of disease activity since the beginning of treatment. Due to the development of flu-like symptoms and poor adherence to the medication, she was switched to teriflunomide (TFN). After one month of treatment with TFN, the patient reported a new-onset right-sided lumbar pain and dyspnea. At the occasion, chest CT angiography showed signs of embolism in the inferior segmental pulmonary arteries, as well as pulmonary infarction in the right lower lobe. The patient was, then, started on rivaroxaban and presented significant improvement of the symptoms over the following weeks. TFN was discontinued and the patient maintained the initial Expanded Disability Status Scale (EDSS) of 2.5. Echocardiogram showed no signs of pulmonary hypertension. **Discussion:** TFN is an oral immunomodulator and a disease modifying therapy for patients with RRMS. Although liver toxicity is the most frequent concern during the treatment with this medication, scarce reports of pulmonary embolism (PE) are present in the literature. Thrombotic and thromboembolic events have been described in clinical trials evaluating TFN for RRMS, with the group receiving TFN having a slightly, but not significantly higher, incidence compared to the placebo group (1.7% vs. 1%). Here, we report the case of a patient that developed PE approximately one month after the beginning of treatment with TFN. Despite the non-significant higher incidence, given the potential association between the use of TFN and development of PE, clinicians should be aware of this serious condition in the follow-up of this set of patients. **Final Comments:** The case presented demonstrates a rare association between the treatment of RRMS with teriflunomide and the development of pulmonary embolism. Although liver toxicity is the most frequent concern, scarce reports of PE are present in the literature and should be considered as a differential diagnosis when evaluating a patient with new-onset respiratory symptoms during the treatment with TFN.

Neuroimunologia

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NEUROMYELITIS OPTICA SPECTRUM DISORDER (NMOSD) ASSOCIATED WITH PRIMARY ANTIPHOSPHOLIPID SYNDROME: A CASE PRESENTATION

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Case Presentation: A previously healthy woman aged 25 years reported that one and a half years ago, left-sided paresthesia up to her breast evolving to the right side in a few days, followed by painful sensations in both legs, urinary retention, and bilateral weakness limiting her to walk. She was diagnosed with a deep vein thrombosis in the left leg and received oral anticoagulation for six months. She was able to walk after 30 days without treatment. One month later, she presented a sudden visual loss in the right eye with progressive improvement after 15 days. After five months, a mild episode of paresthesia and weakness in both legs, with spontaneous improvement after a few days. On neurologic exam during a new severe episode revealed clinical signs of acute transverse thoracic myelitis with no muscle strength in both legs and anesthesia up to T4 spinal level. Intermittent bladder catheterization was required (high urinary residue). Cranial nerves were normal, including visual function. Spinal cord images showed signs of myelitis affecting C7-T1 and T8-T9 levels with slight contrast enhancement. The cerebrospinal fluid analysis showed mildly elevated cell count and protein level with no signs of infections. Oligoclonal bands were not done. There was a positive aquaporin-4 antibodies test (1:80). Oral anticoagulation was indicated again due to lupus anticoagulant positivity. Visual evoked potential showed signs of visual pathway dysfunction in both eyes. She received five days of intravenous methylprednisolone followed by seven sessions of plasmapheresis. Muscle strength improved to grade 3 in both legs, and she recovered truncal and bladder control. She is currently receiving initial doses of rituximab. **Discussion:** Recurrent transverse myelitis, optic neuropathy, and positive aquaporin-4 antibody indicate NMOSD in this patient. Previous deep venous thrombosis in association with a lupus anticoagulant are features of the antiphospholipid syndrome (APS). The association between NMOSD and primary APS is rare, although autoantibodies related to APS had been often reported in patients with NMOSD. Pathophysiological mechanisms are complex, and both diseases may overlap, affecting the nervous system. Cooccurrence of both conditions increases morbidity and unfavorable outcomes. **Final Comments:** NMOSD is a severe disease, and comorbidity with rheumatological diseases may occur and worsen the prognosis. Further studies are necessary to understand this relationship.

Neuroimunologia

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CASE PRESENTATION BELLY-DANCER SYNDROME IN A PATIENT WITH NEUROMYELITIS OPTICA SPECTRUM DISORDER

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Case Presentation: Female, 46 years old, diagnosed with Neuromyelitis Optica Spectrum Disorder (AQP4 positive) in 2010, with episodes of inflammation characterized by optic neuritis, left hemiparesis and hemiparesis. She was treated with methotrexate, azathioprine and, after a new attack, rituximab was initiated until now. The last episode was in July 2020, characterized by paresthesia below T8 vertebral level. In January 2021, she reported abdominal involuntary movements, radiating to the dorsal region at the thoracolumbar transition. It was interpreted as bilateral internal and external oblique muscle contraction. On electroneuromyography, spontaneous discharges of action potentials of arrhythmic activity and variable frequency was found, suggestive of myokymia at rest in the rectus abdominis muscles and more markedly in the left oblique. Considering the diagnostic hypothesis of Belly-dancer Syndrome, levetiracetam 250mg 12/12h was started, with no satisfactory response either when associated with carbamazepine 200mg 12/12h and gabapentin 300mg 8/8h. Opted for the application of botulinum toxin in the left rectus abdominis and left external oblique with partial improvement of movements. **Discussion:** Belly-Dancer Syndrome or diaphragmatic flutter is a rare condition that involves repetitive and involuntary movements of the abdominal wall muscles. The probable etiologies are drug (levodopa, biperiden, quetiapine), postoperative abdominal surgery, encephalitis, stroke, osmotic demyelination, metabolic changes and trauma. Such changes lead to abnormal excitation of the phrenic nerve, via the central nervous system or along the nerve. Another proposed mechanism is irritation of the diaphragm muscle itself. Diagnosis can be made with ultrasound, thoracic videofluoroscopy, or electromyography. Supposed treatment includes phenytoin, carbamazepine, clonazepam, fluoxetine, gabapentin and valproic acid. The use of botulinum toxin is one of the most recent therapies. In general, treatment attempts have been largely ineffective. **Final Comments:** The present case of Belly-Dancer Syndrome in a patient with Neuromyelitis Optica Spectrum Disorder, may be associated with the last episode inflammation of the disease. The syndrome is little known and that more descriptions of cases and treatments are needed to provide a better evaluation of treatment.

Neuroimunologia

TL 1106429

CENTRAL NERVOUS SYSTEM INVOLVEMENT IN SJÖGREN'S SYNDROME: A DIAGNOSTIC CHALLENGE

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Case Presentation: A 26-year-old woman, previously healthy, started in april 2021 with intermittent pulsatile headache, which months later was associated with progressive sensorineural hearing loss on the right. after 4 months, she presented episodes of rotational vertigo, with subacute onset, ataxia and paresthesia in the left foot and right hand, when she was admitted for investigation. magnetic resonance imaging (mri) of the brain was performed, which revealed multifocal lesions with post-contrast enhancement, affecting the basal nucleus, subcortical white matter, cerebellum and pericerebellar leptomeninges. she had spontaneous improvement of symptoms, being discharged for follow-up investigation as an outpatient. however, after 02 months, she evolved with pain in the topography of the right parotid gland, progressive low visual acuity on the right (acuity: 20/200), with pain and no tears on the same eye. she was hospitalized again, performing a new brain mri, which showed regression of the previous lesions without contrast enhancement. a tear gland biopsy revealed a typical pattern of sjögren's syndrome. she started pulse therapy with methylprednisolone 1 g/day for 5 days and the patient regressed from the visual deficit. **Discussion:** Sjögren's syndrome (ss) is an autoimmune disease that affects the exocrine glands and, when primary, is associated with extraglandular manifestations. neurological impairment occurs in about 25% of cases, with peripheral manifestations being well documented in up to 20% of patients with ss, unlike central alterations, which are rarely described. central involvement usually precedes the classic symptoms of sicca syndrome, in addition to the fact that antibodies (anti-ro/anti-la) are not altered in most cases at the beginning of the disease and the central symptoms can manifest in a relapsing-remitting manner with asymptomatic periods, delaying diagnosis and treatment. **Final Comments:** The case demonstrates that despite the predominantly central pattern of involvement, without symptoms and without serological markers of ss, this patient should be periodically reassessed for the possibility of being ss, due to its diversity of presentations and special evolutionary characteristics, requiring time and high level of suspicion to close the diagnosis and direct the treatment in time to reduce morbidities and control the disease.

Neuroimunologia

TL 1106431

54-YEAR-OLD MALE DIAGNOSED WITH ACUTE DISSEMINATED ENCEPHALOMYELITIS (ADEM) AFTER RECEIVING A DOSE OF ASTRAZENECA COVID-19 VACCINE

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Case Presentation: 54-year-old male farmer, from north-east Brazil, previously healthy, presented with headache and non-measured fever 2 days before neurological symptoms. patient received a dose of astrazeneca vaccine against sars-cov-2, and on the following day, he presented with worsening of headache and left-sided weakness. hours later, confusion, altered level of consciousness and seizures. magnetic resonance imaging (mri) revealed white-matter t2/flair hyperintense plaques and diffusion weighted (dwi) alterations on right occipital, temporal, frontal and parietal lobes, as well as basal ganglia. on a second mri study 3 days later, lesions progressed in size and new lesions were seen on the left side. electroencephalogram displayed focal slowing on both right and left frontal and temporal lobes, focal epileptiform discharges on right frontal lobe and disruption of base activity. during the next days, patient developed acute kidney injury. he also needed ventilatory support and was transferred to an intensive care unit. he was submitted to therapy with antibiotics for bacterial meningitis and acyclovir, as well as methylprednisolone in high doses. a progressive improvement was seen on level of consciousness. there was partial recovery of cognition and motor symptoms patient no longer needed renal replacement therapy. he continues to receive anti-epilepsy medications with complete control of seizures. after treatment, cerebral spinal fluid (csf) analysis showed cell count 1, protein 59.1 and glucose 98. viral and bacterial panel was negative for all pathogens on essay. pcr for sars-cov-2 on csf came back negative. **Discussion:** This patient was diagnosed with adem after receiving a dose of astrazeneca covid-19 vaccine. this case figures among rare possible side effects of a recently released vaccine. however, further observation is required to establish causality, since this patient reported prodromic symptoms even before receiving the dose. other demyelinating cns diseases, such as multiple sclerosis, infectious aetiologies, vasculitis, cerebral venous thrombosis and posterior reversible encephalopathy syndrome should be considered in the differential diagnosis for this case. **Final Comments:** Hopefully, this case presentation will help scientific community in face of urgent and recent findings concerning covid-19 pandemics.

Neuroimunologia

TL 1106448

NEUROLOGICAL SYMPTOMS AS INITIAL MANIFESTATION IN A PATIENT WITH BEHÇET SYNDROME

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Case Presentation: Male patient, 44 years old, no comorbidities. He started dysphagia for 3 months and, then, complete right paresis of III cranial nerve, dysarthria and bilateral dysmetria with gradual worsening. On admission, the presented foud were described, in addition with mouth ulcers refractory of oral treatment. An investigation was carried out with CSF suggestive of an inflammatory process, negative serology and neuroimaging with hypersignal on T2/FLAIR in the brainstem in the rostral region of the medulla, midbrain and left thalamus, besides positive pathergy test and biopsies of mucocutaneous and genital lesions with positive findings. Thus, given the central, chronic and progressive neurological condition, with involvement of the brainstem and important sequelae, treatment with methylprednisolone 1000 mg/day for 3 days was chosen, followed by maintenance with azathioprine at a dose of 2 mg/kg/day. The patient evolved with stagnation of sequelae and did not present new neurological manifestations, remaining with cerebellar symptoms and persistence of paresis of the III cranial nerve. **Discussion:** Behçet's syndrome is a rare disease whose main characteristics are mouth ulcers and systemic manifestations that are mostly due to vasculitis. **Final Comments:** Although rare, Behçet's syndrome should be included in the differential diagnosis of patients with progressive cerebellar syndrome, associated with the absence of demyelinating or neoplastic lesions, with negative infectious screening and CSF suggestive of an inflammatory process. Active research for dermatological changes in the patient's clinical history and physical examination should be carried out, as the delay in treatment can lead to the irreversibility of neurological sequelae.

Neuroimunologia

TL 1106449

A NEUROMYELITIS OPTICA CASE FOLLOWED FIRST-DOSE ASTRAZENECA COVID-19 VACCINATION

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Case Presentation: R.S, female, 64 years old, with systemic arterial hypertension, diabetes mellitus, hypothyroidism and smoker. First dose of Astrazeneca vaccine performed on 05/05/2021. On 05/23/2021, she presented with bladder retention associated with paraparesis and hypoesthesia of the lower limbs. On 05/28/2021 was Hospitalized with a hypothesis of Transverse Myelitis, performed pulse therapy with Methylprednisolone 1000mg for 5 days. Data: Oligoclonal Banding (presence of identical IgG bands in CSF and serum), CSF (Cell 281 [98% lymphocytes], pt 94, glyc 59), negative anti aquaporin, non-reactive ADA, serology negative (syphilis; toxoplasmosis; varicella zoster; herpes simplex and CMV). On 07/2021 performed MRI of the brain and total spine, which showed longitudinally extensive myelitis without gadolinium enhancement, without evidence of involvement of optic nerves, CSF (normal range) and serology (without changes). Hospital discharge with neurological examination presenting no visual deficits and paraparesis and hypoesthesia with T10 level. On 10/2021 she presented with optic neuritis in the left eye which corroborates the diagnosis of Neuromyelitis optica. **Discussion:** Neuromyelitis optica (NMO) and neuromyelitis optica spectrum disorders (NMOSD) are inflammatory disorders of the central nervous system characterized by severe immune-mediated demyelination associated with predominant axonal injury of the optic nerves and spinal cord (may arise in different chronologies). **Final Comments:** We found in the literature cases descriptions of patients who developed transverse myelitis (TM) after receiving the Oxford/AstraZeneca vaccine (contains chimpanzee adenovirus antigen as an adjuvant) and only four cases of NMO following COVID-19 vaccination (of a different type than AstraZeneca). Immunization can lead TM through different mechanisms such as a cross-reaction between foreign antigens and self-antigens, overactivation of antigen presenting cells and the subsequent autoimmune response, and polyclonal or bystander B cell activation that can lead to cytokine synthesis and activation of autoreactive T cells. In our case it could be hypothesized that the cytokine storm, leads to cell lysis and release of AQP4 molecules and might trigger humoral response in individuals with genetic susceptibility to autoimmune diseases. The role of adjuvants as contributing factors to the inflammatory and immune adverse vaccine reaction should be emphasized and studied.

Neuroimunologia

TL 1106455

MOG ANTIBODY-ASSOCIATED DISEASE (MOGAD) AFTER VACCINATION WITH AD26.COV2.S

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Case Presentation: A previously healthy 37-year-old woman presented to the Hospital with a 7-day history of progressive worsening eye pain, headache, blurry vision of the left eye, urinary retention, numbness in both hands, weakness in both legs, bilateral Babinski sign and Lhermitte sign. She had received one dose of the Ad26.COV2.S vaccine against SARS-CoV-2 13 days before the onset of symptoms. Laboratory findings included a positive MOG-Ab, negative AQP4-Ab, an elevated CSF leukocyte cell counts and protein levels, and a positive antinuclear antibody. Other results were non-significant. A magnetic resonance imaging (MRI) of the brain and the spine revealed multiple hyperintense lesions on the cerebral cortex, basal ganglia, and from the C5-level to the conus medullaris in FLAIR and T2-weighted images, with no post-contrast enhancement. The patient received high-dose IV steroids, 5 sessions of plasma exchange therapy, and oral steroids. She had a complete remission of the visual symptoms after the treatment, and was discharged in need of a walking aid, with an EDSS of 6,5, and in use of monthly intravenous immunoglobulin therapy (IVIg). Eleven months after the episode, the patient was in a good general state, with only a mild urinary retention. Her EDSS was 3,0. A new MRI showed an almost complete resolution of the previous lesions, and MOG-Ab dosage was negative. She had received IVIg for 6 months and now was initiating Rituximab. Cases of Myelin oligodendrocyte glycoprotein (MOG) antibody-associated disorder (MOGAD) developed after COVID-19 vaccination have been described. Although molecular mimicry between SARS-CoV-2 spike proteins and MOG is unclear, analyses of the cross-reactions between host and microbial proteins show a potential correlation between CNS inflammation and demyelinating diseases. In the reported case, the patient developed MOGAD 13 days after receiving an adenovirus vector-based vaccine against SARS-CoV-2, which is considered sufficient time for the creation of autoantibodies. MOG-Ab-associated disease is related to better outcomes than AQP4-Ab-associated disease, although MOGAD has a higher relapse rate of optic neuritis. Patients who remain seropositive after treatment seem to present more relapses. Therefore, a prospective follow-up of MOG-Ab titres is important for the prognosis evaluation. This case presents a patient with MOGAD post Ad26.COV2.S vaccination with an unusual presentation and excellent recovery.

Neuroimunologia

TL 1106463

POST COVID-19 VACCINATION NEUROMYELITIS OPTICA SPECTRUM DISORDER: CASE PRESENTATION

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Case Presentation: A 40 years old woman presented to the emergency service with left-sided hemiparesis and bilateral numbness in arms and legs progressive over four days. The patient didn't have history of recent infection diseases, she received the first dose of CORONAVAC vaccine 12 days before the beginning of symptoms. Previously, this patient had been diagnosed with Neuromyelitis Optica Spectrum Disorders (NMOSD) after episodes of unilateral optic neuritis, in 2012, and a longitudinal extensive transverse myelitis, in 2013. At that time, she had a negative anti-aquaporin-4 (anti-AQP4). Since the last attack she had been using Azathioprine for recurrence prevention. In the current Hospitalization, MRI showed a diffuse thinning of the right optic pathway and, in cervical spinal cord, longitudinally extensive was identified at levels from C1 to C5, associated with an area of contrast enhancement at C3 and C4, suggestive of an active inflammatory/demyelinating lesion. A new longitudinally extensive cervical myelitis was then confirmed. Treatment was performed with methylprednisolone and plasmapheresis. The patient was discharged from Hospital in good general condition. In a recent outpatient visit, the patient had complete remission of this deficits. In this consultation, azathioprine was replaced by rituximab due to therapeutic failure. This case presentation highlights the temporal relationship between the realization of the vaccine for SARS-CoV2 and an outbreak of NMOSD, aiming at the discussion of possible causality between the vaccine and the occurrence of relapses in patients with autoimmune diseases. SARS-CoV-2 infection has been associated with the development of autoimmune processes. Molecular mimicry has been suggested as a potential mechanism for these associations. Some studies showed that antibodies against the spike protein S1 of SARS-CoV-2 had high affinity against specific human tissue proteins. As this is the same viral protein that the vaccine mRNA codes for, it is plausible that these vaccines may unmask or flare-up autoimmune diseases in predisposed patients. Whether there exists a causal relationship between COVID-19 vaccination and the flare-up of autoimmune diseases it remains to be determined. Only long-term follow-up of large cohorts of patients receiving the vaccine will answer this question. Until then, healthcare providers are encouraged to remain vigilant about potential side effects that will likely emerge with the spreading of vaccination.

Neuroimunologia

TL 1106474

RELAPSING ANTI-NMDA RECEPTOR ENCEPHALITIS: CASE PRESENTATION

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Case Presentation: A previously healthy 16-year-old female patient presented with an episode of tonic seizures. After 05 days, she evolved with syncope. One month later, she had seizures characterized by ocular supraversion, followed by loss of consciousness, limb hypertonia, and drooling. She progressed with behavioral changes, visual hallucinations, gait and language alterations. In addition, she had psychomotor agitation, urinary and fecal incontinence, and oromandibular dyskinesias. Cerebrospinal fluid analysis showed pleocytosis. Brain MRI was normal. Tomographic screening of neoplasms was negative. The electroencephalogram was suggestive of nonspecific encephalopathy. In view of the subacute onset of psychiatric symptoms, associated with new focal neurological findings, the presence of epileptic seizures and CSF pleocytosis, the patient fulfilled diagnostic criteria for autoimmune encephalitis. She performed an immunological panel on CSF, which confirmed the diagnosis of anti-NMDA receptor encephalitis. During Hospitalization, she underwent pulse therapy with immunoglobulin and methylprednisolone, showing a significant improvement in her condition. After 4 years, she evolved with disease recurrence, characterized by agitation, aggression, disorientation, behavioral alteration, visual hallucinations, and generalized tonic-clonic seizures. She underwent pulse therapy with methylprednisolone and immunoglobulin again, showing complete remission. **Discussion:** Anti-NMDA autoimmune encephalitis is an inflammatory condition of the brain triggered by the production of antibodies against the NMDA receptor on the neuronal cell surface. It predominantly affects young women. The initial symptom is usually neuropsychiatric, followed by cognitive dysfunction, movement disorders, decreased level of consciousness, autonomic dysfunction, and hypoventilation. Epileptic seizures tend to occur at any time during the illness. MRI is altered in about a third of patients. The CSF is altered in about 80% of cases. EEG reveals abnormal findings in around 90% of patients. Although it is a disease better known as monophasic, some patients can evolve with recurrence of symptoms. **Final Comments:** The present report aims to describe a case of relapsing anti-NMDA encephalitis, characterizing an atypical evolution of the disease.

Neuroimunologia

TL 1106501

NEUROMYELITIS OPTICA SPECTRUM DISORDER: A CASE PRESENTATION WITH THE SAME ONSET IN IDENTICAL TWINS

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Case Presentation: We describe the case of identical twins sisters with neuromyelitis optica spectrum disorder (NMOSD) with anti-aquaporin 4 antibodies (AQP4-Ab) positive, within a two years difference in the onset of symptoms between them. Sister 1: 19-years-old, with back and neck pain, lower limb weakness and sensory loss and evolved with urinary retention, persistent headache and ocular pain with transitory loss of vision in both eyes. Spinal magnetic resonance imaging (SMRI) revealed T2/FLAIR hyperintensity at the level of C3-C7 and L1-S1. Cerebrospinal fluid (CSF) examination showed a cellular reaction of 499 WBC/mm³, Protein level of 71 mg/dl, Glucose 40 mg/dl. The AQP4-Ab test was positive (1:80). Methylprednisolone intravenous pulse therapy (IVMP) associated with immunoglobulin worked to bring a clinical improvement of the symptoms, and the empiric maintenance regimen were made with Rituximab after the change of Azathioprine due it resulted in hepatotoxicity. Sister 2 20-year-old, that two months prior to her clinic visit also experienced back pain, paresthesia in upper limbs and ascending hypoesthesia localized in the intermamillary region and evolved with weakness of her legs, paraplegia and sphincter defect. On neurologic examination revealed meningeal irritation, and confirmed the sensory loss and paraplegia. SMRI evidenced T2 hypersignal at the level of T10 and longitudinally extended hypersignal in the cervical column. CSF revealed 8 WBC/mm, Protein level of 98 mg/dl, Glucose 54mg/dl. The AQP4-Ab test was also positive. IVMP associated with Plasma exchange was a success and the empiric maintenance regimen was made with Rituximab. **Discussion:** NMOSD is an group of demyelinating syndromes characterized by IgG auto-antibodies directed against aquaporin-4 causing severe attacks in the optic nerve and/or spinal cord, leading to blindness, dysautonomic symptoms and paralysis. Most cases of NMOSD are sporadic, but some familial reports were reviewed, with a prevalence of 0,8%. So, in this case, we observe twin sisters whose onset of symptoms were similar and started within a two years difference between them and had to receive the second line attack therapy with rituximab due to the good response from her sister. **Final Comments:** The treatment refractoriness of the sisters evidence that there may be a prognostic relationship influenced by genetics, especially in the improvement of symptoms after acute treatment.

Neuroimunologia

TL 1106507

REVERSIBLE CEREBELLAR MAGNETIC RESONANCE IMAGING ABNORMALITY AND ATAXIA AS A FIRST MANIFESTATION IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Case Presentation: A 36-year-old woman, admitted to the emergency department complaining cognitive changes, in gait imbalance, progressive incoordination, asthenia and weight loss started 3 months ago. At the neurological examination, the patient presented disorientation with inappropriate behavior, gait ataxia and bilateral dysmetria and dysidiadokokinesia. Initial investigations showed the presence of normochromic normocytic anemia, with positive direct coombs. The FLAIR in brain MRI revealed high signal intensity in the cerebellum symmetrically and bilaterally and the DWI showed restricted diffusion. The study of CSF that had inflammatory characteristics with 35 cells (85%monocytes), proteins 58 mg/dl and glucose 50 mg/dl, the CSF culture did not reveal any signs of infection; the culture was negative at 72 h, cytomegalovirus PCR, human herpesvirus 1 e 2, varicella-zoster virus and Epstein-Barr virus PCR were all negative. Serological profile for hepatitis B, hepatitis C and human immunodeficiency virus were negative. Neoplastic screening with tomography was negative. Autoimmune profile, however, was positive for antinuclear antibody (1:640 titers, speckled pattern), anti-DNA antibody (1:40 titer) and anti-Sm antibody reagent with consumed C3, C4 and CH50. Systemic lupus erythematosus, unequivocally fulfilling the Systemic Lupus International Collaborating Clinics classification criteria, was made and corticosteroid treatment was started with prednisone 60 mg per day, after a week of follow-up the patient has a completely improved of ataxia and the brain MRI abnormality. **Discussion:** Neuropsychiatric manifestation is commonly found in patients with SLE, it can manifest as changes in both the central and peripheral nervous systems. The most common manifestations are cognitive dysfunction, headache, seizures, and psychiatric conditions; aseptic meningitis, stroke, encephalopathy, movement disorders, and myelopathy are also seen. Cerebellar involvement in SLE is estimated at less than 2% of cases and more rarely as the first clinical manifestation. We present a patient that had a cerebellar syndrome and MRI abnormalities as the first presentation of SLE. The diagnosis of inflammatory cerebellitis associated with SLE was made supported by mild inflammatory changes in the CSF, clinical changes and abnormalities in neuro-imaging rapidly reversible. Cerebellar degeneration due to SLE is not common.

Neuroimunologia

TL 1106548

YOUNG PATIENT DIAGNOSED WITH CRION WITH VISUAL IMPROVEMENT AFTER USE OF HUMAN IMMUNOGLOBULIN: A CASE PRESENTATION

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Case Presentation: A 26-year-old female patient with low visual acuity and photophobia in the left eye. The condition evolved progressively, with the appearance of pain on ocular mobilization 2 days after the onset of symptoms. Condition occurred acutely, already waking up with visual changes. The patient had several similar episodes in both eyes since the age of 14, with a milder condition, with no limitation of daily activities in most episodes. The current episode it is the second with an impact on daily activities. In the first severe episode, she underwent pulse therapy with methylprednisolone and her symptoms completely improved. In an ophthalmological evaluation, an important change in the visual field of the left eye was observed by the campimetry. MRI of the brain was performed, with a slight hypersignal on T2 in the left optic nerve. Cerebrospinal fluid was normal. Requested rheumatological tests, anti-aquaporin 4, and anti-MOG research, all negative. The patient was again submitted to pulse therapy with methylprednisolone, without a satisfactory clinical response. Next step was intravenous immunoglobulin infusion, and a significant positive response of improvement in the visual acuity was observed. In this context, in view of the clinical manifestation and the complementary exams, the diagnosis of Chronic Relapsing Inflammatory Optic Neuropathy (CRION) was made. **Discussion:** CRION is a rare form of autoimmune optic neuropathy. Is characterized by vision loss and pain, and the occurrence of symptoms for more than one episode. MRI shows contrast enhancement in the optic nerve, and a good response to corticosteroid therapy is expected. Absence or suspension of treatment can lead to recurrence of symptoms, with possible irreversible visual loss. An atypical aspect of the case described is the therapeutic response only with the use of human Immunoglobulin. Kalish et al. described a series of CRION cases with a favorable outcome in selected cases treated with venous immunoglobulin. **Final Comments:** Patients with CRION are a diagnostic challenge. It is important for neurologists to maintain clinical suspicion. Initiation of early appropriate therapy may promote a better clinical response and a better functional outcome. The treatment of acute CRION events with human immunoglobulin is not the first therapeutic option, but as described in the literature, it becomes a viable option, especially in cases of first-line refractoriness.

Neuroimunologia

TL 1106599

AN ILLUSTRATIVE PHENOTYPE IN ANTI-MOG ASSOCIATED MENINGOENCEPHALITIS

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Case Presentation: A 23-year-old male admitted to our Hospital with a headache three weeks after his first dose of vaccination against SARS-CoV-2. One week later, he was admitted due to generalized tonic-clonic seizure, evolving with fever, encephalopathy, bilateral low visual acuity and left hemiparesis. Cerebrospinal fluid (CSF) examination showed pleocytosis (lymphocyte predominant) and elevated protein. Broad-spectrum antimicrobials were promptly initiated, but specific exams for infectious diseases were all negative. His first brain magnetic resonance imaging (MRI) was normal. He had mental status worsening and a new MRI demonstrated cortical and subcortical hyperintensity on the T2-weighted image and FLAIR in the right frontotemporal region. He was readmitted to our Hospital, where the antimicrobial treatment was suspended and methylprednisolone and plasma exchange were prescribed, considering the possibility of a demyelinating disorder, with optimal response. A new MRI showed frontotemporal cortical and subcortical lesions bilaterally, in the thalamus and brainstem, with areas of subcortical enhancement. Oligoclonal bands in CSF and anti-myelin oligodendrocyte glycoprotein (MOG) in the serum were positive. **Discussion:** MOG antibody-associated disease (MOGAD) is an inflammatory disease of the central nervous system with many described phenotypes still in characterization, including meningoencephalitis. **Final Comments:** We presented a case of MOGAD with meningeal involvement, which occurs in less than 10% of adults. Vaccination may be a risk factor for MOGAD attacks. Some findings of our patient are more specific for MOGAD, like leptomeningeal and thalamic involvement.

Neuroimunologia

TL 1106637

CASE PRESENTATION PSEUDOTUMORAL MULTIPLE SCLEROSIS LESION ASSOCIATED WITH DISCONTINUOUS FINGOLIMOD TREATMENT

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Case Presentation: Female, 26 years old, diagnosed with a Relapsing-Remitting Multiple Sclerosis (RRMS) in 2018 with an episode of right central visual field deficit. She reported two previous episodes in 2012 and 2017, of lower limb paresthesia with progressive improvement and complete remission within thirty days. In May 2021, she had an episode of paresis in the left side and left visual deficit, involvement of VII, IX and X cranial nerves ipsilaterally. She reported having stayed in February and April 2021, without fingolimod. No infectious processes or previous vaccination. Neurological examination showed complete hemiparesis with predominance of the left brachial, AV 20/80 on the left and 20/30 on the right, deviation of the uvula on the right and tongue deviation on the left. EDSS: 8.0 at the time of admission. Complementary tests: cerebrospinal fluid (CSF): 01 cell, proteins: 40, glucose: 53, BOC present. Other exams without changes. Brain magnetic resonance imaging (MRI): tumefactive right nucleocapsar hypersignal with perilesional edema and signs of acute supratentorial inflammatory activity. She received pulse therapy with dexamethasone (200mg/day) for 12 consecutive days with visual and motor deficit improvement: left eye AV (20/40), EDSS: 04. **Discussion:** Multiple Sclerosis (MS) is the main chronic demyelinating inflammatory disease of the Central Nervous System (CNS). Injuries to myelin, oligodendrocytes, and axons, to varying degrees, are caused by an autoimmune response. Abrupt discontinuation of fingolimod is reported to cause a rebound effect, with increased inflammatory activity, severe flare-ups, and eventually pseudotumor manifestations within 4-12 weeks. Treatment is challenging and pulse therapy with corticosteroids is considered an alternative to treat rebound after discontinuation of fingolimod. **Final Comments:** Although a pseudotumoral lesion in MS is always a challenge, in this case presentation its appearance was due to the abrupt discontinuation of fingolimod. Unplanned discontinuation of this medication, caused by interruption of supply, is a risk for MS patients. The rapid recognition of this manifestation is essential for the institution of the correct therapy.

Neuroimunologia

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MILLER FISHER SYNDROME: CRACK USER RECURRENCE

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Case Presentation: A 54-year-old man sought Hospital care due to low fever, productive cough, urinary retention, low back pain, visual alteration, and diffuse paresthesia for 2 days. Miller Fisher Syndrome (MFS) was diagnosed and treated with immunoglobulin. Crack user. Irregular treatment of hepatitis C. On examination, patient in regular general condition, pain facies, eupneic, photoreactive isochoric pupils, extrinsic ocular muscles hampered in the left and bilateral lower gazes, reduced overall strength with difficulty in overcoming resistance, areflexia in lower limbs and hyporeflexia in the upper limbs, appendicular ataxia, other aspects of the neurological examination without abnormalities. Admission exams for infectious screening without changes. Lumbar puncture was performed, clear and colorless CSF, glucose 61, protein 114, 1 cell/mm³, 100% MNM, gram-negative, ink negative, lactate 1.7, VDRL negative. Cranial CT angiography without acute injuries. The patient's general condition worsened, evolving with vomiting, plethoric facies, worsening of left lumbar pain, dyspnea with supplemental oxygen therapy, and need for urinary catheterization. A 2019 electroneuromyography exam showed an old and partially reinnervated demyelinating and axonal process in the upper and lower limbs; signs of diffuse neuropathy involving cranial nerves with signs of old reinnervation, no evidence of active disease. Diagnosed with a relapse of Miller Fisher Syndrome, the patient started with intravenous immunoglobulin for five days. After clinical and neurological improvement, he was discharged from the Hospital. **Discussion:** Considered a rare variant of Guillain-Barré (GBS), MFS is a multifocal neuropathy characterized by the clinical triad of ophthalmoplegia, ataxia, and areflexia. The recurrence of MFS episodes is extremely rare, with only 37 cases reported in the literature until 2016. This fact, associated with the chronic use of crack by the patient, brings curiosity concerning the recurrence of MFS itself. A liquor dissociation appears in 90% of cases as in the case presented. The third cranial nerve is frequently the most affected. **Final Comments:** Clinical history and complementary exams, associated with improvement of the condition with immunoglobulin, strengthen the diagnosis of MFS. The use of crack, considered a risk factor, associated with liver disease and previous acute respiratory infection, may have favored recurrence.

Neuroinfecção

TL 1104949

MITOCHONDRIAL MYOPATHY MANIFESTED AFTER ZIKA VIRUS VIRAL INFECTION: CASE PRESENTATION

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Case Presentation: Female patient, 49 years old, report that for 5 years she has been progressing with urinary incontinence, sleep apnea using Continuous Positive Airway Pressure (CPAP), dysphagia, generalized fatigue, severe pain intensity in lower and upper limbs, mild hearing loss, decreased vocal tone and constipation. Activities like brush the hair, going up and down stairs and walking short distances have become obstacles due to fatigue and pain. She said it all started after Zika virus infection in 2016 (confirmed by IgG antibody). She denied any similar family history. After analyzing the case, two possible differential diagnoses for the case were presented: mitochondrial myopathy and myasthenia gravis and, complementary exams were requested. The electroneuromyography performed in association with the repetitive stimulation test revealed the presence of some action potentials of the motor unit, from the examined proximal muscles, with myopathic characteristics. The lactic acid at rest was 5.8 mg / dl and lactic acid after 15 minutes of walking was 22.2 mg / dl showing an increase of 3.83 times. The acetylcholine anti-receptor antibody was not found. With the history and clinical examination associated with the complementary exams, it was possible to infer as a probable diagnosis of mitochondrial myopathy. Treatment with coenzyme Q10 at a daily dose of 800 mg was instituted and the patient presented a complete remission of the vast majority of symptoms using the coenzyme for 5 months. Discussion Just as infection by the Zika virus has been linked to the occurrence of neurological manifestations, other viruses have also been shown to have similar actions. Reports in the literature prove the relationship between arbovirus infection and the involvement of the nervous system. In the case presented, the Zika virus was associated with a neurological manifestation different from those reported so far in the available literature, mitochondrial myopathy. The relationship can be established since even before the arbovirus infection, the patient denies any symptoms of mitochondrial dysfunction. However, it is not possible to say whether viral infection was the triggering factor of the dysfunction or the environmental activator of the mutation, with the appearance of clinical manifestations. **Final Comments:** Knowledge of the pathogenic mechanisms involved in Zika virus infection and its consequences for adults is still very limited.

Neuroinfecção

TL 1105005

HERPETHIC ENCEPHALITIS WITH FLU-LIKE SYNDROME IN A COVID-19 PANDEMIC CONTEXT: A CASE PRESENTATION

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Case Presentation: An 60-years-old previously healthy women, from a countryside city of Amazonas, was referred to our neurologic service with a family-reported history of flu-like symptoms such as fever and coryza, and both retrograde and anterograde amnesia, starting 45 days before her first consult and, a week before, psychomotor agitation and disorientation in time, space, situation and person. Her neurological examination showed GCS scoring 9, with isochoric photoreactive pupils, no signs of meningeal irritation, no alterations in the cranial nerves and no motor deficit. She had bilateral patellar hyperreflexia, left upper limb spasticity, and ipsilateral Babinski and Hoffman signs were present. She was Hospitalized and tested positive for COVID-19 and, during the Hospitalization, evolved with lowered level of consciousness, seizures and global aphasia. CT showed no signs of hypodensity or bleeding. After discussion, the neurologic team started empirically antibiotic therapy and intravenous Acyclovir and, after 5 days of Hospitalization, PCR in cerebrospinal fluid was requested showing HSV-1, and a MRI presenting signal changes with a slight swelling effect and a component of breakdown of the blood-brain barrier, suggestive of a viral inflammatory process. Patient evolved with improvement compared to admission and referred for outpatient follow-up. **Discussion:** HSV encephalitis represents one of the most devastating forms of encephalitis. Its incidence accounts for up to 20% of all identified viral encephalitis. Little is known about the influence of SARS-Cov-2 infection on the CNS and its neurological alterations. Otherwise, some authors state that a proinflammatory status and release of cytokines are the key to manifest other viral encephalitis, specially the ones caused by reactivation of virus from the Herpesviridae family. Kahwagi et al (2022) reported a case of coinfection including encephalitis caused by Varicella-Zoster virus confirmed by lumbar puncture associated with a swab test positive for COVID-19, reinforcing participation of the new coronavirus in dysregulation of immune system and neuroinflammation. **Final Comments:** The case raises greater vigilance in typical neurological conditions of encephalitis that present SARS-CoV-2 co-infection, because, despite the outcome of this correlation isn't fully established, it is known that the pro-inflammatory state sustained by the coronavirus performs a favorable condition for HSV-1 reactivation.

Neuroinfecção

TL 1105076

VARICELLA ZOSTER MYELOPATHY UNMASKED BY A IMMUNE RECONSTITUTION INFLAMMATORY SYNDROME: A CASE PRESENTATION

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Case Presentation: A 57-year-old woman living with HIV and with previous poor adherence to treatment started regular use of antiretroviral drugs in December 2021 after pneumocystosis. In January 2022, she started a right frontal throbbing headache and vertigo. Afterwards, she developed paresthesia, progressive crural paraparesis, urinary urge incontinence, right hearing loss with pulsatile tinnitus. The neurologic examination showed spastic crural paraparesis, hypoaesthesia and decrease vibratory sense at the T8 level, a left horizontal torsional nystagmus and an abnormal right head impulse test. The cerebrospinal fluid analysis showed a lymphomononuclear pleocytosis and a positive Varicella-Zoster RT-PCR. The magnetic resonance imaging (MRI) demonstrated T2-FLAIR right vestibulocochlear nerve hyperintense signal and spinal cord hyperintensity at the T1-T2 and T4 levels. We started treatment with acyclovir for 21 days, with just mild neurologic improvement. During the investigation, an important increase in the CD4 lymphocyte count was found (12,2 cell/mm3 in December to 300 cell/mm3 in February). Immune reconstitution inflammatory syndrome (IRIS) was suspected, and a 1 g intravenous methylprednisolone pulse therapy was prescribed for 5 days with a significant improvement of bladder symptoms, gait, and proprioception. She was discharged from the Hospital walking with no support. IRIS consists of an increase in T-cell lymphocyte counts resulting in a dysregulated immune response against infecting pathogens and the host, concomitantly. The incidence of this condition changes according to the pathogen, being more common in tuberculosis and Cryptococcosis. Our patient presented an increase of CD4 lymphocyte count associated with new neurological symptoms and abnormal imaging exams and evolved with mild improvement after adequate treatment of the Varicella-Zoster virus. As immune reconstitution inflammatory syndrome was suspected, steroid treatment was prescribed to reconstitute the blood-brain barrier and decrease the response of T lymphocytes. Afterwards, a great clinical improvement was noticed. There is no previous report of a Varicella Zoster myelopathy unmasked in this scenario and with a great response to steroids, resembling the importance of the recognition of this important condition.

Neuroinfecção

TL 1105078

NEUROBORRELIOSIS, MANTAINING A HIGH SUSPICION: CASE PRESENTATION

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Case Presentation: A previously healthy 38-year-old woman presented to an emergency care in São Paulo -Brazil due to severe holocranial headache, nausea and vomiting for the last 7 days. During initial evaluation, she was agitated and disoriented in time and space, but had otherwise normal physical and neurological exams. She denied fever and skin rash and had no signs of meningeal irritation. As for epidemiologic history, she lived in Geneva and was recently vacationing in the USA (Kansas and Washington) before coming to Brazil. Initial tests showed mild neutrophilic leukocytosis and a positive ?-hCG test. Inflammatory and rheumatologic tests were normal and there were no organ disfunctions. Brain MRI showed only non-specific FLAIR hyperintensity in some cortical sulci. CSF analysis showed a slightly elevated opening pressure 21cmH₂O, 38 cells with lymphocytic predominance, 65mg/dL protein, normal glucose and somewhat elevated ADA 4,8mg/dL and lactate 22mg/dL. Serologies for toxoplasma, syphilis, herpes family and bacteriological, micobacteriological and fungal test were negative. Nevertheless, due to clinical suspicious of encephalitis, IV acyclovir and ampicillin were introduced empirically. After 5 days, due to symptomatic maintenance, further analysis of the CSF was conducted with a negative herpes-family PCR and a positive ELISA IgM for Borrelia. Ampicillin and acyclovir were discontinued and intravenous ceftriaxone was introduced and maintained for 28 days with progressive symptomatic improvement. **Discussion:** Lyme disease is a tick-borne illness, endemic in the USA and some parts of Europe. It usually starts within a month of the tick bite by presenting with characteristic skin rash and constitutional symptoms. After a few months, neurologic findings may follow, being the most common presentations cranial nerve palsies, radiculopathy, lymphocytic meningitis and, rarely, encephalomyelitis. For the diagnosis, ELISA and Westernblotting essays for B. burgdorferi s.s. antigens are used. As for the treatment, being the patient pregnant, she was unable to receive oral doxycycline. In these cases possible options are IV cefotaxime, G penicillin or ceftriaxone, which was the therapy of choice in this case. **Final Comments:** In this study we present an unusual case in our country, with non-specific symptoms. Serologic testing combined to an epidemiologic exposure were key for the diagnosis, reinforcing the importance of extensive questioning of travel history during anamnesis.

Neuroinfecção

TL 1105084

ACCIDENTAL TETANUS OF ATYPICAL EVOLUTION, A CASE PRESENTATION

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Case Presentation: A man, 48 years old, rural worker, 210 cm tall, hypertensive and obese, presented for seven days intense pain in the left gastrocnemius. On examination, fixed left gastrocnemius muscle contracture unleashed by light and sound stimulus. Normoreflexia, normoesthesia. Homans sign absent. Trismus/dysphagia absents. Presented two punctate lesions consolidated on the sole of the left foot caused by barbed wire fence, one with 30 days long and the other with 15 days long. No ecographic evidence of deep vein thrombosis, nor phlogistic signs and no signs of clinical severity. Two doses of the dT vaccine, which last one applied 25 years ago. Three visits to the Emergency Room in the last 20 days for trismus as the main complaint, in addition to sardonic laughter, when diazepam being administered for "anxiety". Therefore, patient classified as Tetanus grade I (according to the 1956 Ablett scale) in local phenotype. Treatment with antibiotic therapy, muscle relaxation, analgesia was made, administration of equine tetanus-antibody and a 3rd dose of dT. He was evolved with improvement of muscle spasms after antibodies inoculation, however, remained with muscle pain and required intensive support after showing signs of Respiratory Failure. After discharge from the ICU, he had remission of the initial symptoms. **Discussion:** Recent data from the WHO show an increase in tetanus notifications, a preventable disease and compulsory notification in Brazil. In view of that, it is a case that starts with symptoms of generalized disease and developed to a local clinic. This scenario is against with the mechanism of retrograde axonal transport of tetanospasmin, which implies diffuse involvement, in addition of questioning the bioavailability of the toxin due to the size of the patient. Additionally, there is a low clinical suspicion for intoxication by strychnine, since there was no relaxation of the muscles between spasms or maxillary involvement, and also for conditions such as drug withdrawal or hypocalcemia due to the epidemiology and personal history not suggestive. **Final Comments:** Early diagnosis and treatment are ratified in this scenario and invite specialists to propose a new clinical presentation scale that increases sensitivity for atypical cases of tetanus.

Neuroinfecção

TL 1105092

DENGUE-ASSOCIATED DEMYELINATING LESIONS – CASE PRESENTATION

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Case Presentation: Male patient, 21 years old, presented with myalgia, headache and fever for 3 days and spontaneous resolution. After 7 days of the resolution of the previous condition, he started to have paresthesia in the lower limbs, evolving in a subacute way with urinary retention. Referred to the emergency service with progressive worsening of strength in the upper limbs after 24 hours. After admission, the patient began to present diplopia and excessive sleepiness. In the initial exams, the only laboratory alteration that called attention was a serology for Dengue (positive IgM and IgG). CSF analysis showed pleocytosis (112 cells with lymphocytic predominance) and protein analysis (194mg/dL, presence of a suggestive band in the gamma region). Magnetic resonance imaging of the Thoracic Spine showed a hyperintense centromedullary lesion on T2/stir, affecting practically its entire length with spots enhancement by gadolinium. Brain MRI with signs of infratentorial and supratentorial lesions, without gadolinium enhancement. Lesions were not characteristic of ADEM. It was decided to perform pulse steroid therapy with Methylprednisolone 1g for 5 days, with progressive improvement and complete resolution of the condition. **Discussion:** The Dengue virus can affect the neurological system directly by viral invasion or indirectly with immune-mediated reactions. Complications may arise during a feverish period or during convalescence, and may manifest as Encephalitis, Stroke, Myalgia, Guillain Barré Syndrome, Myositis, Hypokalemic Paralysis, Myelitis and Acute disseminated encephalomyelitis (ADEM). In the case above, the manifestations started 7 days after the febrile period, suggesting a molecular mimicry reaction between the pathogen and the host's myelin. There are few cases in the literature relating longitudinally extensive myelitis associated with dengue, with emphasis on a study in the Amazon region of 51 cases, with favorable evolution and recovery of deficits in 95% of patients. Other cases correlate dengue with brain involvement, most cases occurred during the recovery phase of uncomplicated dengue. **Final Comments:** Dengue is the most prevalent arbovirus in the world, and Brazil recorded an increase of 113.7% in the first months of 2022 compared to the previous year. Consequently, they link an alert to the neurological manifestations associated with this infection that can be serious and potentially fatal.

Neuroinfecção

TL 1105110

CERVICAL INFECTIOUS SPONDYLODISCITIS AS A DIFFERENTIAL DIAGNOSIS OF ACUTE FLACCID TETRAPARESIS

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Case Presentation: A 72-years-old man, previously hypertensive and alcoholic, presented to the emergency room with a five-day history of shortness of breath, cough, fever, muscle pain and four limbs weakness. Neurological examination demonstrated tetraparesis, hypotonia in four limbs, areflexia in upper limbs and hyporeflexia in lower limbs. Laboratory demonstrated leukocytosis. Blood culture was positive for methicillin-sensitive SA. Cerebral spinal fluid analysis revealed important protein elevation, no glucose consumption and cell count within reference range. At arrival of patient, the MRI was not available, and among the differential diagnosis, the main hypothesis was of Guillain-Barré syndrome. Extended investigation with cervical MRI realized as soon as available, showed at FLAIR and T2, inflammatory hypersignal at C3-C4 left facet joint and three isolated collections. The third formation was causing vertebral foramen stenosis with hypersignal alterations at the adjacent spinal cord compatible with compressive myelopathy. All three formations were suggestive of abscess, due bacterial spondylodiscitis. The patient was treated with intravenous oxacillin, with clinical improvement and regression of the masses at a posterior control MRI. **Discussion:** Spondylodiscitis refers to the infectious process that affects the vertebral body, the posterior vertebral arch, and the intervertebral disc. Installed in an insidious way and has a slow evolution, usually manifesting as fever, local pain, changes in sensitivity and strength in limbs, and may present as a radiculopathy as well as a spinal syndrome due to the involvement of the spinal canal. The physical examination of uncooperative patients can make it difficult to discriminate sensitivity changes such as, for example, sensory level in the context of spinal cord syndrome. In this context of flaccid tetraparesis, as in the case above, the acute demyelinating polyradiculopathy syndrome is an important differential diagnosis. This protein-cellular dissociation in the cerebrospinal fluid, uncommon in peri-medullary infections that usually progress with an increased number of cells in the CSF, was notorious confounding factor, alongside to the limb flaccid paresis found in the acute spinal cord injury. **Final Comments:** The acute flaccid tetraparesis has a large number of differential diagnoses and usually demands early complementary exams to the correct evaluation of the lesion, especially in the ER.

Neuroinfecção

TL 1105111

RHOMBENCEPHALITIS SECONDARY TO BRAIN ABSCESS AS A DIFFERENTIAL DIAGNOSIS IN SUBACUTE TETRAPARESIS IN A IMMUNOCOMPROMISED PATIENT

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Case Presentation: R.M, 48 years old, male, with a previous history of untreated HIV infection. Started vomiting, diarrhea, abdominal pain, headache and hiccups in March/2022. After 2 weeks, initiates progressive tetraparesis, starting with the lower limbs and ascending to the upper limbs, preventing ambulation, associated with urinary incontinence, dysphagia, paresthesias that predominated in the lower limbs and unintentional weight loss (8 kg in 2 months). During the initial evaluation, tetraparesis was demonstrated associated with hyperreflexia. The evaluation of the cranial nerves showed a deficit to lift the palate bilaterally, as well as the inability to protrude the tongue. Brain CT was normal, as well as CSF test. Systemic investigation also was negative. Two days after the initial evaluation, the patient had an episode of fever, respiratory failure, and was intubated and started on cefepime. Three days later, the patient started to experience significant clinical improvement in the tetraparesis, as well as in the lower cranial nerves. Brain MRI was performed after clinical stabilization, demonstrating a nodular lesion on T1-weighted gadolinium enhancement in the medulla, with T2 hyperintensity in the medulla up to C5 level in spinal cord. Due to clinical improvement, the hypothesis of brain abscess is considered, and antibiotic therapy is maintained for 6 weeks. A new brain MRI was performed 3 weeks later, demonstrating a significant reduction in the lesion as well as the associated edema. After 6 weeks, the patient was discharged from the Hospital with resolution of symptoms. **Discussion:** Brain abscess does not usually occur in the brainstem region, and the involvement in this location is associated with significant morbidity and mortality. The most likely etiology of a bacterial brain abscess in immunocompromised patients is *Listeria monocytogenes*, and the response to beta-lactam makes this the main hypothesis for our case. *Nocardia* spp is also an important agent, but its clinical course occurs in a more insidious way. **Final Comments:** The immunocompromised patient can be affected by several serious insults, often without the typical manifestations, as we demonstrate a patient with absence of fever and headache in the initial condition. Thus, brain abscess should always be a differential diagnosis in the context of tetraparesis associated with signs of upper motor neurons, especially in immunocompromised patients

Neuroinfecção

TL 1105114

UNUSUAL ETIOLOGY OF TRANSVERSE MYELITIS: CASE PRESENTATION

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Case Presentation: L.S.M., male, 18 YO, was admitted to a Hospital with bilateral paresthesia in the plantar regions, ascending to the lower limbs, associated with sphincter incontinence, in June 2021. The brain MRI showed foci of high-signal on T2/ FLAIR, in the subcortical white matter, variable in size and shape, some with gadolinium enhancement; the cervical spine MRI showed nonspecific focal areas of high-signal on T2. The cerebrospinal fluid (CSF) was clear, with 04 cells mm³ (10% polymorphonuclear, 90% mononuclear); negative bacterioscopy, glucose 64 mg/dl; protein 28.4 mg/dl; negative culture. Blood VDRL was 1/32. He was treated with benzathine penicillin for 3 weeks and pulse therapy with methylprednisolone for 5 days, with no improvement in the symptoms. He was seen in our Service, in July 2021, and new tests were requested that showed VDRL 1/32; Vitamin B12 241, HIV negative, HTLV negative, Hepatitis B and C negative. A new CSF study on 07/27/21 showed 6 cells/mm³, 29 protein mg/dl, 80 lymphocytes, glucose 66 mg/dl, VDRL negative, FTAabs IgG positive, IgM negative, Kappa index 3.7 (normal < 2.9), OCB negative, anti-NMO negative, anti-MOG negative. He was treated with crystalline penicillin for 14 days and pulse therapy again for 5 days with significant improvement of the symptoms. In October, a new VDRL test was performed: 1/16. **Discussion:** Syphilis is a sexually transmitted disease caused by the *Treponema pallidum*. It affects 12 million people a year worldwide. If untreated, the disease can progress up to the tertiary form, which affects the central nervous system (CNS) and can be presented from its different clinical types: meningovascular, general paresis and tabes dorsalis. Transverse myelitis has different etiologies, such as autoimmune diseases, CNS infections, hypovitaminosis B12 and demyelinating diseases Syphilitic myelitis is a form of meningo-vascular syphilis confined to the spinal cord. The diagnosis sometimes is challenging and comprises not only blood tests, but also MRI findings, such as "the flip-flop sign" or "the candle guttering appearance sign" and CSF test. MRI can sometimes be essential for a correct diagnosis. **Final Comments:** This case describes an uncommon presentation of syphilis as a transverse myelitis. Syphilis is a differential diagnosis to look after in transverse myelitis cases specially because of good clinical response to the therapy.

Neuroinfecção

TL 1105136

NEUROTUBERCULOSIS IN AN IMMUNOCOMPETENT PATIENT: A CASE-BASED UPDATE

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Case Presentation: A 35-year-old man was admitted 1 month ago with paraparesis and thunderclap headache. He reports a chronic condition of nausea, vomiting, diarrhea, recurrent fever and monoparesis in the left lower limb for 3 months. A skull Magnetic Resonance Imaging (MRI) was performed, showing important hydrocephalus with diffuse nodules in the leptomeninges. A ventriculoperitoneal shunt was installed, with partial neurological improvement. There was a condition worsening 2 weeks ago, with extension to a tetraplegia associated with ascending paresthesia to the T9-T10 level. Neurological examination revealed global areflexia, central nystagmus, right peripheral facial palsy and partial neck stiffness. Laboratory tests showed negative bacterioscopy, blood cultures, sputum smear microscopy, rapid tests for HIV and VDRL. Chest computed tomography (CT) revealed no lung lesions, and T2 FLAIR MRI of cervical-thoracic spine and skull showed hypersignal in broad sites of telencephalon, brainstem, cerebellum, and C3-T8 spinal cord. Cerebrospinal fluid (CSF) examination revealed cellularity of 60/mm³, proteins of 146 mg/dl, glucose of 28 mg/dl, negative VDRL and bacterioscopy, no fungal growth on India ink stain, and adenosine deaminase (ADA) of 26 U/L, which confirmed the diagnosis of neurotuberculosis. A regimen of rifampicin, isoniazid, pyrazinamide and ethambutol was started, to which the patient has been evolving with paraparesis improvement. **Discussion:** Tuberculosis (TB) is an infectious disease caused by *Mycobacterium tuberculosis* that mainly affects lungs, but can also affect the meninges, causing neurotuberculosis (3% of TB cases in seronegative HIV patients). There is slower subacute evolution than other meningitis. Clinical response to treatment is excellent when it is diagnosed before irreversible neurological damage, so empiric therapy should be initiated immediately in any patient with neurological abnormalities, CSF with low glucose concentration, high protein, lymphocytic pleocytosis and if TB is suspected. Higher ADA levels in CSF are useful in diagnosis. Hydrocephalus and basal meningeal thickening are common findings in CT or MRI. Treatment is initially composed by a four-drug regimen including isoniazid, rifampicin, pyrazinamide and ethambutol daily for 2 months. **Final Comments:** The diagnosis of neurotuberculosis in immunocompetent patients can be challenging and lead to high mortality if not diagnosed soon.

Neuroinfecção

TL 1105151

HOLOCORD SPINAL EPIDURAL ABSCESS – A RARE CASE PRESENTATION

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Case Presentation: male healthy patient, 56 years old, refers back pain started 9 days ago irradiated to lower limbs and perineum with urinary hesitancy and vomit without fever. First diagnosis thought was nephrolithiasis, excluded after normal abdomen computed tomography (CT). The following day, started with weakness in the right lower limb (mainly in quadriceps), constipation and urinary retention. CT of the thoracic and lumbar region was normal. After two days patient evolved with somnolence, flaccid paraplegia, absence of deep sensitivity on lower limbs, peripheral facial palsy in the right, dysautonomia and anisocoria. Cerebrospinal fluid showed pleocytosis (neutrophil and lymphocyte predominance), hyperproteinorrachia and important glucose consumption with lactate level of 9.9. Magnetic resonance evidenced anomalous leptomeningeal enhancement of the cerebellum and skull base cisterns; thickening and anomalous enhancement of the arachnoid and diffusely involving the entire spinal cord, including cauda equina; laminar collection with peripheral contrast enhancement extensive on the epidural posterior region from T1 to L2 with greater thickness in the lumbar. We started empirical treatment with ceftriaxone and meropenem plus dexamethasone 4 mg every six hour. Neurosurgery team indicated conservative management. Serologies for HIV, syphilis and hepatitis and PCR for *M. tuberculosis* in CSF were negative. All CSF and blood cultures (bacteria, fungi and mycobacterium) were negative. On the next day, patient worsened consciousness level requiring orotracheal intubation. After eight days of antibiotic CSF had a significant improvement. We treated the patient during forty days, with mild neurological improvement in consciousness level and quadriplegia sequelae. During Hospitalization patient presented many complications (secondary bacterial infections, acute renal failure and upper gastrointestinal bleeding) progressing to death 44 days after Hospital admission. **Discussion:** Until now less than twenty cases of holocord spinal epidural abscess (SEA) had been published and it is more common in immunocompromised. The commonly symptoms are fever, back pain and neurological deficit and the mortality rate is about 15%. Here we present an immunocompetent man that had a bad evolution. **Final Comments:** SEA can mimic other medical conditions, but have a high morbidity and mortality, so it is essential to be attentive to avoid delaying diagnosis and treatment.

Neuroinfecção

TL 1105208

NEUROBORRELIOSIS, A RARE CONDITION IN BRAZIL – A CASE PRESENTATION GABRIELA DA SILVA CREMONESE, NATHALIA TOMAZONI SILVA, IVANIO ALVES PEREIRA

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Case Presentation: A 37-year-old, male patient presented weakness in the lower limbs that was initially attributed to ciatalgia associated with muscle shortening since 2019. Due to the worsening of the condition with low back pain and cramps, he sought out other professionals who diagnosed him with axial spondyloarthritis and started treatments with non-steroidal anti-inflammatories and sulfasalazine. More than 2 years after the onset of symptoms, consultations with some specialists and progressive worsening of the condition, with associated headache and memory loss, the patient underwent an electroneuromyography that showed axonal neuropathy of the posterior tibial nerves and denervation in activity, and a test of ELISA confirmed with Western Blotting for positive *Borrelia burgdorferi* in different laboratories of excellence. Treatment was started with 28 days of ceftriaxone and continued for two more months with doxycycline. **Discussion:** As evidenced in the case, the clinical manifestations of brazilian neuroborreliosis – Baggio-Yoshinari Syndrome – can mimic multiple diseases and are poorly described so far. Consequently, neuroborreliosis can be overlooked by the clinician, especially in non-endemic areas, and has characteristics of recurrence and a tendency to cause chronic neurological and joint manifestations, especially if antibiotic treatment is instituted late. **Final Comments:** This is a challenging diagnostic zoonosis, with clinical symptoms suggestive of other pathologies and serological tests of limited sensitivity and specificities. Therefore, further studies on the neurological involvement of this syndrome are essential to bring attention to this differential diagnosis for an effective and early treatment.

Neuroinfecção

TL 1105235

OPTIC NEURORETINITIS IN AN HIV-SEROPOSITIVE PATIENT: CASE PRESENTATION

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Case Presentation: Male, 27 years old, white, initially searched an outpatient clinic due to painless and non-pruritic erythematous macules on the soles of the feet and palms, being diagnosed with syphilis, and later as a carrier of the human immunodeficiency virus (AIDS)/HIV, and treatment was started for both diseases (first line for AIDS, and for secondary syphilis according to the protocol of the Brazilian Institute of Health. After 30 days, he began to complain of floaters in the right eye (OD), in addition to fixed scintillating scotomas in the left eye (LE), painless and referred to a secondary Hospital for suspected neurosyphilis. On examination of the eye fundus (FO), papilla edema and signs of retinitis in the RE were observed. During etiological investigation, brain magnetic resonance imaging and cerebrospinal fluid (CSF) were normal (negative VDRL). A diagnosis of syphilis neuroretinitis was defined and the patient received intravenous crystalline penicillin for 14 days, with total regression of symptoms. **Discussion:** Acquired syphilis can be classified as primary, secondary and tertiary, being more frequent and more severe among HIV carriers, and in this case, it may present clinical manifestations of two stages simultaneously. *T. pallidum* is invasive and can affect the central nervous system at any stage of infection. Neurosyphilis is more frequent in HIV carriers, especially in those without antiretroviral therapy. Ocular syphilis, on the other hand, is an uncommon form and early manifestation of syphilis, at any stage of the disease, and can be confused with optic neuritis and neurosyphilis. Uveitis and optic neuritis are common to all patients with ocular syphilis, but some studies suggest that HIV-infected patients tend to have acute posterior uveitis, and particularly posterior chorioretinitis, as we observed in this patient. **Final Comments:** Ocular syphilis has different clinical spectra and can manifest at any stage of the disease, thus, it should be part of the differential diagnoses of retinitis, optic neuritis and especially neuroretinitis in HIV positive patients. Early diagnosis and treatment of neuroretinitis will prevent permanent visual loss.

Neuroinfecção

TL 1105256

NEUROCRYPTOCOCCOSIS AND NEUROSARCOIDOSIS: SIMULTANEOUS PRESENTATION IN AN IMMUNOCOMPETENT PATIENT

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Case Presentation: C.A.J.S. Male, 52 years, underwent restrictive diet (loss of 20 kilos in 3 months), evolving with depressive symptoms associated with the neurological framework of paraparesia, abasia, astasia, urinary and fecal incontinence, constipation and hypoesthesia in lower limbs, being restricted to the bed. After psychiatric treatment with antidepressants and antipsychotics had improved mood and he returned to wander, but only with support. Then there was sudden worsening of strength and equilibrium, being hospitalized for neurological framework. In lumbar puncture was verified the presence of cryptococcus by the direct research of fungi and performed treatment with antifungal for 42 days, with discreet improvement. Concomitant to the previous findings a chest tomography was performed to evaluate pulmonary compromise by the, being evidenced mediastinal lymph node in which anatomopathological suggested the presence of sarcoidosis, raising the idea of neurosarcoidosis in coexistence with neurocryptococcosis. Subsequently, pulse therapy was instituted with methylprednisolone and the patient presented substantial improvement of symptoms. **Discussion:** Neurocryptococcosis is a serious and usually fatal infection caused by two species: *Cryptococcus neoformans* and *Cryptococcus gattii*. In general, it presents as cryptococcal meningitis and affects immunocompromised individuals, with typical symptoms of meningeal infection: headache, lowered level of consciousness and cerebrospinal fluid changes. Neurosarcoidosis (NS) is a granulomatous disease of unknown etiology, whose main manifestations are leptomeningitis, cranial nerve involvement, hypothalamic dysfunction and intraparenchymal masses, corresponding to 5% of the presentations of sarcoidosis. In the case described above, we have a patient presenting with nonspecific systemic manifestations and significant impairment of sensory and motor functions, in addition to associated psychiatric symptoms, with simultaneous presentation of diseases. **Final Comments:** Neurocryptococcosis is a rare fungal disease in immunocompetent patients. This case stands out for its concomitance with another uncommon disease: neurosarcoidosis, in a previously healthy patient, enabling differential diagnoses of rare combinations with a favorable outcome.

Neuroinfecção

TL 1105363

MENINGEAL CARCINOMATOSIS MIMICKING A PSYCHIATRIC CONDITION: CASE PRESENTATION

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Case Presentation: A 45-year-old previously healthy man, who works in an accounting office, comes to the doctor with a complaint of sudden disorientation, delusions, and vomiting for 20 days. The confused mental state caused dependence on others for all activities for one week, then, he partially improved his state for five days and worsened again. He reported a daily headache of strong intensity. He denies seizures, allergies, or comorbidities. His family brought him to the consultation, directing him to a psychiatric condition due to behavioral change, and for having a family history (mother) of severe depression. He denies alcoholism or smoking. On physical examination, the patient does not cooperate, obeys simple commands with difficulty, does not obey complex commands, appears unaware of his surroundings, strength preserved, vivid and symmetrical reflexes, cutaneous-plantar reflex in bilateral flexion, isochoric and photoreactive pupils, preserved extrinsic ocular motricity, mimicry symmetrical face. During the investigation, magnetic resonance imaging (MRI) revealed hypersignal in the sulci of the cerebral hemispheres, associated with gadolinium enhancement in a leptomeningeal pattern. Cerebrospinal fluid (CSF) revealed the presence of neoplastic cells. After screening for neoplasms, non-small cell lung carcinoma was identified. **Discussion:** Meningeal carcinomatosis (MC) is a rare neurological complication characterized by multifocal invasion of the meninges by neoplastic cells. Breast, lung, and melanoma tumors are the main ones responsible for the cases. About 5% of all cancer patients have meningeal carcinomatosis, lung adenocarcinoma is the one that most often affects the meninges when referring to non-small cell cancer. In the present case, the patient presented behavioral changes associated with bradyphasia and headache. MC is a late event in the evolution of the neoplasm and can manifest with the involvement of spinal roots, cranial nerves, and/or lesions of the cerebral hemispheres. The diagnosis is based on clinical findings, neuroimaging, and CSF analysis. It has an unfavorable prognosis, survival is 3 to 6 months in patients undergoing treatment. **Final Comments:** This clinical case is unusual due to the low frequency of MC and the presence of neuropsychiatric manifestations in the absence of a previous diagnosis of neoplasia. In addition, behavioral changes may be a rare and early sign of meningeal carcinomatosis.

Neuroinfecção

TL 1105367

DENGUE AND NEUROLOGICAL SYMPTOMS: AN ATYPICAL REPORT BY GUILLAIN BARRÉ AFTER DENGUE

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Case Presentation: a 52-year-old female patient, 17 days before the arrival of the service, with asthenia, cutaneous rash in thorax and broos, with diagnosis of dengue, evolved 11 days later with hyposthesia and paresis of lower limbs (MMII) with ascence to upper limbs (MMSS), subsequently, the patient presented bilateral facial paresis and constipation. The patient presented Lucidity and well-oriented, with bilateral peripheral facial paralysis, with paresis and hyporeflexia of predominance in IIII, also presented hyperalgesia of them. lumbar puncture showed protein dissociation and cellularity, with 10 mm³ of leukocytes, 95% leukocytes, 540.4 meq/l of protein and 72 mg/dl of glucose, with cervical resonance and cranial tomography without alterations and other general laboratory tests within the normal range. performed as human immunoglobulin treatment 5 g for 5 days. Before discharge, a new cerebrospinal fluid was performed, showing a fall in the protein, 350 meq/l and leukocytes, 4 mm³. After Hospital discharge, the patient presented improvement of the picture with recovery of the strength pattern and improvement of facial paralysis. **Discussion:** Guillain barré syndrome (sgb) is often associated with infections by campylobacter jejuni, cytomegalovirus, Epstein-Barr virus, mycoplasma pneumonia, hiv and the presence of association between arboviroses and sgb is well documented, mainly in relation to zika virus, however cases related to dengue. where only about 5% of the patients present with associated neurological symptoms, they are rare. The symptoms are closely related to infection where antigens stimulate the demyelination process, where most of the symptoms such as ascending paralysis, facial paralysis and areflexia. The only treatment with recognized and recommended clinical response is the application of human immunoglobulin, with a good response, and can be followed after application with prednisone, but with studies with inconclusive results, in the case presented, the patient had a clinical and laboratory condition compatible with the sgb, with a clear indication for immunoglobulin treatment. **Final Comments:** The report shows a case of association between dengue infection and neurological symptoms, in which case the development of sgb is a rare presentation, with a good response to immunoglobulin treatment.

Neuroinfecção

TL 1105394

CENTRAL NERVOUS SYSTEM INVOLVEMENT IN COMPLICATED RHINOSINUSITIS: CASE PRESENTATION

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Case Presentation: Male, 16 years old, illicit drug user, symptoms of headache, mental confusion, bilateral eyelid edema, eye pain, blurred vision and fever after upper airway infection. Exams showing leukocytosis and cerebrospinal fluid indicative of bacterial meningitis. Broad-spectrum antibiotic therapy was started. Pulse therapy with methylprednisone was also performed to help reduce proptosis and visual symptoms. Magnetic Resonance Imaging (MRI) of the brain was requested, which showed intracranial collection starting in the medial temporal region adjacent to the cavernous sinus and extending to the posterior fossa, bypassing the right cerebellar hemisphere, suggestive of subdural empyema. He underwent neurosurgical drainage by posterior fossa craniotomy, without complications. Post-surgical control MRI showed thrombosis of the right sigmoid sinus, probable intraorbital collection on the right, in addition to reduced proptosis and peri-orbital soft tissue edema, and it was decided to maintain antibiotic therapy and start anticoagulation. After a month of antibiotic therapy, a new orbital MRI showed an abscess next to the superior rectus muscle, and ophthalmic surgery was chosen for drainage, which occurred without interurrences. After almost two months of Hospitalization and with clinical improvement, the patient was discharged with mild restriction of right orbit eye movements, being followed up as an outpatient. **Discussion:** the patient in question presented rare and serious complications of rhinosinusitis, both ophthalmic and intracranial. Anatomical factors that explain this infectious dissemination are: close relationship between the orbital contents and the ethmoidal labyrinth, which is separated from the orbit by the papyraceous lamina; the existence of a large number of sutures and neurovascular foramina in the medial wall of the orbit; the optic canal which is closely adjacent to the posterior ethmoid and sphenoid sinuses; and valved ophthalmic veins that communicate with vessels inside the orbit and directly with the cavernous sinus, presenting a direct blood flow, facilitating hematogenous dissemination. The presence of eyelid edema and headache are warning symptoms for a more attentive investigation of rhinosinusitis, in addition to intravenous antibiotic therapy. **Final Comments:** the importance of anatomical knowledge is emphasized for understanding the routes of spread of facial and intracranial infections.

Neuroinfecção

TL 1105397

PSEUDOTUMORAL NEUROCYSTICERCOSIS

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Case Presentation: Woman, 38 years old, no comorbidities, is forwarded to Neurology due to recurrent tonic-clonic seizures on the right, and secondary generalization, with sudden onset one month ago. The patient lives on a rural property, having agricultural and swine farming activities. In the examination, does not present focal signals, however, shows impairment in attention, fixation memory, and basic math operations. Image exam was realized, and brain MRI evidenced a single cystic lesion, easily identified, with 5 cm in the largest diameter, homogeneous and well delimited, with a liquor-like component in all MRI sequences, without contrast capture, in the left parietal lobe with mass effect. It was initiated carbamazepine, albendazole, and dexamethasone, and, after, valproic acid, without modification of the condition, remaining symptomatic and with the same seizures. Parietal craniotomy was realized to aspirate the cystic lesion, obtaining thereabout 80 ml of clear liquid, and capsule and scolex exeresis. In the follow-up, after one year, there has been regression on the image exams, showing residual encephalomalacia area, remission of the seizures and the cognitive deficit, and the anticonvulsants were suspended after three years without attacks. **Discussion:** Neurocysticercosis is an important cause of epilepsy in developing countries, with behavior changes beyond the increased intracranial pressure. Even as the presented case, cerebral lesions are commonly found in brain imaging exams, due to the installation of the parasite in brain tissue and the immune reaction produced. The treatment, occasionally, is a challenge for the professional, needing clinical approach with antiparasitic and anticonvulsants, besides the surgical approach. In the case in question, all the forms of treatment were made, aiming at the remission of the parasite and the symptoms. **Final Comments:** This case evidences a rare form of neurocysticercosis of giant brain cyst, with the differential diagnosis of neoplastic lesions. The treatment of this patient was effective, with efficient results. Thereby, it is necessary to pay attention to the symptoms and the disease incidence in our region, aiming at the improvement of the patient's clinical condition.

Neuroinfecção

TL 1105402

SYPHILITIC OPTIC NEURITIS

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Case Presentation: A 46 years old male, with type 2 diabetes mellitus. He reports sudden decrease in the visual field in the right eye without other associated complaints. After 20 days without improvement, referred to the Ophthalmology service, who found a decrease in visual acuity and papilledema on funduscopy, without impairment in oculomotricity and normal neurological exam. In the investigation, with normal laboratory tests, non-reactive viral serology, VDRL (+); FTA-Abs IgM(-) IgG(+), negative autoantibodies, CSF without pleocytosis and VDRL (-), requested brain MRI, normal, and orbits with hypersignal in the optic nerve in the orbital region on the right. He reports about 3 years ago, a popular lesion on the penis, painless, disappearing later, and intermittent erythematous cutaneous lesions on the extremities, without medical consultation. He remained in the ward for 14 days under treatment with Ceftriaxone – HICC guidance. After discharge, follow-up at an ophthalmic outpatient clinic with a marked reduction in eye symptoms. **Discussion:** Syphilis can affect the CNS at any stage of its natural history. Early neurosyphilis, less common, appears soon after syphilitic infection, and ocular involvement may be the first manifestation. As in the case described, in the presence of isolated inflammatory optic neuritis, there is no clinical finding or specific test that differentiates the syphilis lesion from other causes of optic neuritis. Optic nerve involvement can be unilateral or bilateral, leading to a decrease in visual acuity, with evidence of optic perineuritis, papillitis, neuroretinitis, retrobulbar neuritis and papilledema and the need to perform a lumbar puncture in all cases of neurological or ophthalmological symptoms and the immediately indicated treatment with antibiotic therapy. If treatment is not instituted properly, cranial nerve injuries can be permanent. **Final Comments:** The case shows an atypical manifestation of neurosyphilis. In this way, we emphasize the importance of paying attention to non-specific neuro-ophthalmological disorders and calling the attention to this disease that, unfortunately, still high prevalent in the population.

Neuroinfecção

TL 1105403

SPONDYLODISCITIS ASSOCIATED WITH PSOAS ABSCESS

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Case Presentation: 42-year-old male, smoker, with hepatitis B. One year ago, suffered injury from trampling. Six months ago, presented insidious, diffuse and daily low back pain with progressive worsening related with paresthesia in perineum, buttocks, thighs and feet. Ten days ago, reported decreased strength in the lower limbs and impaired gait. He denies fever and weight loss. The physical examination revealed left paravertebral hypertonia in L3-L4, pain on distal mobilization in bed, anti-algic and parietic gait and ortho support. Proximal strength in right lower limb 5/5 and distal 0/5, proximal strength in left lower limb 4/5 and distal 1/5, hypoactive and symmetrical osteotendinous reflexes, hypoesthesia in the bilateral plantar region. The lumbar spine and abdominal tomography (CT) showed L3-L4 spondylodiscitis, voluminous heterogeneous fluid collection in the left psoas muscle with intrasomatic extension in L2 that reaches spinal canal compressing nerve roots. The laboratory tests were normal and serological tests VDRL, HIV and IGRA were negative. Broad-spectrum antibiotics were started. Ultrasound-guided percutaneous lumbar puncture was performed with aspiration of 100 ml of purulent fluid that was sent for culture and showed no growth of bacteria. The CT control presented abscess remission and reduction of material in the spinal canal, besides improving the pain and gait. At discharge, antibiotic therapy was maintained with clindamycin and ciprofloxacin, putti vest and return for follow-up of spondylodiscitis. **Discussion:** With unspecific symptoms, low back pain due to spondylodiscitis may present with fever, claudication and prostration. Psoas abscess, secondary to infectious hematogenous lesions, tuberculosis, diabetes, tumors, and trauma, was the etiology of disc infection in our case. The diagnosis was confirmed by the history, neurological evaluation and imaging tests, with CT being the gold standard. Although we did not conclusively identify the origin of the abscess, an association with the trauma previously reported is valid. The treatment was based on abscess drainage, antibiotic therapy, and lumbar immobilization for follow-up. **Final Comments:** We would like to emphasize the importance of performing an abdominal CT to the diagnosis of spondylodiscitis caused by the psoas abscess. This condition presents with a complex and little-known diagnosis, related with several other pathologies and which, in our case, triggered a favorable outcome.

Neuroinfecção

TL 1105419

SPINAL TUBERCULOSIS MIMICKING A COMPRESSIVE PARAVERTEBRAL TUMOR: A CASE-BASED UPDATE

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Case Presentation: A 32-year-old immunocompetent male patient developed low back pain that, after 7 months, evolved with paresthesia of the left lower limb (LL), followed by paraparesis, paraparesis, and sphincter incontinence. Neurological exam revealed T10 sensory level, muscle strength grade 2/5 in limbs, patellar and aquileu reflexes grade 4/4, Babinski's and switchblade sign present, and diffuse clonus. Magnetic resonance imaging (MRI) of the spine was performed, which suggested extensive spinal neoplasia, with involvement of the vertebral body on T4 and significant spinal cord compression. A laminectomy was performed, raising the possibility of tuberculosis (TB) vertebral. The regimen for TB was started for 2 months, until the diagnosis was confirmed with a positive sputum test for BK, and the regimen was extended to 12 months. On the 11th postoperative day, the anatomopathological examination revealed chronic granulomatous osteomyelitis with a tuberculoid pattern, without malignancy criterion. After 5 months, he was readmitted with multiple T1 to T6 lytic lesions and extensive subcutaneous collection, consistent with an infectious process. Drainage was performed and antibiotic therapy was started. He had a new infectious condition 2 months after discharge, requiring another decompressive laminectomy, drainage of the collection and an antibiotic regimen for 42 days. Since then, it has evolved with significant neurological improvement, with gains in strength and physiological sphincter functions. **Discussion:** Responsible for 1-2% of TB cases, spinal involvement, more common in immunocompromised patients, results from hematogenous dissemination, with the thoracolumbar junction being the most affected site. The manifestations are nonspecific, insidious and progressive, in order to mimic many other clinical conditions, such as paravertebral tumors themselves, which makes the diagnosis a challenge. MRI is the best imaging test for this extrapulmonary form of TB, but it is worth noting that radiological tests are not always capable of making this diagnosis. As a result, the anatomopathological study is the gold standard. Treatment lasts 12 months, while the prognosis depends, in part, on the degree of established neurological impairment. **Final Comments:** It is important to be aware of the different presentations of spinal TB, which can often mimic a malignancy, in order to avoid diagnosis and treatment delay.

Neuroinfecção

TL 1105453

CEREBRAL SCHISTOSOMIASIS: NA IMPORTANTE DIFFERENTIAL DIAGNOSIS OF BRAIN TUMOR

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Case Presentation: A 24-year old patient, without comorbidities, started to present in March 2021 with seizures. He was admitted to the service with a neurological examination without alterations, but when performing a cranial tomography, hypodensity was observed in the right parieto-occipital region, being hospitalized due to the initial clinical suspicion of a brain tumor, later on a cranial resonance that showed a swollen lesion involving the subcortical white matter. Of the right superior frontal gyrus, characterized by hypersignal on T2/FLAIR sequences and with tiny nodular foci with hypointense and intense post-gadolinium enhancement, without diffusion restriction. In the evaluation of the cerebrospinal fluid, there were no changes in cellularity, proteins and glucose, and it was not possible to perform immunology due to unavailability in the service. Based on the findings of the imaging exam, the possibility of cerebral schistosomiasis was raised and a biopsy was performed, which showed the presence of *Schistosoma mansoni* eggs. Treatment with praziquantel and corticosteroid was performed. Patient evolves with neurological examination without alterations and without new episodes of seizures. **Discussion:** Schistosomiasis of the central nervous system (CNS) is relatively rare, being called neuroschistosomiasis (NE). It occurs mainly in the spinal cord, accounting for about 6 to 13.4% of myelopathies in endemic areas. Intracranial localization, notably in the cerebral hemispheres, is much rarer. NE can be clinically manifested by meningo myelodradiculitis, myelitis, radiculitis, or by the cerebral medullary pseudotumor form. The most common presenting symptoms are cognitive impairment, headache, seizures, and focal neurologic deficits. **Final Comments:** NE with cerebral involvement is very rare, but it should always be suspected in patients with images suggestive of tumors and who live in endemic areas. Early and adequate treatment is of paramount importance to minimize CNS tissue damage and prevent chronic sequelae.

Neuroinfecção

TL 1105469

HUMAN HERPESVIRUS 7 (HHV-7) ENCEPHALITIS: A CASE PRESENTATION

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Case Presentation: Female, 15 year-old, black, previously healthy and immunocompetent, presented to the emergency room with a history of fever, skin rash, headache, diarrhea, feet edema and polyarthritis in January/2022. On the neurological examination, she had drowsiness, mental confusion, psychomotor agitation, memory alteration, generalized motor epileptic seizures, visual field alterations, drooling, urinary incontinence, and objective muscle weakness. A syndromic diagnosis of encephalitis was given, and venous acyclovir was prescribed, with no improvement of the neurological condition. She was then referred to a tertiary Hospital facility. On admission, no changes on the segmental physical examination were present. In the neurological examination, she had a Glasgow Coma Scale of 9 points, mental confusion, memory changes and global aphasia. Visual acuity examination, strength, sensitivity, and reflex were not able to be performed. There were no changes in pupillary reflex and ocular motility. She was then admitted to the intensive care unit and mechanical ventilation was started. Head CT, MRI and EEG were normal. Serum and cerebrospinal fluid (CSF) samples were negative for Zika and Chikungunya viruses and indeterminate for dengue fever virus by PCR method. Multiplex PCR, an exam to simultaneously identify human adenovirus, cytomegalovirus, enterovirus, erythrovirus B19, human herpesvirus 7 (HHV-7), human parechovirus, human herpesvirus 6, Epstein-Barr virus, herpes simplex virus 1, herpes simplex virus 3 and varicella-zoster virus, was performed and positive in the CSF for HHV-7 and negative for the others tested. **Discussion:** Primary HHV-7 infection usually occurs during childhood and presents mainly as a sudden exanthema, with a later latency state and possible reactivation in immunodeficiency. **Final Comments:** However, although rare, cases of encephalitis and post-infectious autoimmune encephalitis related to HHV-7 in immunocompetent patients have already been reported in the literature. This possibility should be considered when the main causes of encephalitis are ruled out. In such cases, multiplex-PCR should be performed, allowing for a rapid and comprehensive etiological diagnosis.

Neuroinfecção

TL 1105531

POST-COVID LEUKOENCEPHALOPATHY WITHOUT SEVERE HYPOXIA WITH EARLY TREATMENT

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Case Presentation: A 44-year-old male patient sought medical evaluation due to concentration and memory deficits initiated three months before. He also referred nausea, excessive irritability, dizziness, and a moderate-intensity headache in the period. His medical history disclosed a previous mild COVID-19 about 5 months before, confirmed by polymerase chain reaction (PCR). The patient did not present hypoxia or great respiratory symptoms in the period that needed further care. He had taken, 1 month before the infection, the first dose of AstraZeneca vaccine and the second dose, 3 months after the disease. There was no evidence of drug and alcohol abuse, intoxication with exogenous substances, radiation exposition or chemotherapy. Physical and mental examination did not show any alteration during tests. An encephalic Magnetic Resonance Imaging (MRI) revealed coalescent hyperintense signal abnormalities in white matter of corona radiata, periventricular, frontoparietal and subcortical temporal lobes in both hemispheres. In addition, in T2/FLAIR sequence, small oval hyperintense abnormalities after contrast in both thalami, cerebellum's white matter, pons, amygdala, and dentate nucleus. There were no hippocampal alterations. Blood count, cerebrospinal fluid analysis and serologies were performed, showing no alterations or plausible causes for the findings. Considering the possible causes and the exclusion of the majority of plausible causes, the diagnosis was given as Acute Diffuse Leukoencephalopathy secondary to COVID-19. Based on previous reports, it was initiated treatment with corticotherapy. After 10 days of treatment, the patient referred great improvement of symptoms, with remission of dizziness. This pathology is mostly related to microhemorrhages, severe hypoxia and, during the pandemic, associated with long-term hospitalization and mechanical ventilation. In this case, the patient showed demyelinating patterns associated with a rare complication of SARS-CoV-2 infection with direct lesions of the nervous cells of the brain. The diagnosis of this pathology and timely treatment is difficult, considering negative findings in physical and mental exams as well as laboratorial testing. Most patients do not have access to MRI and probably will not receive an appropriate diagnostic. The probability of certain medical care is lowered even more if examiners restrict the occurrence of the pathology only in critical ill patients with long exposure to hypoxia.

Neuroinfecção

TL 1105534

ACUTE DISSEMINATED ENCEPHALOMYELITIS (ADEM) AFTER COVID-19 INFECTION: CASE PRESENTATION

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Case Presentation: Woman, 56 years old, with a previous history of infection by covid-19 for a month. Denies smoking, alcoholism or drug addiction, works in an accounting office, goes to the emergency service with a report that 24 hours ago she started left hemihypoesthesia and left rhyme deviation, with no additional complaints. On neurological physical examination, the patient has right peripheral facial palsy and left hemihypoesthesia. During the investigation of the case, computed tomography did not show any changes. Brainstem audiometry (BERA) showed bilateral retrocochlear dysfunction. CSF with a slight increase in protein. Magnetic resonance imaging (MRI) showed a right paramedian image at the medulla-pontine transition, without expansive or atrophic features. Raising the hypothesis of acute disseminated encephalomyelitis (ADEM). We started pulse therapy with methylprednisolone 1g/day for 05 days. The patient evolved in good general condition, eupneic, phasic, conscious and oriented; also reports improvement in the sensation of paresthesia in the left side and of facial paralysis. **Discussion:** Acute disseminated encephalomyelitis (ADEM) is an inflammatory demyelinating condition of the central nervous system, which is usually caused by viral infections, however, it can also be triggered by vaccines or not have an apparent cause. Although its incidence is not fully known, it is known that it is an uncommon disease in adults, occurring more frequently in children. The classic picture of ADEM occurs, on average, 26 days after an infectious condition, manifesting itself with acute multifocal neurological symptoms, often evolving with vertiginous deterioration. Most patients have motor and/or sensory deficits, which may be accompanied by other signs and symptoms such as headache, malaise, ataxia, aphasia and nystagmus. The diagnosis is made from clinical presentation and history, neuroimaging and CSF analysis. The prognosis of the patients with ADEM is mostly favorable, with total or almost complete recovery after approximately 6 months of presentation. **Final Comments:** Thus, this clinical case is uncommon due to the low frequency of ADEM after infections, especially in adults. In addition, this is a rare manifestation resulting from COVID-19 infection, and it can be a condition that leaves sequelae.

Neuroinfecção

TL 1105551

SEPTIC THROMBOPHLEBITIS OF THE CAVERNOUS SINUS AFTER A HYMENOPTERA ACCIDENT: A CASE PRESENTATION

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Case Presentation: A 23 year old male patient, previously healthy, was admitted to the emergency department reporting an insect bite on the nasal dorsum 05 days ago. Since then he had pain, edema and hyperemia in bilateral peri-orbital region, worse to the left, followed by decreased general condition, myalgia, headache, fever, neck stiffness and mental confusion. Laboratory tests showed signs of bacterial meningitis, and treatment with Ceftriaxone was initiated with improvement of the meningeal picture. Computed tomography (CT) of the skull showed signs of thrombosis of the transverse sinus, sigmoid and left internal jugular vein, confirmed by magnetic resonance imaging of the brain. Anticoagulation therapy was started with warfarin. Arteriography showed a carotid-cavernous fistula, but it was decided to maintain anticoagulation due to extensive central venous thrombosis. Besides the neurological picture, cavitary lesions were found on chest CT and bronchoscopy was requested for investigation, however the result of the biopsy showed only signs of inflammation with inconclusive result. After suspension of anticoagulation 24 hours before the bronchoscopy, the patient evolved with motor aphasia and cranial CT showed foci of ischemia. After returning to anticoagulation, there was improvement of neurological deficits. After stabilization and absence of active infectious signs, the patient was referred to vascular neurosurgery for fistula repair. However, a new pre-surgical arteriography showed total and spontaneous correction of the fistula through thrombosis of its path and also revascularization of central venous thrombosis. **Final Comments:** Septic cavernous sinus thrombosis is an uncommon and serious complication of facial infections, and occurs when the infected areas are close to the nose because this region is drained by the ophthalmic veins. About 20 to 30% of cases have fatal outcomes. The importance of this report lies in the rarity of the case associated with the singularity of the event of spontaneous fistula correction.

Neuroinfecção

TL 1105570

ONGOING CENTRAL NERVOUS SYSTEM DEMYELINATING MULTIPLE ACTIVE LESIONS POST MRNA VACCINE: CLINICAL AND SERUM BIOMARKER MONITORING

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Case Presentation: During COVID-19 pandemic, the central nervous system (CNS) involvement was associated with a more severe outcome. In face of these cases, the emerging challenges are the differential diagnosis with inflammatory demyelinating diseases, mainly the multiple sclerosis (MS). Patients with intrathecal synthesis of immunoglobulin G (IgG) and oligoclonal bands (OCB) during the first demyelinating event distributed in the space, excluding other etiologies, fulfill diagnostic criteria of MS allowing us initiate treatment in the more favorable therapeutic window. Otherwise, vaccines can trigger immune response, probably in individuals with genetic susceptibility, and relapsing events need, in specific cases, to be addressed with modifying drug therapies (DMD). Besides clinical and MRI activity serum biomarkers, light chain neurofilament (NFL) can contribute to decision making. Here, we presented a case of a 19-years-old female that has been followed up since December 2021, with neurological exams registers, EDSS, neuroimaging, cerebrospinal fluid and plasma analysis. Single molecule array (SIMOA) was used to analyze neurofilament light level chain (NFL) during the last 6 months. Fifteen days after mRNA COVID-19 vaccine, she presented paresthesia in four limbs, bladder incontinence, appendicular ataxia, visual impairment and mental confuse state. Cranium MRI showed multiple white and grey matter lesions, some of them with mass effect, brainstem and spinal cord lesions, various with gadolinium enhancement. CSF showed 4 cells, protein 38mg%, no viral, fungus or bacterial identification, and Reiber and Felgenhauer diagram showing intrathecal synthesis of Kappa free chain. For the last six months, she remained with active ongoing disease, presented new MRI lesions, several Hospitalization events, receiving intravenous methylprednisolone (IVMPN) treatment and plasmapheresis, leading to a partial improvement. In december-13-2021, NFL level was 247.94pg/mL; in January-17-2022, 216.24pg/mL and, in March-07-2022, 271.38pg/mL. The NFL level is correlated with ongoing disease, new MRI lesions and the EDSS score. In May 2022, we initiated treatment with Natalizumab. Acute demyelinating events post vaccine are generally monophasic. This patient fulfills aggressive MS criteria and differential diagnosis with recurrent ADEM here is a challenge to manage treatment. In this context, NFL level, besides clinical and MRI activity contributed to our decision to treatment onset.

Neuroinfecção

TL 1105581

COMMUNITY-ACQUIRED PSEUDOMONAS AERUGINOSA MENINGOENCEPHALITIS: A CASE PRESENTATION

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Case Presentation: A 66-year old woman with type 2 diabetes mellitus, chronic obstructive pulmonary disease (COPD) and chronic sinusitis was admitted to our institution presenting with a 8-day history of continuous holocranial headache, nausea and vomiting, without fever. She evolved with disorientation and seizures. Computed tomography of the brain was normal and cerebrospinal fluid (CSF) showed pleocytosis with a lymphocytic predominance. Ceftriaxone, ampicillin and acyclovir were started empirically promptly. *Pseudomonas aeruginosa* was identified in CSF culture, and the antibiotic regimen was modified for cefepime, oxacillin and meropenem. Cranial magnetic resonance showed bilateral frontal and sphenoidal sinusopathy, discontinuity of the sphenoid roof, without cerebrospinal fluid insinuation, purulent collections in posterior ventricles, empyema in the left temporal and occipital regions. A new CSF three days later maintained culture with *Pseudomonas aeruginosa*, so meropenem and ciprofloxacin were started. Antibiotic treatment lasted eight weeks, with a control lumbar puncture without bacteria growth. Sphenoid and ethmoid sinusotomy was performed by the otorhinolaryngology team. She was discharged after one month with mild paresthesia on the left hemiface, for outpatient follow-up. **Discussion:** Gram-negative bacilli are an uncommon etiology of community-acquired meningitis in adults, with the majority of the cases occurring in neonates or infants. *Escherichia coli* accounts for the most cases, but there are less frequent pathogens described, such as *Pseudomonas aeruginosa*, *Acinetobacter* and *Enterobacter*. Non-hematogenous bacterial invasion of CSF occurs in situations of compromised integrity of the blood-brain barrier, such as neurosurgery, mastoiditis, sinusitis and malformations. We described a rare case of an elderly patient who has been diagnosed with community-acquired *Pseudomonas aeruginosa* meningoencephalitis and a medical history of chronic sinusitis and COPD. This patient presented with headache and confusional state, the last may be the only clinical feature in the elderly. **Final Comments:** *Pseudomonas aeruginosa* is a rare cause of meningoencephalitis and has a significant morbidity and mortality. Further studies are needed to investigate if it is more common in patients with COPD and chronic sinusitis than in the general population due to the bacterial colonization in this profile of patients.

Neuroinfecção

TL 1105647

CRANIAL NERVE INVOLVEMENT BY NEUROSYPHILIS

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Case Presentation: A 67 yo female patient presented with right peripheral facial paralysis, followed by scotomas in the left eye (LE) and decreased visual acuity. On evaluation, she presented ocular pain and anterior uveitis, low visual acuity (20/100) and relative afferent pupillary defect in the LE, with scotomas in the left temporal hemifield, marked papillitis and bilateral red desaturation. There was also mild peripheral facial paralysis and right sensorineural hearing loss. She also had erythematous, scaly and diffuse infiltrative plaques and papules in the lower limbs, hypogastric, paravertebral and inframammary regions. The serum Venereal Disease Research Laboratory (VDRL) test was positive, with a titer of 1/512, and the Microparticle Chemiluminescence for Syphilis (CMIA) also had a positive result, of 24.21. In the cerebrospinal fluid, mild pleocytosis was observed, with 9 cells/dL, normal total protein (12 mg/dL), reagent FTA-ABS test and negative VDRL. Anatomopathological result of skin biopsy pointed lymphohistiocytic dermatitis, suggesting treponematosis. Brain Magnetic Resonance demonstrated enhancement in the auditory canals, greater on the left. The patient was treated with intravenous crystalline penicillin G for 14 days, with improvement of the skin lesions and visual acuity. **Discussion:** Neurosyphilis can occur at any stage of the disease, as the bacterium *Treponema pallidum* spreads throughout the body early in the infection, including the central nervous system. The patient in question had involvement of the cranial nerves most commonly reported in cases of neurosyphilis (II, VII and VIII), at the secondary stage of the disease. Syphilis diagnosis demands a positive treponemal test and a positive non-treponemal test, while neurosyphilis diagnosis requires a compatible clinical picture and cerebrospinal fluid. CSF in neurosyphilis is marked by pleocytosis and hyperproteinorreaquia. CSF's VDRL is highly specific with a sensitivity between 30-70%, therefore, it does not exclude the diagnosis. CSF treponemal test is highly sensitive but not very specific for the disease. **Final Comments:** Neurosyphilis is a manifestation that can occur at any stage of the disease, however, it is treatable and, if diagnosed and treated early, serious sequelae can be avoided.

Neuroinfecção

TL 1105668

NEUROFILAMENT LIGHT CHAIN (NFL), GLIAL FIBRILLARY ACIDIC PROTEIN (GFAP), UBIQUITIN CARBOXY-TERMINAL HYDROLASE L1 (UCH-L1) AND TAU PROTEIN LEVELS IN A SEVERE COVID 19 PATIENT WITH DOUBLE HOSPITALIZATION AND FAVORABLE OUTCOME

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Case Presentation: Coronavirus disease 19 (COVID-19) is an infectious disease caused by SARS-CoV-2 (Severe Acute Respiratory Syndrome related to Coronavirus 2). Most SARS-CoV-2 infected people experience mild self-limiting respiratory illness. Some, however, become seriously ill and require medical attention. Here we present the case of a 71-years-old male patient with comorbidities (chronic arterial hypertension, diabetes mellitus, previous stroke), and previous history of admission in Intensive Care Unit (ICU), intubation and coma due to COVID-19 in 2020. One year later, in July 2021, he had been admitted to the Clementino Fraga Filho University Hospital ICU for the same reason, when also presented myoclonic seizure; at that time, he was intubated for 2 weeks. Positive PCR for COVID-19 confirmed a second infection. Cranium Computed Tomography (September 2021) showed no acute changes but diffuse atrophy. MRI wasn't possible at the time. During the second ICU admission (2021), his blood sample was collected, so we could investigate neurological and immunological biomarkers (Neurofilament light chain - NFL; Glial fibrillary acidic protein - GFAP; Ubiquitin carboxy-terminal hydrolase L1 - UCH-L1; TAU protein; Interleukin-22 - IL-22), those were measured using SIMOA (Single MOlecule Array), an ultrasensitive technique for biomarkers quantification in peripheral blood. The patient showed the following levels for these biomarkers: NFL- 131,23pg/mL; GFAP- 615,12pg/mL; UCHL1- 115,24pg/mL; TAU- 1,36pg/mL; IL-22- 385,71pg/mL. Most of these values are considered high, when compared to age-matched control group, with the reference values of GFAP- 192,75pg/mL; NFL- 17,52pg/mL; Tau- 1,89pg/mL; UCHL1- 41,56pg/mL and IL-22- 7,49pg/mL. The only biomarker considered at normal levels was TAU protein, in agreement with expected due to his preserved memory and absence of dementia even after the second COVID-19 event. However, the GFAP, NFL, and UCHL1 were significantly higher when compared to the control. These are important markers of neuroinflammation and may be related to the myoclonic seizure and coma. It is important to highlight that he presented high IL-22 levels and that this IL has been considered a protective factor during SARS-CoV-2 infection, and probably an important biomarker for this favorable outcome. These intense variations together with the clinical picture of the patient may be an indication of the relevance of these biomarkers for the progression and outcome of COVID-19.

Neuroinfecção

TL 1105688

MIMICS OF NERVOUS SYSTEM CHAGAS DISEASE REACTIVATION IN HEART TRANSPLANTATION PATIENTS

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Case Presentation: A 48-year old male underwent heart transplantation for heart failure secondary to Chagas disease. On the second postoperative day, he had a tonic-clonic seizure and developed hemiparesis and aphasia. CT revealed superior sagittal sinus thrombosis, venous infarct and subarachnoid hemorrhage. Despite adequate anticoagulation, anti-seizure drugs adjustments and hemorrhage reabsorption, seizures and neurological deficit persisted. MRI revealed a necrohemorrhagic lesion suggestive of chagoma and a hypoplastic superior sagittal sinus. Another 48-year old male with a history of Chagas disease started, 24 days after heart transplantation, a confusional state in the absence of clinical findings that could justify a delirium followed by seizure two days later. Initial cerebrospinal fluid (CSF) analysis was normal and CT scan suggested posterior reversible encephalopathy syndrome (PRES). Tacrolimus was suspended and improvement was noticed afterwards. He was later readmitted confused and hemiparetic. Repeated CT showcased a new lesion with imprecise limits, vasogenic edema, cortical necrosis and hemorrhage indicative of chagoma. *T. cruzi* was identified in both CSF analyses. **Discussion:** Central nervous system (CNS) reactivation in a rare complication of heart transplantation in Chagas disease. It is manifested as meningoencephalitis (altered mental status, seizures, focal deficits) or tumor-like lesions called chagomas (causing seizure, focal deficits). Single or multiple necrotic-hemorrhagic nodular lesions in white matter can be seen in neuroimage. Diagnosis is usually made by a combination of clinical symptoms and detection of the parasite in blood, CSF or cerebral tissue biopsy. Treatment is recommended in asymptomatic or symptomatic patients. Benznidazole is the first line of treatment in the recommended dose of 10-15mg/kg/day, even though there are few studies on its interaction with immunosuppressive drugs. Although there is a relevant risk of chagas reactivation, transplant outcomes of end-stage Chagas cardiac disease are usually similar to other transplantation indications. **Final Comments:** We present two heart transplantation neurological complications that were first diagnosed as central venous thrombosis and PRES, but during follow-up were recognized as CNS manifestation of Chagas disease. This report highlights potential clinical and radiologic findings and the importance of Chagas disease reactivation as a mimic of other neurological diseases.

Neuroinfecção

TL 1105718

TUBERCULOUS PACHYMENINGITIS PRESENTING AS MOTOR NEURON DISEASE

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Case Presentation: A 63-year-old man presented with bilateral frontoparietal headache that progressed within 6 months to generalized weakness, hoarseness, dysphagia, left hemifacial neuralgic pain, unintentional weight loss of 20kg and no history of febrile illness. On examination, there was cranial nerve palsies of nerves V, VII, VIII, IX, X, XI XII on right side and V on left side; tongue and upper limb fasciculation, distal amyotrophy in upper limbs and global hyperreflexia. Brain MRI showed pachymeningeal thickening in the left middle cranial and posterior fossa, nodular lesions with low T2 signal in cerebellar vermis and right cerebellar hemisphere with growth towards cerebellofornix cistern and fourth ventricle. Cervical spine MRI was normal. Cerebrospinal fluid examination showed pleocytosis of 52/mm³ with lymphocytic predominance, hyperproteinorraquia and no glucose consumption. Ziehl-Neelsen, India ink, Gram stain and Xpert MTB negative. Serologies for syphilis and HIV negative. Electroneuromyography (EMG) presented chronic and subacute denervation of preganglionic origin of bulbar, cervical, thoracic and lumbosacral myotomes, without spontaneous activity. Based on clinical and paraclinical data, empiric treatment for tuberculosis was started. Diagnosis was confirmed by bronchoalveolar lavage with a positive Xpert MTB/RIF test. Despite therapy, patient presented clinical complications and died after 2 months. **Discussion:** Tuberculous hypertrophic pachymeningitis usually presents with cranial signs and symptoms such as headache and cranial nerve palsies, or spinal symptoms such as radicular pain. Reported patient showed progressive cranial nerve palsies and on brain MRI, the extensive dural thickening involving the skull base explains all the cranial nerve involvement. He also had signs of upper and lower motor neuron involvement without radicular pain, sensory deficits or sphincter complaints, which is not typical of tuberculous myeloradiculopathy. On cervical spine MRI there was no thickening or enhancement of meninges, but EMG indicated symmetric pre-ganglionic involvement of nerves, probably secondary to a radiculopathy only of the anterior root. Our case shows that this topography should be considered in patients who presents with motor neuron disease and have some alarming features such as new onset headache. **Final Comments:** We presented a case of tuberculous pachymeningitis with extensive cranial nerve involvement and motor neuron symptoms.

Neuroinfecção

TL 1105724

SPASTIC PARAPARESIS IN A 45-YEAR-OLD WOMAN POST MILD SARS-COV-2 INFECTION

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Case Presentation: We present the case of a 45-year-old woman without previous comorbidities, who sought the emergency unit of our university Hospital with a 1-month history of asymmetrical paresthesia in lower limbs, worse in the right side, progressing to both superior limbs after a few days. She had a recent diagnosis of SARS-CoV-2 infection, confirmed through a positive rt-PCR performed ten days before the Hospital admission, with mild respiratory symptoms. Along the following week, she presented ascending weakness (worse on the right side), spasticity and weakness in the lower limbs with necessity of a wheelchair and marked constipation. Physical examination at admission evinced: asymmetrical spastic paraparesis with brisk reflexes, ankle clonus and Babinski sign, diminished sense of vibration and proprioception in lower limbs and absence of sensitive level. We admitted her to the neurology ward, proceeded with brain/whole spine magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) analysis. The MRI did not present any significant alterations and the CSF showed a mild lymphocytic pleocytosis that didn't remain in further evaluation. Specific autoimmune panels and infectious investigations were negative. The hypothesis of autoimmune myelopathy was made and she received high dose (5g) methylprednisolone for five consecutive days, with poor response. After multiple follow-up MRI scans (1, 2, 5 and 6 months from initial symptoms) and a electro-neuromyography with unremarkable findings, we made the diagnosis of MRI-negative post-COVID myelopathy. **Discussion:** After COVID-19 pandemic, there have been rare reports of inflammatory myelopathy after SARS-CoV-2 infection. These patients typically had asymptomatic or mild-moderate COVID-19 disease and absence of radiologic evidence of structural spinal cord abnormality, which differs from previous reports that showed myelopathy in Hospitalized patients with severe COVID-19 disease and needs of critical care. This is a recently described entity, with only a few reports and unknown pathophysiology and prognosis. Up to date there is no effective treatment described, except rehabilitation. **Final Comments:** This case adds with prior reports and reinforces the need to increase clinical suspicion in presentations such as this in which there is a compatible epidemiological history and absence of MRI findings and raises the urge for more studies to better comprehension of pathophysiology and possible therapies.

Neuroinfecção

TL 1105730

MENINGITIS AND SPINAL ARACHNOIDITIS NEUROTUBERCULOSIS PRESENTATION: THE CHALLENGE OF DIAGNOSIS

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Case Presentation: A previously healthy 43-year-old man, who worked as a sand extractor in river benches, was admitted to the Emergency Department, presenting with paraparesis and urinary retention. He had previously sought medical care due to fever and headache four days earlier. Both cranial and thoracolumbar computerized tomography scan were normal. Cerebrospinal fluid (CSF) revealed the following: Glucose <20mg/dL, 96 white blood cells/mm³ (WBC) – 75% lymphocytes, protein 536mg/dL, negative nucleic acid amplification test (NAAT) for tuberculosis (TB), negative Ziehl-Neelsen staining and negative direct microscopy for acid-fast bacilli. Extra neural sites of TB disease were excluded after screening exams as well as neoplastic, inflammatory and other infectious diseases, including HIV. Magnetic resonance imaging was performed and revealed thick diffuse leptomeningeal enhancement, ischemic infarcts in basal ganglia, obstructive hydrocephalus and longitudinal extensive transverse myelitis, from T8 to T11. Due to high suspicion, standard anti-tuberculous therapy was prescribed for presumed TB. During treatment, six CSF samples were collected, showing progressive decrease of Glucose, protein and WBC levels. NAAT, direct microscopy and staining were negative in all samples. Despite medical assistance, patient died seven weeks after symptoms onset. Two weeks after his death, Mycobacterium tuberculosis was detected in CSF cultures. **Discussion:** Central nervous system (CNS) involvement is considered one of the most severe manifestations of extra-pulmonary TB. Immunosuppression is an important risk factor. For instance, HIV/AIDS patients have a five-fold increase in the likelihood of having CNS involvement. Despite being highly sensitive and specific, negative NAAT or any other test should not exclude the diagnosis. The present case presentation is relevant because the rapid TB test, NAAT, which is meant to speed up diagnosis and to guide a better clinical approach, was, in six samples, negative, despite the high suspicious for mycobacterium disease. **Final Comments:** Definitive diagnosis of CNS TB is difficult and may take weeks. This case shows the importance of valuing all the clinical conditions that suggest neurotuberculosis when approaching a patient. Despite the availability of modern and rapid molecular tests, diagnosis should not rely solely on them, and treatment should not be postponed.

Neuroinfecção

TL 1105741

DISSEMINATED HISTOPLASMOSIS AS A DIAGNOSTIC CHALLENGE IN AN IMMUNOSUPPRESSED PATIENT

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Case Presentation: A 38-year-old female patient with a history of systemic lupus erythematosus presented with spastic paraparesis, cognitive dysfunction and urinary incontinence. She underwent head CT and MRI, which showed communicating hydrocephalus. CSF analysis revealed monocytic pleocytosis (45 nc/μl), hypoglycorrhachia (12 mg/dL), significant increase in proteins (733 mg/dL) and was negative for cryptococcus, geneXpert, VDRL and bacterial and fungi cultures. Chest and abdomen CT evidenced calcified perihilar and abdominal lymph nodes, most indicative of tuberculosis (TB) or sarcoidosis. After full workup and multiple blood and CSF tests, TB was ruled out and the patient was treated with corticosteroids and cyclophosphamide, with no improvement. Despite regular immunosuppression, she continued to deteriorate. New CSF analysis kept same previous pattern. Blood radial immunodiffusion was positive for Histoplasma capsulatum. Liposomal amphotericin B was started, followed by oral itraconazole. After three months of treatment, she presents with mild cognitive deficit and discrete gait disturbance. **Discussion:** Histoplasma capsulatum incidence rate has increased with the advance of immunosuppressive therapies in recent years, especially in endemic areas. Up to 10% of patients develop disseminated histoplasmosis via hematogenous dissemination, leading to meningitis, associated vasculitis, focal brain or spinal lesions, or hydrocephalus. Diagnostic tools are mostly nonspecific, and diagnosis relies on strong clinical suspicion, which may be a challenge. For our patient, it came up after ruling out TB, sarcoidosis and lupus hydrocephalus. CSF analysis usually shows monocytic pleocytosis, elevated protein and low glucose. CSF culture has poor sensitivity, and antigen and antibody testing in CSF are not widely available in our scenario. However, urine and blood antigen testing are available, sensitive and highly specific. Pharmacologic treatment should be maintained for at least 1 year, until CSF profile is resolved. If possible, it is ideal to stop immunosuppressive therapies. Relapses are frequent and may require prolonged treatment. **Final Comments:** Untreated histoplasmosis is associated with high morbidity and mortality rates. Diagnosis is critical for adequate treatment implementation and better outcomes. It is even more important to accurately diagnose fungal infection in patients with autoimmune diseases, since treatment plans are opposite.

Neuroinfecção

TL 1105757

FIRST REPORT OF HEPATITIS C PATIENT WITH NEUROMYELITIS OPTIC SPECTRUM DISORDER ANTI-AQP4 ANTIBODIES (+) AND REMARKABLE HIGH LEVEL OF NEUROFILAMENT LIGHT CHAIN

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Case Presentation: Hepatitis-related viruses usually stimulate extra hepatic manifestations in most of the cases. An important representative of these viruses is the hepatitis C virus (HCV), which is also associated to chronic silent infections that can last several years before a diagnosis, leading to cirrhosis and hepatocellular carcinoma. Our aim here is to present a rare phenotype of extra hepatic manifestation involving nervous system, the neuromyelitis optica spectrum disorder (NMOSD), and the correlation with plasma biomarkers level of neurodegeneration. We identified a 68-years-old female patient who has been diagnosed for the hepatitis C in 2014, with a sustained high viral load along the years (~10 UI/mL). In 2021, she received the diagnosis of NMOSD, while presenting HCV detectable in serum and symptoms related to this viral infection. By the time, her blood sample was collected and evaluated for anti-MOG (Myelin-Oligodendrocyte Glycoprotein) and anti-AQP4 (AQUAPORIN 4) antibodies detection by CBA (Cell-Based Assay), which showed positivity for anti-AQP4 antibodies. Neuroimaging showed extensive lesion involving of brainstem and extensive central spinal cord lesion from cervical to l dorsal level. During the onset of NMOSD, she was also presented urinary infection, hemorrhagic gastritis and high HCV viral load, followed by several Hospitalization events, with active disease since then. For a better understanding of this case, we evaluated neurofilament light chain (NfL) levels using SIMOA (Single Molecule Array). The plasma NfL level in 2022 was 365.51 pg/mL (SD=48.39) that compared with matched age control group was very high (~11.73 pg/mL). This result corroborates the severity findings for patients in neuroinflammatory and neurodegenerative processes associated to viral trigger and can contribute to better understanding the large spectrum of NMOSD. The NfL levels can contribute as biomarker for neuroinflammatory and neurodegenerative disease, including those associated to viral trigger. Here, we show the first case of HCV patient positive for anti-AQP4 antibodies, reinforcing that there is a wide neuromyelitis spectrum disorders associated to AQP-4 antibody identified after different viral trigger, suggesting a probable relationship between viral infections and neurological manifestations.

Neuroinfecção

TL 1105771

BILATERAL THROMBOSIS OF THE MIDDLE CEREBRAL ARTERY BY NEUROSYPHILIS: AVCI

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Case Presentation: Male DC 61 years, complaint of right hemiparesis, dysarthria and sudden onset confusion 2 days ago. With progressive improvement, without alteration in the neurological exam already in 1 care. Initially, a protocol was performed for ischemic stroke (ICAVs) with skull resonance: ischemia sequelae in the left temporal and parietal lobe and left postcentral gyrus transthoracic echocardiogram and Doppler ultrasound of carotid and vertebral arteries without significant alterations. In laboratory investigation: fta abs reagent and vdrl 1/64. The study was carried out on cerebrospinal fluid: Leukocytes 70, neutrophils 2, lymphocytes 98, glucose 54 and proteins 75.2. In the face of an etiological finding (neurosyphilis causing vascular injury), a patient submitted to intracranial arterial angio resonance (MMR), showing: Distal subocclusive stenosis of M1 in right MCA, parietal irregularities in M1 segment of left MCA and M2 bilaterally, and parietal irregularities with moderate stenosis in P2 to the right. RMIA confirms the etiological hypothesis, due to its classic pattern of arteritis by syphilis (Heubner's arteritis). DC was treated with crystalline penicillin for 14 days and kept asymptomatic on discharge. The patient unknown the diagnosis of syphilis, and it was impossible to determine the incubation period and showed comorbidities, only smoking. **Discussion:** Syphilis, sexually transmitted, if untreated can reach its tertiary form reaching the central nervous system. DC presents the classic type of meningeal neurosyphilis. It has mainly manifested the AVCI secondary to vasculitis caused by treponema pallidum. The agent causes tapering of the vessel affected by fibroelastic proliferation of the intima, narrowing of the media and fibrosis associated with adventitia alterations, with preference for medium and large caliber arteries. There is appearance and collateral circulation, which explains clinical practice not compatible with image findings. As shown in the report, a poor clinic for an exuberant image. **Final Comments:** Approximately 30% of patients without adequate treatment for neurosyphilis will present CNS involvement, them 70% are asymptomatic. Associated with a non-exuberant clinic and the growing number of syphilis cases, with 72.8 cases/100,000 inhabitants in Brazil, it is always necessary to continue the investigation of these infections in CNS, using the vdrl test as proposed by the Ministry of Health as a screening

Neuroinfecção

TL 1105843

PROGRESSIVE MULTIFOCAL LEUKOENCEPHALOPATHY: AN ATYPICAL PRESENTATION

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Case Presentation: A 45-year-old woman, with no known comorbidities, presents to our service with progressive onset of weakness in her right upper and lower limb. Concomitantly, she developed tingling in her right hemiface and dysarthria. Neurological examination showed right peripheral facial nerve palsy, soft right palatal palsy, in addition to complete and proportionate hemiparesis on the right, with signs of ipsilateral pyramidal release. At investigation, anti-HIV 1/2 reagent serology was found. CT brain scan showed subcortical hypoattenuation in pre and postcentral gyrus on the left. Brain MRI had evidence of hypointense signal on T1 and hypersignal in T2-FLAIR at the topography of the pons and cerebellar peduncles, as well as in the supratentorial topography mentioned above at head CT, involving subcortical U-fibers, and without water diffusion restriction or contrast enhancement. The possibility of progressive multifocal leukoencephalopathy (PML) was then raised, with confirmation after positive JC Virus research in cerebrospinal fluid. **Discussion:** PML is a progressive demyelinating disease of the Central Nervous System (CNS) caused by the JC Virus in immunocompromised patients. Among the conditions most associated with this disease, the best known is HIV infection, in the AIDS phase, however cases of PML have already been described in other contexts of immunodeficiency, such as cancer, organ transplantation, and autoimmune diseases. Concerning the diagnosis, in addition to the clinical history, the patients present, on head CT with asymmetric areas of hypoattenuation in white matter. On MRI, it characteristically presents with hypointensity on T1 and hyperintensity on T2-FLAIR in white matter, as well as the absence of water restriction diffusion and contrast enhancement, typically in supratentorial topographies. The diagnosis is confirmed by the positivity of JC Virus in cerebrospinal fluid of suspected patients. **Final Comments:** Our patient had an atypical presentation of PML, with infratentorial lesions. It is necessary to consider this diagnosis in the face of an adequate clinical context. Despite the atypical presentation, there are similar cases described in the literature, particularly in the context of immunodeficiencies other than those associated with HIV. We hope that this case presentation can help other colleagues in the future who are faced with the same radiological images.

Neuroinfecção

TL 1105859

NEUROLOGIC MANIFESTATION CAUSED BY WEST NILE VIRUS: A CASE PRESENTATION

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Case Presentation: A 39 years old male, physician, traveled to Egypt recently, where he presented odynophagia, myalgia, painful cervical lymphadenopathy, diffuse rash and fever. Symptoms were self-limited for a period of 15 days. Approximately two weeks after the episode, he started feeling burning pain in the scalp, in the left parietal region, associated with dysesthesia. It evolved with pain in the bilateral pectoral region with progression to the back at T4 sensory level, radiating to the thoracoabdominal region on the right to the superior iliac crest and later to the right leg. He denied weakness, mental confusion and dysautonomic symptoms. Cervical ultrasound showed lymph nodes with a reaction pattern. Screening with viral serologies was started and MRI of the spine and brain was performed, which showed areas with hypersignal on T2 in the spinal cord extending from T8 to T11 without anomalous enhancement. CSF was collected with opening pressure of 30cmH₂O; 2 cells/m² lymphocytic predominance; proteins 70mg/dL; ADA 2mg/dL. Positive serum West Nile IgM test. After confirmation of the diagnosis, the patient received 4 days of metiprednisolone 1g/day with satisfactory improvement of symptoms. **Discussion:** West Nile fever is caused by the West Nile virus of the flavivirus class and has been one of the major emerging causes of encephalitis in the US. Transmission occurs mainly through the bite of infected Culex mosquitoes, but vertical transmission and blood transfusion have already been described. Although most individuals who contract the virus are asymptomatic, symptoms such as fever, rash and myalgia are part of the clinical presentation. Neuroinvasion usually occurs in less than 1% of those infected and is characterized by aseptic meningitis, meningoencephalitis, acute flaccid paresis, rhomboencephalitis, myelopathy, polyneuropathy, or radiculopathies. The form of neurological involvement varies according to the seasonality and location of the epidemic. CSF usually shows pleocytosis with a lymphocytic pattern, normal glucose, high protein levels (>900mg%). West Nile virus IgM titers tend to remain high even after acute infection. **Final Comments:** In Brazil, West Nile Fever is not a common cause of myelopathies, and other diagnoses, such as arboviruses, are primarily considered. In the aforementioned case, the epidemiology retrieved during the anamnesis was essential for the investigation and conclusion of the proper diagnosis.

Neuroinfecção

TL 1105863

POST-VARICELLA FOCAL CEREBRAL ARTERIOPATHY: A RARE CAUSE OF ISCHEMIC STROKE

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Case Presentation: A 15-years-old male patient presented to the emergency department complaining of sudden-onset left hemiparesis since the previous day. He also complained about left hemibody paresthesias one week before admission. There was no prior history of illness, vaccinations, travel or trauma. However, the parents said that the patient had contact with the sister who had chickenpox months before the event. Neurological examination revealed complete and disproportionate left hemiparesis, predominantly in the left upper limb, with pyramidal signs. The patient underwent computed tomography, which did not initially reveal any abnormalities, but cranial magnetic resonance revealed signs of ischemia in right basal ganglia and internal capsule and Vessel wall evaluation revealed signs of focal vasculitis in the M1 topography of the right middle cerebral artery. The patient underwent lumbar puncture, which revealed CSF pleocytosis with a predominance of lymphomononuclear cells, serology for Varicella Zoster reagent in the CSF (IgG 1:8) and in the blood (IgG > 5000 and IgM < 0.20). The treatment consisted of high doses of corticosteroid therapy (1g of methylprednisolone per day for 5 days) in addition to Acyclovir (800 mg/kg for 14 days) due to evidence of possible underlying Varicella Zoster infection. In evolutionary control imaging, complete resolution of the findings suggestive of vasculitis in ACM D described above was noted. **Discussion:** Focal cerebral arteriopathy consists of a vasculitis that affects a specific topography of the cerebral circulation, usually affecting the distal portion of the internal carotid artery and proximal to the middle cerebral artery. Despite several described causes, varicella infection is one of the main causes in pediatric patients or those close to this age group. Vascular inflammation can occur in a para-infectious context or even months after the chickenpox condition. This case demonstrates the importance of investigating not only the patient's history, but also the history of indirect contact with the virus. **Final Comments:** focal cerebral arteriopathy is a rare cause of stroke that should be kept in mind in young patients and investigation of varicella infection should be performed especially in the younger and pediatric population

Neuroinfecção

TL 1105874

MENINGOEPITHELIAL MENINGIOMA ASSOCIATED WITH TUBERCULOSIS: A CASE PRESENTATION

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Case Presentation: A 79-year-old woman admitted to the Hospital with a history of seizures and difficulty speaking. She reports thirteen years of infrequent headaches, no seizures or auditory symptoms and a lesion suggestive of meningioma confirmed via computed tomography, also reporting having been oligosymptomatic for 11 years with only mild leukopenia. Three years ago she presented frontal alopecia and after a new CT scan of the skull without contrast that showed an oval lesion, isointense in relation to the cerebral cortex with a regular contour, measuring 2x1.5 cm in diameter, of extra-axial left frontal parasagittal location in direct contact with the ipsilateral superior frontal gyrus without perilesional edema. In January 2020 he had a seizure with loss of voice for about five minutes and two months later he developed a second seizure with the same clinical manifestation. Magnetic resonance imaging showed an oval lesion located in the left frontal parasagittal region with a diameter of 2.5x2.0x2.0 cm and significant perilesional edema. After 6 months, in July of the same year, the patient presented six crises during a period of four days, and the surgical treatment of tumor excision was indicated. **Discussion:** Intracranial tuberculosis has several forms, including tuberculoma and tuberculous abscesses, which often mimic a meningioma radiologically, requiring histopathological and immunohistochemical examination to confirm the diagnosis. The present paper reports a case of coexistence between a meningoendothelial meningioma associated with multiple epithelioid granulomas with foci of caseous necrosis, possibly being the first case presented in anatomopathological images with immunohistochemistry of this association of a tumor with colonization by Koch's bacillus. **Final Comments:** This case study reported a meningoendothelial meningioma associated with multiple epithelioid granulomas with foci of caseous necrosis, thus being one of the first cases with this rare association and that in the anatomopathological examination with immunohistochemistry shows the presence of Koch's bacillus.

Neuroinfecção

TL 1105891

NEUROCRYPTOCOCCOSIS PRESENTED AS ACUTE MOTOR AXONAL NEUROPATHY: AN ATYPICAL MANIFESTATION

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Case Presentation: A 33-year-old man, who worked as air conditioning technician, attended to the emergency department with progressive tetraparesis, ptosis in the left eye, hearing loss and sleepiness, associated with a 2-weeks history of malaise, headache, lumbar and cervical pain. Physical exam revealed arterial hypertension (164/88 mmHg) and tachycardia (116 bpm), with no fever and a normal cardiovascular auscultation. The patient presented with flaccid dysarthria, anisocoric pupils (left 3mm and right 2mm), with direct and consensual reflex preserved, left-ptosis and impaired adduction and upward gaze of left eye. At the motor exams there was tetraparesis, with MRC score of the right arm of 4, left arm 4- and 3 of both legs accompanying global areflexia. CT brain scan was normal and cerebrospinal fluid (CSF) showed pleocytosis (1350 cells/mm³) and hyperproteinorrachia (215 mg/dL), suggesting an infectious process, the opening pressure was 54 cmH₂O. Gram staining was positive for encapsulated yeast, and cultures growth of *Cryptococcus gattii*. VDRL and HIV, HTLV, B and C virus serology were negative. Electromyography confirmed the presence of predominant motor acute axonal neuropathy (AMAN), an Guillain-Barre Syndrome (GBS) variant. It was promptly initialized treatment with immunoglobulin (IVIG), amphotericin B and fluconazole. With the treatment there was improvement of the strength, however he developed complications of the cryptococcal meningoencephalitis with refractory intracranial hypertension (a ventricular shunt was installed), severe hypoacusia and visual impairment. **Discussion:** GBS is an aberrant reaction of the immune system, in genetically susceptible individuals, triggered by typically gastrointestinal infection (*Campylobacter jejuni*). The exact mechanism of the autoimmune reaction is not yet clarified. The association with fungal infection is very rare, there are very few reports of this association. It is not clear if this case was classic Guillain-Barre Syndrome, or a rare manifestation of cryptococcus infection as acute motor neuropathy. **Final Comments:** This case highlighted a very rare manifestation of neurocryptococcosis, and despite the fungal infection IVIG was introduced to treat the AMAN with relative success. Further studies are needed to clearly demonstrate this association.

Neuroinfecção

TL 1105920

PARKINSONISM SECONDARY TO NEUROINFECTION: A FUNCTIONAL LOBECTOMY BY NEUROCRYPTOCOCCOSIS

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Case Presentation: A previously healthy 39 year-old patient started with progressive emotional lability, loss of appetite and hyporexia, associated with a depressive status 2 years ago. The case was initially associated with functional limitation. 12 months ago the initial symptoms worsened, and intermittent holocranial headaches with tremors, slow and clumsy member movements were associated. They were admitted in a specialized service for investigation. A parkinsonian syndrome was detected in the physical examination: plastic hypertonus, bradykinesia, with rest and postural bilateral, symmetric, high frequency and low amplitude tremors of the extremities. Hypomimia, cognitive loss with an amnesic component and bradypsiquism were also found. Routine laboratorial investigation showed no findings. HIV, syphilis and hepatitis serologies were negative, with normal levels of TSH and T4. CSF: OP 19 cmH₂O – Erythrocytes 250; white cells 100 (Lymph 86 | Neutro 15 | Mon 9), Glu 42; Ptn 262; negative acid-fast stain; negative Gram stain; general culture results were also negative. A rapid test for *Cryptococcus neoformans* was positive. Cranial MRI showed a T1 hypointensity signal and a T2 and FLAIR hyperintensity signal in the bilateral nuclear-capsular region, somewhat symmetric. There was also a post-contrast enhancement of these multiple confluent nodular points, with compromise of the ependyma of the lateral ventricles, III and IV ventricles and the basal cisternal leptomeningeal layer. With the diagnose of neurocryptococcosis in mind, treatment started with Amphotericin B, Fluconazole and initial fase corticosteroids, which were later associated with Prolopa and Leveticetam, with substantial improvement of the bradykinesia and bradypsiquism. Ambulatorial follow-up is being made to this day. **Final Comments:** Secondary parkinsonian syndromes are frequently associated with vascular diseases or drug usage, but other differential diagnoses include post-cranial cerebral trauma, neuroinfections, encephalitis, external intoxications and multiple sclerosis. This particular case stood out because of the low frequency of Parkinson syndrome secondary to a neuroinfection; specially a non-HIV infected patient suffering from neurocryptococcosis.

Neuroinfecção

TL 1106063

SIMULTANEOUS OCULAR AND OTOLOGIC SYPHILIS IN A HIV-INFECTED PATIENT: A CASE PRESENTATION

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Case Presentation: A 53-year-old male patient presented with bilateral rapidly progressive painless visual loss for nine days, associated with bilateral hearing loss, was admitted in our emergency service. The comorbidities were HIV infection with a CD4 count of 217 cells/mm³ and undetectable viral load. The neurological examination findings an important reduction of the visual acuity with light perception and vision of counting fingers bilateral. Also a papilledema bilateral with normal photomotor reflex and isochoric pupils. A hearing loss and a tinnitus complaint were present on admission, however no specific tests were performed. No pathological findings were found on cranial CT. CSF analysis demonstrated white cell count of 4 cells/μL, appropriate level of glucose, protein levels 87 mg/dL and a non treponemic test (VDRL) negative. Due to the optic neuritis hypothesis, a brain and orbital MRI was requested and no abnormalities were identified. An ophthalmological evaluation was made with the retinal mapping showing an elevated and hyperemic optic disc with blurred edges 360 degrees, especially in the left eye. A mild macular edema, secondary to papilledema, was present, normal blood vessels and no exudative or hemorrhagic lesions were present. To assess the etiology, the laboratory was screened for infectious agents such as toxoplasmosis and cytomegalovirus, all of which were negative. The serum treponemal test was reactive and the VDRL result was 1: 128. Based on the presence of ocular and otological involvement the hypothesis of simultaneous ocular syphilis and otosyphilis was made. The patient received intravenous therapy with ceftriaxone for 14 days, aqueous crystalline penicillin G was unavailable. A gradual improvement of signs and symptoms were presented with partial recovery of visual acuity and resolution of otological complaints. The patient was discharged for outpatient follow-up. **Discussion:** CNS involvement can occur at any stage of syphilis. Among patients with secondary syphilis, 18% may have neurological symptoms. Syphilis has a wide variety of clinical forms, it can affect the ocular or auditory level. Only one previous case found in the literature presents a clinic similar to this one. Syphilis affecting the auditory system is an uncommon but underdiagnosed condition. **Final Comments:** Although rare, this report is of great value to review the importance of differential diagnosis in neuroinfectology. Ethics Committee No: 5397799

Neuroinfecção

TL 1106123

MYELITIS POST DENGUE

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Case Presentation: A male, 32-year-old patient was admitted to the service due to a history of tetra paresis initiated three days before, progressing with lower limb paresis and urinary retention and inability to walk. Patients denied symptoms such as fever, itching or myalgia. He had no other comorbidities or continued medication. At the patient's examination, she presented a preserved level of consciousness, hypoaesthesia at L4 level and dysesthesia at T5 level and bilateral paresis of the lower limbs. In initial laboratory tests presented plaquetopenia, rapid test for Dengue positive, Liquor initially had leukocytes 99, lymphocytes 89, neutrophils 11, glucose 83 and proteins 118.3, lactate 3.3 and the absence of oligoclonal bands, anti-MOG and anti-aquaporin 4. No changes to initial skull tomography. Then, a column resonance was requested, which evidenced a C6-C7 signal alteration and in the entire dorsal column. MRI of the skull and orbits without alteration. Then initiated pulse therapy with methylprednisolone 1g for 5 days, the patient presented gradual improvement of symptoms, with improvement of sensitivity and strength of lower limbs. **Discussion:** Transverse myelitis is a clinical picture of loss of strength, autonomic and sensory dysfunction, and its relation to dengue is rare. Neurological manifestations resulting from dengue, are atypical, compared to other arboviruses, and within these manifestations are the most frequent encephalitis and Guillain Barré syndrome. The diagnosis is based on the patient's clinical, resonance exams and liquid analysis, and the presentation of the condition can appear about 4 days after classic symptoms of dengue, such as fever and myalgia. In this specific case, the patient did not present classic symptoms resulting from dengue, and the neurological condition was the only manifestation of the disease. The treatment proposed in the literature is pulse therapy with methylprednisolone for 5 days, which was performed in the case with adequate response to medication. **Final Comments:** Post-dengue myelitis is one of the rare neurological manifestations of this arbovirus, with a small sample described in the literature, and it is important to report these cases to better understand the pathological mechanisms related to dengue.

Neuroinfecção

TL 1106134

ATYPICAL PRESENTATION OF NEUROSYPHILIS AS STROKE-LIKE SYNDROME – A CASE PRESENTATION

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Case Presentation: J.C.O, male, 53y, admitted with sudden left hemiparesis, being referenced to the Neurology referral Hospital for thrombolysis window protocol. Upon admission, he had an 11 NIHSS score with temporo-spatial disorientation, mild facial palsy and dysarthria, paresis with hypoesthesia on left hemibody and bilateral amaurosis. He reported optic neuritis diagnosis when he presented bilateral amaurosis in 2013 and carried out an incomplete investigation at the time, by his own choice. In view of the situation, the case was then conducted as a Cerebrovascular Accident (CVA), but the patient was not thrombolysed due to a contraindication related to the time of admission. Etiological investigation exams were performed, showing normal results. Serology with VDRL and rapid test came back positive. The patient was then submitted to cerebrospinal fluid puncture, which showed increased cellularity of 24 and reactive VDRL. During Hospitalization, the family reported that the patient had been experiencing markedly cognitive impairment with memory loss, behavior and psychological changes. He was treated with Crystalline Penicillin according to the protocol and on the 14th day of treatment showed motor, sensory and cognitive improvement, being able to walk and carry out his activities without assistance, maintaining the previous amaurosis. **Discussion:** Syphilis is a sexually transmitted infection caused by the bacterium *Treponema pallidum*. It is estimated the incidence of 12 million cases per year, more than 90% being in developing countries. Neurosyphilis occurs in less than 10% of cases, and its most commonly observed form presents with neuropsychiatric changes. Only 3% of cases are associated with CVA, due to an infiltrative inflammatory process, most commonly of the middle cerebral artery. It is thought that the patient's previous optic neuritis may have been an even more uncommon manifestation of neurosyphilis. **Final Comments:** This case illustrates an atypical manifestations of neurosyphilis. It is important to remember that this condition can be a primary etiology for stroke, especially in patients under 50 years of age, as well as for optic neuritis. Therefore, a detailed evaluation is essential in patients with initial investigation for normal stroke or atypical natural history. Once the diagnosis is made, treatment can prevent an unfavorable evolution and provide a significant clinical improvement.

Neuroinfecção

TL 1106167

SEVERITY OF MYELITIS-ASSOCIATED RHOMBOENCEPHALITIS IN AN IMMUNOSUPPRESSED PATIENT

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Case Presentation: Rhomboencephalitis is rare, potentially serious and fatal, affecting the hindbrain (pons, medulla and cerebellum). The pathology has varied etiologies such as infectious, autoimmune and paraneoplastic. It may be associated with myelitis, causing motor, sensory and autonomic changes. The clinic is characterized by prodromes of fever, vomiting, and headache, followed by neurological manifestations such as cranial nerve paresis, paresthesia, and sphincter dysfunction. To report the case of a patient with Human Immunodeficiency Virus (HIV) without regular treatment, diagnosed with myelitis-associated rhomboencephalitis, by obtaining data from medical records and reviewing the literature on the subject. **Discussion:** R.A.C, 37 years old, admitted to the intensive care unit in April 2020, with a report of vertigo and paresthesia of the left lower limb with 7 days of evolution progressing with crural paraparesis and urinary retention. He has a history of using anabolic steroids, hepatitis B, presenting CD4 lymphocyte 48 and a viral load of 32,000 copies. On admission cranial tomography, he had extensive hypodensity in the subcortical region, cerebrospinal fluid puncture with pleocytosis, proteinorrachia and reagent serology for herpes zoster and cytomegalovirus. A contrast-enhanced magnetic resonance imaging was performed, revealing irregular uptake nodules at the transition of the middle cerebellar peduncles with the hemispheres and at the bulbopontine transition, in addition to poorly defined hypersignal in the left hippocampus, parahippocampal gyrus and ipsilateral inferior temporal, suggesting rhomboencephalitis. The MRI of the cervical and thoracic spine showed hypersignal typical of myelitis. Treated with pulse therapy associated with broad-spectrum, antifungal and antiviral antimicrobials in an intensive care setting with partial improvement of the condition. After 28 days of Hospitalization, he presented worsening and sepsis of pulmonary focus evolving to death. **Final Comments:** The case presented raises the discussion about the involvement of immunosuppressed patients by a pathology little reported in this population, in relation to other pathologies that affect the central nervous system in these individuals, evolving with a severe form, rhomboencephalitis associated with myelitis, a potentially fatal, like the picture described.

Neuroinfecção

TL 1106170

CEREBRAL VENOUS THROMBOSIS (CVT) AND ITS INTERFACE WITH ARBOVIROSES

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Case Presentation: A 50-year-old, female, previously healthy sought medical attention due to progressive, squeezing, diffuse headache associated with persistent fever (38°C), arthralgia and myalgia. Given the suspicion of arbovirus, the patient was transferred to a specialized service in Fortaleza. The admission exams revealed platelets of 20.000 mm³, positive PCR for dengue and positive IgM and negative IgG serology. However, during Hospitalization, headache intensity worsened, associated with nausea, vomiting, visual blurring and unprecedented tonic-clonic seizures in 4 limbs (post-ictal lasting 30 minutes). Due to clinical worsening, underwent Cranial Resonance and Angioresonance (arterial and venous phases) which revealed venous thrombosis in the transverse sinus and left sigmoid sinus. Absence of changes in the electroencephalogram, lumbar puncture with opening pressure of 25 cmH₂O, 10 cells (90% of lymphocytes), no changes in glucose, increase in proteins: 60 mg/dL. It is useful to point out that on this occasion the platelets were 175.000 mm³. Hematological and rheumatological examinations were normal. It was decided to start anticoagulation with low-molecular-weight heparin and later start warfarin 5 mg/day. The patient was discharged without complaints, with oral anticoagulation and follow-up at the CVT outpatient clinic. **Discussion:** Cerebral venous thrombosis (CVT) is a rare and often underdiagnosed condition. It presents with a wide spectrum of signals and with high variability. Headache is present in most cases. The presence of papilledema, vomiting, blurred vision, alteration of the VI cranial nerve, diplopia, focal deficits, seizures and changes in consciousness are highlighted. Hematological and rheumatological exams as well as the study of vessels with angioresonance are essential for the diagnosis. In this approach it is necessary to be attentive to the interface of arboviruses, an example of dengue, with its repercussions on the Central Nervous System in order to better understand the behavior of the disease. CVT is an uncommon complication in dengue. **Final Comments:** It is essential to be aware of the alarm signals (red flags) as a form of suspected secondary causes of headache. In this context cerebral venous thrombosis constitutes an important differential diagnosis, especially in women with persistent headache in the context of arboviroses.

Neuroinfecção

TL 1106187

ATYPICAL NEUROTUBERCULOSIS IN AN ADOLESCENT WITH CLINICAL PRESENTATION OF MENINGOENCEPHALOMYELITIS FORM: A CASE PRESENTATION

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Case Presentation: A previously healthy, 17-year-old woman, with a complete vaccination schedule, in April 2021, presented acute onset of moderate-intensity headache evolving after 2 days to reduced consciousness. Neurological exam: regular general condition, drowsy, uncooperative, and contacting. Presence of meningeal irritation. Ophthalmoplegia in left eye abduction and right eye with nystagmoid jerks. Right central facial. Right hypotonia and left elastic hypertonia (Ashworth-2). Muscle strength-MRC-2, bilateral. Hypoactive deep reflexes globally, bilateral Babinski. Empirical treatment was instituted immediately for bacterial, viral meningoencephalitis, corticosteroid, and therapy for neurotuberculosis. Rheumatological tests and serum angiotensin-converting enzyme negatives. Cerebrospinal fluid (pleocytosis-lymphomononuclear, hyperproteinorrachy and hypoglycorrhachia), negative results to gram, fungus culture and tuberculosis genexpert. Brain MRI with hypersignal of left nucleus-capsular; thickening and enhancement of supra and infratentorial meningeal planes; prominent supratentorial ventricles; focal nodular with peripheral enhancement in right insula. Cervicothoracic and lumbosacral MRI revealed thickening and enhancement of posterior leptomeningeal in D2 and D8; hypersignal D4 to D7 (edema), left enhancement of descending nerve root of S1 (inflammatory neuropathy); increased dimensions and leptomeningeal nodular foci enhancement in C7 and D11 segments. Normal transesophageal echocardiogram. Subsequently, COXCIIP drug-induced hepatitis with suspension, and an alternative tuberculostatic RIPE regimen was replaced, being reintroduced drug by drug each month, for 12 months. The patient presented motor improvement (MRC-4) after 6 months. Maintain outpatient follow-up, with motor rehabilitation. **Discussion:** Central Nervous System Tuberculosis is 1% of all tuberculosis cases and half of these involve the spine. Intramedullary involvement is rare, usually expressed in radiculomyelitis, transverse myelitis, intraspinal granulomas, or anterior spinal artery thrombosis. PCR test for M. Tuberculosis has sensitivity from 56% to 90% and specificity from 88% to 100%. **Final Comments:** As already documented and supported by case series, it is essential, especially in endemic areas, to consider causes of infectious mycobacterial etiology, particularly tuberculosis, in the differential diagnosis of radiculomyelitis, even if the etiological evidence is not confirmed.

Neuroinfecção

TL 1106239

LONGITUDINAL EXTENSIVE TRANSVERSE MYELITIS CAUSED BY SCHISTOSOMIASIS: A CASE PRESENTATION

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Case Presentation: A previously healthy 47-year-old man presented with a history of diarrhea associated with low back pain since January 2022. He evolved in one month with bilateral leg paresthesia and later spastic paraparesis. He also presented urinary urgency and incontinence, intestinal constipation and erectile dysfunction. In the neurological examination there were hyperreflexia in the upper and lower limbs (more overresponsive in the inferior limbs), bilaterally positive Hoffman and Babinski responses, bilateral spinal automatism, and bilateral parietic gait. In addition, there was decreased superficial sensation up to the T7 level, but no alterations in deep sensation. The MRI of the spine revealed an extensive lesion, more evident in the mid-segment of T1-T7, and affecting mainly the central spinal cord denoting probable myelitis. Active infection by *Schistosoma mansoni* was evidenced in a kato-katz stool parasitological examination. The CSF analysis showed increased cellularity including eosinophils and protein level with positive anti-Schistosoma serology. ANA, HIV/HTLV serology, VDRL, rheumatoid factor, anti RO, anti LA, and vitamin B12 level were unremarkable, and anti-aquaporin-4 was negative. He received intravenous therapy with 1g methylprednisolone for 5 days and oral praziquantel. He progressively improved and regained gait after treatment. **Discussion:** We present a case of neuroschistosomiasis as the cause of spastic paraparesis with superficial sensory loss up to thoracic level, and sphincter dysautonomic syndrome. The spinal cord lesion was not only high, but also longitudinally extensive, which is uncommon. Myeloradiculitis is the most common extraintestinal manifestation of schistosomiasis. It is caused by the presence of the parasite egg or the adult worms that migrate through the venous plexus of Batson to the spinal cord and induce a focal granulomatous reaction. Due to its anatomy, the most frequent locations are the lower thoracic and lumbar regions of the spinal cord, unlike the presented case. **Final Comments:** In the context of thoracic, longitudinal extensive spinal cord lesions, it is important that schistosomal myeloradiculitis should be included in the differential diagnosis, especially in endemic areas.

Neuroinfecção

TL 1106264

NEUROCRYPTOCOCCOSIS IN A YOUNG IMMUNOCOMPETENT PATIENT: A CASE-BASED UPDATE

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Case Presentation: A 19-years-old female presented 3 months ago an episode of an intense occipital pulsatile headache with irradiation to the frontal region, associated with vomiting, photophobia, phonophobia, right arm paresthesia and dysarthria. After 20 days, she evolved with a return of this condition, with additional lower limb paraparesis, fever and low back pain, which demanded Hospitalization for 4 days. After partial improvement of the condition, she was discharged, still unable to walk. At her home, she evolved with worsening headache and low back pain, also appearing diplopia and left convergent strabismus. Admitted to the emergency, a skull Computed Tomography and Magnetic Resonance Imaging were performed, both without alterations. The cerebrospinal fluid (CSF) analysis showed hypoglycorrhachia and hyperproteinorrachia, then antibiotic therapy was performed for 12 days, without improvement. A new lumbar puncture was performed, which showed cells suggestive of cryptococcosis, in the strong suspicion of this fungus, multiplex Polymerase Chain Reaction (PCR) was requested, which confirmed the infection by *Cryptococcus Neoformans/Gattii*. **Discussion:** Neurocryptococcosis is a fungal infection caused by two species of *Cryptococcus*, *C. neoformans* and *C. gattii*. It represents an important opportunistic infection related to AIDS, being uncommon in immunocompetent patients. The clinical presentation includes signs and symptoms such as headache, fever, nausea/vomiting, cranial neuropathies, mental disorder, lethargy, diplopia, intracranial hypertension and signs of meningeal irritation. The CSF analysis can include elevated white cell count, with lymphocyte predominance, elevated CSF protein, and low CSF glucose. The definitive diagnosis is made by isolation of *Cryptococcus* or direct detection of the fungus by means of India ink staining of the CSF. Other forms of diagnosis include histopathology, serological and molecular methods. Amphotericin B deoxycholate and flucytosine is the main treatment for severe cryptococcal infection, including meningoencephalitis. Liposomal amphotericin B is a second option with less nephrotoxicity, and other antifungals or combinations can be used in particular cases. **Final Comments:** Early diagnosis of neurocryptococcosis in immunocompetent patients can be challenging. Being aware of this infection in this group of patients is necessary for rapid and appropriate management.

Neuroinfecção

TL 1106311

ONE MORE CASE PRESENTATION OF A PATIENT WITH COVID-19 AND ENCEPHALOPATHY ASSOCIATED WITH POSSIBLE LANCE-ADAMS SYNDROME WITHOUT CARDIAC ARREST

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Case Presentation: A diabetic, hypertensive, with heart and chronic renal disease and ex-smoker 78-year-old man was admitted to the emergency department with acute dyspnea, sweating and fever. A chest CT scan revealed hypo-expanded lungs, atelectasis signs in the bilateral lower lung lobes, right pleural effusion and cardiomegaly. A new chest CT showed diffuse and bilateral ground-glass opacities and a nasopharyngeal swab RT-PCR for SARS-CoV-2 was positive. The patient evolved with lowering of consciousness level associated with left upper limb rapid and repetitive movements described as myoclonus. The electroencephalogram (EEG) showed a diffuse slowing, in the theta/delta range, and periods of generalized attenuation confirming an encephalopathy pattern. There is no previous cardiac arrest. The patient died a few weeks later, after clinical worsening and complications of his renal condition. **Discussion:** We report a rare case of COVID-19 infected patient with severe hypoxia evolving with decrease of consciousness level and focal myoclonus. There are several studies in the literature showing the occurrence of de novo epileptic seizures in SARS-CoV-2 infection, which was not confirmed in this case (DELORME et al., 2020; DIXON et al., 2020; SOMANI et al., 2020). On the other hand, Lance-Adams syndrome is associated with severe hypoxemic states of different etiologies and seems to be reported in very few COVID-19 patients (MUDDASSIR et al., 2021). **Final Comments:** The present case is representative about the severe outcome associated to nervous system involvement and includes the COVID-19 hypoxemic state among the differential diagnosis of Lance-Adams syndrome without previous cardiac arrest. Biological mechanism associated with the severe outcome is still a challenge to be understood and this report may contribute adding up to a few cases describing so far.

Neuroinfecção

TL 1106312

BRAIN FUNCTIONAL CONNECTIVITY IS SEVERELY DISRUPTED IN THE PRESENCE OF ANXIETY AND DEPRESSION SYMPTOMS AFTER MILD COVID-19 INFECTION

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Background: Although most individuals with post-COVID syndrome present symptoms of anxiety and depression, little is known about the associated brain dysfunction. So far, fewer studies have analyzed brain connectivity abnormalities in post-covid syndrome. **Objective:** To investigate the impact of symptoms of anxiety and depression on functional brain connectivity (FC) after mild COVID-19 infection, analyzing 12 large-scale brain networks (NW) with resting-state fMRI (rs-fMRI). **Methods:** We analyzed 154 post-covid subjects who had a mild infection and confirmed RT-PCR test and 90 healthy controls. All individuals were evaluated with BDI (Beck Depression Inventory) and BAI (Beck anxiety inventory) for quantification of symptoms. Patients were divided into asymptomatic-group (ASYMP-Group, no symptoms of depression or anxiety) (84 individuals, 54 women, median age of 36 years), and DOUBLE-group (combined symptoms of anxiety and depression) (70 subjects, 55 women, median age of 41 years). All individuals underwent rs-fMRI scan (along with 3D T1-weighted) on a 3T scanner. Each group was compared to a specific control group paired for age and sex. Images were analyzed with UF2C/SPM12 (<http://www.lni.hc.unicamp.br/aoo/uf2c/>) running on MATLAB2019b. The protocol includes steps of pre-processing (normalization and smoothing) and extraction of time series from 70 Regions of Interest (ROIs) (from 12 resting-state networks: Anterior and Posterior Salience, Basal Ganglia, Dorsal and Ventral Default Mode, Left and Right Executive Control, Auditory, Visual, Language, Sensorimotor and Visuospatial/Dorsal Attention) used to construct Pearson's Cross-Correlation Matrix. Results were considered significant after correction for multiple comparisons at $p < 0.05$. **Results:** While the DOUBLE-group exhibited a widespread, bilateral pattern of hyperconnectivity involving the 12 networks and most of 70 ROIs, the ASYMP-group showed fewer alterations, with hyperconnectivity involving only five networks (posterior salience, ventral and dorsal DMN, and right and left executive control) and nine ROIs. ($p < 0.05$). **Final Comments:** Our results suggest a severe pattern of abnormal cerebral functional connectivity, mainly in individuals with persisting symptoms of anxiety and depression. These abnormalities may be associated with cognitive dysfunction in post-covid syndrome. Symptomatic treatment should be available for these individuals to improve their quality of life.

Neuroinfecção

TL 1106337

NEUROCYSTICERCOSIS WITH EXTENSIVE CERVICAL MYELITIS: CASE PRESENTATION

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Case Presentation: A 59-year-old woman was diagnosed with epilepsy secondary to neurocysticercosis (NCC) at seven years old. Despite the use of albendazole, she developed cysticercal meningitis that was responsive to dexamethasone, but recurred upon withdrawal. The chronic corticotherapy resulted in aseptic necrosis of knees. Then, obstructive hydrocephalus occurred due to a suprachiasmatic cyst, requiring ventriculoperitoneal shunt and cyst removal. One year later, she had an ischemic stroke due to cysticercotic vasculitis, confirmed by arteriography. After 16 years, she presented with asymmetrical tetraparesis, fecal and urinary incontinence and neuropathic pain. Neuroimaging was suggestive of cervicomedullary transition cysts and active diffuse myelitis. Quarterly corticosteroid pulse therapy was started, reducing pain and stabilizing the motor symptoms. However, the condition worsened three years later, not responding to corticotherapy, when treatment was changed to oral methotrexate (MTX) 10mg per week, and then modified to the subcutaneous presentation to minimize gastric intolerance. Six years ago, BIPAP use was started due to severe sleep apnea and restrictive lung disease secondary to myelitis. Currently, epilepsy, neuropathic pain and motor symptoms are stable, but the patient is partially dependent on a wheelchair and needs 10mg of MTX every 21 days, besides carbamazepine, gabapentin and venlafaxine for pain control. **Discussion:** NCC is a disease that presents variable symptoms, being epilepsy the main one, followed by focal neurological deficit, intracranial hypertension and cognitive deficit. Only 2.7% of cases have spinal cord involvement and most of these cases present as radicular pain, sensory and motor deficits. Severe parenchymal or subarachnoid NCC may require long-term immunosuppression, with a steroid-sparing drug, such as methotrexate and etanercept, as prolonged use of corticosteroids can have serious side effects. **Final Comments:** Serious manifestations may result from NCC and myelitis is a rare associated condition. Immunosuppression can stabilize the course of this condition.

Neuroinfecção

TL 1106365

RECURRENT NEUROTUBERCULOSIS WITH RIFAMPICIN RESISTANCE IN IMMUNOCOMPETENT PATIENT: A CASE PRESENTATION

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Case Presentation: Male, 38 years old, previously healthy, in 2018, presented progressive headache, vomiting and fever. After 4 months, diplopia and syncope. On exam, generalized lymphadenomegaly and vertical conjugate gaze limited. Viral hepatitis, syphilis and HIV serologic tests were negative. Cerebrospinal fluid (CSF) showed inflammatory pattern with increased opening pressure, and immunological tests to syphilis, cytomegalovirus, Epstein-Barr, cultures for fungi and mycobacteria were negative. Brain MRI showed solid hypointense nodular images, enhancement in the frontal and bilateral temporal regions. Lung and brain biopsies were inconclusive. Empirical treatment for tuberculous, fungal meningoencephalitis, nocardiosis, and pyogenic brain abscess was started. Antituberculosis drugs were used for 12 months and maintenance doses of fluconazole and prednisone, with improvement of diplopia and headache. After 2-years, he presented again with pain and right axillary lymph node enlargement, anatomopathology realized, suggested ganglionic tuberculosis, and no other changes in new radiological exams. The rapid molecular test for tuberculosis (TRM-TB) was positive with no resistance to Rifampicin and antituberculosis drugs and corticotherapy was reinitiated. Primary immunodeficiencies were searched, with no abnormalities. At the end of treatment, corticotherapy was discontinued; however, diplopia, visual blurriness, dizziness, and headache returned. New brain MRI showed active mesencephalic lesions and MRI-BT of a CSF sample positive for rifampicin resistance. Then, alternative regimen for rifampicin-resistant neurotuberculosis with RIPE regimen, Levofloxacin and Clarithromycin, and adjustment of prednisone were chosen, with improvement of symptoms and outpatient follow-up. **Discussion:** Central nervous system tuberculosis is an infrequent extrapulmonary manifestation. Prior to antituberculous therapy, this condition was uniformly fatal and still results in high morbidity and mortality today. The main risk factors are associated with compromised immune system; therefore, its occurrence in immunocompetent patients is rare. High clinical suspicion and early treatment are associated with better outcomes. **Final Comments:** Neurotuberculosis remains a challenge diagnostic, especially, in immunocompetent patients, the high sensitivity and specificity of diagnostic tests are important tools, however, in some cases, it remains a diagnostic and therapeutic challenge.

Neuroinfecção

TL 1106389

CNS (CENTRAL NERVOUS SYSTEM) PARACOCIDIOMYCOSIS AS A DIFFERENTIAL DIAGNOSIS OF IGG4 – DISEASE RELATED : CASE PRESENTATION

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Case Presentation: Male, 38 years old, healthy, motor, from Belo Horizonte, MG. In Mid 2020, he presented headache, xerophthalmia and perceived two small bilateral cervical nodules in addition to intermittent paresthesias in fingers, hands and face and seizures. He also had type B symptoms (weight loss, frequent sweating and fever spikes). He looked for a neurologist who requested an MRI, EEG and referred him to Hospital. He received pulse therapy with 1g methylprednisolone for five days, with partial improvement in his neurological condition. A single dose of IV cyclophosphamide (1g) was prescribed. The laboratory showed an increase in IgG4 (above 8000), with a brain MRI report suggesting IgG4-related disease. The Rheumatology evaluation ruled out other rheumatologic diseases. The biopsy of cervical lymph node revealed paracoccidioidomycosis (PCM), and treatment with amphotericin B deoxylate was initiated. After treatment, there was significant clinical and radiological improvement, with a drop in serum IgG4 levels. **Discussion:** The IgG4-related disease has various clinical manifestations, with possible involvement of the pancreatobiliary tract, retroperitoneum, head and neck organs and glands. It could mimic neoplastic, inflammatory and infectious conditions. Histopathology remains the key to diagnosis because reliable biomarkers are lacking. IgG4-related disease is treated with glucocorticoids but can even lead to death if not recognized. However, to get accurate diagnosis of IgG4-related disease, it is mandatory to exclude infectious, neoplastic and rheumatologic diseases. On the other side, PCM, a systemic infection caused by the fungus *Paracoccidioides brasiliensis*, affects individuals in all age groups. The lung is the most affected organ, and it may present on the CNS. Santos et al, 2012, evaluated the levels of IgG, IgG 1, IgG 2, IgG 3 and IgG4 in 54 patients with PCM during and after treatment. Only the IgG4 subtype showed a negative correlation between its levels and clinical improvement during treatment. Overall, 65% of patients showed reactivity against IgG 4 when Mexo antigen was used and this reactivity decreased over the course of treatment. The antigen can be used in the measurement of total IgG levels and its subclasses to monitor patients during treatment. **Final Comments:** it's imperative to identify and exclude differential diagnosis on suspecting IgG4-related disease, due its lack of other biomarkers and the clinical and laboratory characteristics may be mimicked by other pathologies.

Neuroinfecção

TL 1106450

NEUROLOGICAL MELIOIDOSIS: A CASE PRESENTATION

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Case Presentation: Male, 54 years old, with SAH and DM 2, on 05/10/2021 she started inappetence, nausea and muscle fatigue. She evolved 7 days later with unmeasured fever, drowsiness and the appearance of a painful nodular lesion on the scalp; after three days she had temporo-spatial disorientation and generalized tonic-clonic seizures. Attended at the HGF emergency room, brain MRI showed changes in tissues from the face to the galea, intradiploal and subdural collections, irregular pachymeningeal enhancement, T2/FLAIR hypersignal and diffusion restriction in the left parietal lobe suggestive of cerebritis, MR angiography with thrombosis sigmoid sinus and CSF study 1182 red blood cells, 9 leukocytes (neutrophils 77%, lymphocytes 22%), proteins 49mg/dl, glucose 55mg/dl, LDH 41, ADA 0.1 and oncotic cells, fungi, BAAR, VDRL and negative cultures. She worsened clinically, requiring mechanical ventilation in the ICU. The pathogen *Burkholderia pseudomallei* was isolated from three blood culture samples and treated with meropenem for 8 weeks. In the eradication treatment, he developed pancytopenia secondary to the use of sulfamethoxazole-trimethoprim. He was discharged with negative blood cultures for outpatient treatment with amoxicillin-clavulanate. Melioidosis is caused by the gram-negative bacillus *Burkholderia pseudomallei*, responsible for 89,000 annual deaths worldwide. In Ceará, the first case was described in 2003. In 2005 it became a notifiable disease. The infection occurs by inhalation/inoculation of the pathogen; is confirmed by culture or PCR. In the neurological form, extracranial lesions occur, may involve contiguous sites, sub or extradural collections, brain micro or macroabscesses, cranial/spinal osteomyelitis, leptomeningitis, meningoencephalitis, brainstem encephalitis with cranial nerve palsy, myelitis and dural venous sinus thrombosis. Risk factors such as DM2, alcohol, COPD, chronic kidney disease can lead to a worse outcome. Intensive treatment varies from 14 days to 8 weeks according to severity, with carbapenems or ceftazidime. Eradication treatment lasts up to 12 weeks, using trimethoprim-sulfamethoxazole to prevent recrudescence. Doxycycline and amoxicillin-clavulanate are options. In endemic areas, it is crucial to maintain high clinical suspicion in cases in which the image suggests the possibility of a diagnosis of the neurological form, helping the rapid institution of therapy due to the risk of severe and fatal evolution.

Neuroinfecção

TL 1106466

A INVASIVE INFECTION BY STREPTOCOCCUS AGALACTIAE CAUSING MENINGITIS OF ATYPICAL PRESENTATION

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Case Presentation: A 33-Year-Old Man, Previously Healthy, Evolved With Severe, Progressive, Persistent Headache Associated With Vomiting, Photo And Phonophobia And 03 Episodes Of Fever (38-39°C). Five Days After The Onset Of The Condition, He Developed Binocular Diplopia When Looking To The Right Side And Worsening Of The Headache Intensity. An Skull Angio-Ct Realized In Another Service That Showed Indirect Signs Of Intracranial Hypertension And Then He Sought Medical Care In Our Service After 15 Days Of Onset Of Symptoms. A Lumbar Puncture Was Performed, Opening Pressure Of 37 CmH₂O, 1.649 Cells (59% Neutrophils), Protein Of 135 Mg/Dl And Glucose Of 39 Mg/Dl. The Sample Was Analyzed By Molecular Test Which Detected Streptococcus Agalactiae. Mri Of Her Brain Demonstrated Various Focus Of Restriction To Diffusion In High Frontoparietal Convexity. The Patient Was Treated With Ceftriaxone 4g/Day For 14 Days And Evolved With Total Improvement Of Symptoms. Repeated Lumbar Puncture After 10 Days Of Treatment: Opening Pressure Of 24 CmH₂O, 160 Cells (78% Lymphocytes And 9% Monocytes), With 46mg/Dl Protein And 52 Mg/Dl Glycorrhachia. **Discussion:** This Clinical Case Demonstrates An Adult Patient With Atypical Presentation Of Bacterial Meningitis With An Unusual Etiologic Agent For The Age Group, Streptococcus Agalactiae (Group B Streptococcus, Gbs). Gbs Is A Normal Floral Bacterium That Colonizes The Gastrointestinal And Genitourinary Tracts, It Is The Main Cause Of Puerperal And Neonatal Infections, Including Meningitis. In Adults, Gbs Meningitis Is Relatively Rare, Accounting For 0.3 To 4.3% Of All Cases Of Bacterial Meningitis. Gbs Meningitis In Adults Usually Occurs In Patients With Underlying Conditions Or In Pregnancy/Puerperium, Diabetes Mellitus Is The Most Associated Comorbidity, During Hospitalization The Patient Was Found To Be Pre-Diabetic. Domingos Et Al Revealed That 86% Of The Patients Had Comorbidities And 50% Had A Distant Focus Of Infection. The Clinical Picture Of Meningitis By Gbs And The Csf Findings Do Not Differ From Other Bacterial Meningitis, However Our Case Revealed Itself With 15 Days Of Evolution And Signs Of Intracranial Hypertension. Although Gbs Are 4 To 10 Times Less Susceptible To Penicillin, Penicillin Is Still The First Choice, As Is Ceftriaxone. **Final Comments:** With The Rate Of Invasive Disease Per Gbs In Adults Increasing, The Physician Must Be Prepared To Recognize And Initiate Effective Treatment To Avoid Complications Of The Disease.

Neuroinfecção

TL 1106478

REPORT OF THREE CASES OF PATIENTS WITH PROBABLE CREUTZFELDT-JAKOB DISEASE IN A PERIOD OF SIX MONTHS IN THE SAME REGION OF BRAZIL

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Case Presentation: A 54-year-old woman presented with psychiatric disorder with persecutory delusions, evolving with focal epileptic seizures for 20 days. On admission, myoclonus was evident in the upper and lower limbs and spasticity in the lower limbs. LCR cultures were negative, VDRL unreactive, oligoclonal bands absent and 14-3-3 protein search present. EEG unchanged. MRI of the skull, signs of restriction and sparse cortical diffusion in the frontal and parietal lobes, and in the striatum on the right. A 57-year-old man presented with progressive cognitive impairment associated with depression, two months ago. On admission, hypovigil, temporo-spatial disorientation, psychomotor retardation, impaired short-term memory, myoclonus in upper and lower limbs and face, spasticity of the limbs. CSF negative for encephalitis. EEG with diffuse slowing of background activity. Cranial MRI with bilateral striated cortex and bilateral lentiform nucleus. A 68-year-old man presented with episodes of spatial disorientation for a month and a half, evolving with mental confusion associated with visual hallucinations, being admitted to psychiatry with a hypothesis of delirium. Assessed by Neurology due to spontaneous myoclonus in the face and upper limbs. The EEG marked disorganized brain electrical activity for age with generalized periodic discharges. CSF with 14-3-3 protein search present. MRI of the skull signs of diffusion restriction involving the caudate and putamen bilaterally and symmetrically. The three cases evolved in less than 30 days of Hospitalization with akinetic mutism, and death from bronchoaspiration pneumonia. **Discussion:** Creutzfeldt-Jakob disease (CJD) is a rare, rapidly progressive neurodegenerative disease, whose diagnosis is based on clinical, radiological, electroencephalographic or cerebrospinal fluid findings, which makes its diagnosis a challenge due to the availability of exams and the low rate of clinical suspicion. **Final Comments:** Despite the rarity, the three cases were identified with a hypothesis of probable CJD, in a period of only six months in the same region of Brazil, with no relationship of kinship and no report of possible acquisition of an iatrogenic form.

Neuroinfecção

TL 1106493

LIMBIC ENCEPHALITIS POS COVID: A CASE PRESENTATION

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Case Presentation: A 57-years-old woman, hypertensive, was admitted to the neurologic emergency with a 10-day history of flu-like symptoms. After 24 hours at admission showed lowering the level of consciousness. Upon neurological exam: spatial disorientation, memory loss, irrelevant speech, Babinski sign bilateral and Brudzinski sign. SWAB COVID-19: positive. Cerebrospinal Fluid (CSF) Analysis: pleocytosis: 400 (63% lymphocytes), glucose 86mg/dL, protein: 36mg/dL, ADA 0,1, LDH 49, PCR multiplex PCR: negative. Therefore, treatment with Acyclovir and Ampicillin, but without improvement after 14 days of therapy. Furthermore, electroencephalogram (EEG) revealed non-convulsive status epilepticus despite phenytoin. Thus, new CSF analysis were performed with panel collection for limbic encephalitis research negative. Also performed magnetic resonance (MRI) revealed limbic encephalitis pattern. Due to clinical and radiological worsening, considered post covid encephalitis. Hence, we started treatment with pulse therapy with methylprednisolone 1g for 5 days and de immunoglobulin for 5 days. She was discharged from Hospital with partial improvement in behavior and interaction with the environment. **Discussion:** The Limbic Encephalitis is a severe inflammatory disease that affects the limbic system. It may have infectious, inflammatory or autoimmune etiology. The diagnosis is realized with clinical, EEG, CSF analysis, Serum antibody dosage and neuroimaging. The clinical is varied, including involvement of memory, cognition, behavioral change, seizures, hallucinations, confusion and agitation. The neurological manifestations of COVID-19 including acute encephalopathies, stroke. Therefore, Immune Mediated Encephalitis post COVID 19 is the possible cause of encephalitis in this case. **Final Comments:** COVID 19 and its complications that need to be studied. The limbic encephalitis as a neurological complication of COVID 19 should be recognized and studied as the new etiology.

Neuroinfecção

TL 1106547

CRYPTOCOCCOSIS AS A CAUSE OF PARAINFECTIOUS CEREBRAL VASCULITIS

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Case Presentation: Female, 56 years old, with sudden onset of difficulty opening the left eye associated with diplopia. She reports a history of severe holocranial headache that started 5 days ago. Known comorbidities: Systemic lupus erythematosus and antiphospholipid antibody syndrome, using Marevan 5mg/day and prednisone 10mg/day. Physical examination: paresis of left eye adduction, left eyelid ptosis, nuchal rigidity. Computed tomography of the brain at admission without alterations. Cranial MRI showed an acute ischemic vascular insult in the left midbrain. CSF study revealed: 240 cells (predominance of mononuclear cells); glucose: 13; proteins: 192; Positive China ink; positive latex test for c. neoformans. In view of the diagnosis of neurocryptococcosis, cerebral infarction secondary to parainfectious cryptococcal vasculitis was suspected and therapy was initiated. **Discussion:** Immunosuppressive conditions increase the risk of CNS infections. CNS vasculitis associated with cryptococcal infection is rare, with clinical manifestations of cerebral infarctions in the internal capsule, thalamus and basal ganglia. The clinical course has a subacute or chronic onset, which can usually lead to delays in diagnosis and adequate treatment. Radiological examinations such as CT and magnetic resonance imaging (MRI) contribute to the visualization of infarcted areas, emphasizing the value of angiography with evidence of vasculitis. The diagnosis is confirmed through confirmation in the cerebrospinal fluid with nanquin ink and culture. Treatment should be instituted as soon as possible, aiming to reduce morbidity and mortality from the disease. **Final Comments:** Although rare, it is important to know this presentation in neurocryptococcosis, since early diagnosis and management provide a reduction in patient morbidity and mortality.

Neuroinfecção

TL 1106563

SUCCESSFUL APPROACH: A CASE PRESENTATION OF A PERIAMYGDALENTAL ABSCESS WITH SPINAL CORD COMPRESSION

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Case Presentation: L.R.N.O., a 60-year-old male, hypertensive and decompensated diabetic, was admitted in July 2021 to the emergency department with ataxia, erythematous patches in upper and lower limbs, edema in large joints associated with odynophagia, and fever. The patient was referred to the Hospital unit for cardiology and rheumatology support, and purulent collections were identified on the topography of the right shoulder and left elbow. Cervical tomography showed retropharyngeal collection extending posteriorly to the left paravertebral space. An etiologic investigation and antibiotic therapy were initiated. In September, the patient developed acute tetraparesis and intense cervicalgia and was referred to the neurosurgery department, where neck MRI showed maintenance of the images, with invasion into the vertebral canal and compression of the anterior aspect of the spinal cord from C1 to C3, causing signs of myelopathy. He underwent laminectomy of C1 and drainage of epidural empyema in that service. After changing antibiotic treatment in which he was admitted to Vancomycin and Cefepime he progressed to the improvement of strength in all 4 limbs, cervicalgia, and headache. Despite cultures collected, the infectious agent was not identified. Deep neck space infections are suppurative infectious processes that can cause abscesses and affect deep visceral spaces of the neck such as the parapharyngeal space and the danger space. A common etiology is the spread of oropharyngeal infections, such as odontogenic infections and tonsillitis, through fascial planes. Deep cervical infections can be fatal due to jugular vein thrombosis, airway obstruction, mediastinitis, and sepsis. This paper aims to report a successful case of a periamygdala abscess with deep cervical dissemination, resulting in tetraparesis and a dismal prognosis. The evolution of antibiotic therapy was responsible for a decrease in the incidence of deep cervical space abscesses. However, in the last decade, the number of this type of abscesses has increased. The patient follows this trend and reveals systemic comorbidities of diabetes mellitus without disease control. Therefore, although tonsillitis is a common infection, it can generate important and fatal complications. In this paper we present a case presentation with significant neurological involvement, such as epidural empyema, with severe headache, tetraparesis, and cervicalgia, giving a dismal prognosis. However, the patient had a positive prognosis.

Neuroinfecção

TL 1106603

FUNGAL BRAIN ABSCESS AS A DIFFERENTIAL DIAGNOSIS OF NEUROINFECTION IN IMMUNOCOMPETENT PATIENTS

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Case Presentation: Patient in 56 years old, hypertensive, presented changes cognitive and behavioral, followed by progressive impairment of the level of consciousness with a gradual evolution in 3 weeks, associated with fever, deficits neurological focal and march slowed down. At admission, appeared lethargic, ECG 10, no abnormalities in the examination of the cranial nerves and with motor deficit on the left and without stiffness in back of neck. The Cranial nuclear magnetic resonance (MRI) showed multiple lesions dispersed in the brain parenchyma, more evident in the nucleocapsular, with annular enhancement after intravenous infusion of the contrast, associate The edema vasogenic of parenchyma brain adjacent, and restriction of peripheral diffusion the lesions, presenting a discrete amount of exudative material in the occipital horns of the lateral ventricles, suggestive of ventriculitis. Treatment was started for bacterial coverage initially with ceftriaxone and Oxacillin, then in Meropenem and vancomycin, for the suspected brain abscess. The liquor collection not performed due to injuries with edema cerebral and Detour in line average. THE investigation for HIV and immunosuppression were negative. Despite antibacterial therapy, patient remained with clinical and laboratory tests worsening, and it was decided to start in therapy with amphotericin B thinking in abscess fungal, although gives renal dysfunction presented by amphotericin B, patient evolved with improvement of the clinic status, being discharged from the intensive care unit. At Hospital discharge, the patient is alert, no language deficit, no motor deficit, mild gait slowing, without stiffness. Control cranial MRI examination after Hospital discharge evidence decrease of edema and of injuries presented. **Discussion:** Fungal brain abscess is a life-threatening condition, more common in immunocompromised individuals, but with increasing reports in the literature in immunocompetent patients. This case highlights the importance and need for a higher level of suspicion and sensitive diagnostic methods to optimize therapy and improve case outcomes. **Final Comments:** Despite of limitations of service for realization of liquor analysis, the importance of evaluating images that suggest fungal abscess were important for outcome positive of case.

Neuroinfecção

TL 1106632

THE REFRACTORY TREATMENT IN A PATIENT WITH SYPHILITIC UVEITIS: A CASE PRESENTATION

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Case Presentation: A 23 year old man was admitted in emergency with progressive bilateral vision impairment, which started one month ago with ocular hyperemia and dark spots. Also reported history of adynamia, hyporexia and fever. Upon neurological exam: pupils with light-near dissociation, fundoscopy: bilateral posterior synechiae and right eye with optic nerve with infiltrative-appearing papilledema. The laboratory tests revealed a venereal disease research laboratory test (VDRL) 1: 1024 and HIV reagent, confirmed by two positive samples. Cerebrospinal fluid (CSF) analysis showed non-reactive VDRL and increased protein. A diagnosis of neurosyphilis was made. Treatment with crystalline penicillin and eye drops (corticoid and tropicamide) was instituted. After 21 days of crystalline penicillin with persistent lesions in the eye, and evidence of capture contrast in basal ganglia in cranial magnetic resonance (MRI), empirical treatment for toxoplasmosis neuroretinitis was discussed and sulfadiazine was prescribed to treat neurotoxoplasmosis. The patient was later reassessed by the ophthalmologist, showing improvement in the retinal edema. **Discussion:** Syphilitic uveitis comprises less than 1-2% of all uveitis cases, however an increase in the number of cases over the past decade was observed. The manifestations are diminished visual acuity, blindness, optic neuropathy, interstitial keratitis and it can occur at any state of natural history of untreated syphilis. In the present case, even with CSF non-reactive, the signs of active uveitis and the VDRL titulation suggested neurosyphilis. Neuroretinitis is a phenotype of syphilis uveitis and is described as a clinical triad of unilateral vision loss accompanied by optic disc edema and the formation of a characteristic macular star. Patients should test for HIV infection either, because the co-infection are frequent. A few cases of neuroretinitis by *Toxoplasma gondii* have been reported, however the presence of hiperdense lesions in basal ganglia in MRI associated with lack of improvement of ocular lesions support the treatment choice for neurotoxoplasmosis. **Final Comments:** Immunosuppression by HIV may contribute to the appearance of opportunistic infections with atypical involvement, such as neuroretinitis in toxoplasmosis and syphilitic uveitis. For the adequate management of these patients a thorough etiological investigation should be performed, as well as the observation of improvement after treatment.

Neuroinfecção

TL 1106635

LEGIONNAIRE'S DISEASE ASSOCIATED – SEVERE ENCEPHALOPATHY

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Case Presentation: A 57-year-old Brazilian man presented to the emergency department with a 3-day history of diarrhea and hiccups. On the next day, he developed incoordination followed by change in speech and vomiting episodes. Symptoms worsened, until he was unable to stand. While waiting for medical attention, he became confused, reporting delusions and hallucinations. He had a past history of smoking, cocaine use and moderate alcohol consumption. On examination, he was drowsy, disoriented in space and time, dysarthric, apparent restriction of vertical upward gaze bilaterally. He had astasia and abasia. In the emergency room, he became agitated and developed tachycardia, tachypnea, fever (40 C) and was intubated and sent to ICU. Blood screening revealed elevated CK of 18.306 (normal: 39-308), thrombocytopenia high creatinine levels of 1,55, hyponatremia of 124, hypokalemia of 3,3, low ionized calcium of 1,08, mild elevated ALT/AST, elevated troponin of 50 and elevated CRP of 244. Chest TC showed an extensive consolidation affecting almost the entire right lower lobe and minimal bilateral pleural effusion. CSF studies revealed 9 leukocytes (88% lymphocytes, 12% monocytes), 5 erythrocytes, glucose of 88 mg/dL, lactate of 27,5. Culture and viral panel were negative. Brain MRI was normal. Hydration for rhabdomyolysis, Ceftriaxone and metronidazole were the initial treatment. Because of lack of response Levofloxacin was started with positive response. Positive Legionella Urine confirmed Legionnaire's disease. After recovering, he was discharged home but with residual ataxia and dysarthria in improvement. Legionnaire's disease is an infection caused by Legionella pneumonia that usually manifests with pneumonia. Extrapulmonary manifestations are rare, but meningitis, encephalopathy, which can be from mild to coma, cerebellar dysfunction and focal deficits. Brain MRI can present with hyperintensities at multiple localizations CSF findings are normal in the vast majority of patients, raising the possibility that Legionella does not directly affect the central nervous system. Therefore, pathophysiology is uncertain and it is unknown whether the mechanism is an immune, toxin-mediated or a direct damage by the bacteria. The patient was followed and persisted with ataxia and dysarthria, in slow recovery three months after discharge.

Neuroinfecção

TL 1105522

POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME AFTER CHRONIC OBSTRUCTIVE PULMONARY DISEASE : A CASE PRESENTATION

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Case Presentation: ERS, male, 61 years old, born in Itacoatiara, Amazonas. He entered the emergency department on January 20, 2022 being immediately intubated and sedated. His family tells a history of chronic obstructive pulmonary disease (COPD) with exacerbation in the last 2 weeks causing respiratory failure due to community-acquired pneumonia. He had systemic arterial hypertension and heart failure. Somatoscopy showed normal color, hydrated, anicteric, acyanotic, and afebrile. Physical examination revealed edema of the lower limbs and the respiratory system showed diffusely abolished breath sounds, with no other complaints. On January 27, the patient had a seizure being medicated with phenytoin and, later, escalated with an association of valproate, phenobarbital and clonazepam. On this same day, a cranial tomography (CT) was performed, which showed hypodense lesions in the posterior fossa, in addition to partial emptying of the sella turcica, which correlated with intracranial hypertension. In view of the clinical findings and the imaging exam, posterior reversible encephalopathy syndrome (PRES) due to COPD was observed and in the same period the patient complicated with a subarachnoid hemorrhage which was reabsorbed later. Lastly Patient evolved to death due to cardiac arrest due to the lack of possibility of long-term treatment. **Discussion:** PRES is a neurotoxic event which results a fluid extravasation marked often by the presence of vasogenic edema in the parieto-occipital region in CT or magnetic resonance imaging (RMI) with clinical features like altered mental status, headache, seizures and vision loss. This syndrome is due mainly from secondary chronic diseases such as hypertension. In addition, it may be complicated by subarachnoid hemorrhage which, despite its not fully understood mechanism, progresses in most cases with bleeding reabsorption within a few days. Finally, due to the reversible behavior of PRES, improvement in the condition generally occurs after controlling the underlying disease. Comentários finais. In this case, we present a curious and rare manifestation of PRES correlated with COPD exacerbation. In patient care, all possible measures to be used in the service were adopted, however, some limitations can be mentioned, such as the lack of long-term therapeutic possibilities, which culminated in the patient's death. In this context, further discussions on future advances and research in the field are necessary.

Neurointensivismo

TL 1105576

CASE PRESENTATION ACUTE CEREBROVASCULAR EVENT WITH COVID-19 INFECTION, A RARE AND SERIOUS COMPLICATION

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Case Presentation: A 60-year-old male patient, diabetic, hypertensive and previously vaccinated with one dose of COVID-19 vaccine (ChAdOx1), was isolated at home for COVID-19, with a positive antigen test and first symptoms 13 days before admission. He was admitted presenting aphasia and right hemiplegia, with no respiratory distress. His cranial computed tomography (CT) had an extensive commitment at the territory of irrigation of the middle cerebral artery (MCA). Another finding was internal carotid and MCA occlusion in CT angiography. Chest CT showed a 40% parenchymal commitment with ground-glass opacification, and bronchopneumonia. He was treated with antibiotics, corticotherapy. During Hospitalization he presented with delirium and acute respiratory distress, afterward, thromboembolism was diagnosed by CT angiography. Then, mechanical ventilation was needed for 6 days. The patient had a 2 months stay in Hospital, due to urinary tract infection, persistent fever and pseudomembranous colitis. Finally, he was discharged with no oxygen supplementation and was prescribed anticoagulation. Moreover, he maintained his motor impairment and improved his aphasia. **Discussion:** COVID-19 is known as a proinflammatory state associated with thrombophilia. There is an increased risk of thrombotic events, even though stroke is considered rare. The incidence of ischemic stroke associated with COVID-19 patients has ranged from 0.4 to 2.7 percent. A study showed that COVID-19 patients were more likely to have a cryptogenic stroke. Outcomes were worse, including longer length of stay, intensive care unit care (ICU) stay, greater rate of neurological worsening, and in-Hospital death. In comparison, our patient had a long length stay, with ICU care needed. Nevertheless, our patient had a atherothrombotic event and evolved with a neurological improvement. Similar to our case, another article demonstrated the association between ischemic stroke and pulmonary embolism. Besides, the median time between the onset of COVID-19 symptoms and diagnosis of stroke was 2 weeks. **Final Comments:** Stroke appears to be a rare but serious event associated with COVID-19 patients. Considering the poor outcome described in literature, our patients, despite all complications, had improvement of his conditions. However, the characteristics of COVID-19 patients with stroke vary in the current literature, therefore more description is needed.

Neurointensivismo

TL 1105854

CANDIDA ALBICANS AND STENOTROPHOMONAS MALTOPHILIA: ATYPICAL CASES OF MENINGITIS IN A NEUROINTENSIVE UNIT

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Case Presentation: 1. Female, 85 years old, previously hypertensive and diabetic, presented, on July 15, 2021, HSA-fisher 3, being admitted to the Neurointensive Unit. He evolved with hydrocephalus and intracranial hypertension. On July 16, an external ventricular shunt was placed with an intracranial pressure monitor. On the third post-operative day, he showed fever and worsening inflammatory patterns. In the CSF study, it had an opening pressure of 20cmH₂O, with 18773/mm³ cells (75% neutrophils), 181 mg/dL protein, 20 mg/dL glucose. CSF culture, with growth of Gram negative *Stenotrophomonas maltophilia* sensitive to levofloxacin and tigecycline. After one month of treatment, he died, despite the medication; 2. A 34-year-old male with no previous comorbidities is admitted to the Neurointensive Unit to investigate a seizure. The patient had no history of epilepsy. It was reported that, a week ago, he had started with a mild headache and tremors of the extremities, which was followed by a fever measured between 38-39°C and blurred vision. In emergence, kernig and brudzinski signs are present. Laboratory tests and magnetic resonance imaging of the skull were performed, without alterations. In neurointensive care, liquor was collected, which showed PCR detecting *Candida albicans*. He was then treated with liposomal amphotericin B and fluconazole. About two weeks later, the patient's symptoms improved significantly and he was discharged from the Hospital. **Discussion:** Meningitis caused by *S. maltophilia* is rare, with only 18 cases reported so far. Populations at risk for *S. maltophilia* meningitis are typically immunocompromised patients, those who have had neurosurgical procedures, premature infants, and patients requiring prolonged Hospitalization. Multi-agent resistance is a hallmark of *S. maltophilia*. Regarding the fungal meningitis described, its prevalence began to decrease in the last decade, while the frequency of detection of *C. albicans* species gradually increased. It presents manifestations similar to those of bacterial meningitis, making the initial use of antibacterials not uncommon. It has a good prognosis after the institution of the correct therapy. **Final Comments:** Although these atypical meningitis remain rare in the population, physicians should be aware of them as possible meningitis-causing organisms for better therapeutic follow-up, even in the absence of their known risk factors.

Neurointensivismo

TL 1105872

ADULT ONSET OPSOCLONUS-MYOCLONUS-ATAXIA SYNDROME WITH SEIZURE AND RELATED TO EPSTEIN BARR VIRUS

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Case Presentation: A 49 year-old-man previously healthy started with fever, headache, malaise, swallow pain and was diagnosed with pharyngitis and treated with azitromycin and amoxicillin clavulanate, without fever resolution. Twelve days after initial symptoms, he had a generalized tremor and presented a tonic-clonic seizure. It was described anxiety and irritability of the patient. On neurological examination, he had generalized tremor with head and axial titubation, GCS 15, dysarthria, ocular motricity with saccades intrusions and opsoclonus, cerebellar ataxia, tremor with kinetic, postural and intention characteristics and myoclonus. He was started on acyclovir. Brain MRI was normal. CSF with pleocytosis and high protein. CBC had leukocytosis with lymphocytosis. HIV, HBsAg, VDRL and HCV serologies were negative. Cytomegalovirus (CMV) IgG serology was positive and IgM negative. HSV was positive for IgM and IgG. Opsoclonus Myoclonus Ataxia Syndrome (OMS) diagnosis was given. Methylprednisolone was given, with significant clinical improvement. The case presented all four symptoms of the syndrome: opsoclonus, myoclonus of trunk and extremities, step ataxia, and behavioral change. In adults, there is a strong paraneoplastic association, in particular with small cell lung carcinomas, breast and ovarian neoplasia, leading us to perform CT screening of chest, abdomen and pelvis, all negative. Brain MRI was made, also normal. Also relevant in adults are infections, being the most related EBV, CMV, HCV, HIV, HSV, post-streptococcal. In our case, the onset was compatible with infectious mononucleosis. The presence of IgM and IgG antibodies for HSV is probably a cross reaction with EBV. Echocardiogram shown a mild pericardial effusion. A remarkable characteristic is the seizure. We didn't find any case presentation of OMS in literature associated with tonic-clonic seizure. The investigation of the patient rule out encephalitis. Our explanation for the seizure was a cortical disfunction without patterns of encephalitis. Thus, our case presentation is the unique to literature with this specific type of presentation. OMS is a rare entity with difficult diagnosis. Recurrence is common, and there are few studies on prognosis of this population. In our case, a detailed history and neurological examination was crucial to suspect OMS, using opsoclonus as an important feature to accurate diagnosing. We ruled out tumors and stabilized early treatment, resulting in good clinical improvement.

Neurointensivismo

TL 1105901

POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME ASSOCIATED WITH POST-VACCINATION GUILLAIN-BARRÉ SYNDROME, WOULD IT BE POSSIBLE?

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Case Presentation: 70-year-old female with hypertension, obesity and anxiety, taking atenolol and citalopram. Admitted with weakness after 7 days of receiving the Janssen-COVID19 vaccine. She evolved with hyperactive delirium, in addition to paraparesis and hyporeflexia that were difficult to assess, admitted to the ICU for delirium control. Head CT with signs of subarachnoid hemorrhage (SAH) in cortical sulci in the frontal right convexities. Cerebral angiography initially without any vascular. She had been presenting with tachycardia and hypertension despite treatment. She presented worsening of the motor condition and after 7 days of control of the delirium. Brain MRI was performed showing hyper-signal in T2/FLAIR cortico-subcortical in the gyri of the convex encephalic convexities, symmetrical, notably frontoparieto and occipital, without restriction on diffusion; suggesting as posterior reversible encephalopathy syndrome (PRES). The RCVS2 score was 5. Nimodipine was started, and increased intracranial artery velocities were observed on transcranial Doppler. After 2 weeks, presented with sudden cortical blindness; then submitted to arteriography showing diffuse vasospasm in addition to areas of subocclusion in P3 left segments, and received intra-arterial milrinone. For 15 days, she had continuous milrinone and noradrenaline and nimodipine until the vasospasm improved. Control MRI showed areas of right frontal, bilateral parieto-occipital and right cerebellar subacute/chronic ischemic events. CSF with protein-cytological dissociation, was submitted to plasmapheresis, due to a post-vaccination Guillain-Barré Syndrome (GBS) confirmed by an electroneuromyography study. Evolved with improvement of the delirium, treating neuropathic pain and motor rehabilitating. **Discussion:** Recognition of non-aneurysmal SAH causes is still a challenge for the clinician. Systemic assessment of the patient and not just the neurovascular condition is extremely important to recognize the PRES and its possible triggering factors; despite uncertainties regarding the pathophysiology of a minority of cases evolving with unfavorable outcomes. **Final Comments:** The association of GBS with a post-vaccination condition is already well established in the literature, but with few reported cases of association with PRES of severe evolution. Despite the use of medication known to be associated with some antidepressant medications; its association with GBS is poorly described.

Neurointensivismo

TL 1106155

OCULAR MOTILITY IN THE DIAGNOSIS OF COMA

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Case Presentation: A 52-year-old male, obese, hypertensive and non-dialytic chronic renal patient underwent coronary artery bypass graft surgery and wide endarterectomy, with sixty-five minutes of cardiopulmonary bypass (CPB) and forty-five minutes of aortic clamping time, without intercurrents. He progressed in the post-operative period with weaning from vasoactive drugs, maintenance of renal function and reduction of mediastinal drain output. Sedation was turned off 48 hours after the surgical procedure, without awakening or adequate respiratory driving after five days. On physical examination, the patient remained in a comatose state (Glasgow coma scale 3/15), with mild alternating conjugate ocular deviations. A cranial tomography was requested, which showed an increase in the density of the basal ganglia symmetrically and of the thalamus. In the intensive care unit, ocular signs may have prognostic, diagnostic and therapeutic value. Changes such as ptosis, nystagmus and pupillary dilation have different diagnostic values and can help in a faster and more adequate management of the case. In postoperative cardiac surgery units, especially in procedures with cardiopulmonary bypass, cerebrovascular accidents (CVA) should always be included in the list of differential diagnoses, since up to 14% of patients can develop CVA, as well as intraoperative hypoxic lesions. The roving conjugate eye is a slow, conjugated, lateral and symmetrical movement, characteristic of coma and suggestive of ischemic or metabolic lesions. Imaging exams may present without significant changes, which does not exclude brain injury. In the case in question, despite the reduced CPB time and the absence of significant metabolic changes, the risk factor of obesity and the need for endarterectomy may have led to small ischemic lesions, in addition to cardiopulmonary bypass, which constitutes a risk factor isolated for hypoxic and ischemic injuries. Ophthalmological examination is an important step in the physical examination of patients in intensive care and should be a widespread knowledge among attending physicians. Although there are imaging tests that complement and guide, sometimes they become insufficient in the diagnosis, and the clinical examination is an indispensable tool for better management of the patient.

Neurointensivismo

TL 1105026

VERY EARLY-ONSET BEHAVIORAL VARIANT FRONTOTEMPORAL DEMENTIA ASSOCIATED WITH FUS GENE IDENTIFIED VARIANT: A CASE PRESENTATION

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Case Presentation: A 24 year-old male developed progressive neuropsychiatric symptoms following a mild depressive episode, with compulsive behavior, hyperorality, disinhibition and, finally, psychotic symptoms such as delusions and psychomotor agitation, leading to a psychiatric Hospitalization nearly 10 months after symptom onset. The patient was initially diagnosed with schizophrenia and received several treatment attempts, including neuroleptics, SSRIs, electroconvulsive therapy and empiric high-dose corticosteroids, all of which were ineffective. He was discharged after 6 months. The patient progressed with severe cognitive and functional decline within months. At 26 years old, he was referred to our outpatient clinic. On examination, he was globally aphasic and only vocalized a repetitive grunt. He had frontal release signs, mild generalized muscular atrophy and paratonia. Of note, he did not have motor neuron disease. The patient had 11 years of schooling and normal childhood development. He had no history of CNS infections or trauma, nor previous psychiatric disorders. Family history included a paternal uncle with schizophrenia. Extensive investigation ruled out systemic diseases. Neuroimaging performed 1-2 years after symptom onset showed severe atrophy in the hippocampus and head of the caudate bilaterally. Genetic testing identified a variant of uncertain significance (VUS) in the FUS gene: p.Gly229_Gly231dup. **Discussion:** This is a case of very early-onset behavioral variant frontotemporal degeneration (bvFTD) clinical phenotype with an unusual rapidly progressive course. The clinical history and neuroimaging support a hypothesis of FUS pathology, which might be related to the found variant. The fact that the same VUS was found in the patient's asymptomatic 58 year-old father raises uncertainty for establishing this relation, although most FUS gene mutations show incomplete penetrance. Importantly, the identical VUS has been reported with essential tremor and ALS. Other FUS gene variants have been reported, but without pathological confirmation. If neuropathology demonstrates FUS protein deposition, this could be the first case of FUS mutation potentially causing FTLD-FUS pathology. **Final Comments:** FTLD-FUS pathology must be suspected in sporadic, early-onset and rapidly progressive bvFTD. The found VUS could represent a novel FUS gene mutation related to bvFTD, although additional evidence is needed to determine its pathogenicity.

Neurologia Cognitiva e do Envelhecimento

TL 1105144

"DYSMETRIA OF THOUGHT": CEREBELLAR STROKE AS A POSSIBLE CAUSE OF STRATEGIC INFARCT DEMENTIA

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Case Presentation: A 71-years-old woman, 8 years of schooling, presented to Hospital with intense vertigo, vomiting and headache. Brain CT showed cerebellar (CB) infarct in vermis and CB right hemisphere, diagnosed CB stroke. After discharge, the patient developed a non-progressive cognitive decline with amnesic complaints, with executive, attention and social cognition declines. After 9 months, she presented to our service to cognitive investigation. The neurological examination showed right skew deviation and horizontal nystagmus, besides right dysmetria and dysidiadochokinesia. In cognitive tests, she scored 22/30 in the Mini Mental State Examination; 19/30 in the Montreal Cognitive Assessment, with Clinical Dementia Rating Scale (CDR) score equal to 1 and Frontal Assessment Battery scale score of 14/18. In the Schmahmann Syndrome Scale, a specific office to evaluation of cognitive impairment in CB lesions, the patient scored 52/120 points, with fail in 9/10 tests. Laboratory tests were unremarkable. Brain MRI showed increased T2/FLAIR signal in area of previous CB stroke. A diagnosis of probable cerebellar vascular cognitive decline was made. **Discussion:** The cerebellar cognitive affective syndrome (CCAS), was described first in 1998, also named Schmahmann's syndrome, is characterized by declines in executive functions, linguistic processing, spatial cognition, and affect regulation. The "dysmetria of thought hypothesis" attributes the cerebellar-related cognitive declines to changes in pathways connecting the CB with the limbic circuitry, prefrontal and parietal association cortices. Part of patients with CB lesions and cognitive complaints present normal ranges in tests as MoCA and MMSE. Thus, in 2018 was published the CCAS Scale, that recently in 2022 was translate and validated to Portuguese. In our case, we used the validated version of CCAS scale and we detected significant decline in CB functions associated to a related subtle decline in functional daily activity, based on CDR scale, indicating probable dementia. In addition, symptoms were reported to have started soon after acute CB injury and there was no significant report of progressive worsening of these symptoms. **Final Comments:** Cerebellar stroke can be a cause of vascular cognitive impairment, which in this case fulfills the criteria for the diagnosis of a dementia syndrome, thus also fulfilling the criteria for the diagnosis of dementia due to strategic infarctions.

Neurologia Cognitiva e do Envelhecimento

TL 1105155

ATYPICAL DEMENTIA WITH LEWY BODIES: A CASE PRESENTATION

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Case Presentation: Female, 65-year-old, refers restless sleep, visual hallucinations and sporadic forgetfulness for 1 year. Family reports spatial disorientation, with an intensification of the hallucinatory condition for 1 month, period in which persecutory delusions were presented. Developed, after administration of risperidone, difficulty in motor control and, after replacing it with aripiprazole, evolved with worsening of disorientation and delusions. Her education attainment level was college and the Mini-Mental State Examination (MMSE) scored 20, suggesting cognitive impairment. Brain magnetic resonance imaging showed rare foci of microangiopathy, as well as small foci of gliosis in the right thalamus, with no evidence of parenchymal atrophy. Fludeoxyglucose positron emission tomography (FDG-PET) demonstrated bilateral parieto-occipital hypometabolism, with preservation from medial portions of occipital lobes. **Discussion:** This report describes a case of dementia of Lewy bodies (DLB). History establishes cognitive changes impacting function consistent with dementia, a required criterion, with early deficits in executive function and visuospatial ability. Plus, the patient presents core features of DLB: fluctuating cognition, rapid eye movement (REM) sleep behavior disorder and recurrent visual hallucinations, meeting criteria for probable DLB, established by the presence of two or more core clinical features or one core clinical feature plus one indicative biomarker. Antipsychotic hypersensitivity and systematized delusions are supportive clinical features. Further testing can be helpful to establish the diagnosis, and the presence of occipital cortex hypometabolism on FDG-PET imaging is an essential tool in differentiation between DLB and Alzheimer's disease, the latter with typically temporoparietal hypometabolism. The parieto-occipital pattern seen in this case, when aligned with the characteristic patient's clinic, suggests an atypical image of DLB. Treatment is symptomatic to address cognitive, motor and other non-motor symptoms, and involves avoiding medications that may exacerbate symptoms, besides palliative care. **Final Comments:** DLB is diagnosed in the context of dementia with clinical core features of DLB on history and physical examination, with or without indicative biomarker. DLB leads to a decline in functional outcome and quality of life, requiring a multidisciplinary approach.

Neurologia Cognitiva e do Envelhecimento

TL 1105295

AGRAMMATIC PRIMARY PROGRESSIVE APHASIA DUE TO GRN MUTATION: A CASE PRESENTATION

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Case Presentation: Male, 70 years old, caucasian, aircraft mechanic and 11 years of schooling. Patient sought care for behavioral and speech changes that had been progressing for one year. Patient was recently sadder, quieter, with decreased social interaction and functional decline. He'd previously no neurological symptoms. His mother died at the age 86 due to dementia without specific diagnosis. His older brother is diagnosed with cortico-basal syndrome and his younger brother has no diagnosis but he has similar symptoms to the patient. Neuropsychological assessment was notable for significant deficits in the Boston Naming Test (-3.8 SD), MOCA test (7/30), along with impaired comprehension of complex sentences, non fluent speech and altered memory, executive function and attention. Brain MRI showed atrophy especially in the left frontal lobe. A genetic investigation found the GRN T272fs mutation (exon 8, c.813_816del), establishing a genetically confirmed case of non-fluent variant frontotemporal dementia (nfvFTD). **Discussion:** Frontotemporal dementia (FTD) is a clinical and pathologically heterogeneous syndrome characterized by behavioral, language and executive abnormalities. The spectrum of presentation includes behavioral variant frontotemporal dementia (bvFTD), non-fluent variant primary progressive aphasia (nfvPPA) and semantic variant primary progressive aphasia (svPPA). Unlike the clinical syndrome, the term frontotemporal lobar degeneration (FTLD) is reserved to describe pathological changes of the disease that encompasses the syndrome. So FTD is characterized by a disorder with distinct clinical phenotypes that can coexist and are associated with multiple neuropathological substrates. NfvPPA is characterized by degeneration of frontal language networks in the brain. It usually manifests in late middle age with minor difficulties with speech but with preserved executive and cognitive functions that, with disease progression, are also impaired. In etiological terms, it should be noted that mutations in MAPT, C9ORF72 and GRN are the main causes of FTLD, accounting for 30% of the cases. Besides, the most common GRN mutation is T272fs, with a known founder effect in Italy, where the patient's family is descended. **Final Comments:** PPA is an uncommon and underdiagnosed disease often misdiagnosed with Alzheimer's. It's important to highlight that genetic investigation has a main role to confirm diagnosis, allowing better therapeutic approach and even genetic counseling.

Neurologia Cognitiva e do Envelhecimento

TL 1105395

POSTERIOR CORTICAL ATROPHY: VARIANT OF ALZHEIMER'S DISEASE RELATED TO PSEN1 MUTATION MANIFESTING AS RAPIDLY PROGRESSIVE DEMENTIA

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Case Presentation: A 44yo right-handed female artisan, with university degree, was brought to emergency care due to a history of approximately 1 year of progressive memory deficit (losing objects, messages and appointments), which rapidly evolved in 6 months to prominent visuospatial impairment preventing her to drive her car, and dyspraxia hindering her to paint or performing manual tasks and to carry on her work, significant global dysphasia, and urinary incontinence, leading to severe functional limitation. In the month before admission, she no longer handled money or was able to cook. On neurological examination, she scored 5 out of 30 points on the MMSE. Insight and social adequacy were preserved. The patient was disoriented in time and space, ideomotor apraxia was observed, as lack of verbal fluency with anomia, and inability to repeat complex sentences. Also, an ocular apraxia was noticed. Tendon reflexes were symmetric and no frontal releasing signs were observed. Gait was normal. Magnetic Resonance Imaging (MRI) of the brain showed diffuse brain reduction, with marked bilateral parietal atrophy. Genetic testing detected a pathogenic variant in the PSEN1 gene. She was treated with donepezil up to 10mg daily. Her deceased mother had a history of early-onset dementia without a confirmed diagnosis. **Discussion:** The patient's clinical manifestation, with accelerated evolution and dramatic impairment of functionality, draws attention to a rapidly progressive dementia, whose main causes include prion diseases (Creutzfeldt-Jakob), infectious, inflammatory and neurodegenerative diseases, such as corticobasal degeneration, dementia with Lewy bodies and atypical Alzheimer's disease. The proposed diagnosis was posterior cortical atrophy (PCA), an uncommon type of dementia that is often neglected or subdiagnosed. **Final Comments:** Posterior cortical atrophy (PCA) or progressive posterior cortical dysfunction syndrome (PPCD) is a rare but disabling dementia syndrome. This progressive dementia initially presents with visual disturbances and is characterized by visuospatial and visual perceptual impairment, visual agnosia, with a later onset of cognitive decline generally associated with typical Alzheimer's disease. Furthermore, PPCD can sometimes be mistakenly attributed to ophthalmological disorders. Its diagnosis remains challenging, with differential diagnoses including rapidly progressive dementias.

Neurologia Cognitiva e do Envelhecimento

TL 1105440

NEUROPSYCHOLOGICAL ALTERATIONS AND PET/CT ABNORMALITIES AFTER COVID-19 INFECTION: CASE PRESENTATION

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Case Presentation: A 75-year-old woman, diabetic, less than 4 years of schooling, no past or family history of dementia or psychiatric disorder, presented with sudden behavioral change after COVID-19 infection in April of 2021. Throughout the infection, the patient had fever, anosmia, with no need of Hospitalization. After 14 days, exhibited infantilized behavior, dysarthria and did not recognize her family members, mistaking her daughter for her own mother. The Addenbrooke's Cognitive Examination-Revised (ACE-R) and Neupsilin were administered, revealing executive dysfunction, visuospatial impairment, deficits in working memory, language and sustained attention. Patient remains stable after one-year follow-up. Cerebral Magnetic Resonance Imaging (MR) with MR angiography, CSF analysis and laboratory studies identified no abnormalities. Positron Emission Tomography (PET/CT), after F18-FDG injection, showed discrete diffuse cortical hypometabolism, with bilateral frontal, posterior temporal, bilateral parietal, precuneus and cingulate gyrus predominance, not suggestive of vascular dementia. **Discussion:** Cognitive impairment is a complication of COVID-19, that usually initiates during the disease's acute or subacute phase and improves a few months afterwards. Memory and attention deficits are the most common symptoms. In this case, the impairment persists after a one-year course and, unlike late-onset Alzheimer Disease (AD), exhibits disexecutive predominance. Identification of vascular and inflammatory lesions in MR images has assisted in comprehending its pathophysiology, however they were not present in this patient's exams. On the other hand, it needs to be clarified whether COVID-19 can influence the development of dementias, such as AD, accelerating its course, as it is suggested in this case. Cerebral PET and SPECT findings, despite the variety of possibilities, always show, after COVID-19, hypometabolism with frontoparietal or bilateral frontal predominance. The recent finding that SARS-CoV-2 infection induces hypometabolism in brain areas that are affected by neurodegenerative diseases will contribute to better comprehend the neuropsychological profiles in mixed cases of COVID-19 and AD. **Final Comments:** It was presented probably new dementia syndrome characterized by the sudden beginning, prolonged and fluctuating course after COVID-19.

Neurologia Cognitiva e do Envelhecimento

TL 1105521

CREUTZFELD JAKOB DISEASE AND COVID 19: A CASE PRESENTATION

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Case Presentation: We describe a case of a male patient, 48-years-old, whose started 20 days after the diagnosis of coronavirus disease 2019 (COVID 19) with progressive fatigue, binocular diplopia, headache and slowing of thought. After a few weeks, he began to experience difficulties in walking, with ataxia and incoordination. The symptoms has a dramatic evolution over three months with onset of diffuse myoclonus, significant language and movement impairment and finally evolved to akinetic mutism. Complementary investigation whit brain magnetic resonance imaging (MRI) showed restricted diffusion in the cortical and subcortical posterior regions and deep nuclei gray matter (left caudate nuclei). The electroencephalogram (EEG) revealed intermittent generalized periodic discharges (GPD) and diffuse-symmetric slowing. The cerebrospinal fluid (CSF) analysis and the 14-3-3 research were negative. In view of the findings, we agree with the Crutzfeld-Jacob disease hypothesis, supported by University of Edinburgh diagnostic criteria (2017) to probable sporadic Jakob-Creutzfeldt disease. Patient complicated with bronchoaspiration pneumonia with needing of mechanical ventilation. Nowadays he persist in akinetic mutism with tracheostomy and gastrostomy. **Discussion:** Prion diseases are a heterogeneous group of diseases caused by prions -abnormally shaped proteins, and it can be acquired, hereditary or sporadic form. CJD is a more common form of prion disease that affects humans and courses with rapidly progressive dementia associated with behavioral changes, ataxia, extrapyramidal features and myoclonus. Its diagnosis is based on the clinical history and interpretation of the clinical features, CSE, EEG and MRI, in addition to the genetic tests supported by polymerase chain reaction (PCR-RT). There are some reports of patients who developed CJD with a temporal relationship to COVID 19 that were initially interpreted as a coincidence because the rare incidence of CJD and the COVID 19 pandemic that we have faced in recent years. Although, there are descriptions of simultaneous clinical presentations of COVID-19 and CJD, that can indicate some correlated mechanism between them. **Final Comments:** There is still no definitive explanation for the relationship between COVID 19 and CJD, but it is questioned whether the inflammatory cascade caused by the coronavirus infection could accelerate or even precipitate the neurodegeneration characteristic of CJD.

Neurologia Cognitiva e do Envelhecimento

TL 1105568

A "VARIANT" IN THE DIAGNOSIS OF SPORADIC CREUTZFELDT-JAKOB DISEASE

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Case Presentation: M.P.J., 73 years old, presented with a history of progressive gait ataxia since Jan 2021, evolving with hypophonia, memory loss, executive dysfunction, agitation and dysphagia over a few months. In July 2021 the patient became wheelchair bound and totally dependent to perform daily activities. Exam: echolalia, did not follow commands, pupils were miotic and poorly responsive to light. Marked ataxia, unable to stand. Action tremor, myoclonus, mild paratonia and brisk reflexes in the upper limbs. Brain MRI showed restricted diffusion in the left frontoparietal cortex, bilateral pulvinar and caudate nuclei, with the "double hockey stick sign". 14-3-3 protein testing in CSF was negative. EEG was nonspecific. Investigation for other causes was unremarkable. A diagnosis of probable sporadic Creutzfeldt-Jakob disease (sCJD) was made. The patient died within 13 months of symptoms onset. **Discussion:** sCJD is a fatal neurodegenerative disorder. Diagnosis is made through clinical presentation and at least one supporting test: periodic sharp wave complexes in EEG; elevated 14-3-3 protein in the CSF (not sensitive nor specific). In 2009 the criteria were updated to include typical MRI findings. The patient in question filled all clinical criteria for probable sCJD. Despite negative CSF protein 14-3-3 and EEG, she had one positive supporting test, an altered MRI, showing the "double hockey stick sign", reported to be up to 100% specific for the variant form of the disease. Although this sign has been rarely seen in the sporadic form. Correlation with others MRI findings suggestive of sCJD could be useful in the differentiation of vCJD such as: pulvinar involvement with lower signal intensity than putamen and caudate best seen in DWI compared to FLAIR and prominent cortical diffusion restriction [5]. **Final Comments:** This case illustrates how a patient with probable sCJD may present as a mimic of a much rarer and epidemiologically concerning form of the disease, variant CJD (vCJD), with an MRI sign reported to be highly specific to it. Thus, clinicians should be wary when encountering "classical" clinical or imaging findings, and remember to consider the full picture in context. In this case, the patient did not fill the criteria for vCJD besides the MRI finding - she did not live in a risk area, did not have predominant psychiatric symptoms and was not in the age group most commonly affected. [1]

Neurologia Cognitiva e do Envelhecimento

TL 1105620

FAHR'S SYNDROME: A CASE PRESENTATION

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Case Presentation: M. M. V, male, 76 years old, presented symptoms consistent with parkinsonian syndrome and convulsive symptoms associated with fever for two years. Later he progressed with worsening of motor symptoms, presenting chorea and neuropsychiatric alterations such as irritability, excessive anxiety, and mental confusion. One year ago, after a seizure event, the patient presented decreased level of consciousness, was admitted to the Hospital, and requested a CT scan. It revealed calcifications affecting the basal ganglia, dentate nuclei of the cerebellum, and white matter bilaterally, suggestive of Fahr's disease. Laboratory tests were requested and demonstrated hypocalcemia, vitamin B12 deficiency, and low PTH levels. The patient started his therapy during Hospitalization with supplementation of calcitriol, calcium carbonate, and cholecalciferol. After two months of Hospitalization, clinical treatment was supplemented with quetiapine, phenytoin, levodopa, biperiden, and zolpidem, demonstrating significant improvement in neuropsychiatric and motor symptoms. **Discussion:** Fahr's disease is a rare neurodegenerative disorder characterized by idiopathic, symmetrical, and bilateral calcification of the basal ganglia. Symptoms include movement disorders, with parkinsonian syndrome being the most common, in addition to seizures, severe headache, and neuropsychiatric changes. Treatment aims to control symptoms and improve quality of life. **Final Comments:** The literature is still outdated when it comes to how to manage these patients, and studies are needed to control the symptoms and improve the patient's quality of life since it is an irreversible condition and there is no definitive treatment according to the guidelines.

Neurologia Cognitiva e do Envelhecimento

TL 1105621

THE IMPORTANCE OF BIOMARKER TO IDENTIFY THE ETIOLOGY OF POSTERIOR CORTICAL ATROPHY SPECTRUM: A CASE PRESENTATION

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Case Presentation: A 66-year-old female patient with a Law degree was evaluated at an outpatient clinic due to a series of cognitive deficits for the past 2 years: difficulty reading, spatial perception impairment and environmental agnosia. In the past year, she would forget the password for her credit card, developed apraxia, apraxia of dressing, acalculia and impaired perception of objects. The ophthalmological clinical evaluation was unremarkable. She more recently has been repeating stories and had difficulty establishing a line of reasoning within them. On physical examination, she had no pathological signs, except for simultanagnosia. In the Addenbrooke test, she was unable to perform the task of counting dots without pointing at them. In the MoCA test, she achieved a score of 9 and performed notably worse in visual tasks. Brain MRI showed parieto-occipital cortical atrophy mainly on the left hemisphere. In the neuropsychological evaluation, difficulties in verbal and semantic fluency and in visuomotor coordination were observed, as well as executive dysfunction. Brain SPECT showed hypoperfusion in frontotemporal and parietal regions of the left cerebral hemisphere, extending to the hippocampus and posterior cingulate, with a nonspecific scintigraphic appearance. Finally, the PET-CT with Florbetaben-F18 indicated accumulation of β -amyloid plaques predominantly in the prefrontal, orbitofrontal, parietal, temporal, cingulate and occipital regions. **Discussion:** In 1988, Frank Benson and colleagues described five unusual patients with progressive visual difficulties that they termed posterior cortical atrophy (PCA). Subsequent autopsies of cases of posterior cortical atrophy showed a predominance of Alzheimer's neuropathology, but other diagnoses such as prion disease and Lewy body dementia were also reported. Currently, we can use biomarkers that help diagnose the etiology of PCA. This case, the patient had clinical criteria for PCA, that could be related to Alzheimer's disease through PET-CT performed with Florbetaben-F18. **Final Comments:** The use of functional neuroimaging as a biomarker, especially for amyloid deposition, is a helpful tool for identifying the etiology within the spectrum of PCA. Consequently, new clinical trials will be able to investigate atypical phenotypes within the same etiology. Getting to a more specific diagnosis using biological markers is the first step towards finding effective targeted therapies for PCA patients.

Neurologia Cognitiva e do Envelhecimento

TL 1105641

ANAPLASTIC ASTROCYTOMA CAUSING RAPIDLY PROGRESSIVE DEMENTIA – A CASE PRESENTATION

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Case Presentation: Patient M.B.S., 50 years old, male, hypertensive. Two months ago he started with memory and executive function alterations, with an impact on his functionality and his work. In addition, with severe left hemispheric headache, stabbing type, lasting 2 minutes, intermittent, without photophobia, phonophobia, nausea or vomiting. After 2 months, he presented with seizure of unknown onset with evolution to generalized tonic-clonic, without associated fever. No weight loss. Physical examination revealed mixed aphasia of motor predominance, with phonemic paraphasia and agraphia. No change in strength, sensitivity or coordination. Cranial resonance with T2/FLAIR hypersignal affecting the caudate nucleus, putamen, globus pallidus, hippocampus and left insula, as well as cortical surface of the frontal and homolateral parietal regions, with mass effect, without DWI restriction and without gadolinium enhancement. Ascopsy with choline peak and n-acetyl-aspartate reduction. Biopsy showed grade III anaplastic astrocytoma. Descompressive craniectomy and radiotherapy were performed. **Discussion:** Analyses of the global disease burden have shown that 43.8 million people lived with dementia. Assuming that a substantial percentage of these patients presented with rapidly progressive dementias (RPD), it is evident the importance to consider superimposed reversible conditions in the differential diagnosis of RPD. Besides paraneoplastic immunemediated encephalopathies, primary CNS neoplasias are also potential causes of RPD. These neoplasias can affect cortical and subcortical structures through space occupation, infiltration and destruction of brain tissue, secondary edema or disturbance of CSF circulation resulting in intracranial hypertension. However, treatment options that prolong survival and might improve cognitive performance are becoming available. **Final Comments:** In RPD, the chances of treatment success are better the earlier the diagnosis is made. Mainly when potentially treatable pathologies are identified.

Neurologia Cognitiva e do Envelhecimento

TL 1105726

WHITE MATTER HYPERINTENSITIES IN A SPORADIC CASE OF SEMANTIC VARIANT PRIMARY PROGRESSIVE APHASIA: A CLUE TO GRN GENOTYPE

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Case Presentation: A previously healthy 66-year-old woman presented with a four-year history of progressive difficulty in communication. The patient's chief complaint was a "very bad voice". She initially had occasional problems in understanding single words and often asked for sentences to be repeated. Gradual worsening of the symptoms led to severe impairment in comprehension. She eventually stopped reading and praying. There were no memory, orientation or behavioral problems. There was no family history of neuropsychiatric disorders. On the neurological examination, she had no signs of motor neuron involvement, nor parkinsonism. She exhibited aphasia with severe loss of fluency, very poor object naming and single-word comprehension combined with surface dyslexia. The patient scored 14 points at the Mini-Mental State Examination (MMSE) (16 years of education), 10/20 points at the Boston Naming Test and 6 and 3 points at Verbal Fluency Test for animals and fruits, respectively. Brain magnetic resonance imaging (MRI) revealed asymmetric left temporal atrophy, prominent left parietal atrophy and white matter hyperintensities. The clinical diagnosis was semantic variant primary progressive aphasia (svPPA). A next generation sequencing panel for dementias identified a pathogenic variant in GRN gene (c.1252C>T – p.Arg418*). **Discussion:** The hallmark of the Primary Progressive Aphasia (PPA) is the insidious deterioration of language, with relative preservation of other cognitive functions. This group of disorders is clinically heterogeneous, and comprises at least three subtypes: nonfluent, semantic and logopenic. SvPPA is characterized by anomia associated with loss of semantic knowledge, and severe impairment in single-word comprehension. Familial cases of svPPA have been linked to pathogenic variants in GRN gene, which encodes the protein progranulin. However, sporadic cases of svPPA harboring pathogenic variants in this gene are extremely rare. Our patient had a clinical phenotype consistent with svPPA, and no family history. White matter hyperintensities, in combination with asymmetric temporoparietal atrophy have raised the suspicion for a GRN genotype, which was subsequently confirmed by the molecular study. **Final Comments:** GRN genotype is rare among sporadic svPPA cases, but white matter hyperintensities and asymmetric temporoparietal atrophy should reduce the threshold for molecular investigation.

Neurologia Cognitiva e do Envelhecimento

TL 1105754

CASE PRESENTATION LATE PRESENTATION CREUTZFELDT-JACOB DISEASE

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Case Presentation: An 88-year-old female patient, previously functional, presented with gait disturbance, progressive involuntary movements, dysphagia, dysarthria and altered cognitive status with an evolution of only 3 months. In the evaluation, the patient presented with eye opening to verbal stimuli, non-contacting, non-responsive to commands, in akinetic mutism, with spasticity and myoclonus in the 4 limbs, in addition to signs of frontal release. In view of the rapid cognitive decline and loss of important functionality associated with myoclonus, investigation for rapidly progressive dementias (RPD) was initiated. Cranial magnetic resonance imaging (MRI) showed areas of restriction to diffusion in the cortical region, bilateral, predominantly on the left, in addition to an electroencephalogram with a periodic sharp-wave complexes, filling in, after ruling out other causes of PRD, diagnostic criteria for Creutzfeldt-Jacob disease (CJD). **Discussion:** CJD belongs to a group of degenerative diseases caused by infectious protein particles (prions), which are pathogenic forms caused by the conversion of normal prion protein into its abnormal isoform. This process is replicated exponentially, which explains the rapid development of the disease. Clinical diagnosis is based on a set of symptoms and complementary exams, including CSF, EEG and MRI. The classic phenotype is rapidly progressive dementia with behavioral changes, ataxia (usually in gait), extrapyramidal signs and myoclonus. The EEG demonstrates periodic complexes of sharp waves, whereas the MRI may present with diffusion restriction in the cortex or in the basal ganglia. CSF is usually normal and the usefulness of biomarkers such as 14-3-3 is still controversial due to the variability of sensitivity and specificity. The disease is inevitably fatal, with an average survival of 6 months, as there is no specific treatment to date. **Final Comments:** CJD affects many areas of the brain, so it can present itself as a multiform and it can also mimic, particularly early on, many other neurological and psychiatric conditions, which can delay diagnosis. However, despite having no specific treatment, it is highly stigmatizing, and its suspicion and diagnosis are important for a better management of symptoms.

Neurologia Cognitiva e do Envelhecimento

TL 1105793

RAPIDLY PROGRESSIVE DEMENTIA IN A PATIENT WITH NON-HEMORRHAGIC CEREBRAL AMYLOID ANGIOPATHY: A CASE PRESENTATION

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Case Presentation: A 84 year-old male patient with a history of follicular low grade non-hodgkin's lymphoma treated, evolving for two months with loss of self-care, significant difficulty in walking, total dependence for daily life activities associated with cognitive impairment, especially recent memory. He denied urinary incontinence. Infectious, paraneoplastic, immune-mediated processes were ruled out. MRI of the brain showed diffuse encephalic volumetric reduction, without lobar predominance. Hyperintense lesions on T2/Flair in the white matter of the cerebral hemispheres, without expansive character and a tendency to confluence in a periventricular situation, suggestive of moderate degree microangiopathy, as well as enlarged perivascular spaces, mainly affecting the basal ganglia, this finding being a marker of small vessel disease. For diagnostic clarification a brain biopsy was performed with characteristic findings of amyloid angiopathy with an inflammatory pattern. Treatment was performed with methylprednisolone 1g daily for 3 days with significant improvement in the patient's quality of life

Discussion: Cerebral amyloid angiopathy (CAA) is characterized by acellular thickening of the walls of small and medium arteries due to accumulation of amyloid. CAA is often asymptomatic but usually manifests as dementia and vascular syndromes, and there is an important association between CAA and spontaneous intracerebral hemorrhage. In this sense, we present a case of non-hemorrhagic CAA, with an inflammatory pattern, presenting as rapidly progressive dementia. Histologically analyzed lesions can be of amyloidoma, CAA with primary angiitis of the central nervous system (SPAC) and, as in this case, they consisted of isolated CAA. Its pathogenesis is not very clear, but amyloid deposition is accompanied by inflammation or edema, in contrast to the usual presentation of patients with CAA, involving acute intracerebral hemorrhage. **Final Comments:** The literature emphasizes the diagnosis of CAA and its association with hemorrhages and dementia, however rare cases of non-hemorrhagic brain lesions are associated with CAA, as exposed above. Little information is available regarding their treatment. Non-hemorrhagic AAC has shown a satisfactory response to corticosteroid treatment in patients with evidence of inflammation, as we also evidenced in our case. Thus, although rare, it should be considered in the differential diagnosis of lesions of the central nervous system.

Neurologia Cognitiva e do Envelhecimento

TL 1105816

A LATE PRESENTATION OF FRONTOTEMPORAL DEMENTIA (FTD) ASSOCIATED WITH PSYCHIATRIC SYMPTOMS: A CASE PRESENTATION

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Case Presentation: A 73-years-old patient, male, from Manaus, Amazonas, arrived to our service with a long-standing sleep disorder, and about 2 years ago the clinical condition was associated with anterograde amnesia, in addition to episodes of disorientation and mental confusion, according to his wife. He progressively evolved with behavioral changes, characterized by agitation, akathisia, constant speech of persecutory content and worsening of sleep disorders, followed by episodes of visual hallucination. He denies a history of neurological or psychiatric diseases in the family, and at the time of the first consult was using Risperidone and Rivastigmine. On the neurological examination was slightly disoriented in time, space, person and situation. CGS 14, restless, with constant talk of self-destructive content, echolalia and akathisia. Osteotendinous reflexes abolished in upper limbs, Patellar present, Achilles absent and Glabellar tap was fatiguable. Cognitive tests were distorted, with MMSE score of 15 out of 30 and clock test score of 1. MRI showed moderate atrophy, without hippocampal predominance and without signal changes or ventricular dilatation, leading to the probable diagnosis of Frontotemporal dementia (FTD). After discussion by the neurologic team, Periciazine and Olanzapine were introduced and Risperidone was suspended. In the following months, patient returned with support of the clinical picture, in addition to the appearance of essential tremor in upper limbs, maintenance of sleep disorder and he became dependent on family members for basic-life activities around 6 months since the first consult, and Citalopram was added to the prescription for symptomatic control. The follow-up continued at our service to assess the progression of the cognitive-behavioral status, and he died at home in 2022 after approximately 3 years of follow-up. **Discussion:** The case reveals a late presentation of FTD, since the most affected age group is under 65 years, and helps to emphasize that the presence of this condition in older ages should not be ignored. The presence of hallucinations and sleep disorders are not as common as behavioral and language changes, fact that may have directed examiners to hypothesis such as Lewy-body dementia in this case. **Final Comments:** Healthcare providers should order routine tests and deep examination to rule out other conditions that cause similar symptoms and advised on the reserved prognosis of this disease.

Neurologia Cognitiva e do Envelhecimento

TL 1106058

THE CEREBELLAR COGNITIVE AFFECTIVE SYNDROME AS MANIFESTATION OF ISCHEMIC STROKE: CASE PRESENTATION

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Case Presentation: The cerebellum is the control center for voluntary movements and coordination contributing to planning, execution and modulation of movements. However, research reveals a wider role of the cerebellum in cognitive, emotional, and behavioral functions because of widespread cerebellar connections to the cortex and associated areas. Clinically, Cerebellar Cognitive Affective Syndrome (CACAS), also known as Schmahmann's Syndrome, is characterized by language deficits, visual and spatial executive functions, and personality changes seen in patients with focal cerebellar lesions. Some studies with maps of lesions with clinical diagnosis in patients with CACS and normal motor performance suggest the posterior cerebellar injury involving the lobes VII and VIII, being a risk factor for cognitive deficits. We report the case of a male patient, 82 years old, admitted to a semi-intensive therapy presenting with a delta time greater than 5 hours with speech impaired and understanding, inappropriate laughter and presence of logorrhea in the eyes of his relatives (wife and son). Patient referred for Hospitalization to the care of neurology as a diagnostic hypothesis of Cerebral Vascular Accident. Neurological physical exams show a conscious and cooperative elderly man with cautious gait, reduced grammatical speech, lack of fluency and inadequate understanding and laughter; tests for coordination (nose finger test, heel test and dysdiadochokinesia) were normal, sensory and motor systems within normality, cranial nerves without alterations. Mental functions reveal impaired attention and concentration, recent memory, and altered perception. MRI Brain showed a peripheral intraparenchymal focus in the right cerebellar hemisphere presenting restriction to the diffusion of water in the Ecoplanar sequence, with a weak hypersignal in the T2 / FLAIR sequence. After extensive diagnostic and exclusionary investigation, the diagnosis of Cerebellar Vascular Accident with cognitive manifestation and without motor impairment was instituted. The Cerebellar Cognitive Affective Syndrome conceptualized by Schmahmann and Sherman, seen in a patient with cerebellar damage, was clinically prominent in patients with cerebellar posterior lobe disease and worms, and there may be several causes of cerebellar damage, such as viral infections and drugs, but no specific etiology can be detected in our patient, which evidenced cause of ischemic vascular etiology of cerebellar damage.

Neurologia Cognitiva e do Envelhecimento

TL 1106268

LEUKOENCEPHALOPATHY WITH EVANESCENT WHITE MATTER PRESENTING WITH SUBCORTICAL DEMENTIA, A CASE PRESENTATION

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Case Presentation: A 48-year aged woman was attended at the SCMSP Cognitive and Behavioral Neurology outpatient clinic in May 2020. She was graduated on high school and had a job as a supermarket cashier. She failed 1 year at high school and was fired on one occasion when she argued with a customer. On 2017 she had an isolated episode of tonic-clonic seizure, evolving with a progressive picture of incoordination of gait and cognitive processing speed impairment. She also had chronic visual blurring and early menopause at age 45. On neurological examination, she was inattentive, disoriented in time and space, with optic atrophy and cerebellar ataxia. She presented significant impairment on cognitive scales MMSE: 18/30 and functionality scales, KATZ: 6/6 and LAWTON: 18/27. She underwent neuropsychological assessment, which showed a 50 IQ, severe orientation and recent memory impairment, significant emotional lability and executive dysfunction, little initiative and flexibility, and a lot of confabulation. Neuroimaging revealed cystic degeneration of the white matter of both cerebral hemispheres with extension to the U-fibers and diffusion-symmetric curvilinear periventricular restriction. She underwent genetic testing, which revealed homozygosity in the EIF2B5 gene, with a p.Arg113His mutation. **Discussion:** LEWB is an autosomal recessive, chronic and progressive leukodystrophy caused by mutation in any of the 5 genes encoding eIF2B. It typically presents with ataxia, spasticity and optic atrophy that typically follow an initial stressor event. On adults, a case series with 16 patients (Labauge et. Al, 2009) showed cognitive decline in 8 subjects, in addition to spasticity and/or cerebellar syndrome, seizures, dementia, psychiatric symptoms, and ovarian failure (Ovarioleukodystrophy). The correlation between LSBE and its neuropsychiatric presentations has already been studied by authors such as Van der Knaap, 1997, in a report of 9 children with diffuse cystic degeneration of BS, relatively preserving the cerebral cortex; Prass, 2001, in a report of 1 patient with presenile dementia syndrome; Van der Knaap, 2004, in a report of 1 case of a 25-year-old woman who presented paranoid behavior, evolving with a decline in cognitive functions. **Final Comments:** Our patient presented with a subcortical dementia profile with spatial disorientation, slow cognitive processing, attentional impairment, dysnomia, visuoconstructive disorder with preserved recognition and cortical function

Neurologia Cognitiva e do Envelhecimento

TL 1106291

ADULT-ONSET LEUKOENCEPHALOPATHY WITH AXONAL SPHEROIDS AND PIGMENTED GLIA

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Case Presentation: A 45-year-old man consulted his doctor with a year's onset of memory problems and behavioral changes. He worked as a delivery man for a company and began to mix up his deliveries and get lost in previously known streets. His wife also noticed he was forgetting to pay the bills, a task he had always done without problem, and that he was using the money for other purposes. Because these problems were getting worse and affecting the life of the patient and his family, the wife chose to take him for a medical examination. He underwent cognitive testing and his results were indicative of mild cognitive impairment, especially in the memory and visual-spatial areas. Screening tests were ordered. His blood tests were normal and the CSF showed only a slight increase in total protein. The MRI revealed atrophy and white matter lesions in the frontal lobe of the cerebrum and around the lateral ventricles, making those appear enlarged; a thinning of the anterior corpus callosum; and small calcifications in the white matter around the semi-oval centers, radiated crowns, and knee of the corpus callosum. These findings suggested Adult-Onset Leukoencephalopathy with Axonal Spheroids and Pigmented Glia (ALSP) and a genetic test was done for further research. A Variant of Uncertain Significance, c.2522A>G (p.Tyr841Cys), was identified in the colony-stimulating factor-1 receptor (CSF1R) gene, which is associated with autosomal dominant hereditary diffuse leukoencephalopathy with spheroids (HDLS) and pigmentary orthochromatic leukodystrophy (POLD). Once both disorders were linked to CSF1R gene variants, they became known as ALSP. Treatment was initiated with a cholinesterase inhibitor and antipsychotic drugs, with a partial response from the patient, mostly in behavioral symptoms. Non-pharmacological approaches were also established. The family was oriented about the diagnosis, and the disease's progressive nature, and they were sent to genetic counseling. ALSP is a rare, progressive neurological disease that causes leukodystrophy, forming lesions in certain brain areas due to disease-causing variants in the CSF1R gene. It is estimated to occur in 10 to 25% of adult-onset leukodystrophies. Lesions of the white matter lead to major changes in personality, cognition, and muscle function. There are currently no approved treatments for ALSP and current treatment options do not reverse brain damage but instead are meant to manage symptoms.

Neurologia Cognitiva e do Envelhecimento

TL 1106315

CREUTZFELDT-JAKOB DISEASE HEIDENHAIN SUBTYPE: CASE PRESENTATION

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Case Presentation: J.T.M., female, 67, alcoholic. Social isolation begins and evolves with a bilateral visual deficit, abdominal pain, and headache. She presented with dental septicemia. At examination: miotic pupils, photoreactive pupils, upper limbs with spasticity, paratonia, and myoclonus. She didn't interact and didn't obey commands. In addition, patellar hyperreflexia and bilateral flexion on plantar reflex. Evolution with gait ataxia, changes in motor coordination, ideomotor and speech apraxia, and behavioral changes were present. During Hospitalization, she evolved with bilateral extension at plantar reflex. 14.3.3 protein was dosed due to the hypothesis of spongiform encephalopathy. Its result showed a value of 33,712 (0-19,999). There has been an intense investigation of differential diagnoses. The patient began to have anticipated complications and progressed to support. **Discussion:** Regarding CJD, about 80 to 85% of cases are caused by the sporadic form, with prevalence in the elderly population, between the 5th and 6th decade of life, with no sex differentiation and absence of distinct transmission patterns. In Heidenhain subtype CJD, the Parieto-Occipital cortex is affected, affecting the primary visual area and causing image changes such as cortical hyperintensities on T2/FLAIR MRI and basal ganglia (striatum and thalamus). The electroencephalography (EEG) shows periodic, three-phase acute waves, predominantly in the posterior areas. 14.3.3 protein can also be dosed from the cerebrospinal fluid. Prognosis is one year of life after the onset of symptoms. The classic presentation of this variant involves cortical blindness capable of generating various types of visual changes, visual field restriction, hallucinations, and visuospatial recognition. Neuropsychiatric symptoms culminated in agitation and aggression. The symptoms progressed in a short time (60 days) until presenting akinetic mutism, typical of the disease. Measurement of 14.3.3 protein in the CSF, among the presence of at least two supportive clinical criteria, and the association with progressive dementia were crucial for defining a probable diagnostic hypothesis. **Final Comments:** The signs and symptoms described in the literature were compatible with those presented in this case, representing the Heidenhain subtype of sCJD. There is no specific treatment. Currently, the basis of the therapeutic approach is on supportive measures.

Neurologia Cognitiva e do Envelhecimento

TL 1106502

AMYLOID- β -RELATED ANGIITIS: A CASE PRESENTATION

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Case Presentation: 76 years old female, previously diagnosed with systemic arterial hypertension and diabetes mellitus 2, presented episodic falls, chronically progressing to cognitive impairment, due mainly to apraxia and loss of motor functionality, without rigidity or other parkinsonian symptoms. After 2 months of worsening of symptoms, patient had new seizure, with tonic posture in four limbs, leading to seek medical care. On admission, cognitive assessment showed ideomotor apraxia, but, due to reduced level of consciousness, the exam was jeopardized. The patient underwent a non-contrast CT scan, which demonstrated subcortical diffuse hypodensity. For further investigation, brain MRI was performed, revealing T2 and FLAIR hyperintensity of subcortical white matter and SWI several microbleeds in right parietal lobe. Besides, serology, biochemical exams and cerebrospinal fluid did not present any alterations. Diagnosis of amyloid angiitis was proposed. High dose IV methylprednisolone was started, with significant improvement of cognitive symptoms, however, after 4 days, the patient had a seizure, with worsening of awareness. After two weeks of Hospitalization, patient was discharged with mild to moderate improvement of cognitive function, but locomotion was disabled. 60-day re-evaluation at neurological clinic showed betterment of walking and cognitive impairment. **Discussion:** Cerebral amyloid angiitis, a rare condition, given the evidence of 50 cases in the literature, linked to aging and resulting from the pathogenic mechanism of autoimmunity to β -amyloid, insoluble nature, which are deposited in cerebral vessels, causing a local inflammation with eventual fibrinoid necrosis and, later, cerebral infarction, being demonstrated on MRI a hyperintensity and cortical/subcortical microhemorrhages. Symptoms include altered mental status, focal neurological deficits and seizures which, in a complete analysis of the clinical case, may lead to a diagnosis of amyloid angiitis. **Final Comments:** Amyloid angiitis is a difficult pathology to diagnose and staging due to its clinical presentation, which matches other etiologies, usually being a differential diagnosis. Finally, even in the face of published studies, it is essential to conduct further research on this subject, since there is little current research, besides the lack of probable diagnostic measures.

Neurologia Cognitiva e do Envelhecimento

TL 1106557

CREUTZFELDT-JAKOB DISEASE AND THE CHALLENGES OF THE DIAGNOSTIC INVESTIGATION: A CASE PRESENTATION

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Case Presentation: A previously healthy 68 yo male initiated symptoms of imbalance and gait disturbance, followed by anterograde amnesia and impaired functionality. In the following weeks, he progressed with decreased verbal fluency and social interaction, worsening of gait and became completely dependent to perform daily activities. A month after the initial symptoms he was Hospitalized with akinetic mutism, myoclonus, severe dysphagia, and dystonic upper limb posture. The initial investigation with electroencephalogram (EEG) and brain magnetic resonance imaging (MRI) showed no characteristic signs of the disease, despite the study of cerebrospinal fluid (CSF) showing the presence of 14.3.3 protein. The third performed EEG showed signs of diffuse encephalopathy and periodic generalized discharges that infer a high propensity to epileptic seizures, suggesting the possibility of prion disease. A posterior brain MRI showed cortical restricted diffusion and FLAIR hyperintensity on occipital lobes, head of caudate nucleus and putamen and DWI/ADC abnormalities in temporo-parietal cortex. Signal abnormalities were subtle on the previous MRIs but became more pronounced with the progress of the disease as well as the cerebral atrophy. Given the patient's clinical condition, associated with typical EEG and MRI and the protein 14.3.3 on CSF, the patient had diagnostic criteria for probable sporadic Creutzfeldt-Jakob Disease (sCJD). He was discharged after four months in Hospital, taking anticonvulsants to control myoclonus. **Discussion:** Prion diseases are rare and fatal degenerative diseases of the central nervous system, with an estimated annual incidence of 1 to 1.5 cases per million inhabitants. They can occur sporadically, genetically or acquired. sCJD is the most common form of them and its median survival is about six months. Although clinically variable, neuropsychiatric decline, cerebellar manifestations, myoclonus, and extrapyramidal signs are present in most cases. MRI has a diagnostic accuracy of 97%. EEG sensitivity increases in more advanced cases, requiring repetition throughout the clinical course. The sensitivity and specificity of the 14.3.3 protein is variable. CSF prion detection is the gold standard test for lifelong diagnosis. Treatment consists of clinical support. **Final Comments:** This report describes a rare disease, emphasizing the importance of persisting in the complementary investigation, even in the face of uncharacteristic initial tests.

Neurologia Cognitiva e do Envelhecimento

TL 1105116

MULTIPLE SCLEROSIS IN A 13-YEAR-OLD GIRL

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Case Presentation: Female, 13 years old. Asthma was mentioned as the only pre-existing disease. Reported that three months ago had started with paresthesia and loss of sensitivity in both lower limbs, but in the last month he remained only in the LLL, which culminated in the difficulty to extend and flexing the thigh, leg and left foot, this being the only reason for seeking the health service. Presented negative Brudzinski and Kering signs, isophotoreactive pupils and no cognitive alterations; grade II strength in the LLL, weak contractions, producing movement with elimination of gravity; and degree V of strength in the RLL, in addition with presence of varicose veins in the medial region of the thigh. Underwent complete blood count, EchoDoppler of the LLL and carotid arteries, echocardiogram and cerebrospinal fluid examination, speech-language analysis and pediatric evaluation – normal results. However, initial CT showed small hypodense area in left frontal lobe with an aspect unspecific to method. Brain MRI showed focal areas with signal alteration in white matter bilaterally, compromising periventricular, juxtacortical and optical radiation topography bilaterally, without contrast enhancement. Extending the scan, MRI of thoracic spine showed characteristic lesion of recent inflammatory activity, leading to sclerosing demyelinating disease (MS) hypothesis, which was confirmed by focal lesion identification with enhancement through the contrast at the T5 vertebral body. Pulse therapy with synthetic steroids was already started and this follow-up was necessary on an ongoing basis. Closed diagnosis of MS-with recurrent outbreak, re-presenting at two-month intervals. Etiology remained unknown. According to DATASUS, state of SC is in 2nd place in relation to cases of disease in the southern region, having that, information pertinent to this situation in young patients, exposing the diagnostic protocol, can help more effective therapeutic outcomes. Therefore, rapid diagnostic, management and treatment of young patients with MS is increased through detailed anamnesis and neuroimaging. As a is chronic inflammatory disease of an autoimmune nature, adequate treatment increases and improves the patient's quality/expectation of life. Knowledge of the epidemiological profile associated with a detailed neurological anamnesis supported by neuroimaging exams can reduce the interval between diagnostic confirmation and treatment, defining a more accurate and decisive prognosis.

Neurologia Infantil

TL 1105441

INFANTILE NEUROAXONAL DYSTROPHY: REPORT OF TWO SISTERS

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Case Presentation: We present a case presentation of sisters with infantile neuroaxonal dystrophy (INAD), one of them was diagnosed. Both are daughters of non-consanguineous parents and there were no complications during pregnancy and childbirth. Case 1: female, 7 years and 7 months, the onset of symptoms at 9 months, with difficulty in crawling, convergent strabismus, and hyperopia. Progressive worsening of symptoms: at 2 and a half years she developed loss of cephalic support and at 5 years started clonus in all four limbs. Physical examination without interaction with the environment, the neck was in hyperextension, without cephalic support. She had nystagmus, temporal optic nerve pallor, muscle atrophy, reduced strength, and generalized spastic hypertonia. MRI revealed changes in cerebellar volume and subtle hyper signal on T2 and FLAIR. Molecular analysis by exome sequencing was performed and two heterozygous variants in the PLA2G6 gene were identified. The association of exams with the clinical picture confirmed the diagnosis. Case 2: female, 3 years and 10 months, the onset of symptoms at 2 years, with difficulty in walking. Progressive worsening of the condition resulting in motor incoordination and speech impairment, and inability to walk and sit without support. Physical examination with interaction with the environment, nystagmus, convergent strabismus and no changes in the optic disc. She had muscle atrophy, generalized hypotonia, and appendicular and axial dysmetria. Absence of tremors and clonus. Laboratory tests without changes. The patient awaits exome performance to clarify the diagnosis but receives the same treatment as her sister. **Discussion:** INAD is an autosomal recessive neurodegenerative pathology associated with mutation of PLA2G6 gene. It is rare and has a poor prognosis, whose symptoms are marked by regression of neuropsychomotor development and neurological impairment, gait disorders, extrapyramidal symptoms, and ophthalmological abnormalities, with rapid and progressive evolution, usually fatal. Both cases have characteristics like those found in the literature: onset of symptoms (average of 14 months, with a range of 12-22), characteristic clinical picture and progressive worsening, no type of complications during pregnancy and childbirth, and non-consanguineous parents. **Final Comments:** Even presenting similar characteristics to other reports, the cases bring important data that aggregate INAD study, in search of better prognosis and conduct.

Neurologia Infantil

TL 1105575

REPORT OF A RARE CASE OF PONTOCEREBELLAR HYPOPLASIA TYPE 6 IN A CHILD IN THE AMAZON REGION OF NORTHERN BRAZIL

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Case Presentation: EGM, female, 2 years old, presenting with recurrent and uncontrolled seizures since 4 days of life, in addition to spasms and myoclonus. Neuropsychomotor and hypersecretive delay. Cranial Computed Tomography showed prominence of the retrocerebellar cerebrospinal fluid space in the posterior fossa, suggesting the possibility of megacisterna magna. He had frequent seizures during Hospitalization. The diagnostic hypothesis of Pontocerebellar Hypoplasia was then raised. The Exome Post Expanded Panel, from the DNA extracted from nasal swab, was requested. In the molecular analysis by exome sequencing, two heterozygous variants were identified in the RARS2 gene, position chr6: 87,589,923, T > C variation, concluding the diagnosis of Pontocerebellar Hypoplasia Type IV, associated with Double Hemiparesis, Epilepsy and Global Delay in the Neuropsychomotor Development. For better quality of life and refractoriness of epileptic seizures, the drugs OXCARBAMAZEPINE, PHENYTOIN, DIAZEPAN, PHENOBARBITAL, LEVETIRACETAM, CLOBAZAM were scheduled, in addition to general support. Due to neurodegenerative dysphagia, a gastrostomy was made to maintain the diet. Genetic counseling and general guidelines were carried out. The patient was referred for outpatient, multidisciplinary follow-up and rehabilitation. **Discussion:** Pontocerebellar Hypoplasia (HPC) is a group of rare, hereditary, progressive neurodegenerative disorders that start in utero. It is a heterogeneous group of rare neurodegenerative diseases caused by genetic mutations and characterized by progressive atrophy of various parts of the brain, cerebellum or brain stem. Seven subtypes of mutations have already been identified, and in all of them there are characteristics of severe cognitive and motor deficiencies. When known, these disorders are inherited in an autosomal recessive manner. Treatment for all types is only symptomatic and supportive, as the prognosis is of death during early childhood in most of the few cases described in the literature, and this was established for the patient in question. **Final Comments:** The reported case and cited publications raise the complex situation that is the described disease and that despite the poor prognosis and the absence of specific treatment, the correct diagnosis and stabilization of a symptomatic and supportive therapy can improve the quality of life of patients, patients and family members.

Neurologia Infantil

TL 1105689

LEUKOENCEPHALOPATHY WITH VANISHING WHITE MATTER: A CASE PRESENTATION

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Case Presentation: CRS, male, 2 years and 2 months, from Goiânia-GO. A previously healthy child, 15 months of age, daughter of first cousins, after receiving a booster vaccine (triple viral vaccine, dTP, hepatitis A, OPV) developed high fever, pain in the lower limbs, edema and hyperemia at the injection site. He evolved with progressive tetraparesis with difficulty sitting and supporting the trunk, inability to walk, spastic hypertonia in four limbs, presence of abnormal reflexes, macrocephaly and seizures. A genetic test was performed showing EIF2B5 alteration and cranial magnetic resonance imaging suggestive of leukoencephalopathy with vanishing White matter (VWM). **Discussion:** VWM, also known as diffuse central myelinopathy and Cree leukoencephalopathy, is a chronic and progressive white matter disorder exacerbated by infection, head trauma or other stresses. These stresses can trigger the first symptoms of the disease or worsen existing symptoms and can cause affected individuals to become lethargic or comatose. The inheritance of VWM is autosomal recessive. Mutations in any of the five genes encoding the eukaryotic translation initiation factor EIF2B subunits are the cause of VWM and its phenotypic variants. The partial loss of EIF2B function makes it more difficult for the body's cells to regulate protein synthesis and cope with changing conditions and stress. Glial cells appear to have a selective vulnerability to decreased eIF2B activity, which explains the predominant involvement of white matter in the brain. The most distinctive clinical feature of VWM is progressive neurological deterioration, often with prominent ataxia and spasticity. There are five clinical subtypes of VWM based on age of onset. Patients with an earlier age of onset have a more severe course and decline more rapidly. Prevalence is unknown. Although it is a rare disease, it is believed to be one of the most common hereditary diseases that affect the white matter. **Final Comments:** The findings in the literature are compatible with the clinical picture of the patient described here. Elucidating the clinical course of VWM will be useful to facilitate early diagnosis and avoid specific situations that could trigger the deterioration of the disease, as well as to better understand the pathogenesis and provide subsidies for possible therapeutic targets and treatment strategies in the future.

Neurologia Infantil

TL 1105752

MUTIPLE UNUSUAL CONDITIONS ASSOCIATED WITH CEREBRAL PALSY IN A PRETERM WAS BORN IN BAHIA

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Case Presentation: A 3-year-old male was diagnosed with epileptic encephalopathy, cerebral palsy, autism, Autosomal Recessive Deafness 77 and identification of genetic variants (CAXNA1C, FKTN, SLC19A3), with delayed neuropsychomotor development. According to the mother's report, during pregnancy, she had constant symptoms of severe headache (10/10), agitation, failure to maintain sleep, a seizure episode that was treated with carbamazepine in the second trimester, preeclampsia, hematemesis and gestational diabetes. In the prenatal exams, the patient was, in most cases, without intrauterine movements and at other times with heart arrhythmia. Therefore, an emergency caesarian section was performed at 34 weeks in the state of Bahia. The patient was unconscious and cyanotic, required orotracheal intubation and an exchange transfusion, was diagnosed with epilepsy in the first days of the ICU, the transfontanelar ultrasound examination identified grade 3 of peri-intraventricular hemorrhage, in addition to early neonatal sepsis. After his condition improved, he became jaundiced and was treated with phototherapy. At 7 months, the first case of coma, when the mother noticed a cyanotic appearance, leading to the child's second Hospitalization. On return to the ICU, with oxygen saturation of 60%, Glasgow exam of 4, magnetic resonance imaging of the skull was performed and results were observed with cerebral atrophy and increase in the interpeduncular cistern. At two years for age, he had 3 episodes of cardiorespiratory arrest. The electrocardiogram performed for investigation showed right atrial and biventricular overload. One year later, he was Hospitalized due to the presence of urine calculus and severe UTI, grade 3 phlebitis in the lower limb, pericardial effusion, constipation and convulsion, with the latter having been treated with diazepam and phenytoin. Holter confirmed ectopic atrial rhythm. Currently, on an outpatient basis, he is stable, using topiramate 50 mg, phenobarbital drops, risperidone 1 mg, clobazam 10 mg, levetiracetam 100 mg/ml, pericyazine 1 mg/drop and biotin 10 mg/ml, all via gastrostomy tube. This case portrays the consequences of common underlying genetic predispositions and deleterious events in the prenatal and neonatal periods in a patient with cerebral palsy, which is already a condition associated with an increased risk for the occurrence of other medical comorbidities that interfere with the quality of life of the patient and caregiving family.

Neurologia Infantil

TL 1105899

AUTISM SPECTRUM DISORDER (ASD) ASSOCIATED WITH HIPPOCAMPAL SCLEROSIS AND MESIAL TEMPORAL LOBE EPILEPSY

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Case Presentation: Female, 4 years old, had her first consultation 1 year ago due to epileptic seizures and difficulties in cognitive development. The characteristic episodes of focal seizures started at 9 months of age, the last one occurring when the patient was 2 years old. Anamnesis: antisocial behavior, echolalia, and stereotypes. She had previously used depakene (suspended due to jaundice observed in the physical exam) and valproate. Valproate was replaced by carbamazepine, and clonazepam was introduced to avoid possible withdrawal crises. In the return visit, in regular use of the medications, the absence crisis did not reoccur, but the cognitive and behavioral symptoms, compatible with the autistic spectrum, still persistent. Magnetic resonance imaging showed right hippocampal hypotrophy with hypersignal on T2, suggestive of medial hippocampal sclerosis. Electroencephalogram (EEG) showed slow-wave spike complexes of 5 to 6 Hz in central and parietal regions intermittently with slowing of regional rhythms and negative spikes in right temporal (T4) and left central (C3). There is also excessive slow and fast activity without Alpha-Theta predominance, typical of the patient's age range. The patient is on a combined follow-up with speech therapy, psychopedagogy and occupational therapy. **Discussion:** Medial temporal lobe epilepsy is commonly manifested by focal seizures, with EEG manifestations as spike-like epileptiform activity and sharp wave, which may be followed by slow wave, with maximum electronegativity in leads T3-F7/T4-F8, especially in sphenoidal leads, having already been reported by Andrade-Valença (2006) in concomitance with hippocampal sclerosis. However, the association of these pathologies with the ASD is an expensive record in the literature. In the meantime, besides the patient in question having clinical imaging and exams proving the three conditions, the diagnosis of these conditions was made early. Thus, therapy with carbamazepine and clonazepam was efficient in controlling the epileptic seizures. As for the behavioral changes, it is expected that with advancing age and multiprofessional intervention they will evolve with improvement. **Final Comments:** The case is unique because of the diversity of symptoms stemming from the combination of two distinct diagnoses that complement each other in the patient's clinical picture. The therapy used in the case, along with the multiprofessional follow-up has had good results.

Neurologia Infantil

TL 1105937

SÍNDROME DE RETT: RELATO DE CASO SOBRE AS COMPLICAÇÕES DE UMA DOENÇA NEURODEGENERATIVA RARA

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Case Presentation: EDMA is a 4 years and 11 months old female, born by Cesarean section. Her mother presented with asthma and fibromyalgia while pregnant. When EDMA was 6 months old, she started presenting myoclonic-atonic seizures. At 16 months, she was diagnosed with Autism Spectrum Disorder (ASD). At 17 months, EDMA began to walk, but, after 3 months, she presented with ataxic gait. At 23 months, a molecular study for Rett Syndrome (RS) was realized, detecting a variance of uncertain meaning of heterozygosity on the gene MECP2. When she turned 2 years old, she presented Neuropsychomotor developmental delay (NDD), manual stereotypies, global hypotonia, aggressiveness and intractable epilepsy. An electroencephalography evidenced suggestive findings of diffuse encephalopathy. 2 months later, a MRI exhibited discrete encephalic sulci and fissures global emphasis. At 3 years and 11 months, EDMA underwent a gastrectomy and, at 4 years and 1 month, she had an episode of bronchoaspiration during a seizure, which mostly occur during her sleep, with an interval of 2 days between episodes. **Discussion:** this case describes complications related to RS, a rare neurodegenerative disease caused by a mutation on the X chromosome gene MECP2. This condition affects neurodevelopment and the patient usually has a typical development until 6 months, when symptoms, such as NDD, smaller brain growth, sleep disorders, global hypotonia, irritability, intractable epilepsy, gait dysfunction, disfagia and gastroesophageal reflux. Furthermore, this progressive encephalopathy, by presenting absence of protein 2 (MeCP2) methyl-cytokine ligand, generates a discrete encephalic atrophy, since MeCP2 is important for its adequate development. In addition, the presence of manual stereotypies, similar to handwashing, and lower social and environmental interest may lead to the equivocate diagnosis of ASD at early stages of the disease. A gastrectomy is also an important strategy for preventing bronchoaspiration caused pneumonias, some of the most prevalent causes of death in SR. **Final Comments:** in face of RS complications, more studies on this encephalopathy are necessary, since the correct diagnosis and multiprofessional intervention can contribute to the improvement of social abilities and to the attempt of preserving motor skills.

Neurologia Infantil

TL 1106148

BILATERAL OPEN LIP SCHIZENCEPHALY, A CORTICAL MALFORMATION: A CASE PRESENTATIONAna Beatriz Vieira Pacheco, Isabela Pires Porto
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Case Presentation: A 3-year-old female patient was diagnosed with bilateral open-lip schizencephaly at 4 months of age. The pregnancy was discovered at 6 months, with placental abruption and loss of amniotic fluid. The patient was delivered by cesarean section, without complications. At 20 days, she had the first seizure and the second at 2 months, when the investigation was started. At 4 months, the patient was diagnosed with open-lip Schizencephaly through tomography, with the presence of three clefts that communicate the ventricular system to the surface of the pia mater, showing clefts in the temporal region on the right, and in the frontal and temporal lobes on the left, with initiation of carbamazepine, but without resolution, being Hospitalized in a pediatric ICU with ventilatory support due to repeated crises. After changing the medication, the condition stabilized for a period, with the use of phenytoin, valproic acid and phenobarbital. In this year, 2022, there was a return of seizures, associated with a drop in oxygen saturation, carbamazepine was prescribed again and neuleptil was started, with the medications already prescribed. On physical examination, the patient has a delay in neuropsychomotor development, does not hold her head, does not sit without support, does not walk and does not speak, but has preserved speech understanding. Currently, the condition has improved with the medications in use and he is undergoing continuous medical follow-up. **Discussion:** Schizencephaly is a rare malformation of the cerebral cortex (1.48/100,000 births) that can be open or closed, bilateral or not. The etiology is still not well understood, as well as the period of embryonic development that is involved in the development of the malformation. The clinical manifestations of the disease are associated with the degree to which the brain area was affected, ranging from individuals who lead a normal life without manifestations, to severe cases of neurological impairment, with epilepsy being the most common manifestation. **Final Comments:** This study brings the symptomatic management of a cortical malformation, showing the importance of continued monitoring so that there is an improvement in the quality of life of the child and parents, reducing the damage caused by clinical manifestations and allowing the child to have a better life, as close to normal as possible.

Neurologia Infantil

TL 1106308

PAEDIATRIC-ONSET OF HTLV-1 ASSOCIATED TROPICAL SPASTIC PARAPARESIS, DUE TO VERTICAL TRANSMISSION – A CASE PRESENTATIONSarah Diógenes Alencar, Paula Caracas Barreto,
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Case Presentation: M.L.F.O, 13 years old, female, previously diagnosed with neurogenic bladder, started at age 10 with an insidious condition of mechanical arthralgia in the right knee, associated with difficulty in walking and non-progressive or ascending asymmetrical lower limb paresis. Due to a family history of dermatopolymyositis in a first-degree relative, she was admitted to the Hospital for investigation of inflammatory myopathies in a pediatric rheumatoid department. On examination, hyperreflexia was identified in the lower limbs with an increased reflex area on the left, inexhaustible clonus on the left, grade IV strength distally in the left lower limb and paretic gait on the left, with no other clinical findings. Initial tests showed CPK 211, TGO 24, TGP 17, DLH 249, non-reactive Anti-Jo1. During investigation of infectious causes, the presence of antibodies to HTLV-1 was detected by Western Blot, with the presence of GD21, p19, p24 and rgp 46-I bands. Electroneuromyography of the lower limbs and resonance of the right thigh did not show any changes, however spinal resonance showed apparent thinning of the spinal cord of the dorsal segment. The exclusion of acute infectious, metabolic, and demyelinating pathologies and the positivity of the serology for the HTLV-1 virus led to the diagnosis of tropical spastic paraparesis, pyramidal motor sequelae caused by HTLV infection, resulting from vertical transmission, whether via breastfeeding, transplacental or birth canal, considering maternal serology also positive for this virus. **Discussion:** The human T cell lymphotropic virus type I (HTLV-1) was the first retrovirus isolated in humans, in a patient with cutaneous lymphoma. Currently, its prevalence in Brazil is 0.47%, and its manifestation as early-onset myelopathy has been described in 27 cases in the country. According to Bittencourt et al, in a study carried out in Salvador, the infection is acquired mainly through breastfeeding, being the main form of vertical transmission, and then through sexual intercourse. **Final Comments:** Spastic paraparesis is a severe and disabling myelopathy, with a chronic and progressive presentation, involving the pyramidal tract, with proximal leg weakness, low back pain and neurogenic bladder. When it begins in childhood, it is manifested by pyramidal signs followed by gait disturbances and sensory symptoms. The evolution is usually rapidly progressive, making early diagnosis essential.

Neurologia Infantil

TL 1106484

WEST SYNDROME ASSOCIATED WITH AUTOSOMAL DOMINANT INTELLECTUAL DEVELOPMENT DISORDER 13: CASE PRESENTATIONLuan Monte Barroso, Marina Mayara Pereira,
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Case Presentation: Infant, 8 months and 13 days old, at 5 months of age, he started to have partial myoclonic seizures that presented with the upper limbs in repetitive extension movements. Initially, the seizures lasted for seconds and had a frequency of 2 to 3 episodes per day. An electroencephalogram (EEG) was performed, which showed a pattern of hypsarrhythmia, and West Syndrome (WS) was diagnosed due to the EEG pattern, spasms and developmental delay. His first Hospitalization was at 7 months, when he had a crisis that lasted more than 5 minutes. During Hospitalization, he performed several complementary exams to investigate the etiology of the condition, EEG, magnetic resonance and exome. This confirmed Autosomal Dominant Intellectual Development Disorder 13, which presents with neuropsychomotor developmental delay, epilepsy, cortical changes in CNS migration and hypotonia. She currently performs therapies with valproic acid, gabapentin, levetiracetam, nitrazepam and ACTH for the treatment of epilepsy and WS. **Discussion:** The WS is the most common encephalopathy in the first two years of life, consisting of: epileptic spasms, hypsarrhythmia on encephalography, and delay or arrest in neuropsychomotor development. In this aspect, the diagnosis of WS is made by combining clinical characteristics and electroencephalographic (EEG) findings. The most prevalent characteristic EEG finding is a hypsarrhythmic pattern. In addition to the EEG, imaging tests and genetic tests are necessary for the etiology of the syndrome. The patient's exome report indicates pathogenic variants in the DYNC1H1 gene, which would correlate with the presented phenotype of Autosomal Dominant Intellectual Developmental Disorder 13, clinically characterized by intellectual disability, behavioral changes, epilepsy, central nervous system abnormalities and variable dysmorphisms. **Final Comments:** The WS constitutes 2% of all childhood epilepsies, being, therefore, a rare syndrome and despite the clinical signs, it is essential to perform imaging tests for diagnosis. It is a pathology with a poor prognosis, as it has no cure, evolving largely into difficult-to-control epilepsy associated with delayed neuropsychomotor development. Therefore, early diagnosis is necessary to start pharmacological treatment and strict follow-up with a multidisciplinary team, including speech therapy, physical therapy and neurologist to control the crises.

Neurologia Infantil

TL 1106605

ATYPICAL OCULO-CEREBRO-RENAL PRESENTATION – A REPORT CASE OF LOWE'S SYNDROME

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Case Presentation: JJS, male, 10 months old. Born by vaginal delivery, APGAR 9/9, presented significant hypoglycemia and respiratory distress in the ward, and was referred to the ICU, where he stayed Hospitalized for 15 days. After ICU discharge, he was admitted to the maternity ward due to frequent regurgitations. During Hospitalization, regression of the sucking reflex and global hypotonia were observed, and he was referred to a neuropsychiatrician for evaluation for SMA investigation, with negative molecular results for neuromuscular diseases. During the multi-professional evaluation, congenital bilateral cataract and renal tubular acidosis were diagnosed; serologies came back negative. Thus, Lowe's Syndrome (oculo-cerebro-renal syndrome) was investigated and diagnosed. Patient evolved with neuropsychomotor developmental delay (NPDM), visceromegaly, dysphagia, requiring gastrostomy. Neurological examination: pupils punctiform and leukocoria, nystagmus, global hypotonia and preserved cutaneous extension. **Discussion:** Lowe's syndrome has an incidence of 1:500,000 live births, being a disease clinically characterized by congenital cataract, renal tubular acidosis and changes in the central nervous system, such as neuromuscular symptoms, intellectual disability, epilepsy and neurodevelopmental delay. Lowe's is a disease with an X-linked inheritance pattern, and it's associated with mutations in the OCRL gene, a gene involved in multiple intracellular processes involving endocytic trafficking and actin's skeleton dynamics. Lowe's Syndrome can be attributed to a de novo variant in about 70% of the cases. So far, there is no treatment for this genetic condition, but symptomatic treatment such as cataract surgery and physical therapy can be performed to preserve existing motor function. **Final Comments:** Lowe's Syndrome is a rare condition, predominantly affecting boys. The present case is compatible for the condition, with expected clinical history and presentation. It is suggested that further investigation with genetic testing be performed and follow-up of this child with therapies and interventions aimed at a better prognosis.

Neurologia Infantil

TL 1106630

MACROCRANIA, PHENOTYPIC ALTERATIONS AND INTELLECTUAL DEFICIT: A CASE PRESENTATION OF SOTOS SYNDROME

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Case Presentation: AOD, male, 9yo. Genitor reports asymmetric skull growth of the infant at 3 months of age. **Background:** TPAL 1-0-0-1, HC: 37cm at birth, delayed NPMD (first words at 1y2m), no family history of neurological disease. He reports a moderate to strong and pulsatile holocranial headache that worsens with physical activity and noise. Neurological examination: turriccephaly, HC: 62cm, symmetrical typical facies with micrognathia, hypertelorism, distant teeth, atypical gait, speech without impairment, sensitivity and tone preserved, MF 5/5 globally, osteotendinous reflexes 2/4 globally, symmetrical, without pyramidal signs, no changes in the cranial nerves. Segmental examination: syndactyly between the 2nd and 3rd left podotactiles, increase of the 2nd right podotactile, presence of multiple surgical scars from lipoma's removal and multiple verrucous lesions in the cervical and inguinal region. The suspicion of hydrocephalus was ruled out after skull MRI, which showed macrocrania, multiple enlargements of the perivascular spaces in the subcortical regions, especially in the frontoparietal region, cystic lesions in the bilateral fronto-temporo-parietal and periventricular left white matter. Clinical presentation compatible with Sotos Syndrome. **Discussion:** Sotos Syndrome is clinically characterized by macrocephaly, phenotypic changes, intellectual deficit, autistic spectrum disorder, hyperextensibility of joints, scoliosis and epilepsy. It is a genetic disease marked by a pathogenic variant in the heterozygosity in the NSD1 gene, and most mutations are de novo, as we could see in the presented case, in which there is no family history of similar neurological conditions. The diagnosis is made by identifying the mutation in a genetic panel. So far, there is no treatment for the genetic condition, but symptomatic treatment can be done, such as the performance of adjunct therapies like speech and physical therapy. The literature has not yet demonstrated an association with migraine or the presence of lipomas. **Final Comments:** The present case is highly suggestive for the condition, with compatible clinical history and presentation. Further investigation with genetic testing is suggested for full diagnoses confirmation. Besides that, follow-up of this child with therapies and interventions for a better prognosis are suggested.

Neurologia Infantil

TL 1105323

ACUTE CEREBRAL THROMBOSIS CAUSED BY FLOW DIVERSION STENT AND RESISTANCE TO TICAGRELOR – CASE PRESENTATION

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Case Presentation: A 52-year-old female presented to the interventional neurology team for treatment of saccular aneurysm located in the ophthalmic segment of the left internal carotid artery. As comorbidities she reported having Systemic Arterial Hypertension (SAH). She reported using losartan 50mg and two weeks before the procedure the neurologist prescribed prasugrel and acetylsalicylic acid 100mg. Cerebral angiography was requested for treatment of the aneurysm. In the cerebral angiography a flow diversion stent was positioned in the left middle cerebral artery. The final control showed incomplete opacification of the internal carotid due to platelet aggregation, denoting subsequent stenosis. Balloon angioplasty was performed, but was unsuccessful. It was necessary to use a hyperglide balloon, showing improvement of the left internal carotid artery, but presenting occlusion of the upper trunk of the left M1 associated with the avascular area. A Trevo stent progression was performed in the same catheter, released in the m3-m2 segment of the left middle cerebral, and thrombus was removed. Being requested Computed Tomography of the skull for follow-up. **Discussion:** Taking into consideration that the patient in question takes the platelet antiaggregant ticagrelor 90 mg, it is necessary to point out some information about it. Ticagrelor is a platelet antiaggregant medication that acts differently from clopidogrel and prasugrel. The 3 drugs have the same mechanism of action, which is to inhibit the platelet receptor P2Y₁₂, but while the thienopyridines act irreversibly, ticagrelor has a reversible effect (Li et al., 2022). Based on the case of the patient in question and according to a study published by the American Journal of Neuroradiology, several factors can influence intracranial aneurysm thrombosis, such as: flow-diverter stenting, which causes reduced blood flow. However, due to the lack of detailed measurements of intra-aneurysmal flow, its real impact remains uncertain (BRINA, 2019). **Final Comments:** The reported case and exposed publications bring the discussion succession of infrequent events. The patient used prasugrel and acetylsalicylic acid within two weeks prior to the angiography and at the time of the procedure was administered ticagrelor. Conduct after placement of a flow diversion stent in the left middle cerebral artery caused cerebral thrombosis, which was resolved with thrombectomy and the use of tirofiban hydrochloride.

Neurologia Intervencionista

TL 1106072

CEREBRAL ISCHEMIA DUE TO SPONTANEOUS DISSECTING ANEURYSM OF THE ANTERIOR CEREBRAL ARTERY TREATED WITH A FLOW DIVERTER STENT SILK VISTA BABY

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Case Presentation: A 68-year-old man admitted with sudden onset of weakness in his right leg, with no other neurological symptoms. He did not report headache. There were no illnesses in his medical history. Head computed tomography did not show ischemia or hemorrhage, while CT angiography revealed aneurysmal dilatation of left anterior cerebral artery (ACA), partially thrombosed. Subsequent cranial magnetic resonance imaging showed acute ischemia in the left ACA territory. Digital subtraction angiography confirmed aneurysmal dilatation in the A1-A2 transition of the left ACA measuring 10 x 7 x 6 mm, with a wide implantation base and the presence of a small stenosis in the post-aneurysmal segment, suggesting a dissecting etiology. Initially, he was treated with dual antiplatelet therapy. After 14 days of ictus, he underwent endovascular treatment with the flow diverter stent (FDS) Silk Vista Baby – SVB (Balt, Montmorency, France), without complications. Patient was discharged 2 days later with minimal right leg weakness, modified Rankin Scale 2. **Discussion:** Spontaneous dissections of intracranial arteries are rare and there are few reports of spontaneous dissections of ACA, whose optimal treatment remains uncertain. Endovascular treatment of ACA dissection has been reported as a feasible alternative, especially in patients in whom subarachnoid hemorrhage is the main clinical presentation. However, in the reported cases of ACA dissection with ischemia alone, conservative treatment showed good outcome in most cases. FDS were initially designed and constructed for the treatment of aneurysms that could not be easily treated using more conventional endovascular approaches. The SVB is the first FDS that can be delivered via a 0.017 inch microcatheter and is designed to target aneurysms of the distal vasculature for which existing FDS are not optimized. By implanting the densely woven mesh in the parent vessel, the aneurysm neck is covered and blood flow is directed away from the aneurysm orifice. Despite the lack of studies, SVB has achieved sufficient occlusion rates of intracranial aneurysms originating from peripheral segments, which are comparable to conventional FDS with acceptably low complication rates. **Final Comments:** Dissecting aneurysm is a rare cause of cerebral ischemia in the ACA territory and patients tend to have a good clinical outcome, as in the case presented. The SVB is a new promising device as a possible treatment.

Neurologia Intervencionista

TL 1105130

EARLY DIAGNOSIS OF HEREDITARY AMYLOIDOSIS

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Case Presentation: Asymptomatic individuals carrying the hereditary amyloidosis gene are screened and submitted to annual assessments in our service and it include a neurological examination, a questionnaire on symptoms of dysautonomia, complementary tests that include electroneuromyography and screening for dysautonomia, such as Quantitative Sensory Test (QST), heart rate variability (HRV) and reflex sympathetic response (RSR). We identified abnormal findings in two young women carriers of the hereditary amyloidosis transthyretin. A 32-year-old female carrier of Phe84Leu gene presented symptoms of dysautonomia (blurred vision, dry eyes, increased sweating in her hands, orthostatic intolerance and constipation). The electroneuromyography showed mild carpal tunnel syndrome bilateral. The reflex-sympathetic response was abolished in the four limbs. She was then submitted to a salivary gland biopsy which revealed deposit of amyloid material. Her father was the index case and at the age of 64 he started a condition of polyneuropathy and dysautonomia, receiving a late diagnosis and treatment was not possible, dying from disease complications. The second case was a 22-year-old female carrier of Val50Met gene. She had reflex sympathetic response abolished in the 4 segments and the quantitative sensory test (QST), revealed warm detection and warm pain thresholds were higher and the cold detection and cold pain thresholds were smaller when compared to a sex and age matched healthy control. She also underwent salivary gland biopsy, which also confirmed the deposition of amyloid material in the tissue. Her aunt was the index case and presented polyneuropathy and dysautonomia at the age of 38, and treatment was not possible because it was in an advanced stage. **Discussion:** Hereditary amyloidosis transthyretin is a rare, autosomal dominant systemic disease caused by extracellular deposition of insoluble amyloid fibrils formed by the mutated transthyretin protein. The clinical presentation and the age of onset varies. The disease is often rapidly progressive, so an early diagnosis is important for an effectively treatment. **Final Comments:** Systematic and regular monitoring of an asymptomatic carrier is necessary to detect early signs of hereditary amyloidosis transthyretin.

Neuropatias Periféricas

TL 1105182

PERIPHERAL POLYNEUROPATHY AFTER COVID-19 INFECTION: A CASE PRESENTATION

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Case Presentation: Patient, 42 years old, male, no comorbidities, attended at an outpatient neurosurgery service complaining of burning pain in the plantar region of both feet, a condition that started after 3 months of mild infection of the airways by COVID-19. On physical examination, bilateral symmetrical paraparesis was identified, and a grade 3 strength rating for plantar flexion and dorsiflexion and for hip flexion-extension. Was also identified change in orthostatism and in the stimulation of ambulation, with balance deficit due to alteration in proprioception. Paresthesia in both heels and distal phalanx of the hallux in both feet. Also reported intense pain in both lower limbs, graded in 10 on the visual analogue pain scale. The current clinical condition directly affects their work and daily life activities. Was reported previous use of opioids to control symptoms, being refractory to treatment and without improvement. With the progressive worsening of the clinical condition and considering that the patient has a significant kinetic-functional deficit, treatment with antidepressants and anticonvulsants at optimized doses and spinal electrode treatment was indicated for neuropathic pain. **Discussion:** The post-COVID-19 syndrome brought with it peripheral polyneuropathy. One of the main symptoms reported by patients who suffer from this syndrome is pain, which can be associated with several other systemic disorders. The wide range of neurological manifestations resulting from the immune response to the virus has led to studies on the pathophysiology of this it. The main mechanism associated with the onset of this varied condition is associated with the invasion of the blood-brain barrier by the virus, in addition to the systemic pro-inflammatory action due to the release of numerous cytokines. Peripheral polyneuropathies in patients post-COVID-19 are frequent and associated not only with toxicity resulting from immunological aspects, but also due to the neurotoxic action of the main drugs used to combat the disease. It was noted that COVID-19 does not have a direct influence on the peripheral nerve branches, and no infectious neuropathy caused by this virus were established. **Final Comments:** In view of the cases already reported in the literature and the frequency of neurological complaints after COVID-19 infection, there is a relevance of these studies for future clarifications on the subject and treatment measures for it.

Neuropatias Periféricas

TL 1105189

NEUROPATIA RADIAL SECUNDÁRIA À PRÁTICA DE CROSSFIT

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Case Presentation: Patient GDL, male, 29 years old, previously healthy, reports that after a rope climbing CrossFit activity on 12/01/21, presented with left humerus fracture, resulting in wrist fall (Strength 0/5) and elbow extension deficit (Strength 1/5). Submitted to surgery for fracture correction with application of locking plate on 03/12/21, without complications. After the procedure, the patient persisted with clinical presentation. An electroneuromyography done in 02/01/22 shows accentuated fibers impairment of radial nerve, probably located above the spiral sulcus, with present denervation and no signs of reinnervation. Patient is currently being followed up with physical therapy with a strength grade of 1/5 for left wrist extension and 2/5 for ipsilateral elbow extension.

Discussion: Crossfit is a program developed to promote the improvement of physical conditioning with exercise variability and high intensity¹. It has gained a lot of popularity and several studies have emerged with the purpose of highlighting the physical benefits and answering questions about safety in its practice, mainly related to the higher prevalence of injuries.² Feito et al evaluated groups of Crossfit practitioners for 04 years and showed injuries to the shoulders (39%), back (36%), knees (15%), elbows (12%) and wrists (11%) were the most common³. Costa et al revealed that among injuries associated with Crossfit, mild/moderate ones represented 89.1% of the cases, while severe/very severe injuries 10.9% and injuries that required surgical treatment represented 2.6%. Humeral stress fractures have been described in activities with excessive use of the upper extremities and weight-lifting⁴, aspects present in Crossfit practices. Since there is a close anatomical relationship between the radial nerve and the humerus and the fact that it promotes motor innervation of the triceps and wrist, with wrist drop being the hallmark of a radial nerve injury⁵, the context and consequences of the patient's injury significantly reflect the findings found in literature. **Final Comments:** Although Crossfit's rising popularity demonstrates an interest in physical exercise, its practice without guidance can result in serious harm to the body. The episode described brought consequences that affect the patient's life as a whole due to the current coexistence with a neuromotor dysfunction of indefinite duration, shining a light on the importance of discussions about the safety of the practice.

Neuropatias Periféricas

TL 1105209

POEMS SYNDROME, ALTHOUGH RARE, PRESENTS A CONSTELLATION OF CLINICAL FINDINGS THAT ALLOWS ITS EARLY DIAGNOSIS – A CASE PRESENTATION

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Case Presentation: A 55-year-old male patient presented acrocyanosis with intense pain in the hands and paresthesia in boot-glove pattern associated with a significant weight loss, asthenia and hyporexia, in 3 years of evolution. On clinical examination, in addition to acrocyanosis, he had telangiectasias on the upper body and face, bilateral gynecomastia, palpable hepatomegaly, global hyporeflexia, 4+ muscle strength in the extremities, reduced pain sensitivity in the lower limbs, reduced vibration in the upper and lower limbs. Complementary exams showed a polycythemia and monoclonal peak between beta 2 and gamma globulin fractions, a chronic inflammatory demyelinating pattern polyneuropathy and osteosclerotic and lytic areas in the body of the sternum and osteosclerotic lesions in the skull. A bone marrow aspirate was performed, which revealed 9% of plasma cells and in the sternal biopsy bone marrow hypocellularity with discrete polyclonal reactive plasmacytosis. The set of such findings allowed the final diagnosis of POEMS Syndrome, an Osteosclerotic Multiple Myeloma. Treatment was based on cycles of dexamethasone, disodium pamidronate, amitriptyline and gabapentin for neuropathic pain management, followed by a VTD protocol (Bortezomib+Doxorubicin+Thalidomide). **Discussion:** This report aims to expose a rare case, with an estimated prevalence of 0.3 per 100,000 people, which had an evolution of more than 3 years, and with a constellation of clinical presentations that may have made early diagnosis difficult. POEMS Syndrome should be analyzed as a differential diagnosis of several diseases, such as polyneuropathies, osteosclerotic lesions, plasma cell dyscrasias, neoplasms, infections and vasculitis. **Final Comments:** The case in question portrays that the identification of the disease can be a challenge, but a good anamnesis and physical examination followed by appropriate tests can distinguish this Syndrome from its differential diagnoses.

Neuropatias Periféricas

TL 1105223

COVID-19 INDUCED NEUROPATHY

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Case Presentation: Male patient, 35 years old, hypertensive and obese, was admitted in ICU by COVID-19 infection, spending 14 days intubated and 7 days in the ward. While in the Hospital, the patient started to feel lower limb weakness, paresthesia and neuropathic pain, which until the day he was discharged the symptoms were assigned to be related by both collateral effects of medications administered and the time he spent at the ICU itself. Already at home the lower limb weakness got worse, especially in the proximal musculature of the right leg (iliopsoas and quadriceps). Thus, was solicited an electroneuromyography that showed serious injury and prominent denervation of both right femoral and left sciatic nerve, in addition to sensory axonal polyneuropathy in the lower limbs, post COVID-19. Therefore treatment was started with pulse therapy of 1g methylprednisolone for 5 days and pregabalin with progression up to 150mg 12/12h. Upon return, the patient reports improvement in neuropathic pain and paresthesia, but maintains a motor deficit in the lower limbs. The electroneuromyography was repeated, which showed signs of femoral and sciatic reinnervation, but still with severe injury in these nerves. **Discussion:** COVID-19 is a disease with tropism for the respiratory system, but as a reflection of serious infections by SARS-COV-2, patients can develop neurological deficits, such as motor and sensory ones. Among the main pathophysiological mechanisms hypotheses are: ischemic phenomena; development of autoimmunity; and the neuropathy of the critically ill patient. Thus, the development of neuropathy as a symptom of COVID-19 is reinforced by the temporal relationship with the onset of the disease and by the absence of other factors that could predispose to this condition, such as diabetes mellitus or Guillain-Barré syndrome. **Final Comments:** A rare case of neurological manifestation of COVID-19 was presented here, which differs from the pattern of neuropathies caused by other viruses, which usually have a distal polyneuropathy pattern with sensory predominance. Thus, it is important for health care professionals to be alert for manifestations similar to those described in this case, as well as other possible neurological manifestations caused by SARS-COV-2. Finally, treatment with pulse therapy of methylprednisolone, pregabalin, and physical therapy showed some degree of efficacy in improving the condition.

Neuropatias Periféricas

TL 1105224

INTERMEDIATE CHARCOT-MARIE-TOOTH DISEASE TYPE C: A CASE PRESENTATION

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Case Presentation: A 31-year-old male with a history of progressive muscle weakness in four limbs for more than 20 years. He has 2 sisters and 1 maternal uncle with a similar condition. He denied dysphagia, respiratory complaints, or continuous use of medications. On examination, muscle strength was assessed in proximal and distal grade IV in all four limbs and deep tendon reflexes were hypoactive globally. Artresthesic and vibratory sensitivities were reduced in the lower limbs, while the superficial was reduced in the four limbs at the 10 Hz monofilament test. He had Gowers maneuver and gait with weakness of the gluteal muscles. There was no calf pseudohypertrophy, nervous hypertrophy or myotonia. Electroneuromyography (ENMG) revealed a moderate reduction in motor and sensory conduction velocities in the four limbs, with F waves abolished in ulnar and tibial nerves. In electromyography, voluntary contraction showed signs of chronic reinnervation with moderate rarefaction of motor units in the proximal muscles of upper limbs, and marked rarefaction in distal muscles of upper limbs and in proximal and distal muscles of lower limbs, describing a primarily demyelinating, sensory-motor, symmetrical, chronic polyneuropathy, with proximal and distal involvement, worse in the lower limbs. Vitamin B12, folic acid, infectious disease serology, metabolic panel, aldolase, ANA and anti-acetylcholine tests were normal. CPK was increased (990U/L). Genetic testing confirmed the diagnosis of intermediate type C Charcot-Marie-Tooth Disease (CMT) by mutation in the PLEKHG5 gene. **Discussion:** CMT is a group of genetic diseases that present as chronic and progressive motor and sensory neuropathy. The CMT1 and CMT2 subtypes are the most frequent and represent the demyelinating and axonal forms, respectively. The most common clinical presentation is distal weakness, atrophy, and sensory loss in a boot-and-glove pattern. In the present case, the patient has proximal weakness and intermediate motor nerve conduction velocity. The genetic test allowed the diagnosis of the intermediate type C form, an uncommon subtype characterized by mixed, axonal and demyelinating involvement. **Final Comments:** CMT is described as the most common hereditary neuromuscular disease, and the intermediate type C form, a rare variant of the disease, was identified in this report. In many cases, the diagnosis still focuses on ENMG, but genetic testing is critical for confirmation.

Neuropatias Periféricas

TL 1105255

ASSOCIATION OF TWO DISEASES AND THEIR CLINICAL MANIFESTATIONS: CHARCOT-MARIE-TOOTH AND HEREDITARY TRANSTHYRETIN AMYLOIDOSIS

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Case Presentation: A 41-year-old man presented a history of difficulty walking since he was 5 years old, with stumbling and frequent falls. The condition remained stable until 5 years ago, when he noticed a progressive worsening of gait, with muscle weakness and atrophy in the upper and lower limbs. On neurological examination, he had decreased thermal and pain sensitivity in the lower limbs and hands, in addition to decreased vibratory sensitivity in the lower limbs. He had grade 3 hand strength in the lower limbs (MRC scale) and generalized areflexia. In addition, the patient had signs of dysautonomia, such as erectile dysfunction, orthostatic intolerance, and palpitations. Electroneuromyography revealed severe sensorimotor polyneuropathy. Genetic testing revealed two genetic mutations, PMP22 causing Charcot-Marie-Tooth disease and Val142Ile, a gene responsible for hereditary transthyretin amyloidosis. **Discussion:** Charcot-Marie-Tooth disease is a group of hereditary diseases that manifests itself in adolescence or early adulthood, with loss of sensitivity, weakness, deformity and muscle atrophy mainly in the lower limbs, with dysautonomic manifestations not being common. Hereditary amyloidosis courses with progressive polyneuropathy, often associated with dysautonomia, also causing multisystem symptoms such as gastrointestinal impairment, cardiomyopathy, nephropathy, or ocular deposition. The genetic test provided the opportunity to diagnose two genes that cause peripheral neuropathy, with hereditary amyloidosis being a potentially treatable disease depending on the stage. As the patient was still able to walk with bilateral support, patirisan was indicated and he is awaiting treatment. **Final Comments:** it is important to be aware of atypical signs of a disease looking for other diagnostic alternatives or even concomitant diseases as was this case.

Neuropatias Periféricas

TL 1105277

CHARCOT-MARIE-TOOTH TYPE 1A AND POEMS SYNDROME: WHEN TO INVESTIGATE ACQUIRED CAUSES IN HEREDITARY NEUROPATHIES

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Case Presentation: The proband was diagnosed at age 40 with CMT1A. Her symptoms were mild and slowly progressive, as expected for most CMT1A. At age of 56, her walk deteriorated fast, she started with difficulty to climb stairs and frequent falls, associated with positive sensory symptoms. Additionally, she lost about 17 kg in 8 months. On examination, first came to our attention the changes in her skin color. She also had hepatosplenomegaly, lymph adenomegaly, papilledema, and generalized edema. NCS revealed a marked worsening in the velocities. A monoclonal IgA lambda peak was detected. Lytic lesions were found, and bone marrow biopsy revealed 25% monoclonal plasma cells. Based on clinical and laboratory findings, a diagnosis of POEMS syndrome was made. Systemic chemotherapy followed by autologous bone marrow transplantation was performed. She evolved with dramatic improvement of her motor and sensory symptoms. In the setting of an inherited neuropathy of slow progression, a sudden clinical decline and a rapid progression, should raise attention for an associated condition. In the case we present, the systemic manifestations and the laboratory investigations permitted a fast diagnosis and treatment of POEMS syndrome. Significant changes in the natural history of an inherited neuropathy should be considered as a red flag for an associated condition. CMT1A is the commonest type of hereditary neuropathy, presenting as a slowly progressive length-dependent sensory and motor neuropathy, areflexia, pes cavus and uniform reduction of NCV. POEMS is a paraneoplastic disorder of plasma cells frequently associated to a sensory and motor demyelinating neuropathy. Distinction of both diseases is important, as early treatment significantly changes POEMS prognosis.

Neuropatias Periféricas

TL 1105296

GASTROINTESTINAL DYSMOTILITY AS THE LEADING COMPLAINTS IN SENSORY NEURONOPATHY PATIENTS

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Case Presentation: A – A 43-year-old male patient reported a 2-month history of paresthesia on calves. Examination demonstrated asymmetric loss of vibration and position sense (including pseudoathetosis), Romberg's sign, diffuse hyporeflexia, and tabetic gait. The patient developed constipation followed by acute abdominal pain. Intestinal pseudo-obstruction was diagnosed, and an exploratory laparotomy was performed but no mechanical obstruction was found. B – A 58-year-old woman reported pain and paresthesia in her right hand initiated 8 years before, with gait imbalance. Neurological evaluation showed diffuse loss of position and vibration sense, Romberg's sign, and global areflexia. She also reported urinary incontinence and severe constipation with onset in the previous 4 years. HTLV-1 serology was positive. C – A 53-year-old female patient was admitted for investigation of dysphagia associated with post-meal vomiting and paresthesia in the upper limbs for the past 6 months. Neurological examination showed pseudoathetosis and sensory ataxia in the upper limbs, tactile and painful hypoesthesia in the four limbs, diffuse hypoparesis, and diffuse hyporeflexia. A diagnosis of achalasia was made and was surgically corrected. NCS and EMG of all three patients demonstrated a severe reduction in SNAPs and preserved motor conduction, compatible with the SN diagnosis. **Discussion:** The frequency and pattern of dysautonomic features in SN are still poorly understood, despite being remarkably present in clinical practice. A study performed on patients with non-paraneoplastic SN demonstrated the involvement of various subdomains of the autonomic nervous system (ANS), with about half of the patients presenting orthostatic hypotension and three-quarters with cardiovascular autonomic neuropathy. From the 12 cases of intestinal pseudo-obstruction associated with SN reported in literature, only 3 were idiopathic. Autopsy studies of the patients revealed severe damage of myenteric plexus. The patients discussed here differ from the other reports since they are non-paraneoplastic forms of SN. Furthermore, the association between SN and achalasia is also quite rare in the literature, with few cases reported. **Final Comments:** Gastrointestinal dysautonomia leading to dysmotility is a substantial burden to patients with SN. Understanding the involvement of the ANS in SN is of special interest both for earlier diagnosis of atypical presentations and for improvement in quality of life.

Neuropatias Periféricas

TL 1105314

CANVAS: EXPANDING THE DIFFERENTIAL DIAGNOSIS OF SENSORY POLYNEUROPATHIES/NEURONOPATHIES: POSITIVE SENSORY MANIFESTATIONS AS THE MAIN COMPLAINT

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Case Presentation: A 38 year male patient started with severe paresthesia in the feet that interfered with sleep and daily activities. Over the years, there was a progressive worsening, with involvement of the lower limbs up to the knees, and more recently, after 6 years of evolution, of the hands and forearms. There is also a report of chronic cough. This is a previously healthy patient, that also complained of reflux and rhinitis and no other comorbidities. No family history of peripheral neuropathy. On neurological examination, the patient had preserved muscle strength, decreased osteotendinous reflexes globally, tactile, thermal and painful hypoesthesia up to the bilateral ankles and distal hypoparesis. Electroneuromyography revealed a length-dependent sensory neuropathy/neuronopathy. The patient was extensively investigated and, due to the severity of his symptoms, he repeatedly sought different health services. Autoimmune, infectious, metabolic diseases and research for amyloidosis, which were normal, were excluded. The exome showed no pathogenic variants. The CANVAS search revealed a bilateral AAGGG expansion at the RFC1 gene. The propaedeutics of vestibular tests and brain MRI showed no alterations. **Discussion:** The typical cerebellar ataxia, neuropathy and vestibular areflexia syndrome (CANVAS) is a progressive neurologic disease characterized by imbalance, sensory neuropathy and, occasionally, chronic cough and autonomic dysfunction. Its etiopathogenesis is associated with a biallelic (AAGGG) expansion in an intronic region of the RFC gene. Recently, the phenotypic spectrum associated with this mutation has been expanded and more limited phenotypes involving predominantly or exclusively one of the systems involved in balance control were described. Our patient complained essentially of a neuropathy and retrospectively we identified cough. As that pathogenic RFC1 AAGGG repeat expansions cannot be routinely detected by sequence-based multigene panels or exome sequencing, we need to be aware for this condition to order the appropriate genetic test. **Final Comments:** Sensory neuropathy/neuronopathy is a challenge in clinical practice, especially when the most common acquired diseases are ruled out. Our case shows that incomplete and phenotypes of CANVAS syndrome should be considered in this scenario.

Neuropatias Periféricas

TL 1105326

MULTIPLE MONONEURITIS SECONDARY TO VASCULITIS AS A DIFFERENTIAL DIAGNOSIS OF THE GUILLAIN-BARRÉ SYNDROME

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Case Presentation: A 46-year-old female patient with a history of burning pain and paresthesia on the left foot and progressing to the right foot. Subsequently, the worsening of sensory complaints, besides motor involvement, with ascending progression of symptoms and involvement of the upper limbs. In the physical exam, edema and ecchymosis on the lower limbs; hypoperfused podotactiles; predominantly distal flaccid tetraparesis, with an asymmetrical pattern; normoactive deep tendinopathic reflexes, except for abolished right Achilles; and aparesis on the right podotactiles, suggesting the hypothesis of Guillain-Barré Syndrome and was maid guided pulse therapy with immunoglobulin without clinical improvement. In the investigation, serologies were negative, laboratory evaluation of suspected vasculitis and connective tissue was reagent for rheumatoid factor, ANA, and lupus anticoagulant, polyclonal gammopathy detected by serum protein electrophoresis. CSF did not present alterations. Electroneuromyography disclosed severe asymmetrical sensory-motor axonal neuropathy and signs of denervation. Lumbar spine magnetic resonance didn't present alterations. Diagnosis of multiple mononeuritis secondary to systemic vasculitis was suspected, and pulse therapy with methylprednisolone was initiated, with an improvement of peripheral nervous symptoms. The patient evolved during Hospitalization with cardiac and central nervous system involvement, and died due to the severity of the disease. **Discussion:** The mononeuritis multiplex clinical presentation is linked to progressive motor and sensory deficits through the distribution of specific peripheral nerves, with irregular and asymmetrical characteristics. Ordinarily, it's caused by vasculitis, due to an inflammatory process that results in multifocal ischemia or peripheral nerve infarction. It presents a predilection for the extremities of lower limbs and has acute and subacute rhythms. The acute mononeuritis multiplex being similar to the Guillain-Barré Syndrome. The early diagnosis and proper treatment can prevent comorbidities and reduce mortality, namely to reduce the damage and consequently improve the quality of life. **Final Comments:** Guillain-Barré has a clinical presentation similar to acute-onset multiple mononeuritis. Thus, it is important to evaluate the differential diagnoses of the cases with acute evolution which can have high mortality if there is delay in diagnosis and treatment.

Neuropatias Periféricas

TL 1105348

WILSON'S DISEASE WITH PERIPHERAL NERVOUS SYSTEM INVOLVEMENT: ATYPICAL PRESENTATION OF A COPPER DEPOSIT DISEASE

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Case Presentation: Male, 21 years old, known to have Wilson's Disease for 1 year. Two months after admission, he started to lose strength in the lower limbs after starting treatment with D-penicillamine. On examination of the lower limbs, he presented proximal grade IV and distal grade II strength, in addition to areflexia. He also had hypopalesthesia and hypoesthesia distal. Examination of upper limb motricity, cranial nerves and coordination showed no changes. The slit lamp examination showed the presence of Kayser-Fleischer rings and the cerebrospinal fluid did not show protein-cytological dissociation. Electroneuromyography showed marked distal axonal polyneuropathy, predominantly motor, and a diagnostic hypothesis of a polyneuropathy of metabolic/toxic cause was raised due to the use of medication. The treatment instituted was the exchange of the copper chelator in addition to symptomatic medications for peripheral polyneuropathy, resulting in improved motor strength. **Discussion:** Wilson's disease is an autosomal recessive condition caused by a mutation in the ATP7B gene, responsible for copper metabolism. Its global prevalence is estimated at 1/10000 to 1/30000. About 70% of patients have neurological manifestations, which are mainly represented by movement disorders, such as dystonic, ataxic and parkinsonian syndromes, with peripheral nerve involvement being a rare event. The disease associated with polyneuropathy is generally characterized by distal and symmetrical sensory and/or motor involvement. Furthermore, drugs such as D-penicillamine may be associated with polyneuropathies, inferring a toxic cause for this condition in the context of Wilson's disease. The patient under study, despite the diagnosis of Wilson's disease 1 year ago, presented symptoms after the introduction of D-penicillamine therapy. It is not yet possible to establish a precise cause and effect relationship with the initiation of this drug, but this highlights the importance of new studies aimed at evaluating this condition. **Final Comments:** In the context of symptoms compatible with peripheral neuropathy in patients with Wilson's disease, it is important to consider storage disease itself as etiologies, as well as the use of copper chelating medications such as D-penicillamine.

Neuropatias Periféricas

TL 1105423

EXPANDING THE DIFFERENTIAL DIAGNOSIS OF POLYNEUROPATHIES: AN ATYPICAL MANIFESTATION AFTER GASTRIC BYPASS

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Case Presentation: Woman, 26 years old, started to experience several episodes of nausea and vomiting 15 days after performing a Roux-en-Y gastric bypass, due to grade III obesity. During this period, she did not ingest prescribed vitamin supplementation. After two months, she developed weakness of the lower limbs, with difficulty walking without assistance. She was admitted to the neurology service with symmetrical grade III strength in the lower limbs, in addition to burning-type paresthesias in the distal extremities of the four limbs. Complementary investigation of cerebrospinal fluid, serology and vitamin B12 were normal. Homocysteine levels were elevated. The neurophysiological study showed polyneuropathy with an accentuated sensory and moderate motor axonal pattern in the lower limbs. A presumptive diagnosis of deficiency polyneuropathy was performed and vitamin B complex and folic acid replacement was performed. The patient evolved with considerable improvement, walking without assistance after 2 months. **Discussion:** Bariatric procedures are an alternative to the treatment of morbid obesity, offering benefits in relation to weight reduction and management of associated comorbidities. However, they can contribute to the evolution with important neurological complications, due to the reduction of the absorption surface, the decrease in the production of intrinsic factor and gastric juice by the parietal cells, in addition to the possibility of dietary restriction associated with events of vomiting in the postoperative period. This leads to vitamin and mineral deficiencies, such as the commonly described vitamins B1, B6, B9, B12, A, D, E, folate and copper. Neurological complications may arise after 3 to 20 months of the procedure and involve nerves, spinal cord and brain, as in polyneuropathies, myelopathies and cognitive syndrome. Treatment consists of vitamin replacement, with a strong role in remyelination and brain glucose metabolism, which may contribute to the gradual improvement or clinical stabilization of the patient. **Final Comments:** Polyneuropathy is a possible neurological complication of bariatric procedures. The urgent approach with correction of nutritional disorders is mandatory to avoid the irreversibility of the condition.

Neuropatias Periféricas

TL 1105432

NEUROPATIA COMPRESSIVA DO NERVO RADIAL POR PRÓTESE DE SILICONE

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Case Presentation: MFV, 26 years old, female, past bariatric surgery on 01/2010, current plastic surgery for placement of silicone prostheses (breasts, gluteus, calves, arms: biceps and triceps) on 10/2010. The patient evolved with loss of motor strength in left wrist extension/left wrist drop (0/5 strength) with preservation of elbow extension. An MRI was performed, identifying the well-positioned prostheses, except for one in the left arm, which maintains close contact with the radial nerve between the triceps heads. Furthermore an electroneuromyography on 09/01/2010 which showed a severe, but partial axonal mononeuropathy of the left radial nerve in the arm (axonotmesis). She underwent surgery to remove the prostheses on 11/2010, in which the anatomical integrity of the nerve was identified with a weak response to intraoperative electrical stimulation. There was full functional recovery after 1 year of rehabilitation. **Discussion:** Silicone implants have been used in Medicine for over 60 years. Historically, interest in aesthetic procedures to alter body contour has increased greatly during the 1980s due to the popularization of body building as a sport and the fitness culture¹. Since the beginning of the century, due to the increasing accessibility of information about the procedures a new rising wave of demand for silicone implants for body contouring has been installed¹. In particular, prostheses for previously unexplored areas are gaining more and more space, as is the case of implants to increase the biceps and triceps. Despite the relative scarcity of data and publications², the most expected complications in upper limb implants include local infection, seroma, implant extrusion, asymmetry, compartment syndrome, muscle and nerve damage, with neuropathia being the most common immediate complication¹³. In agreement with the reports in the literature, the present case presented neuropathy as a result of the implant performed in the posterior region of the arm. The triceps is innervated by the radial nerve. Radial nerve damage can occur either from intraoperative damage during dissection of the muscle or from compression by positioning the prosthesis. **Final Comments:** Currently available data on complication rates in cosmetic procedures for upper limb implants are scarce and in small samples, ranging from 26.7%² to 47.6%³ – the latter encompassing only biceps implants. Further studies about complications of implants are necessary.

Neuropatias Periféricas

TL 1105475

MULTIPLE MONONEURITIS SECONDARY TO RHEUMATOID VASCULITIS

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Case Presentation: A 60-year-old male patient, diabetic, hypertensive and rheumatoid arthritis without treatment, started about 2 months ago with multiple cutaneous lesions on lower members without pruritic purpuric appearance with dissemination from upper limbs and face. Beside that, the patient presents pain and paresthesia on the plantar region. In the physical exam it was observed: ulnar deviation of the fingers on symmetrical hands with bone prominences on metacarpophalangeal joints. Neurological examination: oriented, tactile and painful hypoaesthesia on the left lower limb, strength grade 2 on the left lower limb and grade 3 on the right lower limb and upper limbs with global hyporeflexia. The serologies for HIV, hepatitis B and C were negative with evidence of thrombocytosis, reactive rheumatoid factor (1280 UI), no-reactive FAN, decreased C4 dosage and normal C3 dosage. The electroneuromyography showed a peripheral sensory-motor involvement, of axonal nature, asymmetrical involvement, subacute, acute grade, compatible with multiple mononeuritis. A diagnosis of rheumatoid arthritis complicated with rheumatoid vasculitis was made. Pulse therapy was initiated with methylprednisolone for 5 days and associated with gabapentin for neuropathic pain treatment. Patient evolved with improvement of skin lesions, strength of the lower limbs, plantar and hands paresthesia after the pulse therapy. Opted for Hospital discharge and maintenance treatment with rituximab. **Discussion:** The rheumatoid arthritis is a chronic and systemic inflammatory disease progressive that involves joints, but also can show extra-articular manifestations, such as vasculitis. Rheumatoid vasculitis is a rare and serious complication that must have an early diagnosis, given the great interference in the evolution, treatment and prognosis of the base disease. The peripheral nervous system is affected in 50% of the cases. The worst prognosis of multiple mononeuritis occur with acute installation, axonal degeneration and presence of extensive and severe angitis. **Final Comments:** Rheumatoid vasculitis is a complication rare and severe of rheumatoid arthritis, that can be associated with others comorbidities, in especial the acute multiple mononeuritis that needs an aggressive early treatment for the reduction of the morbidity and mortality.

Neuropatias Periféricas

TL 1105516

LATE-ONSET CHARCOT-MARIE-TOOTH DISEASE WITH LEUKOENCEPHALOPATHY DUE TO MFN2 GENE VARIANT

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Case Presentation: A 73-year-old Austrian man presented with a long-standing clinical course of 20 years of slowly progressive unsteady gait, imbalance, tingling in the legs and hands, and complained about difficulty in speech and hand tremors for 5 years and the need of unilateral support for ambulation for 2 years. Medical history disclosed: bilateral cataracts and glaucoma surgery; chronic migraine with aura; left sinusotomy at age 35; chronic allergic rhinitis; three episodes of motor aphasia lasting about 30 minutes; generalized tonic-clonic seizures at 46 and 56 years; progressive hearing loss since age 58 years; chronic diarrhea; moderate hepatic steatosis. Family history was unremarkable. Examination disclosed diffuse palatal tremor, thoracic hyperkyphotic posture, slight saccades decomposition at horizontal and conjugate gaze, reduced deep tendon reflexes, flaccid quadriparesis (predominantly distal); hypopalesthesia, painful hypoaesthesia, and distal-proximal thermal gradient in the lower limbs. Electroneuromyography disclosed signs of severe axonal sensorimotor polyneuropathy, predominantly sensory, distal. Brain MR imaging disclosed diffuse leukoencephalopathy. Muscle biopsy showed chronic nonspecific neurogenic amyotrophy (10% of COX-negative muscle fibers). Cerebrospinal fluid analysis was unremarkable. Audiometry showed moderate bilateral sensorineural hearing loss. Genetic testing for Fabry's disease was negative. NGS-based multigene panel for neuropathies disclosed the definitely pathogenic missense variant c.2119C>T (p.Arg707Trp) in single heterozygosity in the MFN2 gene. **Discussion:** MFN2 (mitofusin 2) gene (1p36.22) is associated with mediating mitochondrial fusion and fission. MFN2 variants are the most common cause of axonal CMT, generally presenting with optic atrophy, hearing loss, vocal cord paralysis, severe early-onset axonal neuropathy (SEOAN) and even spasticity. Most cases have onset during the infancy, childhood, adolescence, or early adulthood, however our patient presented with late-onset compromise, as well as with palatal tremor, kyphosis, seizures, and leukoencephalopathy, which are not the most typical signs associated with MFN2 pathogenic variants. **Final Comments:** Genetic screening for MFN2 variants in late-onset neuropathies with complex clinical pictures is important in the diagnostic work-up, including cases with marked leukoencephalopathy.

Neuropatias Periféricas

TL 1105535

PIDC CASE AFTER VACCINATION FOR COVID-19 IN A PATIENT FOLLOWED UP AT HOSPITAL UNIVERSITÁRIO GETÚLIO VARGAS AND AMBULATÓRIO ARAÚJO LIMA

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Case Presentation: Vaccines against COVID-19 have mainly been associated with adverse events of little clinical importance. The profile of these side effects is still undetermined and more serious adverse events have been reported in a minority of cases. One such event is immune-mediated inflammatory polyneuropathy. There are some reports in the literature of cases of acute-onset inflammatory polyradiculoneuropathy in patients after COVID-19 infection and vaccination. Although little described in the literature, CIDP has already been reported in the post-vaccination period for influenza, tetanus and other common vaccines. We describe the case of a 52-year-old patient, who performed the first dose of vaccination for Covid-19 (Astrazeneca) on 06/12/2021 and, on June 16, 2021, started tingling in his hands and feet. The following day, he developed weakness in the lower limbs and, one day later, he was no longer able to walk. He was taken to the emergency room, submitted to Hospital admission on the third day of illness; during this Hospitalization, therapy with human immunoglobulin (1.65 mg/kg dose) was performed, with partial improvement of symptoms. After three weeks of Hospital discharge, he developed dysphagia and respiratory distress, being Hospitalized again in the emergency room on 08/03/2021. He was transferred to HUGV for follow-up with Neurology on 08/07/2021. On 08/13/2021, an electroneuromyography of the four limbs was performed, which showed sensory and motor neuropathy, axonal, symmetrical, with denervation of very serious intensity and intense signs of activity. During this Hospitalization, he performed seven sessions of plasmapheresis, observing significant clinical improvement. Two weeks after Hospital discharge, the weakness worsened, initially in the upper limbs and later in the lower limbs, and he was Hospitalized again on 10/14/2021 for clinical compensation, when immunoglobulin therapy was performed at a dose of 2 g/kg and this therapy was maintained. Monthly for six months, with significant improvement. Due to the clinical characteristics, ruling out other diagnoses and the form of disease progression, the diagnostic hypothesis of CIDP was proposed. In view of the above, it can be seen that there is temporality between vaccination and the onset of symptoms presented by the patient. The adverse effects of vaccines for Covid-19 are still being known, so it is essential to publish this report as a form of contribution to the scientific community.

Neuropatias Periféricas

TL 1105553

CASE PRESENTATION OF NON-SYSTEMIC VASCULITIC NEUROPATHY WITH IMPROVEMENT AFTER THERAPY WITH METHYLPREDNISOLONE AND CYCLOPHOSPHAMIDE

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Case Presentation: Vasculitic neuropathies are a heterogeneous and rare group of autoimmune disorders of the peripheral nerves. In primary vasculitic neuropathies, there is no clinical evidence of involvement of other organs. They usually have a multifocal distribution and develop acutely or subacutely. The most commonly affected nerves are the common peroneal nerve in the leg and the ulnar nerve in the arm. Electromyography reveals a predominantly axonal, asymmetric sensory and motor neuropathy (multiple mononeuropathies). Immunosuppression is the cornerstone of disease-modifying therapy. We describe the case of a 37-year-old patient with tingling paresthesia, shock-like pain, and paresis in the right hand, presenting after two weeks a similar picture in the topography of the left foot. She is initially evaluated by orthopedics and rheumatology for carpal tunnel syndrome. Neurological evaluation characterized multiple mononeuropathies. In a complementary investigation, asymmetric sensory and motor neuropathy of axonal nature and with signs of activity was confirmed, considering the possibility of vasculitic neuropathy. It was carried out an extensive investigation of the causes of systemic vasculitis, with negative results. The biopsy of the left sural nerve performed revealed a normal histological and immunohistochemical appearance. It was decided to perform therapy with methylprednisolone and cyclophosphamide due: first, to worsening of sensory and motor function, and second, a sustained increase in inflammatory tests. Excellent response with this treatment has been obtained. The importance of this case presentation lies in the fact that non-systemic vasculitic neuropathy is a challenging diagnosis. Usually, it is hard to perform a nerve biopsy. The adequate response to the instituted therapy reaffirms the relevance of the clinical evaluation to determine the beginning of the therapy, despite the negativity to the performed biopsy.

Neuropatias Periféricas

TL 1105560

ADULT-ONSET CHARCOT-MARIE-TOOTH DISEASE DUE TO SAMD9L GENE VARIANT

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Case Presentation: A 49-year-old Brazilian woman presented with lower limb weakness for 8 years, associated with tingling and sensory disturbance in both hands and feet. Medical history disclosed chronic diarrhea, self-limited episodes of blurred vision and abdominal pain mimicking appendicitis. Family history was unremarkable. Her cognitive function and general development were normal. Examination revealed inability to perform tandem gait, positive Romberg test, pes cavus, diplopia during bilateral horizontal gaze. Reduced deep tendon reflexes in upper limbs and brisk reflexes in lower limbs. Paraparesis (predominantly distal). Muscle biopsy disclosed mild subsarcolemmal mitochondrial proliferation and nonspecific chronic neurogenic amyotrophy. Nerve conduction studies disclosed distal and uniform demyelinating sensorimotor polyneuropathy. Brain MR imaging disclosed mild leukoencephalopathy, mild cerebellar atrophy, and the hot-cross bun sign. Laboratory tests revealed normal values. Whole-exome sequencing disclosed a heterozygous pathogenic variant in the SAMD9L gene. **Discussion:** Variants in Sterile Alpha Motif Domain-Contain Protein 9-Like (SAMD9L) gene have been associated with ataxia-pancytopenia syndrome (ATXPC), spinocerebellar ataxia type 49, myelodysplasia and leukemia syndrome with monosomy 7, and SAMD9L-associated autoinflammatory disease. The main clinical presentation in our case is a late-onset inherited demyelinating neuropathy, mimicking features of Charcot-Marie-Tooth (CMT) disease. Despite the presence of mild cerebellar atrophy, our patient presented mainly with sensory ataxia and her other clinical features resulted from chronic neuropathic compromise. **Final Comments:** Our report provides new evidence linking SAMD9L variants to primary peripheral nerve compromise, expanding the clinical spectrum associated with this gene. SAMD9L variants must be included in the differential diagnosis of late-onset demyelinating CMT phenotypes.

Neuropatias Periféricas

TL 1105717

ACUTE AUTONOMIC NEUROPATHY POST COVID19 VACCINE: A CASE PRESENTATION

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Case Presentation: A 26-years old woman presented with a four-days history of recurrent postprandial vomiting, diarrhea, headache and fainting. Her symptoms started within 24 hours after the second dose of COVID-19 vaccine AstraZeneca/Oxford. Her symptoms were mild at presentation, including headache, lightheadedness and vomiting. Then she had a syncope while taking shower and started experiencing recurrent episodes of palpitation and dizziness. On neurological examination her gait was normal. She was able to walk on heels and toes. Romberg sign was absent. Coordination was normal. There was an anisocoria and light-near reflex dissociation. Pinprick evoked pain, tactile and thermal sensation was abnormal throughout, but worse on the left side. Strength was normal in all muscles. Deep tendon reflexes were normal. There were no upper motor neuron signs. Her blood pressure lying was 100x60 mmHg and seated, 76x50mmHg. She did not tolerate standing. Tilt test was performed and at the 3rd minute her heart rate was 180 bpm, she had near fainting symptoms, causing it to be interrupted. CSF was normal at the 10th day of symptoms. Brain MRI was unremarkable. NCS performed at days 10 and 20 were within normal limits. Small-fibre evaluation revealed decreased RR variability. QST was abnormal. Sympathetic skin response to sound stimulus was absent in the upper limbs. Considering her clinical presentation and the electrophysiological abnormalities found, a diagnosis of acute autonomic neuropathy was made. She was treated with high dose of IV methylprednisolone and IgIV with excellent response. She was discharged after 2 weeks with mild tachycardia. **Discussion:** Acute autonomic neuropathy (AAN) is an extremely rare syndrome and may manifest with abnormalities in cardiovascular (orthostatic hypotension), gastrointestinal (anorexia or early satiety) and urogenital systems (erectile dysfunction), dry eyes and mouth, and also visual impairment due to disturbance of pupils constriction. AAN is often immune mediated. The vast majority of cases have good prognosis. It is known mild abnormalities on autonomic system are common following COVID-19 infection. This is the second case presentation that associates COVID-19 vaccine and AAN. **Final Comments:** The case we present, describing the development of acute autonomic neuropathy after COVID-19 vaccination and favorable response possibly associated with IVIg and corticosteroids, supports previous reports of this association.

Neuropatias Periféricas

TL 1105749

GUILLAIN-BARRÉ SYNDROME POST VACCINE: A CASE PRESENTATION

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Case Presentation: A 23 year old, male patient on March 6th started to feel paresthesia in both hands with later progression to his feet at night. After two days, he progressed with dysphagia and quadriparesis and worsening of paresthesia, later being admitted in an emergency room and being transferred within 01 day to an ICU of a tertiary Hospital. He denied any infections prior to his symptoms and said that he was vaccinated with his third dose of SARS COV 2 vaccine (Astrazeneca) within 14 days before his symptoms. On physical examination, he presented flaccid quadriparesis with abolished reflexes in lower limbs and diminished in upper limbs with facial diplegia and bulbar palsy, with no sensitive impairments. Electromyogram was performed on the fourth day of onset, with no significantly findings. CSF was analyzed and showed albuminocytologic dissociation. MR showed no abnormalities. He was treated with IVIG for 05 days and two days after the IVIG course he developed respiratory insufficiency and disautonomia and had cardiac respiratory arrest during rapid sequence intubation. He was resuscitated after eleven cycles and after two days he developed diffuse brain swelling and elevated ICP, being monitored with an intraventricular ICP device, and after one week he developed refractoriness to clinical treatment, being submitted to an EVD. He died two days after. The Guillain-Barré syndrome is an acute autoimmune disorder after infection or rarely to immunization in susceptible individuals. Autoimmune antibodies that recognize gangliosides are found in a large proportion of GBS patients and are thought to contribute to the ultimate pathology by inducing complement-mediated axonal injury and demyelination.¹ The first case of GBS following COVID-19 vaccination was reported in February 2021 in the USA in an elderly female who presented 2 weeks after the first dose of the vaccine.² Also, it has been reported GBS syndrome after SARS COV2 infection, with approximately ten days between viral infection and the development of GBS syndrome.³ One narrative review showed nine articles reporting 18 cases of SARS COV2 Vaccine associated GBS Syndrome.⁴ We described a male patient of 23 year old who developed a flaccid areflex quadriparesis and bulbar palsy with albuminocytologic dissociation within 14 days of vaccination with Astrazeneca, who had refractoriness to IVIG and died after complications from cardiac arrest. This report is importante due to the rarity of GBS induced by vaccination.

Neuropatias Periféricas

TL 1105768

AUTOSSOMAL DOMINANT IFIH1-DISORDER ASSOCIATED WITH SENSORIMOTOR POLYNEUROPATHY

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Case Presentation: The IFIH1 gene encodes a cytoplasmic receptor for viral double-stranded RNA, that activates type I interferon signaling and antiviral response. Gain-of-function mutations in IFIH1 are related to increased and inappropriate exposure to type I interferon resulting in a distinct set of inborn errors of immunity, Aicardi-Goutieres, and Singleton-Merten syndromes (SMS). SMS is a rare genetic disease characterized by calcifications in the aorta and cardiac valves, delayed primary tooth eruption, dysregulated mineralization, teeth loss, osteoporosis, dysmorphism, muscle weakness, psoriasis, and recurrent infections. This is the first description of polyneuropathy in an autosomal dominant IFIH1-Disorder presenting with incomplete Singleton-Merten syndrome. A male patient by the age of 2 started presenting joint retractions, hypodontia, fatigue, exercise intolerance, and large joint pain. He had severe bone fractures following minor traumas. At the age of 12, he was diagnosed with psoriasis. Clinical examination revealed hypodontia, broad forehead, anteverted ears, malformation of helix and goblin, and trophic alteration of skin. We observed distal weakness and retractions. Lower limb deep tendon reflexes were absent and upper limb reflexes were hypoactive. There was hypoesthesia in the lower limbs up to the knee and in fingertips in the upper limbs, associated with reduced vibration sense. Cognition was normal. Brain MRI showed foci of calcification in the left frontal radiated crown. Echocardiogram, EKG, and CSF analysis were normal. Bone densitometry revealed severe osteoporosis. Nerve conduction studies showed a length-dependent sensorimotor polyneuropathy. The nerve ultrasound was normal. Muscle ultrasound showed neurogenic changes in lower limbs sparing the rectus femoris, and tibialis anterior. Upper limb muscles were normal, as were paraspinal and respiratory muscles. Whole exome sequencing revealed a heterozygous mutation in the IFIH1 gene that was not previously described and is absent from population databases. We classified the variant presented here as likely pathogenic according to the ACMG criteria. This case confirms that IFIH1 gain-of-function mutations cause a spectrum of diseases. The sparing of the tibialis anterior muscle is of particular interest, as it is a commonly affected muscle in other hereditary polyneuropathies and this pattern may help in the differential diagnosis. Finally, polyneuropathy could play a role in patients' phenotype.

Neuropatias Periféricas

TL 1105778

EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS AS A CAUSE OF POLYNEURORADICULOPATHY

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Case Presentation: Patient 57 years old, male, with a history of asthma and no other comorbidities, with a report of COVID in JUNE 2021 evolving about 2 weeks later with palpable purpuric lesions in the lower limbs associated with initial crural sensory-motor deficit insidious and progressive character. On admission, he had facial diparesis and areflex quadriparesis (grade IV-). In the clinical investigation, CSF collection was performed, which showed 05 cells and 35 proteins, electroneuromyography that came with suggestive signs of acute immune-mediated inflammatory polyneuroradiculopathy and axonal nature (AMSAN). Laboratory tests showed significant eosinophilia (> 5000), positive rheumatoid factor and anti-DNA, P-ANCA 1/40, protein electrophoresis with increased gamma globulin and alpha-1-globulin. Such findings close the diagnosis of Eosinophilic Granulomatosis with Polyangiitis (GEP). Imaging examinations showed pansinusopathy. During Hospitalization, he underwent immunoglobulin for 5 days followed by pulse therapy with methylprednisolone, evolving with excellent clinical response and absence of eosinophilia. He is discharged with azathioprine and prednisone and remains asymptomatic in outpatient follow-up. **Discussion:** PEG, formerly called Churg-Strauss disease, is a systemic necrotizing vasculitis of small and medium-sized vessels characterized by extravascular granulomas, eosinophilia and eosinophilic tissue infiltrate. The annual incidence is estimated at 1 to 3 cases in 1000,000 people. The mean age of onset is 48 years, with a slight predilection for females. According to the American College of Rheumatology criteria, of the 6 possible clinical criteria (asthma, eosinophilia greater than 10%, mononeuropathy or polyneuropathy, non-fixed pulmonary infiltrate, paranasal sinuses abnormality, biopsy with extravascular eosinophils) the patient had 5, confirming a case of PEG. **Final Comments:** We present a case of a rare cause of polyneuropathy that had a good response after adequate therapy, showing the importance of including it among the hypotheses when the patient presents any of the suggestive symptoms for PEG, as it was diagnosed with early intervention, provides a significant improvement in the patient's quality of life and disease prognosis.

Neuropatias Periféricas

TL 1105787

LEFT BRACHIAL PLEXOPATHY AS A CONSEQUENCE OF THE NEUROLOGICAL SEQUELAE OF COVID-19

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Case Presentation: Male patient, 46 years old, diagnosed with COVID-19, remained Hospitalized in the ICU for 38 days, and after being discharged, he presented with flaccid paresis in MSE, loss of strength and sensitivity and pain. The patient underwent an electroneuromyography (ENMG) which indicated a plexopathy in the lower left trunk and a cranial Magnetic Resonance (RNM), which showed abnormalities in right semi-oval center. Subsequently, a plexus MRI was performed, which demonstrated a regular diffuse thickening of the left brachial plexus. Dorene TABS 75mg, Benerva and FST were prescribed. After 5 months, there was improvement in paresis, sensitivity, maintaining difficulty in moving the MSE. **Discussion:** The brachial plexus is a complex nervous structure, and is formed by the roots C5, C6, C7, C8 and T1, and which can also receive contributions from C4 and T2, and is a critical region of the peripheral nervous system, due to the fragility of its anatomical region and its relationship with neighboring mobile structures. Commonly, brachial plexus injuries are divided into: open, closed, perinatal paralysis and after radiation. Closed injuries can be further classified into supra, infra and subclavicular. The symptomatology presented by the patient is wide, as it is closely related to the type of nerve that was injured, whether it is sensory, motor or mixed. If it is a lesion of an exclusively motor nerve, the symptomatology will be a paralysis of the musculature innervated by it. If it is a lesion of an exclusively sensory nerve, the symptomatology will be a loss of sensation. And if it is a lesion of a mixed nerve, the symptoms presented by the patient will be characteristic of sensory and motor losses. To make the diagnosis, the patient's history and neurological examination must be taken into account. Electroneuromyography is an essential test to confirm the diagnosis, and imaging tests may also be requested. In general, in less severe injuries, the nerve can heal on its own, and physical therapy or occupational therapy is usually indicated. In more severe injuries, surgical intervention is usually required. **Final Comments:** Brachial plexus injury is a common pathology that compromises the individual in numerous spheres, such as social and economic. Therefore, it must be recognized early in order to reduce the risk of sequelae that this injury can cause.

Neuropatias Periféricas

TL 1105805

SEVERE IMMUNE NEUROPATHY WITH RESPIRATORY DISTRESS AND LOCKED-IN SYNDROME

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Case Presentation: A 58-years-old man, presented with tingling and needles in both hands, progressed to weakness. Over 5 weeks his symptoms progressed to the lower limbs, and he also complains of difficulty to go up and down stairs. In a few months, he was unable to walk without support. On neurological examination he presented with proximal and distal asymmetrical weakness, areflexia and sensory impairment distally. CSF was normal. NCS revealed a sensory and motor demyelinating polyradiculoneuropathy. A hypothesis of CIDP was made and he was put on high doses of intravenous methylprednisolone for 5 consecutive days. Despite that he rapidly deteriorated, and he was treated with IVIG with no response. He evolved with respiratory insufficiency requiring invasive mechanical ventilation. In addition, he had bilateral eyelid ptosis and restriction of eye movement in all positions. A bedside test with pyridostigmine was performed, with a slight improvement in ptosis in one eye. Then, PLASMEX was done and he started to recover. Based on the poor response to treatment IgG-4 nodo-paranodopathy was considered and rituximab was done, and he is improving. **Discussion:** CIDP is an acquired immune inflammatory polyneuropathy characterized by a progression over at least 2 months with a progressive or relapsing course. Our case had several unusual features that raised the possibility of a different diagnosis, including onset of weakness from upper limbs, cranial neuropathy, poor response to IVIG and methylprednisolone. The incidence of ventilatory failure in CIDP is very rare (1-9%), its mechanism is believed to be via phrenic nerve demyelination. Cranial neuropathy, ptosis and ophthalmoplegia are not common in patients with CIDP, raising the possibility of an overlap of myasthenic syndrome. **Final Comments:** We identified a patient with a distinct phenotype characterized by severe sensorimotor neuropathy presenting with tetraplegia, cranial nerve involvement, respiratory failure and locked-in syndrome and poor response to steroids and IVIG. There are some case presentation of anti-pan-neurofascin-associated neuropathy with similar phenotype. Unfortunately, we were not able to test specific antibodies.

Neuropatias Periféricas

TL 1105824

AN INFLAMMATORY LUMBAR RADICULOPLEXOPATHY POS SEVERE COVID-19 INFECTION ASSOCIATED WITH HETEROTOPIC OSSIFICATION

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Case Presentation: Neurological complications due to covid-19 infection have been linked to severe cases of COVID-19 infection since the onset of the pandemic. However, the clinical and pathophysiological characterization of peripheral nervous system involvement is still unclear. We describe a case presentation of a 57 years old man, previously healthy, who presented an asymmetric lumbar radiculoplexitis after a severe presentation of COVID-19 infection responsive to corticotherapy. In December of 2020, the patient was admitted to the Hospital with a respiratory failure related to covid-19 infection. He was intubated and placed on ECMO for 26 days. During this time, the patient had 3 secondary infections, multiple blood transfusions, and drug-induced hepatitis. He was tracheostomized and in February of 2021, after sedation was removed, the patient was quadriplegic and areflexia, and the diagnosis of critical ill polyneuropathy was made. In the following months, the patient was under rehabilitation, but no motor improve was noted on the left foot. In September 2021 came to our peripheral neuropathy center. At physical examination, the patient presented a plegia on the left foot with areflexia on left Achilles tendon. The nerve conduction study and the EMG showed an important partial axonal involvement of the left tibial and peroneal nerves with few functioning motor units. Partial axonal involvement of the right peroneal nerve. Lumbosacral MRI with neurography showed signs of inflammation in the roots of L4 to S1 and hyperintensity in T2 STIR with gadolinium enhancement bilaterally, but more intensive on the left, with extension to left sciatic nerve. A heterotopic ossification was identified between the left gluteus minimus muscle. Heterotopic ossification on left iliac cortical bone related to the previous Hospitalization period with edema located in the deep muscle fibers of the left gluteus minimus. Inflammatory edema affecting the soft tissues between the iliotibial tract and the greater femoral trochanter bilaterally, as well as the insertional portions of the gluteus minimus and medius, being slightly more evident on the right, may correspond to a change related to friction/pressure by decubitus. Signs of neuropathy/neuritis of L4 to S1 bilaterally more evident on the left. **Final Comments:** Severe cases of COVID-19 infection can lead to an inflammatory presentation of the peripheral nervous system as a radiculoplexitis presented here.

Neuropatias Periféricas

TL 1105831

CSF PATTERN, SEVERE PROTEINORRACHI, IN GUILLAIN BARRÉ SYNDROME AFTER SARS-COV-2 VACCINE, ASTRAZENECA – REPORT IN 3 CASES

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Case Presentation: 1. RF, 22 years old, ten days after the 3rd dose of astrazeneca vaccine, develops bilateral paresthesia in hands and feet, the following day the deficit increases to limbs. After 4 days, there was loss of strength in proximal limbs, hypoesthesia in the tongue and mouth and dysphagia. In cerebrospinal fluid (CSF): protein 476 and 1 leukocyte. After 2 days, the deficit worsened with new 938 protein orrachia. 2. R, 52 years old, 1 week after the first dose of astrazeneca, developed paresis in the right lower limb, on the 8th day he presented the same complaint in the left lower limbs, both ascending, in addition to bilateral upper limbs. On the 10th day, a vesical and nasoenteral tube was needed. In lcr: 453.9 proteins and 1 leukocyte. 3. FD, 56 years old, ten days after the 3rd dose of astrazeneca vaccine, he develops paresthesia in the hands and feet and 1 day after the deficit ascends to the legs bilaterally. It evolves with loss of strength in feet and legs, in addition to urinary retention. In lcr: 1 leukocyte, 306.6 proteins. All patients had MRIs of the skull and cervical spine, without changes and also without complaints of recent viral infection. In addition to an electroneuromyography study demonstrating demyelinating impairment. They also have a severe course with the need to repeat human immunoglobulin monthly. **Discussion:** The vaccine in question is composed of deficient replication of the adenovirus in chimpanzees, which stimulates the S glycoprotein of COVID 19, producing T cells and antibodies, which generates a molecular mimicry generating the attack on the myelin sheath, which culminates in guillain-barré syndrome (GBS). In all the cases presented, a closed diagnosis and clinical, cerebrospinal fluid and Enmg criteria. A common fact that draws attention is the high increase in protein, creating a CSF pattern for GBS post-ChAdOx 1 vaccine, not yet evidenced in the literature. **Final Comments:** Even though it is a rare occurrence, post-AstraZeneca vaccine GBS, totaling 27 cases in Brazil until 07/2021, being included as an adverse event in the immunization package insert. Given this fact, added to the importance of vaccination, since it reduces about 80% Hospitalizations in the elderly. It is extremely important to create a pattern for this new etiology of GBS, studying not only a new cerebrospinal fluid pattern, seen in an important proteinorachia report, but also clinical and electroneuromyography.

Neuropatias Periféricas

TL 1105850

MEDIAN NERVE ABSCESS DUE TO LEPROSY REACTION AFTER COVID-19 IMMUNIZATION: A CASE PRESENTATION

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Case Presentation: A 58-year-old woman, previously hypertensive, diagnosed with dimorphic-virchowian leprosy in 2017, underwent multidrug therapy (MDT) for 12 months with multibacillary MB regimen, but had to go through a new treatment cycle due to therapeutic failure at the end of MDT. After 19 months of this second treatment approach, the patient presented with fever, chills, intense arthralgia, edema, and paresthesia in the limbs, besides significant pain in hands and feet and cutaneous erythema nodosum leprosum. A sensory-motor evaluation revealed hyposthesia and paresis, especially in the left hand. No systemic co-infections were identified. However, the patient reported that she received the Oxford/Covishield vaccine against COVID-19 10 days before the onset of the condition. Upper and lower limb peripheral nerve ultrasonography identified marked thickening of the left median nerve. Doppler analysis demonstrated the presence of flow associated with a small hypochoic image measuring 0.5x0.3 centimeters near the perineurium, compatible with an abscess, in addition to thickening of the tibial, left fibular, and left ulnar nerves. **Discussion:** This report discusses a case of a type 2 reaction with neural abscess formation in a patient being treated for dimorphic-virchowian leprosy after receiving Oxford/Covishield immunization. Immune reactions can occur due to an immunological imbalance in leprosy. These reactions are divided into type 1, or reverse reaction, and type 2, or erythema nodosum leprosum and are acute and potentially serious and can constitute a medical emergency due to their morbidity and the worsening of already existing damage. There are some risk factors related to the triggering of those reactions such as co-infections and vaccines. The pathophysiology of leprosy reaction type 2 is still not completely understood, but previous studies show an intense inflammation mediated by humoral immune response, with increased levels of TNF-?. These same mechanisms are possibly involved in the immune response against COVID-19 and may be related to this post-vaccine reaction episode. **Final Comments:** The reported case serves as an alert to the possible consequences of the pandemic of COVID-19 for leprosy patients. It is necessary to ensure access to immunization for these patients, with increased vigilance after immunization, especially those with multibacillary forms and recurrent leprosy reactions.

Neuropatias Periféricas

TL 1106084

POLR3B VARIANT CAUSING AN AXONAL CHARCOT-MARIE-TOOTH DISEASE

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Case Presentation: An 18-years-old male patient presented with a history of daily vomiting, delayed puberty, and weakness. He complained of gastrointestinal tract symptoms since his first year of life. Frequent falls were noted when learning to walk. At age seven, he could no longer handle small objects, and was unable to climb stairs. The patient also complained of painful paresthesia. On physical examination, muscle atrophy was perceived, strength in opposition had a MRC grade 4 in proximal muscles and MRC grade 0 in distal muscles. Deep tendon reflexes were absent. Pinprick and vibration were reduced in upper and lower extremities. Electrodagnostic studies (EDX) revealed a severe axonal sensory-motor polyneuropathy. Median and ulnar motor conduction studies were within normal range. Neuromuscular ultrasound showed neurogenic changes in distal muscles. Whole-exome sequencing identified a heterozygous de novo mutation in POLR3B – chr12: 106.444.552 G>T (p.Arg682Lys). A final diagnosis of Charcot-Marie-Tooth type 2 (axonal) was made. **Discussion:** Polymerase RNA 3 B is the largest subunit of RNA polymerase III, an enzyme involved in transcription of small non-coding RNA. Classical disease descriptions were related bi-allelic mutations causing leukodystrophy. In 2021 this gene was associated with peripheral neuropathy. To our knowledge, only nine patients presenting with this pathogenic variant were described worldwide (six in original publication, two in Japan, and one in China). There are no descriptions of this variant causing neuropathy in Latin America. Although all cases reported had a EDX with predominantly demyelinating features, the case here presented is of a clear axonal phenotype, as demonstrated by EDX and nerve ultrasound. Delayed puberty was present in our patient, like classical leukodystrophy type. Other features such as abnormal dentition and ataxia were not present. **Final Comments:** Despite widespread use of Next Generation Sequencing in Hereditary Neuropathies, many patients remain without a final genetic diagnosis. This number is even larger in axonal types. Newly described variants associated with disease as the one here presented are of great interest in improving diagnostic rates and expanding phenotypes.

Neuropatias Periféricas

TL 1106181

MYASTHENIA GRAVIS IN ASSOCIATION WITH GUILLAIN BARRE SYNDROME: A CASE PRESENTATION

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Case Presentation: A 48-year-old male was diagnosed with Myasthenia Gravis (MG) five years ago, when presenting paresis in the all four limbs, eyelid ptosis and difficulty in swallowing, symptoms currently controlled with the use of pyridostigmine bromide and corticosteroid therapy. Concomitantly, thymoma was also detected and treated with thymectomy, radiotherapy and chemotherapy, getting complete remission. In April 2022, the patient sought an emergency care unit (ECU) due to fever and diffuse joint pain, being diagnosed with Chikungunya and medicated with analgesics. After a week, he evolved with paresis in the lower limbs, difficulty in locomotion, and paresthesias in the feet and hands, looking again for the ECU, where he was medicated with betamethasone. However, he evolved into urinary incontinence and progression of paresthesia to the trunk and upper limbs, being referred to the State General Hospital, where he was diagnosed with Guillain B arré syndrome (GBS), treated with plasmapheresis for 10 days. He currently has grade 5 strength in all limbs and preserved tactile, painful, and vibration sensitivities, only complaining of diffuse paresthesias. **Discussion:** GBS and MG are autoimmune neurological diseases. MG is caused by the action of anti-acetylcholine receptor antibodies, causing neuromuscular junction dysfunction and culminating in fluctuating muscle weakness. MG is often associated with thymomas, as described in the case, especially in the presence of positive antibody to AChR (AChR-MG). GBS is an acute inflammatory polyneuropathy, caused by the action of autoantibodies that act against myelin proteins or against components of peripheral nerve axons, often triggered by infections, in this case Chikungunya. It is estimated that the frequency of co-occurrence of MG and GBS, as in the case in question, is less than 1 in 10 billion. **Final Comments:** Although comorbidity of GBS and MG is extremely rare, early recognition of this combination of inflammation of peripheral nerves and the dysfunction of neuromuscular junction is of great importance for both initial treatment and a better prognosis.

Neuropatias Periféricas

TL 1106506

POEMS SYNDROME: CASE PRESENTATION

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Case Presentation: A 70-year-old woman was admitted to our Hospital with a 2-year history of complaints of lower limb paresthesia, imbalance, ascending paresis to the upper limbs culminating in bedridden and weight loss of 30 kg. Physical examination revealed hippocratic facies, diffuse muscular atrophy, severe bilateral hand contracture, non-painful hepatomegaly and lower limb edema with acrocyanosis. Neurological examination showed distal weakness, generalized hyporeflexia and paresthesia, hypoesthesia of the legs. Laboratory tests results showed a normal blood count and renal function, proteinuria and elevated TSH and VEGF. Electroneuromyography revealed sensorimotor peripheral polyneuropathy with a demyelinating pattern with axonal loss and Lumbar puncture had albuminocytological dissociation. Computer tomography showed bilateral pleural effusions, ascitis and osteosclerotic lesions at the vertebral body of L3, L4, and sacroiliac. Further investigation exhibited protein electrophoresis with a distortion in gamma zone and immunofixation elevated free light chains. Bone marrow biopsy showed a reactive and hypercellular marrow with interstitial plasmacytosis and polyclonal free light chain elevation. **Discussion:** POEMS syndrome is defined by the presence of a monoclonal plasma cell disorder, peripheral neuropathy, and one or more of the following features: osteosclerotic myeloma, Castleman disease, increased levels of serum vascular endothelial growth factor (VEGF), organomegaly, endocrinopathy, edema, typical skin changes, and papilledema. The pathogenesis of syndrome is not well understood, VEGF is the cytokine that correlates best with disease activity. Our patient exhibited most of the diagnostic criteria for POEMS. The most disabling feature of POEMS is the demyelinating neuropathy, frequently misdiagnosed as CIDP (chronic inflammatory demyelinating polyradiculoneuropathy). A typical POEMS neuropathy is a symmetrical, sensorimotor, length-dependent painful neuropathy, which progresses proximally over time. Treatment is aimed at the clonal hematologic disease and the cytokine drive and supportive care for the neurological and endocrine consequences of the disease. **Final Comments:** The aim of this study was to report a case of POEMS syndrome, emphasizing the importance of this condition, which, although rare, it should be part of the differential diagnosis of polyneuropathy.

Neuropatias Periféricas

TL 1106537

MILLER FISHER SYNDROME (MFS) ASSOCIATED WITH HEPATITIS B VIRUS (HBV) INFECTION: A CASE PRESENTATION

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Case Presentation: A 55-year-old female presenting an acute onset of bilateral ptosis, ophthalmoparesis, bilateral fixed mydriasis, limb ataxia and neck pain. Serum serology demonstrated B virus antigen. A ENMG and CSF analysis were normal in the first week. Because of the acute onset and pupil involvement, anti botulinum toxin serum was administered. However, in 3 days, the patient still presented worsening symptoms with new onset bulbar signs. Another CSF analysis identified protein-cytologic dissociation. Plasmapheresis was started suspecting MFS. Five sessions were performed and, after that, the patient improved from symptoms and remained in Hospital for dysautonomia surveillance. Treatment with tenofovir was also initiated for Hepatitis B, considering the patient's clinical condition was an extrahepatic manifestation. Due to improvement of dysautonomia and neurological symptoms, the patient was discharged from Hospital, after another ENMG, that showed typical signs of Miller Fisher Syndrome. **Discussion:** In MFS there is an acute polyneuritis, characterized by a triad of areflexia, ophthalmoplegia and ataxia. These last two signs were present in the patient in question. Anti-ganglioside antibodies are found in CSF analysis of patients with MFS in more than 90% of cases. This anti-GQ1b inflicts immune-mediated damage on target tissue found in cranial nerves III, IV and VI, which causes the characteristic ophthalmoplegia. Plasmapheresis, and intravenous human immunoglobulin are first line treatments. There are few case-reports of Hepatitis B Virus and MFS association in literature, with no evidence of direct cause-effect correlation. However, Guillain Barre syndrome has been reported as extra hepatic manifestation of hepatitis B, with viral antigen present in CSF and nerve biopsy. **Final Comments:** The relative rarity of MFS and the variety of diagnoses make it difficult to plan and implement procedures and treatments. It is assumed that the anti-GQ1b antibody is the mediator of such syndrome. Thus, early recognition and prompt plasmapheresis therapy can decrease the severity and duration of the disease. Furthermore, there is still a need for unpublished treatments that are evidenced in terms of their effectiveness and availability.

Neuropatias Periféricas

TL 1106617

DEVELOPMENT OF NEUROMA SECONDARY TO DECOMPRESSIVE SURGERY FOR CARPAL TUNNEL SYNDROME

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Case Presentation: A 42-year-old female with a previous diagnosis of carpal tunnel syndrome and two months of surgical decompression evolution median nerve endoscopically. According to the information collected, during the procedure (CIS) there was a possible cutting injury to the aforementioned nerve. After detecting the lesion, the physician would have made a longitudinal incision in the region for exploration, followed by primary raphy of the nerve. In the late postoperative period, the patient evolved with intense pain and paralysis of the muscles of the median nerve territory. The physical examination showed limited flexion and abduction of the wrist, weakness in thumb opposition, painful palpation of the region and positive Tinel's sign. A MRI of the wrist was requested, which showed a neuroma on the topography of the median nerve with a probable total lesion. The patient underwent microsurgery to expose the lesion and exeresis of the neuroma, with subsequent sural nerve grafting to try to improve pain and return movement of the muscles innervated by the affected nerve. One year postoperatively, the patient evolved with a functional hand, grade 4 strength in the muscles innervated by the median nerve and significant pain improvement (VAS 1). **Discussion:** Intraoperative complications of carpal tunnel syndrome treatment can occur during superficial skin dissection or ligament release. Endoscopic decompression surgeries of the nerve in question, can lead to the development of certain complications. The median nerve, having both motor and sensory fibers can have a variable pattern of deficit. Lesions repaired with neural raphy, can evolve with damage to the internal structure of the nerve fiber, allowing fascicular escape and disorganized nerve repair of axons in the circulating tissue. Added to this, there may be deposition of fibroblasts, schwann cells, which usually evolve with neuroma formation. After neuroma formation internal neurolysis may be indicated, with subsequent nerve autograft to supply the motor and sensory needs related to the median nerve damage. **Final Comments:** The case presented and the publications raised bring to light the discussion of the therapy of a complex situation that is postoperative neuroma and show that although adopted in a minority of cases, when well performed and in properly selected patients, internal neurolysis associated with autogenous graft is able to obtain satisfactory and lasting results regarding symptomatic relief.

Neuropatias Periféricas

TL 1105639

THE IMPACT OF INTRAOPERATIVE NEUROPHYSIOLOGICAL MONITORING ON THE POST-SURGICAL CLINICAL REPERCUSSION OF AN EPENDYMOA: A CASE PRESENTATION

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Case Presentation: Patient, male, 59 years old, in 2019, started with neck pain along with paresthesia in an irradiated band for the upper limbs (UL), muscle strength in the upper limbs grade IV and normal coordination. Cervical magnetic resonance imaging showed an intramedullary expansive lesion at the level of C4-C5 with a neoplastic appearance, ependymoma. Surgery was performed through posterior cervical access with laminectomy and fixation. Direct stimulation of medullary tissue was also used for precise location of the physiological midline and determination of the best myelotomy site. There was complete resection of the lesion during microsurgery with the use of intraoperative neurophysiological monitoring, which demonstrated importance in the functional and clinical outcome of the patient, given the sensory, which is potential more than expected, and without alterations in motor potentials. Postoperatively, the patient presented ataxic gait, hypoesthesia in the lower limbs (LL), mild paresthesia on the right side, and altered proprioception. Two months after surgery, the patient progressed with significant motor improvement, without worsening muscle strength. The sensory deficit reduced significantly and gradually in postoperative evaluations. At six months after the surgery, there were no more complaints. **Discussion:** Among the main subtypes of gliomas, ependymomas stand out, accounting for 60 to 80% of the cases. They are the most common intramedullary tumors in adults, whose peak age varies between 30 and 40 years. Due to the intramedullary location, the patient's symptoms resulted from progressive myelopathy due to compression of normal neural pathways. Intraoperative neurophysiological monitoring (IONM), used in surgery, is performed using techniques for analyzing the integrity of the neural system through somatosensory evoked potentials (SEPs), motor evoked potentials (MEP), D-wave and electroneuromyography. This technology has been the crucial factor in the post-surgical clinical repercussions, as it helps in the real-time feedback of critical pathways, preventing neurological deficits, enabling a rapid clinical evolution, in the absence of sequel, accelerating its functional status, as it happened with the patient. **Final Comments:** Neurophysiological monitoring is a crucial factor to avoid nervous involvement with postoperative motor and sensory repercussions, this been, a factor for reducing the risk of sequelae in the patient's clinic.

Neurorreabilitação

TL 1106379

EFFECT OF NON-INVASIVE NEUROMODULATION ON LANGUAGE REHABILITATION IN ACUTE STROKE

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Case Presentation: A 58-year-old male patient in the late postoperative period of mitral valve replacement associated with infective endocarditis. It evolved suddenly with motor and language alterations (onset of symptoms 14 days prior to Hospital admission date). On examination, NIHSS 16 (right hemiplegia and motor aphasia). Brain imaging showed hypodensity in the left middle cerebral artery territory associated with ECASS 2 hemorrhagic transformation. The patient was clinically managed, and rehabilitation measures were initiated while in the Hospital. There were 4 sessions of Transcranial Direct Current Stimulation – tDCS (20 minutes/session; 2mA; 35cm2)+ 8 sessions of speech therapy. Semantic and phonological processing were prioritized in a functional communicational context. After the interventions (discharge from Hospital), an improvement in fluency, access to the lexicon, and spontaneous and written language was observed, affecting mood, adherence to other opportunistic therapies (psychology and physical therapy), and a reduction in the NIHSS to 12. **Discussion:** Changes in neuroplasticity and cortical excitability may contribute to regulating neural activity. Both could be modified by applying direct electrical current in the sensorimotor cortex, with results dependent on the type and modality of the polarity of the current, and its effect would last for days after the end of the stimulation. To describe language skills achieved from Hospital bedside language rehabilitation in acute stroke. The cascade effect on therapeutic improvement has been well documented in the literature; however, the use of tDCs in the acute phase of stroke is still controversial, despite several studies having presented consistent data. In the Hospital environment, it is highly relevant to provide therapies that shorten rehabilitation since most patients come from accessible health networks (health prevention). **Final Comments:** The semantic and phonological networks were improved by using tDCS in the Hospital environment, providing an opportunity to enhance mood and adherence to the proposed multidisciplinary therapies.

Neurorreabilitação

TL 1105373

IMPRESSIONS FROM MEDICAL STUDENTS ON THE IMPACT OF THE COVID-19 PANDEMIC ON SLEEP AND QUALITY OF LIFE

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Background: The new Coronavirus, known as SARS-CoV-2, had its first case recorded in Wuhan, China in December 2019. However, there are already more than 500 million confirmed cases of COVID-19 in 220 countries around the world, with about 6 million deaths due to the novel coronavirus. There are many repercussions generated by the pandemic established as a result of this virus, especially consequences for people's quality of life and sleep. Medical students are a target group of studies on quality of life and sleep due to the stress and pressure they go through during academic life. **Objectives:** To evaluate the impact generated by the COVID-19 pandemic on the quality of life and sleep of medical students in Teresina-PI through their impressions. **Methods:** This is an observational, quantitative, and cross-sectional study, performed by filling out an online form by medical students and sent through social networks, from June to December 2021. **Results:** A sample of 198 participants was obtained, most of the participants were female (66%), with an average age of 23 years, most of the students were in the last two years of medical school (56.5%) and were from a public university (64%). Less than half (40%) of respondents said they had a good quality of sleep. When students compared poor sleep quality before and during the COVID-19 pandemic there was a 13% increase. Regarding quality of life, 70% of respondents said they had been negatively impacted. In addition, most students classified as negative the impact generated by the COVID-19 pandemic on sleep, academic performance, continuity of their studies, family income, physical and mental health. On the other hand, the vast majority of students (71.7%) reported improvement in family relationships. **Final Comments:** COVID-19 has significantly impacted in a negative form many areas related to the quality of life and sleep of medical students. Perhaps due to the measures of distancing, interruption of various activities and uncertainty about their continuity.

Neurosonologia

TL 1104934

ARTERY OF PERCHERON TERRITORY INFARCT: AN UNUSUAL CAUSE OF EXCESSIVE DAYTIME SLEEPINESS

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Case Presentation: A 23yo female comes to the emergency department complaining of excessive daytime sleepiness (EDS) starting in the last three days. Before, the patient went to bed around 11 pm, slept at 1:30 am, and woke up at 8:00 am. Currently, the patient sleeps around 18 to 20 hours per day. The patient refers one episode of visual hallucination three days ago. After that, permanent mild dizziness and sleep attacks. Her Epworth sleepiness scale was 14/21. The neurology exam was normal, except by subjectively drowsy but aroused easily to voice. The general labs, cerebrospinal fluid, serum tests, rheumatological and hypercoagulability panels were normal. Axial FLAIR and T2-weighted MRI show symmetrical hyperintense lesions on the paramedian thalamus with abnormal restriction of water diffusion on DWI and a low ADC. The findings above confirmed the diagnosis of an acute infarction of the artery of Percheron (AOP). **Discussion:** EDS is a common complaint in neurologic practice and has a wide spectrum of differential diagnoses. We describe an unusual cause of EDS caused by stroke in the bilateral thalamus. Thalamus is a large egg-shaped mass of grey matter situated at the core of the diencephalon and lies between the midbrain and forebrain, acting as a central hub. The medial thalamus is involved in memory and is essential for regulating arousal because it represents the superior extension of the midbrain reticular activating system. Bilateral lesions of the medial thalamus can cause hypersomnia and decrease consciousness. Paramedian arteries usually supply the medial thalamus. AOP is a rare anatomical variation presented in 4-12% of the population when a single arterial trunk originating from the posterior cerebral artery irrigates the paramedian regions of the thalamus and, sometimes, the rostral midbrain. Bilateral paramedian thalamic infarcts account for approximately 0.1% to 2% of all ischemic strokes and 27% of thalamic infarcts. People with AOP infarct have vertical gaze paresis (65%), neuropsychiatric symptoms, coma (42%), and memory impairment (58%). The EDS was the initial complaint in our patient and improved gradually after two months. **Final Comments:** Clinicians must be aware of AOP's infarction as a differential diagnosis of EDS, especially in the presence of sudden EDS onset or neuropsychological and ophthalmological signs.

Sono

TL 1105878

HYPERSOMNIA, BEHAVIORAL AND COGNITIVE ALTERATIONS: REPORT OF KLEINE LEVIN SYNDROME

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Case Presentation: R.C.C., 29 years old, started episodic hypersomnolence at age 15. Sleep attacks lasted between days to three weeks. The first was preceded by bacterial pharyngitis. During these periods, he only slept and only fed or ingested liquids when he was awakened. During awakenings, aggressive behavior, disinhibition, polyphagia, visual and auditory hallucinations, as well as memory changes were observed. There were no mood disorders, no sleep disturbances, substance abuse, or previous developmental delays. The first physical examination performed after the third episode of hypersomnia was normal. However, there was a history of spontaneously appearing hematomas on the skin. During this period, magnetic resonance imaging of the brain, examination of cerebrospinal fluid, digital electroencephalogram and polysomnography were performed, with normal results. Other complementary tests were also requested. There was a change in the white series in the blood count. Bone marrow biopsy showed changes consistent with chronic myeloid leukemia (CML). The intensity and frequency of hypersomnia episodes, which reached four in a single year, decreased over the years. The last crisis was in 2020, lasting 1 week. Thus, the diagnosis of Kleine-Levin Syndrome (KLS) was made. **Discussion:** KLS is a rare sleep disorder, characterized by recurrent episodes of hypersomnia that last from days to weeks associated, during the period of awakening, with transient cognitive and behavioral changes. The frequency and intensity eventually decay over 8 to 12 years, and may become extinct. The estimated prevalence is 1-5: 1,000,000 of cases, being more common in men. The average age of starting SKL is 16 years. The pathophysiology is still unknown. Autoimmune disorders, among others, appear as candidates for precipitating factors of the syndrome. However, we did not find CML among them in the scientific literature. The diagnosis is based on criteria established by the 3rd International Classification of Sleep Disorders (ICSD). Because it is a rare syndrome, there are no clinical trials and treatment guidelines. Currently, exclusively symptomatic therapy is chosen. **Final Comments:** In addition to the rarity of the syndrome, the never mentioned coexistence with a lymphoproliferative disease and the occurrence of CML in an unusual age group may provide new insights into the pathophysiology of the disease.

Sono

TL 1106305

HYPNIC HEADACHE AND COVID-19 PANDEMIC: IS IT A COINCIDENCE?

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Case Presentation: Hypnic headache (HH) is characterized by attacks that occur only during sleep with awakening. Patients with HH complain of pain between fifteen minutes to four hours, bilateral, and varies from mild to moderate intensity. The pathophysiology is poorly understood, but circadian disruption can be involved. We describe a case of HH during the COVID-19 pandemic. A 22-year-old patient from São Paulo complains of headaches exclusively during sleep (at night and napping), without autonomic symptoms, that started during the quarantine in the first wave of the COVID-19 pandemic in Brazil. The patient had significant circadian disruption during that period of life due to the pandemic. Neurological exams, magnetic resonance imaging, magnetic resonance angiography of the brain, and sleep studies were regular. He had three PCR tests negative for SARS-CoV-2 and no flu symptoms. Treatment with lithium carbonate 450mg at night led to the remission of the pain attacks. Circadian rhythms interfere in many aspects of physiology and pathophysiology of many biological systems and headaches. The association between the sleep patterns caused by the restrictions during the COVID19 pandemic can influence the incidence of a few kinds of headaches such as HH. The disrupted sleep and circadian rhythm could be contributed to the beginning of HH. Indeed, the medication that adjusted the circadian cycle was effective. This case highlights the possible impact of the COVID-19 pandemic and the lockdown on the incidence of HH and other headaches due to sleep and circadian disruption.

Sono

TL 1106328

WHOLE-GENOME ANALYSIS OF MONOZYGOTIC TWINS DISCORDANT FOR TYPE 1 NARCOLEPSY

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Case Presentation: Narcolepsy type 1 (NT1) is a rare and chronic neurological disease characterized by sudden sleep attacks, overwhelming daytime drowsiness, and cataplexy. To contribute to the understanding of NT1 genetic basis, here we describe a whole-genome analysis of a monozygotic twin pair discordant for NT1. Our whole-genome analysis revealed that both twins have identical pathogenic mutations in NT1 associated genes (such as HLA-DQB1*06:02:01, HLA-DRB1*11:01:02/*15:03:01) and therefore the affected twin has the expected clinical manifestation while the unaffected twin has an unexpected phenotype. This suggests that the unaffected twin might have protective alleles outside the HLA complex. The unaffected twin has significantly more frameshift mutations as compared to the affected twin (108 versus 75) and mutations that affect stop codons (61 versus 5 in stop gain, 26 versus 2 in start lost). Frameshift mutations and stop codon mutations are generally associated with loss-of-function effects and protective alleles are almost always loss-of-function rare alleles. Also, overrepresentation analysis of genes containing variants with potential clinical relevance in the unaffected twin shows that most mutations are in genes related to immune regulation function, Golgi apparatus, MHC, and olfactory receptor. These results support the notion that NT1 has an immunological basis but that protective mutations in non-HLA alleles might interfere with the expression of the NT1 phenotype and consequently with the clinical manifestation of the disease. Discordant monozygotic twin studies provide a unique opportunity to observe the effects of very rare alleles that might not be picked up in population studies.

Sono

TL 1106414

SLEEP PATTERNS AMONG PATIENTS WITH CEREBROTENDINOUS XANTHOMATOSIS

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Case Presentation: Two adult patients with CTX were evaluated with sleep anamnesis, physical exam and full-night type 1 polysomnogram. Case 1: J.R.A, male, 42 years old, presents generalized epilepsy and neuropsychomotor development delay since childhood. He had cataracts in his left eye, ataxia and tendon xanthomas. C.M.C, female, 32 years old, presents alterations in neuropsychomotor development in childhood, in addition to diarrhea. She had bilateral visual acuity impairment, ataxia, pyramidal signs and tendon xanthomas. Both patients had genetic confirmation. Sleep events were scored according to standard clinical criteria according to the American Academy of Sleep Medicine guidelines. The patients reported no specific sleep complaint or daytime consequence. Physical examination of those subjects revealed cranial deformities including high-arched palate and maxillary hypoplasia. No soft tissue abnormalities were identified. Patient 1 polysomnography showed reduced sleep efficiency (37.2%), high apnea hypopnea index (AHI) (15.5 events/h), high periodic leg movement index (PLMI) (77.4 events/h) and presence of alpha-delta pattern in NREM sleep. Patient 2 exam showed reduced sleep efficiency (80%), normal AHI (2.2 events/h) and presence of alpha-delta pattern. **Discussion:** Cerebrotendinous xanthomatosis (CTX) is a rare autosomal-recessive lipid storage disease caused by pathogenic variants in CYP27A1, which lead to deficiency of the mitochondrial enzyme, sterol 27-hydroxylase, resulting in the accumulation of cholestanol in the serum and many organs. Clinical presentation is variable and characterized by cholestasis, refractory diarrhea, juvenile cataracts, tendon xanthomas, osteoporosis, coronary heart disease, and progressive neuropsychiatric disturbances. Neuropsychiatric abnormalities include intellectual disability or dementia, psychiatric symptoms, cerebellar signs, pyramidal signs, progressive myelopathy, peripheral neuropathy, extrapyramidal manifestations, and seizures. To our knowledge, sleep disorders have not been studied in these patients. **Final Comments:** Patients with CTX may present higher prevalence of sleep disorders when compared with the general population. The absence of sleep complaints could be related to an overlap of symptoms with other neuropsychiatric disorders. Other studies with higher number of patients are necessary to understand sleep disorders in these patients.

Sono

TL 1106472

CENTRAL NERVOUS SYSTEM GANGLIOGLIOMA PRESENTING WITH SEVERE CENTRAL AND OBSTRUCTIVE SLEEP APNEA IN A CHILD: A CASE PRESENTATION

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Case Presentation: we report the case of a four-year-old girl who has experienced, according to her mother, snoring, breathing pauses lasting for 8-10 seconds, and a breathing pattern suggestive of Cheyne-Stokes ventilation during sleep since she was two years old. At the age of three, she presented headaches, vomiting and mental confusion, being diagnosed with Grade I ganglioglioma in brainstem and cerebellum. She was submitted to tumor resection and maintained under vinblastine chemotherapy. The sleep complaints got progressively worse, and she was referred to evaluation in the Neurology Sleep Medicine service. This child was obese (BMI = 37.6 kg/m²), usually sleeping from 11 p.m. to 9 a.m., waking up multiple times to urinate, but without any complaints of daytime sleepiness. She also had a small oral cavity (Mallampati 4, Brodsky tonsil scale 3, and an ogival palate). Polysomnography (PSG) evidenced severe sleep apnea [Apnea Hypopnea Index (AHI) = 84.1 events/h] with both obstructive (AHI = 13.3 events/h) and central (AHI = 70.8 events/h) components. Baseline SatO₂ was 90% and it decreased until 68% during apnea episodes. The central sleep apnea (CSA) showed up in the second half of the night, and Cheyne-Stokes breathing was observed. CPAP titration was effective to stop obstructive sleep apneas at the pressure of 8cm of H₂O, yet central apneas remained. In this second PSG, CSA also appeared in the second part of the night. **Discussion:** This case is about a rare condition, associated to severe central (also rare in children) and obstructive sleep apnea, not to mention the Cheyne-Stokes breathing. The prevalence of CSA is 4-6% in children with underlying conditions (using a cutoff > 5 events/h). Brainstem lesions may lead to several sleep modifications, such as reduced sleep efficiency, excessive daytime sleepiness, decreased stage 3 and REM sleep, and narcolepsy symptoms. These lesions might cause alveolar hypoventilation, having an impact on sleep, which may be severe enough to cause respiratory insufficiency and require mechanical ventilation, especially when lesions extend to the medulla. Brainstem lesions are a rare yet possible cause of CSA and the prevalence of central nervous system (CNS) tumors is higher in individuals with CSA when compared to controls. **Final Comments:** The presence of brainstem lesions increases the risk of multiple sleep pathologies. CSA is a rare condition in children and CNS tumors should be investigated as a possible cause.

Sono

TL 1106601

REFRACTORY HYPERSOMNOLENCE IN A PATIENT WITH MULTIPLE AUTOIMMUNE MORBIDITIES AND PSYCHIATRIC DISORDER

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Case Presentation: 35-year-old female presenting with excessive daytime sleepiness (EDS), insomnia, bruxism, snoring, fatigue, and fluctuating muscular weakness. She also complained of periods of hypersomnia that lasted for 3 to 7 days, without food compulsion, hypersexuality, or worsening of cognitive impairment. There was also worsening of muscular weakness in events of acute distress, but without complaints of sleep attacks, hallucinations, or sleep paralysis. She had the additional diagnosis of Mast Cell Activation Syndrome (MCAS), Ehlers-Danlos Syndrome subtype 3, chronic migraine, depression, Asperger syndrome, and a history of thyroidectomy followed by radioactive iodine therapy due to papillary carcinoma. She also referred to anasarca, drowsiness, and generalized muscle weakness in extremes of climate or acute distress and with consumption of histamine-rich foods. On physical examination, she had retrognathia and ogival palate, and there was also unilateral sustained palpebral ptosis. Muscular fatigability tests were positive for diplopia and led to discrete appendicular weakness. Polysomnography showed moderate obstructive sleep apnea (apnea-hypopnea index of 21 events per hour) and periodic limb movement syndrome (periodic limb movement of 49 per hour). Multiple latency sleep tests showed no sleep-onset REM period and average sleep latency of 19 minutes. Electroneuromyography with single fiber was compatible with myasthenia gravis, and the autonomic study was suggestive of autonomic neuropathy of tiny fibers. Thoracic computerized tomography did not demonstrate thymoma. Laboratory workup showed iron deficiency and a positive dosage of anti-acetylcholine receptor antibody. Renal and thyroid function and dosage of vitamin B12 were normal. She sustained EDS (Epworth sleepiness scale of 12/24) after treatment with CPAP, intravenous iron supplementation, and use of lisdexamfetamine, venlafaxine, trazodone, escitalopram, aripiprazole, coenzyme Q10, levothyroxine, and zolpidem. In the following steps, we will look for the prevalence of the HLA-DQB1*0602 allele and hypocretin level in cerebrospinal fluid. **Final Comments:** The differential diagnosis of EDS is complex and multifactorial. Proper workup is essential for a better clinical approach, although polytherapy is not always sufficient to entirely manage the symptoms.

Sono

TL 1104981

ACERULOPLASMINEMIA: A SEVERE NEUROMETABOLIC DISORDER

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Case Presentation: 59-year-old woman presented with recent onset depressed mood and emotional lability. After 6 months, forgetfulness, impairment of activities and speech difficulties were observed, followed by gait imbalance. On examination, she was dysarthric, disoriented in time, and also had saccadic intrusions, rigidity, oral dyskinesic movements, cervical dystonia, and ataxic gait. Investigation included complete blood counts, liver, thyroid and kidney function tests, HIV, syphilis and hepatitis serologies; and they were unremarkable. Additional tests revealed elevated ferritin levels (2540ng/ml), low serum iron (27 µg/dL), reduced plasmatic ceruloplasmin and low urinary copper. Brain MRI showed T2 hypointensity and SWI low signal on basal ganglia, thalamus and dentate nucleus. NGS-based gene panel testing was performed showing the pathogenic variant c.1149G>A (p.Tr383) as well as a variant of uncertain significance (VUS) c.31118T>C (p.Cys1040Arg) in the ceruloplasmin (CP) gene, in heterozygosity. The VUS is expected to disrupt CP protein function, although not yet reported on CP-related conditions. Despite not being able to determine if the CP variants are in cis or in trans, our final diagnosis was aceruloplasminemia (ACP). The patient initiated the treatment with deferoxamine combined with fresh frozen plasma (FFP) which controlled the disease progression. **Discussion:** ACP is an adult-onset autosomal recessive disorder caused by homozygous or compound heterozygous mutations in the CP gene (responsible for iron homeostasis). This mutation can cause systemic and cerebral iron overload. The clinical features are diabetes, retinopathy, liver disease and progressive neurological symptoms: movements disorders and cognitive dysfunction. An early sign of the disease is an atypical anemia with microcytosis, low transferrin saturation and hyperferritinemia. Brain MRI can show dentate nuclei of cerebellum, basal ganglia and thalamus with diffuse hypointensity in T2 and SWI. The diagnosis of ACP is established with typical clinical findings and the identification of pathogenic CP gene variants. The treatment is focused on iron chelators therapy (ICT). ICT is more effective in reducing systemic iron overload but less effective on neurological symptoms and it can be combined with FFP administration. **Final Comments:** ACP is a severe neurometabolic disorder that needs a prompt diagnosis and therapy to prevent neurological complications.

Transtornos do Movimento

TL 1105061

CLINICAL REASONING IN THE DIAGNOSIS OF MULTIPLE SYSTEM ATROPHY: A CASE PRESENTATION

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Case Presentation: A 53-year-old female complained of sudden and temporary loss of consciousness episodes after assuming the orthostasis position at the first admission. 2 years later, she had worsening symptoms, associated with dysarthria and insidious paresis of the right side. Currently, she had a clinical deterioration, with multiple fainting spells; severe dysarthria; insomnia and urinary incontinence. Upon examination, she was alert and oriented; had dysmetria, movement decomposition, hypermetric saccade and nystagmus; right-sided hemiparesis; plastic hypertonia; bradykinesia 3+/4+. Tilt test with positive result of type 3 confirmed postural hypotension and brain MRI showed tenuous T2/Flair hyperintensity in the topography of the median raphe of the pons. Follow-up neuroimaging showed a "Hot cross burn" signal, cerebellar volumetric reduction and bilateral putaminal atrophy. In the clinical course, Levodopa was used, with an unsatisfactory response and a possible extrapyramidal reaction, reinforcing the probable diagnosis of Multiple System Atrophy (MSA). In addition, she used fludrocortisone and had no clinical response to baclofen. **Discussion:** MSA is a neurodegenerative disorder that affects approximately 2 to 5 people per 100,000 population and has an adult-onset with a mean age of 55-60 years old. Although the cause of MSA is unknown, the described mechanism accounts for the abnormal accumulation of α -synuclein mostly in oligodendroglial cells. The clinical features include autonomic failure, parkinsonism and ataxia. MSA has two motor phenotypes: parkinsonian and cerebellar (MSA-C). This patient has an MSA-C phenotype, presenting as gait ataxia, limb ataxia, ataxic dysarthria and cerebellar disturbances of eye movements. A poor response to levodopa is highlighted as a characteristic that differentiates Parkinson's disease from MSA, as well as cerebellar and autonomic signs. MRI may reveal putamen, pons and middle cerebellar peduncles atrophy, as was described in the case here. The "Hot Cross Burn", a transverse and anteroposterior T2 hyperintensity in the pons region, is a characteristic sign result of the pontocerebellar fibers degeneration. **Final Comments:** MSA is a rare and severe disease, critical clinical reasoning associated with neuroimaging is extremely important for early diagnosis, since there is a symptomatological correlation mainly with idiopathic Parkinson disease and other atypical parkinsonian syndromes.

Transtornos do Movimento

TL 1105185

A TEENAGER WITH CHOREA AND TOOTH AGENESIS: A CASE PRESENTATION

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Case Presentation: A 16 year old female patient began 7 years earlier with involuntary movements in superior limbs and trunk, being medicated with Clonazepam and Topiramate, but with no improvement of the symptoms. Previously use of Levothyroxine and SSRI, there was no history of delivery complications or abnormal neurodevelopment. She had no family history of movement disorders or other neurologic diseases. The neurologic exam showed gait ataxia, myoclonus on upper limbs, generalized chorea and partial tooth agenesis. At this point, a trial with Levodopa three times daily was initiated and the symptoms ameliorated. She underwent several complementary investigations, such as brain MRI, EEG and blood laboratory tests, which were all unremarkable. At last, whole exome sequencing was performed and the results pointed microdeletion on chromosome 14 (14q.13.3q.21.1), with 5,4Mb, heterozygosis and variation on copy numbers, therefore, involving the following genes: NKX2-1 and PAX-9. Hence, this patient was diagnosed with hereditary benign chorea within NKX2-1 spectrum disorder and, notably, the tooth agenesis was due the mutation on PAX-9 gene, both located in proximity. **Discussion:** The mutation on NKX2-1 gene is related to systemic manifestations, such as benign hereditary chorea, congenital hypothyroidism, and neonatal respiratory distress (also known as brain-lung-thyroid syndrome). Additionally, the PAX-9 gene mutation, associated to hypodontia, is not expected to be involved, though, our patient presents molar tooth agenesis and we concluded that this phenotypic presentation is due the proximity between the NKX2-1 and PAX-9 genes on chromosome 14. **Final Comments:** PAX-9 mutation was not expected with NKX-2's, as both doesn't have any phenotypic or genetic association. But, in this particular case, genetic sequencing showed that one mutation may lead to subsequently alterations in other genes located nearby. Furthermore, it is of great importance to remind that benign hereditary chorea is the only form of chorea that improves with Levodopa treatment, contrarily with other choreas, which worsen with this medication.

Transtornos do Movimento

TL 1105195

PARKINSON'S DISEASE EMERGED DURING ACUTE COVID-19 ILLNESS: A CASE PRESENTATION

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Case Presentation: A 47-year-old female was evaluated in an outpatient setting with a complaint of asymmetrical rigidity, rest tremor and slowness. She had been Hospitalized two months prior due to mild COVID-19, notably with anosmia and ageusia. The motor symptoms were noticed by the patient on the third day of Hospitalization, with progressive worsening. She has a medical history of chronic constipation and was taking multivitamins and pantoprazole. She lives in an urban area and works at a Hospital laundry. She declined psychiatric or cognitive problems. Her family history was negative for neurological disorders. On MDS-UPDRS part III she scored 37 points, presenting bradykinesia grade 4 in the right upper limb (LUL), grade 3 in the right lower limb (RUL) and grade 1 in LUL. She had unilateral grade 2 rest tremor in RUL, bilateral postural and intention tremor, and unilateral grade 2 rigidity in LUL. The rest of the neurological exam was unremarkable. Laboratory assessment and brain MRI were normal. Functional study with SPECT with TRODAT showed bilateral putamen reduced caption, with right and left basal ganglia/occipital relation of 0.39 and 0.58, respectively (reference value: 1.98 ± 0.24). She was started on levodopa/benserazide 100/25mg TID. After 1 month, she noticed an improvement with a slight objective reduction of the MDS-UPDRS part III to 34 points (8.1% decrease). An increase in medication was considered, but the patient did not desire dose modification. **Discussion:** Numerous neurological symptoms have been described in acute and long COVID-19. Among the theories involving the invasion of the CNS, there are the hematogenous route and axonal transport through the olfactory neuroepithelium. To date, only three COVID-19-related parkinsonism cases have been reported. Same as our patient, two of them presented hyposmia. The SPECT with TRODAT was also abnormal in all three cases, and the motor symptom onset ranged between 10 and 32 days after PCR-confirmed diagnosis. **Final Comments:** Although a latent hypokinetic rigid syndrome may be unmasked in the context of great stress or infection, possibly by depleting compensatory mechanisms, the acute presentation during viral illness may implicate a more direct causal relation. Thus, this case could represent either one of secondary parkinsonism, with a more benign prognosis, or of a neurodegeneration exacerbated or even triggered by COVID-19. Follow-up will help differentiate between the two scenarios.

Transtornos do Movimento

TL 1105315

PARANEOPLASTIC HEMICHOREA ASSOCIATED WITH SMALL CELL LUNG CARCINOMA

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Case Presentation: A 69-year-old male, with a previous history of hypertension and smoking, and no relevant family history, presented with a report of dry cough with progressive worsening over several months, evolving with mild dyspnea on exertion. He was admitted to the Pulmonology service for investigation, and a lung nodule was detected on chest CT. During Hospitalization, neurological evaluation was requested due to a painless involuntary movement in the right lower limb, which had started about 2 months before. On physical neurological examination, the patient had right hemichorea with no other abnormalities. He underwent an extensive etiological investigation; a brain MRI showed no significant abnormalities, laboratory tests (including blood glucose, thyroid function, infectious serology, rheumatology screening tests) were also performed with normal results. He underwent biopsy of the pulmonary nodule, and small-cell lung cancer was detected. Symptomatic treatment with a haloperidol was initiated, with partial improvement of the condition. **Discussion:** Chorea is characterized by involuntary random movements, erratic and migratory, with an abrupt onset, large and short duration, which are repeated with variable intensity. Immune-mediated chorea encompasses several etiologies including paraneoplastic, post-infectious, and idiopathic. Paraneoplastic chorea (PC) is a rare entity, caused by nerve tissue damage by the effects of onconeural autoimmune factors, mediated by T cells. PC is more common around adult age, and small cell lung carcinoma (SCLC) is the most prevalent association. Chorea may predate the discovery of the neoplasm by months to years and may present in conjunction with other movement disorders and encephalopathy. The antibodies most commonly identified in PC are CRMP-5 (CV2) and ANNA-1 (Hu) IgG. In SCLC, approximately 70% are associated with CRMP-5. Tests being false negatives in a minority of cases, which may imply discouraging neoplastic screening in less classic cases. The expected brain MRI finding is basal nuclei hypointensity on T1-weighted sequences and hypersignal on T2/FLAIR. The result and prognosis depend on the diagnosis and early initiation of therapy. **Final Comments:** Although rare, a paraneoplastic etiology must be considered as a differential diagnosis in patients presenting with chorea. Screening for neoplastic conditions should be performed, and early treatment for the underlying malignancy may help decrease morbimortality.

Transtornos do Movimento

TL 1105408

ORTHOSTATIC TREMOR PLUS ASSOCIATED WITH PARKINSONISM: A CASE PRESENTATION

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Case Presentation: Female patient, 79 years old, hypertensive, family history of Parkinson's disease. Five years ago, she started presenting tremors in the lower limbs only when in orthostatic position. It progressed until last year, when it evolved to difficulty in standing. Four months ago, she started treatment with Primidone 100mg/day, without significant improvement. She denied symptoms of REM sleep behavior disorder, anosmia, or constipation. On physical examination, high-frequency tremor was observed in orthostatic position in all four limbs, more significant in lower limbs, grade 2 bradykinesia on the upper limbs and 1 on the right lower limb, rigidity grade 1 on the right and 2 on the left, reduced arm swing during ambulation and postural instability. Previous MRI and ENMG were normal. Based on the patient's clinic, the diagnosis of orthostatic tremor (OT) was defined. The patient showed a good response to the increase of Primidone to 250mg/day and subsequently started treatment with Levodopa 200mg/day for Parkinsonism. **Discussion:** In this report, the patient presented a movement disorder characterized by tremor while in an orthostatic position associated with parkinsonism, suggesting a diagnosis of OT-plus. OT is a rare condition of unknown origin that usually occurs in the lower limbs and occasionally involves the trunk and abdomen. The tremor is characterized by its high-frequency and low-amplitude. It begins immediately or a few minutes after assuming orthostatic position and disappears or decreases with ambulation or rest. When associated with another neurological disease, in this case parkinsonism, it is called OT-Plus. Other neurological manifestations reported in the literature are Parkinson's disease, dyskinesias, neuropathies and others. **Final Comments:** In order to make the OT diagnosis, muscle recording performed by surface electrodes while standing is essential, since high-frequency tremor 13 to 18 Hz is characteristic. It is important to emphasize that the conventional ENMG of the lower limbs does not present significant changes. Regarding treatment, drugs known to be effective in reducing tremors, such as primidone and propranolol, should be considered. Levodopa has also shown a modest response to OT-Plus associated with parkinsonism, suggesting involvement of the dopaminergic system. Among the differential diagnoses, it is worth mentioning other causes of postural instability, such as Parkinson's disease, ataxias and other types of tremor.

Transtornos do Movimento

TL 1105447

POTOMANIA AS SYMPTOM IN WILSON'S DISEASE: A CASE PRESENTATION

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Case Presentation: It is a report of a 22-year-old male patient who, since March 2020, has evolved with depressed mood, apathy, social isolation and speech reduction, anterograde amnesia and dyscalculia. In one month, he presented slowing of speech and hypophony, progressively worsening with dysphagia, imbalance and tremors, especially in the lower limbs. He was restricted to the bed in November 2020. No comorbidities or similar family history and non-consanguineous parents. On physical examination, a palpable liver and free tube. He had depressed mood and bradypsychism; alert, oriented, with micrograph, without other language changes. He presented hypophony and the kayser-fleischer rings were clear, associated with parkinsonism and cognitive impairment. He was diagnosed with Wilson's disease and started treatment, progressing with potomania, with intense fluid intake and vomiting. Psychiatric symptoms in Wilson's disease occur in almost 100%, but potomania are less common, with no cases reported in the disease, so far.

Transtornos do Movimento

TL 1105488

MYOCLONUS-DYSTONIA OF PROBABLE GENETIC ETIOLOGY: CASE PRESENTATION WITH MOTOR SYMPTOMS RESPONSIVE TO THE USE OF LEVODOPA

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Case Presentation: Female, 21 years-old, without relevant family history, present to our service of movement disorders due tremor. Around 7-years-old, she had tremors exacerbated in moments of mood changes and it was common to drop objects, especially in morning. After ten years the symptoms were more evident in the left upper limb and interfered with her daily life, and since then there has been progression to the contralateral upper and lower limbs. In addition, she initiated movements described as "muscle spasms", which were often painful. In our evaluation, she had increased tone and trophism in the left sternocleidomastoid and trapezius and in the extensor and flexor muscles of upper limbs. Stereotyped involuntary movements more intense on the left in upper limbs, of low frequency and high amplitude, less evident on lower limbs, bilateral oromandibular dyskinesia. Ultra-rapid movements, especially in shoulders and arms. No other alterations on neurological examination. MRI and laboratorial tests were normal. Electroneuromyography showed postural and intention tremor, bilateral and asymmetric, predominance in the left upper limb and frequency of 6Hz. Subsequently, Levodopa+Benserazide 100/25mg tree doses daily was started, with substantial improvement in symptoms. **Discussion:** Myoclonus-dystonia (M-D) is an autosomal-dominant movement disorder caused by mutations in Epsilon-Sarcoglycan (SGCE) gene, located on chromosome 7q21. The onset of symptoms usually occurs in the first decade of life. It manifests with myoclonus that most often affect the trunk, cervical region and upper limbs; and dystonia also in the upper limbs. There is a description of improvement in motor symptoms after alcohol libation. The diagnostic criteria proposed by Grünewald et al. (2008) classify M-D, according to the presence of family history and classic symptoms, as Definitive, Probable or Possible. Drugs that should be considered for treatment are benzodiazepines and anticholinergics, including in combination. Several other drugs need more robust studies. **Final Comments:** M-D's definitive diagnosis is limited by unavailability of genomic sequencing in most services. There is still a lack of therapies that change the prognosis of a disease that leads to significant functional impairment. And, in this sense, Levodopa may appear as a viable option, something little described in the literature.

Transtornos do Movimento

TL 1105506

COVID-19-ASSOCIATED MYOCLONIC ENCEPHALOPATHY – REPORT OF A 79-YEAR-OLD PATIENT

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Case Presentation: A 79-year-old male patient with RT-PCR confirmed COVID developed jerky movements in his trunk, face, and limbs on the eighth day of symptoms, preventing him from walking. He developed dysarthria and dysphagia, which led to the need for a feeding tube. Fourteen days after COVID symptoms started, he was transferred to the Hospital, for supplemental oxygen, without intubation. The neurologic examination showed mild somnolence, temporal disorientation, and generalized myoclonus, with positive and negative jerks, predominantly involving all four limbs, trunk, and proximal musculature. CSF analysis and brain MRI were both unremarkable. Under the hypothesis of COVID-associated myoclonus, methylprednisolone (MP) 1000mg/day for five days was used, with mild improvement of dysphagia and, to a lesser extent, dysarthria. Immunoglobulin IV 0,4g/Kg/day for five days was used with improvement in walking, confusion, and myoclonus. He was discharged with Levetiracetam, Clonazepam, and Valproate. He gradually improved, weaning off his medications over the following months. He was released from the outpatient service without any prescription, remaining without myoclonus and with complete recovery of motor and cognitive function. **Discussion:** Myoclonus is a condition with multiple etiologies and topographies, most commonly secondary to medical illness. SARS-Cov-2 potential to cause myoclonus has been explored in recent reports and probably shares mechanisms with other viruses under the hypothesis of an immune-mediated condition. Other myoclonus causes were extensively sought but were dismissed. The patient didn't undergo severe hypoxia nor use myoclonus-inducing drugs, and there were no metabolic disorders. **Final Comments:** In this case, we present an unusual manifestation of a severe generalized myoclonic encephalopathy due to COVID. Some limitations should be cited: despite the significant improvement, it's unclear whether this was a result of immunotherapy or the natural evolution of this poorly understood disease; finally, EEG and EMG weren't performed due to logistical difficulties aggravated by the pandemic.

Transtornos do Movimento

TL 1105617

CASE PRESENTATION HEMICHOREA IN A PATIENT WITH NON-KETOTIC HYPEROSMOLAR HYPERGLYCEMIC STATE AFTER KIDNEY TRANSPLANTATION

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Case Presentation: A 76-year-old woman was admitted to the emergency room with involuntary movements in the left hemibody that started suddenly four days ago. Medical history revealed diabetes mellitus since 2005, systemic arterial hypertension and renal transplantation 5 months before admission, after nephropathy due to the underlying diseases. Pre-transplantation tests showed 6% glycated hemoglobin in oral hypoglycemic use, but after transplantation there was decompensation of glycemia, attributed to the use of corticosteroids, with the need for insulin therapy. On neurological examination, she presented continuous choreoathetoid movements in the distal region of the upper and lower limbs on the left. She had a note of blood glucose tests, most of them above 250mg/dL. Glycated hemoglobin was 11.6% and venous blood gas was normal. Brain MRI showed mild T1 hyper-signal in the globus pallidus and putamen on the right. The diagnosis of hemichorea secondary to a non-ketotic hyperosmolar hyperglycemic state was established and the patient was managed with intensive diabetes control and haloperidol 2.5mg orally twice daily. There was significant improvement in the following days and the patient was discharged with almost no symptoms. **Discussion:** This case describes the clinical presentation and imaging findings of hemichorea in a non-ketotic hyperosmolar hyperglycemic state. This condition is relatively rare, with an estimated prevalence of 1 in 100,000 people, and it occurs more frequently in women with uncontrolled type II diabetes. Brain MRI typically demonstrates a hyperintense signal in the putamen and caudate nuclei on T1-weighted images, usually more prominent on the side contralateral to the hemichorea. Proposed etiologies include petechial hemorrhage, temporary ischemia (with or without hyperviscosity), or decreased synthesis of GABA and acetylcholine secondary to metabolic changes. Treatment is based on blood glucose control, but chorea may continue for months after hyperglycemia resolves and require specific medication. **Final Comments:** The present case illustrates a condition that, although rare, is treatable and, therefore, must be recognized. A thorough physical examination and compatible clinical history and imaging are essential tools for diagnosing and treating metabolic chorea. In this case of symptoms secondary to diabetes decompensation due to corticosteroid use after transplantation, glycemic control and central dopaminergic inhibitor were effective.

Transtornos do Movimento

TL 1105619

PARKINSON'S DISEASE ASSOCIATED WITH PARK7 MUTATION: CASE PRESENTATION OF CLINICAL ONSET AT 20-YEARS-OLD

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Case Presentation: Female patient, at 20 years, presented with rest tremor in right hand, evolving during 5 years with continuous tremor, worse in right side, associated with walking difficult. Approximately 2 years ago, the patient refers the onset of vocal disturb. The patient was admitted in our service in 2022 presenting spastic and hypophonic speech, severe bradykinesia and postural instability associated with high-frequency, moderate amplitude tremor, with right hand intention tremor and bilateral kinetic tremors of the hands. The patient was submitted to neuroimaging investigation of brain and cervical spine, as well as metabolic screening, with no pathological findings. The patient was submitted to Levodopa overload test, showing objective improvement in symptoms, and subsequently to genetic investigation, that showed PARK7 mutation. Initial treatment with levodopa at 400mg daily developed orofacial dyskinesias, so long-acting Pramipexol was started, with significant improvement of bradykinesia, vocalization and postural instability. **Discussion:** Although it is known that the majority of PD is a combination of complex genetic susceptibility and environmental factors, around 5–10% of PD may be attributed to monogenic forms. Mutations in the PARK7 gene are a rare cause of autosomal recessive PD, with at least 20 mutations in its gene identified. The majority (83%) of PARK7-linked PD cases have a clinical onset between the second and third decades of life, whereas 4% have late onset and 13% have juvenile onset, and the youngest youngest case presented was in a 5-year-old patient. Clinically, PARK7 mutations has been described as a levodopa-responsive parkinsonism with early or even juvenile onset and slow progression. In contrast to others neurodegenerative diseases that manifests itself at such a young age, autosomal recessive PD caused by PRKN, PINK1, and PARK7 mutations typically combines young age at onset with very slow progression, as illustrated by our current case. **Final Comments:** Although gene mutations in PD are rare, atypical cases in young patients with adequate diagnosis implies in significant recovery in quality of life. Furthermore, studies of the genetic and neuropathological features of rare genetic forms of it has been described PARK7 as potentially key role in new therapeutic targets and interventions.

Transtornos do Movimento

TL 1105629

WILSON'S DISEASE: PROGRESSIVE DYSTONIC PRESENTATION

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Case Presentation: A 16-year-old male patient, previously healthy, presented with involuntary movements that started 2 months ago in the 5th finger of the left hand, later compromising the entire hand. He had non-painful sustained contractions, that occurs at rest, in action and worsened with stress. One month after the onset of the condition, he developed cervical tremor, facial contractions and speech alteration. There were no family history for neurological diseases. On neurological exam he had mild hypotonia, dystonia of the left upper limb, cervical and facial, cervical tremor that worsened with rotation of the head to the right, and alteration in speech due to muscle contraction (dystonia) of the facial/orobuccal muscles. Ophthalmological evaluation showed the presence of Kayser-Fleischer rings. MRI revealed hypersignal in the head of the caudate nucleus and putamen bilaterally. Electroneuromyography showed dystonic movements of the left upper limb. Laboratory tests: hepatic function without alterations, ceruloplasmin < 9 (21–53mg/dl), serum copper: 32 (70–150 µg/dl). **Discussion:** Wilson's disease is an autosomal recessive genetic disorder of copper metabolism, resulting from the mutation in the ATP7B gene that encodes ceruloplasmin (transmembrane copper transport protein), leading to the accumulation of the metal in the liver, brain, cutaneous tissues, as well as in the cornea (Kayser-Fleischer rings). Typically manifesting by early adulthood, characterized by tremor, dystonia, chorea, parkinsonism, cerebellar and psychiatric symptoms, liver disease (tends to be seen in early-onset presentations). MRI shows hypersignal on T2 and hypointensity on T1 in the basal ganglia, thalamus, midbrain (face of the giant panda sign), pons and cerebellum. Laboratory tests show reduced levels of ceruloplasmin, normal or increased serum copper, urinary copper >100 g/24h. The gold standard for diagnosis is liver biopsy. Treatment is based on drugs that chelate or reduce copper absorption. **Final Comments:** In this case, the patient showed predominantly neurological involvement, with the presence of Kayser-Fleischer rings without hepatic/psychiatric involvement. Progressive dystonic condition was observed with early diagnosis based on laboratory and neuroimaging alterations. Early diagnosis is important, and appropriate treatment should be promptly initiated to prevent progressive and irreversible damage.

Transtornos do Movimento

TL 1105710

CORTICOBASAL DEGENERATION DIAGNOSTIC CHALLENGES IN A PATIENT WITH WERNICKE-MANN'S POSITION

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Case Presentation: A 52 year-old male with a prior history of hypertension and well controlled epilepsy was admitted to the outpatient neurology department with severe pain and rigidity in the right upper limb. The symptoms had an insidious onset and gradual progression over the course of 2 years, assuming a Wernicke-Mann position. The patient reported depressive mood, irritability and self-injurious behavior mainly against his right arm. On neurological examination, hemiplegic gait, allodynia in the right upper limb, weakness and hypertonia in the right side were observed. Evaluation of the affected limb was limited by the patient's pain. Brain MRI was normal without noticeable vascular lesions that could explain the motor symptoms. He was started on citalopram and was referred to psychiatry that suspected of conversion disorder. Over the next years, he presented with worsening of the cognitive symptoms with memory impairment, ideomotor apraxia and mild dysarthria and developed loss of balance and bradykinesia in the four limbs, unresponsive to Levodopa. A new MRI showed mild corpus callosum atrophy, leading to the hypothesis of corticobasal degeneration (CBD). **Discussion:** CBD is a rare neurodegenerative disease characterized by intracellular accumulation of misfolded tau proteins which presents as an asymmetric rigid-akinetic parkinsonism with dystonia and cortical deficits. Frontal behavioral-spatial syndrome, nonfluent/agrammatic variant of primary progressive aphasia and progressive supranuclear palsy (PSP) syndrome are also possible phenotypes of this disorder. Similarly, multiple etiologies may mimic its symptoms, namely PSP, Alzheimer disease, Pick disease and dementia with Lewy bodies. This overlap represents a challenge to the diagnosis of CBD with an antemortem prediction in only 25 to 56% of cases. Also, as seen in this case, symptoms like dystonia and pain may hinder the assessment of apraxia and bradykinesia. In this context, new biomarkers are important to distinguish similar etiologies. Unfortunately, their high cost is impracticable for use in the general population, especially in developing countries. **Final Comments:** Misdiagnosing CBD is frequent and can lead to harmful and unnecessary interventions. Even without a specific treatment, the prognostic perspective to the patient and its family is invaluable. Also, it is important for neurologists to be able to distinguish CBD from its mimics because of potential future treatments.

Transtornos do Movimento

TL 1105716

EXCESSIVE SMILE CAN BE WILSON DISEASE

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Case Presentation: A 22-year-old male patient with no previous disease with a 6-month history of deep sadness, anhedonia and recurrent crying episodes. Three months ago, he presented with dysarthria, dysphonia and dysphagia, losing 6 kg since then. Neurological examination evidenced an excessive and maintained smile, despite the depressive symptoms, associated with difficulty in opening the mouth, mild instability of gait and bradykinesia predominantly on the left side, without rigidity or tremor. Magnetic resonance evidenced extensive signal alteration spreading through thalamus, pons, putamen, caudate nuclei and a questionably alteration of globus pallidus, in addition to volumetric loss of basal nuclei and cerebellum. Serum ceruloplasmin was 3,6 mg/dL (normal: 20,0 – 60,0 mg/dL) and urine copper was 184,8 (normal: < 80 mcg/dL). Slit-lamp examination revealed Kayser-Fleischer ring and the diagnosis of Wilson Disease was established. **Discussion:** This report highlights that excessive smile can be the major clue to Wilson Disease (WD) at initial presentation. The association of this sign with depression, bulbar symptoms, bradykinesia and unstable gait in a young patient lead to the hypothesis of WD that was confirmed with laboratorial tests and ophthalmological examination. Abnormal face expression is a common presentation of dystonia in WD, but can vary in its appearance, ranging from classical presentations to more unspecific cases, where a smile can lead to a differential diagnosis with psychiatric diseases or pseudobulbar palsy. The classic manifestations of facial dystonia in WD are “risus sardonius”, a forced smile due to dystonic spasm of the risorius muscle, and “vacuous smile”, an open-mouth smile due to dystonic dropped jaw. Excessive smiling, elicited to trivial stimuli is also described. Recognize these signs is important, not only for diagnosis, but also because it interferes with eating, speech and increase mortality of the patients, needing correct management. **Final Comments:** Identify the myriad of signs and symptoms of WD is essential, once it is a potentially treatable disorder, especially if early diagnosed. The facial manifestations include “risus sardonius”, “vacuous smile” or excessive smile. In the context of a young patient and associated with suggestive symptoms, these signs must lead to the appropriate investigation.

Transtornos do Movimento

TL 1105740

TETRABENAZINE USE IN TREATMENT OF MOVEMENT DISORDERS IN ANTI-NMDAR ENCEPHALITIS – A CASE PRESENTATION

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Case Presentation: 14-year-old female presented with acute behavioral changes, confusion, excessive worry and short-term memory impairment. Later on, she exhibited manual automatisms, fever and seizures. CSF exam showed 75 cells with lymphomonomorphic predominance and normal glucose. Intravenous acyclovir was prescribed. She was intubated after an episode of bronchoaspiration and still showed continuous ocular, facial and limb dyskinesias and dystonic posture, tachycardia and fever after sedation. She was diagnosed with encephalitis anti-NMDA based on typical clinical presentation and positive CSF antibodies. Brain MRI with mild hypersignal in FLAIR and contrast enhancement in encephalic cortical sulci. FDG-PET scan showed diffuse asymmetry of cerebral metabolic activity, with increased metabolism in the basal frontal cortex specially on the right hemisphere and marked deficit of metabolism in the bilateral occipital cortex. Electroencephalogram showed extreme delta brush pattern, without epileptic paroxysms. Tumor screening with pelvic MRI and full body pet scan was negative. Treatment with methylprednisolone 1g/daily for 5 days and intravenous immunoglobulin (IVIG) was initiated. Dyskinesias, dystonic posture and dysautonomia persisted. There was no symptomatic response to oral benzodiazepines, ketamine or clonidine relying on IV benzodiazepines. Initiation of tetrabenazine made significant improvement of movement disorders, which resurface upon drug half-time clearance. **Discussion:** Anti-NMDAR encephalitis is characterized by limbic encephalitis, fever, seizures and dysautonomia. Movement disorders are present in 75% of young children and 95% of adults, appearing as oral, facial or lingual dyskinesias, dystonia, chorea, athetosis, myorhythmia, opisthotonus, ballismus, blepharospasm and oculogyric crisis. Immunotherapy generally improves this movement disorders. Benzodiazepines, valproate, haloperidol, ketamine and propofol have been reported for symptomatic treatment for refractory cases. Tetrabenazine is a reversible inhibitor of the human vesicular monoamine transporter type 2 (VMAT-2) with scarce data about use in anti-NMDA encephalitis. The case presented presented time-related and dose-related improvement of dyskinesias and dystonia with tetrabenazine. **Final Comments:** Tetrabenazine is shown as a possible alternative symptomatic treatment to high doses of benzodiazepines with less adverse effects in movement disorders of anti-NMDAR encephalitis.

Transtornos do Movimento

TL 1105744

ATYPICAL PRESENTATION OF THE CHOREA ASSOCIATED WITH PRIME DIAGNOSIS OF DIABETES: A CASE PRESENTATION

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Case Presentation: A 71-year-old female patient presents with choric movements in the left hemibody that has initiated 3 years before after a hyperglycemic episode diagnosed as Diabetes Mellitus (DM). This diagnosis of DM happened after an episode of impaired strength of the lower limbs, mental disorder and whole body paresthesia, that has led her to seek for an emergency servicer where it was diagnosed the hyperglycemic condition (522mg/l). Head CT scan and the other laboratorial exams showed no changes. She was Hospitalized for one week and after a glycemic control she maintained outpatient treatment with metformin. Despite of the treatment of the high serum glucosis, the patient maintained choric movement lasting more than three years. She used Aripiprazole, that was switched to Risperidona and after that it was noticed significant improvement of chorea. **Discussion:** The chorea is a hyperkinetic movement disturb presenting rapid and unpredicted muscle contractions. It affects mainly the distal portion of the limbs, besides the face and the torso and can be classified as hereditary, as in Huntington Disease, or acquired, when associated with other etiologies as traumatic brain injuries, drugs and metabolic changes (hyperglycemia among others). According to literature, metabolic features appears as second main etiology of acute chorea, present in 30% of all cases. Generally there is slow remission of the chorea after glycemic control and abnormal movements may last for more than one year. **Final Comments:** This case shows the association between an acute choric event after hyperglycemia that remains even after glycemic stabilization, reinforcing the non-remission of the symptoms acutely after the etiological treatment. Furthermore, it is relevant to perform an adequate etiological investigation in order to have early recognition of the cause and prompt treatment avoiding additional complications.

Transtornos do Movimento

TL 1105764

WILSON DISEASE AFTER A TRAUMATIC INJURY: A CASE PRESENTATION

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Case Presentation: A 23-years-old male suffered traumatic injury at 2019, he was beaten until he passed out needing to go to an emergency room and after a month presented a symmetric, progressive and slowly weakness in lower limbs, movement disorders in hands and, posteriorly, dysphagia, dysarthria and loss of trunk control, being restricted to bed after 3 years of the disease onset. Parental consanguinity (1th degree cousins) and similar family history negative. Had abuse of marihuana, cocaine and crack. On physical examination, alert, thin, exophoria (left eye), symmetric reduced lift of soft palate, dysarthria and dysphagia. Symmetric cogwheel hypertonia on upper limbs and spasticity (Ashworth 1) on lower limbs. Force-MRC V. Present axial face reflexes, normal osteotendinous reflexes and bilateral Babinski sign. Exhaustible Achilles Clonus. Apraxic and ataxic gait. Index-nose maneuver with bilateral movement decomposition. Dystonia and myoclonus in hands were presented. Eyes inspection with dark brown halo on irises. Brain MRI revealed supra and infratentorial atrophy, hypersignal foci (T2-weighted sequence) in frontal periventricular white matter, subinsular, claustrum, cerebral peduncles, midbrain tegmentum, pons and middle cerebellar peduncles, diffusion restriction on splenium of corpus callosum and hemoglobin breakdown products on caudate and lentiform nucleus. Laboratory showed reduced ceruloplasmin level (5mg/dL) and elevated 24 hour urinary copper (239.7µg/24h). Liver ultrasound showed steatosis and eye evaluation confirmed Kayser-Fleischer ring. Thus, it was consistent with Wilson's disease (WD) and was started D-penicillamine with good tolerance. **Discussion:** WD is a autosomal recessive inheritance disease due a mutation on ATP7B gene on chromosome 1, leading to an accumulation of copper in the body, including the central nervous system. Triggers are not usually found, but recently has been described WD onset after traumatic injury. In our opinion, it is difficult to separate the direct effects of trauma from those resulting from neurodegenerative processes, but some degenerative diseases are being initiated after trauma, without a clear explanation. **Final Comments:** The recognition of this relationship between traumatic injury and WD onset is still poorly described. We alert that early diagnostic recognition and treatment for WD, is essential for the best outcome of the patient, even with unlikely triggers and diagnostic confounders.

Transtornos do Movimento

TL 1105770

ACUTE DYSKINESIA AND STROKE IN A PATIENT WITH THYMIC CARCINOMA

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Case Presentation: An 80-year-old woman under chemotherapy for thymic carcinoma and no history of movement disorder (MD) or antipsychotic use evolved with fever and focal seizure at home. Once admitted at emergency room, she presented a secondarily generalized seizure that led her to orotracheal intubation. Phenytoin was started in parallel to piperacillin/ tazobactam (for pneumonia) and acyclovir (for viral encephalitis). She was extubated and upon evaluation on 3 rd day she was torporous, obeyed simple commands despite plegic right hand and had orolingual dyskinesia and choreic movements of lower limbs. MRI disclosed left frontal leptomeningeal hypersignal with frontoparietal cortical hypersignal and restricted DWI in hand motor area. CSF revealed 373 cells (73% lymphomononuclear), 102mg/dL protein and no neoplastic cells. On 4 th day, CSF PCR and hemoculture were positive for *Listeria monocytogenes* and ampicillin was started. The hyperkinetic movements remitted in the 2 nd week of treatment but right hand remained plegic. Patient recovered conscious level but died after 3 months of internment. **Discussion:** Listeriosis incidence is around 3-6 cases/million/year globally, although the prevalence is higher in the elderly and immunocompromised patients, and the mortality rate is about 30% whether neurological involvement. In a Chinese systematic review, dyskinesia was mentioned only in 3 cases of 163 non-perinatal patients with listeriosis. In other review, just 10 from 139 MRI lesions showed basal ganglia affection, and no dyskinesia was cited. We report an elderly oncologic patient with meningoencephalitis and no clear radiological substrate for hyperkinetic MD that evolved with acute orolingual dyskinesias and LL chorea that remitted during ampicillin employment. Moreover, the ischemia endorses the role of meningoencephalitis in vasculitic thrombotic infarction. Differential diagnosis would be a paraneoplastic syndrome (PS). An example of possible PS is anti-NMDA encephalitis that may present with dyskinesia but is not associated to thymic neoplasms (TN). Besides, thymic carcinomas, unlike thymoma, is not related to PS and dyskinesia was solved after ampicillin use. **Final Comments:** We present a patient with thymic carcinoma undergoing chemotherapy who developed dyskinesia secondary to a neuroinfection. Despite MRI findings being isolated to frontal lobe, the MD allied to the cortical stroke suggest a diffuse vasculitis due to *Listeria meningoencephalitis*.

Transtornos do Movimento

TL 1105815

MACHADO-JOSEPH DISEASE AGGRAVATED BY HIV THERAPY ABANDONED AND COVID-19 INFECTION: CASE PRESENTATION

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Case Presentation: 45 years-old female, presented with incoordination and drunken gait for 15 years, with progressive worsen along the years. She has positive familiar history of spinocerebellar atrophy type 3 (SCA-3) with positive genetic test in father and sister. Her genetic test for SCA-3 was also positive. After multiple pharmacological attempts, reached the symptomatic control with risperidone, baclofen, buspiron and clonazepam, restoring her functionality for daily basic activities. On September 2021, she has evolved with worsen of the ataxic condition and weight loss, even with regular medication use, becoming bedridden. Thus, she was admitted for investigation and clinical compensation. On neurological exam alert, communicative, thin, horizontal gaze decomposition, severe dysphagia, severe high amplitude tremor in limbs and head, movement decomposition, dysmetria, dysdiadochokinesis, scant speech and positive Babinski torso-thigh test. She had symptoms of a respiratory infection, with laboratory positive for COVID-19 e HIV infections. She claimed to know of HIV infection since 2011 and abandoned the treatment nearly 12 months before the ataxic decompensation. Brain RMI ruled out opportunistic infection in CNS and showed moderate atrophy of pons, cerebellum and middle cerebellar peduncles, with T2/FLAIR hypersignal in the same location. After an initial improvement of the respiratory infection, she evolved with cardiorespiratory arrest in pulseless VT for 26 minutes, returned in coma vigil and died after a few weeks in the ICU. **Discussion:** The SCA 3 is the prototype of autosomic dominant spinocerebellar atrophies. It stems from the CAG expansion in the ATXN3 gene in chromosome 14. There is no specific treatment, and symptomatic management being performed. The decompensation is common in systemic abnormalities, such infection. **Final Comments:** We emphasize that atypical exacerbation of spinocerebellar atrophy disorder is linked principally to infections, and their control is essential in the prognosis and survival of patients with neurodegenerative diseases.

Transtornos do Movimento

TL 1105861

GPI DBS IN PATIENT WITH DYT28 ASSOCIATED WITH KLIPPEL-FEIL SYNDROME: A ONE YEAR FOLLOW-UP CASE PRESENTATION

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Case Presentation: A 14-year-old female with history of congenital scoliosis, Klippel-Feil Syndrome (KFS), Sprengel's anomaly and generalized dystonia status post bilateral Globus Pallidus Internus (GPI) Deep Brain Stimulation (DBS). Patient was diagnosed with congenital scoliosis, KFS and Sprengel's anomaly at age 1. At age 6, she developed focal left lower extremity dystonia, that progressed to trunk, both upper extremities and oromandibular region over the years. Patient was posteriorly diagnosed at age 11 with KMT2B-related dystonia (DYT28) associated to KFS. It was confirmed by genetic testing that revealed microdeletion on chromosome 19 between the 19q13.11q13.12 bands, consistent with the cytogenetic location of KMT2B (OMIM 606834). She was wheel-chair bound for most part of her day, presented with severe spasmodic dysphonia and responded partially to levodopa. She underwent bilateral GPI DBS at age 13 and returned to our clinic for a 1-year postoperative follow up, presenting a 22% improvement in the Burke-Fahn-Marsden Dystonia Rating Scale when compared to pre operative assessment, especially in neck, trunk and lower extremities. We also noticed a significant improvement on gait, which was reflected in a subjective improvement of functionality and quality of life (QoL). **Discussion:** Mutations in KMT2B were recently associated with an early-onset, progressive and often complicated dystonia (DYT28), commonly presenting with an initial lower-limb dystonia that progresses to generalized dystonia. Among generalized early-onset dystonias, DYT28 may account for up to 10% of the cases. DBS is indicated for patients with generalized or segmental dystonia, refractory to drug treatments. This surgery improves about 42% of the clinical conditions in DYT-KMT2B cases, significantly improving motor symptoms. KFS is a congenital syndrome marked by the fusion of two or more vertebrae. It may be associated with other disorders, but the co-occurrence of typical KMT2B-related dystonia has not been reported in association with KFS to our knowledge. This case was previously presented by the authors and now we present the post DBS follow-up. **Final Comments:** This is a patient status post GPI DBS as stated in the literature, there was a relevant improvement on patient's symptoms, especially on gait. More studies are necessary to better understand the benefits of this procedure and to get broader and more concrete results.

Transtornos do Movimento

TL 1105885

NBIA-5 WITH WDR45 GENE MUTATION – CLINICAL AND NEUROIMAGE FINDINGS

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Case Presentation: We report the case of a female 33 years old who developed seizures at eight months of age, being treated with anticonvulsants suspended at one year and eight months of life. In the past pathological history, she had delayed development, gait, and speech. At school, presented cognitive development delay without other motor alterations. At 32, she complained of sleep disorders and significant bilateral parkinsonism, treated as a Dandy-Walker variant with parkinsonism. Initially, she had an excellent response to levodopa. In the following months, she developed headaches, psychiatric involvement with depressive mood, and a worsening of the neurodegenerative condition with an intensification of parkinsonism, cervical dystonia, cognitive deterioration, and disturbances in sleep cycles. The Complete Exome sequencing identified a heterozygous mutation in the WDR45 gene, confirming the diagnosis of neurodegeneration with brain iron accumulation type 5 (NBIA5). The patient was submitted to a brain MRI that was considered normal. However, a posterior neuroimaging analysis revealed a hypersignal on T1 and hyposignal on T2 in the substantia nigra, typical findings of NBIA5. The patient still has progressive balance worsening, difficulty in gait, cognitive decline, parkinsonism, and cervical dystonia. **Discussion:** NBIA5 is a group of inherited neurodegenerative diseases characterized by iron deposition in the basal ganglia. Patients with mutations in the WDR45 gene are characterized by delayed development in early childhood, mainly in language and motor skills, followed by a period of stability and later neurodegenerative manifestations. As in the literature, the patient showed an excellent response to levodopa in the initial adulthood period, but these effects were of short duration. NBIA5 syndrome still lacks data in the literature on the subject. **Final Comments:** Based on the case presented and the literature review, it is suggested that NBIA5 be inserted as a differential diagnosis for neurological disorders in patients with developmental delay in childhood and parkinsonism in early adulthood. We present the typical neuroimage findings that must bring to mind this rare diagnosis.

Transtornos do Movimento

TL 1105909

CASE OF GENERALIZED MYOCLONIAS CAUSED BY SEVERE HYPONATREMIA INDUCED BY TOPIRAMATE

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Case Presentation: Female, 73 years old, rural worker, admitted to the movement disorders clinic for investigation of generalized and progressive myoclonus since 2016 with cognitive decline, and worsening 1 year ago. The movements started in the foot or hand and spreading to the other limbs, sparing the face. It started only in the left upper and lower limbs, but progressed to the right hemibody. The myoclonias lasted up to 3 days, including during sleep, but with decreased intensity. She had type 2 diabetes mellitus and systemic arterial hypertension and was using topiramate, chlorthalidone, valsartan-hydrochlorothiazide, metformin, biperiden, acetylsalicylic acid, and simvastatin. On neurological examination, she presented bilateral tabetic gait, inability to perform tandem gait, and bilateral anarthria. Dysmetria and decomposition in the index-nasal and index-finger in the upper limbs were also evident. Ideative and ideomotor apraxia and visuospatial dysfunction. During the investigation, severe hyponatremia (serum Na of 104) was found, and the patient became drowsy; intravenous NaCl 3% replacement was performed. The main diagnostic hypothesis was a syndrome of inadequate ADH secretion (SIADH) caused by topiramate. A liquor analysis revealed an acellular pattern, with normal glucose and protein. During the investigation of the myoclonus picture, the electroencephalogram (EEG) results were awake and drowsy without abnormalities and the magnetic resonance imaging (MRI) of the skull showed no relevant alterations. At clinic follow-up, after correction of hyponatremia and suspension of topiramate, there was improvement of movements. **Discussion:** SIADH has many etiologies, through physiological, pathological or drug alterations. In particular, oxcarbazepine and carbamazepine act in part by increasing sensitivity to ADH. Similarly, it was observed in a control study that older patients taking the anticonvulsants valproate, phenytoin, and topiramate had an increased risk of Hospitalization for hyponatremia, as observed in this case. Therefore, hyponatremia should be noted as a possible etiology of metabolic myoclonus, among others such as hepatic encephalopathy and uremia. **Final Comments:** Authors describe a rare case of SIADH caused by topiramate, a drug poorly correlated as a cause of hyponatremia in the medical literature. Hyponatremia resulting from this disorder can lead to severe consequences, such as movement disorders, which incapacitate for daily activities.

Transtornos do Movimento

TL 1106119

FUNCTIONAL TIC SIMILAR TO TOURETTE'S SYNDROME ASSOCIATED WITH DIGITAL MEDIA CONSUMPTION DURING THE COVID-19 PANDEMIC

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Case Presentation: An 15-years-old woman, started body movements six months before the admission in the neurology service, after emotional stress with acutely present complex and diverse movements. After one month, she started nonspecific vocalizations, such as whistling and coprolalia, which worsen with anxiety and presence of self- and hetero-aggressive movements. Such movements can be suggested and get worse if suppressed. She started risperidone and sertraline. Important consumption of digital media. Cranial MRI and electroencephalogram did not show any changes that would justify the condition by secondary causes. **Discussion:** Tics are sudden, rapid and repetitive movements or sounds that are preceded by an urge sensation and can be temporarily suppressed. Motor tics can be clonic, dystonic or tonic movements and vocal tics are projected mostly as irrelevant sounds. The tic can be considered complex with more elaborate movements like kicks, copropraxia, speech of words or phrases, with coprolalia and echolalia. Tourette's Syndrome (TS) occur mostly in the pediatric population and males. Characterized as motor tics and one or more verbal tics present at some point in the disease. The pandemic caused by Covid-19, there has been an worldwide increase in cases of people between 12 and 25 years old, mostly female, with a rapid onset and development of complex motor or vocal tic-like behaviors, causing emergency department visits, which was named of Functional Tic-like Behaviors (FTLBs). A despite the geographic diversity, most cases shared common denominators, such as exposure to specific content on online media platforms such as TikTok and YouTube. Associated with the content of TS and FTLBs. As main differences between TS and FTLBs is an absence of progression from initially simple to complex tics; less suppression capacity and presence of psychosocial stressors such as tension between parents or family members, intensified by the lockdown, in addition to a higher prevalence with anxiety disorder or depression. **Final Comments:** Therefore, attention to the symptoms presented and the chronology of the disease are important, in order to avoid the automatic diagnosis of TS in patients with motor and verbal tics and also consider the presence of a functional disorder. In these last cases, modifying inappropriate social media exposures, psychotherapy, and addressing psychiatric disorders such as anxiety and depression can all be considered.

Transtornos do Movimento

TL 1106127

NEUROLOGICAL WILSON DISEASE IN A YOUNG ADULT: A CASE PRESENTATION

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Case Presentation: A 28-years-old male, mixed race, bricklayer, residing in Paraisópolis, Minas Gerais State, Brazil. History of alcoholism and use of narcotics. On December 3rd, 2021, the patient suffered the first generalized clonic tonic seizure while sleeping. Magnetic resonance imaging (MRI) of the brain this month detected a brain tumor. Levetiracetam was orally initiated on December 21st. He was diagnosed with Covid 19 on February 03rd, 2022, but with a benign evolution. The wife noticed from February 9th speech and gait disturbance, difficulty mobilizing the right hand, and urinary incontinence. On February 16th, the anticonvulsant was switched to Phenytoin orally and associated with Dexamethasone orally. Cerebrospinal fluid (CSF) on March 21th revealed a cell count of 0 units; glucose 89 mg/dL, lactate 15.7 mg/dL, gram without staining bacteria, and CSF culture without bacterial growth. He was Hospitalized on March 24th for social reasons. The patient presented to the neurological examination risus sardonius and right-hand flexor dystonia and ataxic gait. Magnetic resonance imaging (MRI) of the brain on April 01st, 2022 showed bilateral and symmetrical involvement of the basal ganglia and crus posterius, in addition to the midbrain and pons, and cerebellar peduncles. T2 FLAIR demonstrates a focus on diffusion restriction without significant expansive effect or paramagnetic contrast enhancement. Laboratory workup on March 24th revealed total serum copper of 24.6 mg/dL, serum ceruloplasmin of 7.0 mg/dL, and, finally, a 24-hours urine copper test of 187.4 mg/dL which confirmed the diagnosis of Wilson disease. Penicillamine was started at low dose, however, he is evolving with neurological worsening. **Discussion:** Incipient neurological symptoms are subtle and nonspecific and begin on average between 20 and 40 years. Neurological impairment occurs about a decade after liver failure and, therefore, are signs of advanced disease. Clinical worsening may occur after starting Penicillamine. **Final Comments:** It is a rare disease whose epidemiology in Brazil lacks data and complementary tests have reduced specificity. Diagnosis at an advanced stage of the disease compromises the neurological prognosis.

Transtornos do Movimento

TL 1106176

LATE CLINICAL MANIFESTATION OF DANDY-WALKER SYNDROME: CASE PRESENTATION

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Case Presentation: The Dandy-Walker malformation was described by Virchow in 1863 and Fusari in 1891. However it was Dandy and Taggart and Walker who described the clinical and radiological elements necessary for the perfect characterization of the syndrome. This is an embryonic defect in the posterior fossa with absence from of the 4th ventricle, agenesis of the lower part of the cerebellar vermis, very high situation of the lateral sinus and cerebellum tent, and dilation of the ventricular system. A case of late manifestation of the disease is reported. Female patient, 44 years old, complaining of insidious impairment of evolution of more than 15 years with progressive ataxia, dysarthria, scant speech, incoordination. Patient with incomplete higher education. **Discussion:** Dandy-Walker Syndrome is a rare clinical and radiological disease that most commonly is the subject of study in the pediatric age group. However, this case is presented with a patient who arrived at the university until the last year of advocacy. Because the cerebellum is a vital part of movement, balance and body coordination, many people with Dandy-Walker syndrome have problems with their voluntary muscle movements and coordination. They may also experience difficulties with their motor skills, mood, and behavior, and may have limited intellectual development. About half of people with Dandy-Walker syndrome have a below-average IQ. Despite the classic symptoms, the 44-year-old G.M.O. patient was evaluated with a complaint of insidious ataxia, cognitive impairment, nystagmus, incoordination and scant speech. He dropped out of college a few months before completing it. It observed losses in the performance of its domestic activities. When, already in his fifth decade of life, he sought medical assistance. Magnetic resonance imaging examination of the brain, at the time of the first visits in December 2019, revealed Dandy-Walker variant in the posterior fossa with limited development of the cerebellar vermis, right and left side of the cerebellum, enlargement of the fourth ventricle, development of a large, cyst-like formation at the base of the skull, where the brain stem and cerebellum are located. **Final Comments:** For most people, the signs and symptoms of Dandy-Walker syndrome are evident at birth or in the first year, but 10 to 20% of individuals may not develop symptoms until late childhood or early adulthood.

Transtornos do Movimento

TL 1106184

BROWNELL-OPPENHEIMER VARIANT OF SPORADIC CREUTZFELDT-JAKOB DISEASE: A CASE PRESENTATION IN RECIFE

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Case Presentation: 61 years-old female, farmer, presented with progressive weight loss (nearly 20 kg in 3 months), ataxic gait, imbalance and falls in October 2020, that associated along 3 months with symmetric tremor in upper limbs, visual hallucinations (with animals), behavior change, persecutory delusion, incoordination, aphasia non-fluent, urinary incontinence and dysphasia. In the end of this period, she became confined to bed and her family sought medical care, being transferred to a tertiary neurology service to investigate encephalitis of unknown etiology. Relatives deny infectious symptoms before the disease onset neither similar cases with closed relatives. On physical examination, alert, no interaction with the environment, pupillary reflex slowed down, cogwheel hypertonia and spasticity in upper limbs, associated with Gegenhalten paratonia. Presence of axial face reflexes, osteotendinous reflexes increased and indifferent cutaneous-plantar reflex. Myoclonic tremor (postural and Kinect) in upper limbs and head. Startle reflex. Laboratory without abnormalities. Cerebral MRI showed hypersignal in frontal cortex, caudate nucleus head, putamen, medial wall of the III ventricle and pulvinar of the thalamus (hockey stick sign), with DWI restriction. EEG showed diffuse disorganization of the base rhythm. CSF showed mild pleocytosis, with normal glucose and protein. With the exclusion of the most common causes of encephalitis, the main diagnostic hypothesis was Creutzfeldt-Jakob disease (CJD). Thus, 14-3-3 protein was measured in the CSF; with positive result. After about a month of Hospitalization, the patient died of aspiration pneumonia. **Discussion:** CJD is the prototype of prion diseases, with rapid progression and leading to death in less than a year of onset the disease. The Brownell and Oppenheimer form starts with cerebellar ataxia and dementia during the course of the disease. EEG findings of this variant usually reveal absence of periodic sharp-wave complexes. Most cases are sporadic, as in the case presented, but acquired and genetic forms have already been described. It's an exclusion diagnosis and there is no specific treatment. **Final Comments:** Prion disease must always be considered in case of rapidly progressive dementia without defined diagnosis, especially when associated to myoclonus and eventually, ataxia. We emphasize that early diagnosis and identification of people in contact with the patient is essential to prevent the spread of the disease.

Transtornos do Movimento

TL 1106236

ACUTE CHOREA ASSOCIATED WITH THYROTOXICOSIS

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Case Presentation: A 30 year-old with a three-month history of progressive generalized hyperkinetics movement. She had a prior history of severe anxiety, followed by a diagnosis of depression with psychotic symptoms. The condition persisted even after medical prescription of antipsychotics. Due to significant clinical deterioration and loss of functionality, the family sought neurological care. On admission, was agitated, hypervigilant, disoriented, tearful, feverish, tachycardic and presenting generalized choreoathetotic movement. She also had exophthalmos. After much questioning, family members reported that one year prior to admission, she also had unmeasured hyperthermia episodes, sweating, hair loss, asthenia, significant weight loss and low amplitude tremor of the extremities. ECG showed atrial fibrillation. Burch and Wartofsky index scored 75 points. Initial exams revealed TSH: < 0.01 and T4L > 6. Thyrotoxic crisis was diagnosed and the patient was treated with propylthiouracil, beta-blockers, and corticosteroids. She also received low doses of haloperidol and clonazepam to symptomatic control of chorea. Subsequent examinations showed TRAB > 40. There were positive rheumatologic markers, but no target organ damage, so immunosuppressants were not performed. The patient experienced substantial and progressive improvement of choreic movements after normalization of thyroid function. At the follow-up evaluation choreic movements had completely disappeared. **Discussion:** We report a case of a young female patient, with no previous comorbidities, who presented to the Hospital with generalized choreic movements due to thyrotoxicosis. She was further diagnosed with Graves disease. Chorea is described as a very rare manifestation, present in less than 2% cases of thyrotoxicosis. Mechanism of chorea in these cases is still poorly established. Previous reported cases suggest that it might be related to dopamine effect on basal ganglia, and to functional modification of adrenergic receptors. Association with autoimmunity has also been described. **Final Comments:** In conclusion, we enforce the importance of considering hyperthyroidism as a main differential diagnosis of acute chorea. This etiology is identifiable using simple laboratory tests and has a good response to drug treatment. Further studies are needed to elucidate pathophysiology of chorea related to hyperthyroidism and autoimmunity

Transtornos do Movimento

TL 1106247

STATUS DYSTONICUS IN WILSON DISEASE

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Case Presentation: We describe a young woman, born from consanguineous parents, who presented at the age of 23 years with a 9-month past of anxiety and depression and a 2-month history of left upper limb dystonia. She progressed rapidly and severe generalized dystonia. She was admitted to the emergency room with status dystonicus (video). Laboratory investigation showed elevated creatine kinase, hepatic dysfunction, and low serum level of ceruloplasmin. Brain MRI demonstrated a bilateral T2-W hyperintensity in basal ganglia, pons, and midbrain (figure). Whole-exome sequencing revealed a homozygous pathogenic variant in the ATP7B gene, c.547C>T: p.(Arg183Trp). Initially, management of dystonic status was performed with orotracheal intubation, high doses of intravenous benzodiazepine associated with baclofen, amantadine, and gabapentin via enteral tube. Regarding a poor response to the therapy instituted so far, it was decided to implement intermittent intrathecal baclofen, but the treatment had to be interrupted due to infection of the central nervous system. Therefore, we chose to perform unilateral craniotomy as an adjuvant treatment of status dystonicus. **Discussion:** Wilson disease (OMIM #277900) is an autosomal recessive inherited disorder of copper metabolism, caused by a mutation in the copper-transporting gene ATP7B. The disease can manifest as chronic liver disease, a progressive neurologic illness, or a psychiatric disorder. Movement disorder is one of the main symptoms of neurologic manifestation and dystonia is a common feature of WD. Dystonia can be focal, multifocal, segmental, or generalized. Status dystonicus is rare and is commonly associated d-penicillamine introduction, but Teive et al suggest that this presentation may be part of the natural history of this disorder. **Final Comments:** We report a case of Wilson disease with a rare presentation of status dystonicus without any treatment introduction.

Transtornos do Movimento

TL 1106277

SPORADIC ADULT-ONSET ATAXIA IN A PATIENT WITH PLATYBASIA AND BASILAR INVAGINATION

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Case Presentation: A 77-year-old man, white, born in Alagoas, with a negative family history of neurological disease, came to our observation complaining of gait disturbances since 2020. His medical history included systemic arterial hypertension, a depressive disorder and benign prostatic hyperplasia. He reported that his complaints began after a fall from a height in 1982, when he suffered head trauma and that resulted in dizziness with progressive worsening associated with gait imbalance. Physical examination revealed enlargement of the anteroposterior skull diameter and downbeat nystagmus, ataxia, intention tremor, and wide-based gait. MRI showed evidence of platybasia characterized by a Welcker basal angle of 149.1°. The tip of the odontoid process is located approximately 19 mm from the Chamberlain line, indicating basilar invagination (BI). **Discussion:** Platybasia, defined by a Welcker basilar angle greater than 140°, is the flattening of the skull base and may occur as a single finding or in association with other forms of skull base or craniovertebral deformity. Basilar impression (secondary/acquired) and BI (primary) occur when the mastoid process of C2 invades the foramen magnum and causes compression of the brainstem and upper cervical spine. The top of the mastoid process typically lies below the Chamberlain line (from the hard palate to the opisthion) and is abnormal if it protrudes more than 5 mm above this line. Platybasia alone does not usually cause symptoms unless it is associated with BI. When it is present, the main symptoms are paraparesis, quadriparesis, ataxia, nystagmus, dysphagia and dyspnea. The cause of BI is more commonly associated with congenital anomalies such as Klippel-Feil syndrome. Basilar impression is more commonly associated with conditions that cause softening of the bone, including Paget's disease, hyperparathyroidism and osteogenesis imperfecta. Remarkably, trauma was a precipitating factor for symptoms in patients with BI, as in our patient. BI is a progressively debilitating disease that can lead to severe neurologic deficits and death if left untreated. Treatment is usually surgical and includes differential decompression followed by posterior stabilization. **Final Comments:** Our goal is to emphasize the importance of careful clinical observation, in this case, the apparent platybasia and the evaluation of the characteristics of this rare disease, allowing early diagnosis and offering the best possible treatment.

Transtornos do Movimento

TL 1106278

CAPN1 MUTATION ASSOCIATED WITH SPASTIC PARAPLEGIA, A RARE PROGRESSIVE GAIT DISORDER WITH FEW REPORTS IN LITERATURE

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Case Presentation: Two young patients attended our clinic presenting difficulties to walk at the same age, 30 years old, with progressive worsening. A woman, with a history of two ischemic strokes, the first at 24, and later with 40 years of age, struggles to walk at 30 years of age, associated with weakness in the lower limbs, hypertonia, signs of pyramidal release and ataxia. Neuroimaging showed only an area of previous ischemic stroke at the right frontoparietoinular region. She had no similar family history or consanguinity but is of German descent. She improved symptoms with thiozidine. The second case, a man, started the symptoms with difficulty in walking, progressively worsening, associated with dysarthria, global spasticity, ataxia, significant postural instability and club foot. Neuroimaging showed atrophy of the brainstem and cerebellum. He had no pathological antecedents and had Portuguese ancestry. The man has family members with confirmed disease, a sister who started symptoms at 20 years old (homozygous) and another asymptomatic sister (heterozygous). Both cases were diagnosed after genetic testing, confirming the CAPN1 mutation. **Discussion:** CAPN1 is a gene involved in protein coding and related to nervous system development and the integrin pathway, also associated with autophagy-lysosomal pathway in cerebral ischemia scenario. Structural alterations in this gene are associated with Autosomal Recessive Spastic Paraplegia 76, spasticity and possibly with neuronal damage secondary to impairment in autophagic flux during a cerebral ischemia. Autosomal recessive spastic paraplegia spastic type 76 usually begins in adulthood with slow progression, leading to spasticity and hyperreflexia, as well as dysarthria. Other findings that may be found are foot deformities (clubfoot), plantar extension reflex and ataxia. **Final Comments:** The finding of gait disturbances with progressive worsening in youth or early adulthood should raise as a suspicion, among other etiologies, of genetic diseases. Often seen as sporadic forms of spastic paraplegia, only with genetic testing is a correct diagnosis possible.

Transtornos do Movimento

TL 1106375

PAROXYSMAL NON-KINESIGENIC DYSKINESIA (PNKD): CASE PRESENTATION

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Case Presentation: MCD, 4 months. Parents report that at 2 months of age, their child has been presenting dystonic episodes characterized by forced semiflexion of the left upper limb, elevation and hyperextension of the right upper limb (posture in swordsmanship), accompanied by hypertonia of the lower limbs. During episodes, the patient presents pain, facial redness and groaning, and at the end of the crisis, she demonstrates crying and irritability. Episodes are usually triggered when placed in the supine position and during the child's sleep. Its beginning was concomitant with the use of orthosis for aesthetic correction of positional plagiocephaly at 2 months of age. Exome and EEG without alterations. Female, born at 38 weeks and 5 days, vaginally, without gestational or neonatal complications. A 30-year-old father has a history of right temporal lobe epilepsy with spontaneous remission with oxcarbazepine use. Autistic maternal cousin – with tetrasomy of chromosome 15. At 4 months of age, oxcarbazepine 0.1 ml twice a day was started – increasing weekly to 0.5 ml 12/12h – 10 mg/kg/day. The patient evolved within a month with an 80% decrease in the number of events with oxcarbazepine use. **Discussion:** PNKD is part of a range of presentations of dyskinetic movement disorders. The typical presentation of this condition involves involuntary movements, resulting from sustained or intermittent muscle contractions. Triggers that can start an attack are caffeine use, alcohol use, stress, or a specific position. Because it is a rare condition, it is poorly studied and its pathophysiology is still not understood. Theories involve disinhibition of certain brain regions, such as the basal ganglia, inadequate GABAergic response in regions related to fine motor skills, structural changes in the brain, and are often associated with genetic alterations. There is debate as to whether these types of dyskinesia can be categorized as having an epileptogenic etiology, since in most cases there is no electroencephalographic alteration. Treatment usually consists of the use of anticonvulsants, with carbamazepine being the drug of choice. **Final Comments:** The patient had an early onset of dyskinetic condition which is triggered by the supine position. There is a family history of epilepsy and autism, but the exome did not find known alterations. Laboratory tests and EEG without changes. The good response to the use of oxcarbazepine guided the therapy in this case.

Transtornos do Movimento

TL 1106467

PROPRIOSPINAL MYOCLONUS POST-SARS-COV-2 INFECTION AND EXPOSITION TO COVID-19 VACCINATION: CAUSAL RELATIONSHIP OR COINCIDENCE?

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Case Presentation: A previously healthy 34-year-old man presented with episodes of focal spasms affecting the right shoulder and causing right-sided flexion of the trunk along with abdominal muscle contractions evolving with progressive worsening for 20 days. He observed that the abnormal movements increase with stressful events and disappear when he was sleeping. Thirty days before, he presented flu symptoms associated with anosmia and ageusia that recovered in seven days. He did not search for medical assistance and received Aztrazeneca COVID-19 vaccination 15 days after improvement. Magnetic resonance images of the spinal cord, laboratory tests including autoantibodies, and neoplastic screening were unremarkable. SARS-CoV-2 IgG antibodies were positive. Electromyography showed motor unit potential bursts between 50-150ms without rhythmic pattern indicating myoclonus in the right-side trunk flexor musculature that changed with body positioning. During polymyography, the electroencephalogram showed no epileptic paroxysms before or after myoclonus, and the Bereitschafts evoked potential was not obtained. The patient received benzodiazepines associated with anticonvulsants and improved. **Discussion:** The clinical features and the electrodiagnosis were indicative of propriospinal myoclonus, which is a rare condition characterized by repetitive, often arrhythmic spasms in the flexor muscles of the trunk, hips, and knees following a regular pattern. It is possible that a spinal cord generator causes this kind of abnormal movement. Diagnosis is challenging due to the possibility of a functional movement disorder, elevated frequency of normal images, and a broad range of triggers (such as viral infections and drugs). There are no biomarkers for this disorder. Among all post-COVID-19 movement disorders previously published, two-thirds are reported as myoclonus, including one case associated with mRNA vaccination. It is possible that they occur due to either neuronal hyperexcitability or post-viral autoimmune syndrome. There is one case presentation of propriospinal myoclonus post-SARS-CoV-2 infection, but not after any COVID-19 vaccination. **Final Comments:** Propriospinal myoclonus is a rare movement disorder. Relationship with immune triggers such as SARS-Cov-2 infection or COVID-19 vaccines may contribute to the pathophysiological knowledge of the disorder, especially when there are no other usual causes.

Transtornos do Movimento

TL 1106473

NEUROACANTHOCYTOSIS WITH PSYCHIATRIC ONSET: A CASE PRESENTATION

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A 12 years old female, previously healthy, presented progressive neuropsychiatric symptoms, characterized by psychotic behavior. Antipsychotic medications were started and patient had complications of its uses, like malignant neuroleptic syndrome extrapyramidal syndrome. Evolved with choreoathetotic movements disorders and postural instability, being referred to the neurological department in a tertiary Hospital. First neurological evaluation showed cognitive impairment, with jeopardize in comprehension, temporo-spatial disorientation, bizarre behavior (allotriophagy), aggressive posture, bradykinesia and rigidity. Tardive dyskinesia and Schizophrenia were the first hypothesis due to prolonged use of neuroleptic medications (10 years). No improvement of symptoms were observed, albeit optimized therapeutic drugs were prescribed. Further investigation was made, with brain MRI showing only hippocampal atrophy. Serum copper level and ceruloplasmin were normal. Peripheral blood analysis showed acanthocytes. **Discussion:** Neuroacanthocytosis is a rare, late onset genetic disorder in which progressive neurological decline associates with acanthocytes (spiculated red blood cells) in the blood, being prevalent in less than one case per one million. The family history, in most cases, is positive, with the main cause being abetalipoproteinemia, an autosomal recessive disease. Common signs and symptoms besides chorea are stereotypic gestures, epileptic seizures, cognitive deficit, amyotrophy, absence of osteotendinous reflexes, high serum creatine kinase level, feeding dystonia, and self-mutilation by biting the lips and tongue. Neuroacanthocytosis was attributed to several mutations in the CHAC (chorea-acanthocytosis) gene, which was later renamed as VPS13A and mapped on chromosome 9q21.2. **Final Comments:** Studies and research on Neuroacanthocytosis are scarce and, when present, old. This condition is still poorly reported in the medical literature, and is therefore underdiagnosed. There is a need for further research on the subject in order to improve the diagnosis and treatment of these patients.

Transtornos do Movimento

TL 1106555

PALIDOTOMY IN PARKINSON'S SURGERY: IS THERE STILL A PLACE?

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Case Presentation: Patient, 48 years old, female, resident and born in Ibiara-PB, presented in 2011 symptoms of movement disorder with tremor at rest in the right upper limb, being diagnosed with Parkinson's Disease (PD). In 2020, she had worsening symptoms with disabling dyskinesias due to levodopa therapy. Pallidotomy was indicated with instant improvement of symptoms and no side effects. **Discussion:** PD is a chronic, progressive and degenerative disease of the Central Nervous System (CNS), with different clinical manifestations, resulting from the deficit in the production of dopamine by the neurons of the substantia nigra. The most effective pharmacological therapy is with levodopa, aimed at maintaining normal levels of dopamine in the CNS. However, its long-term use can cause adverse effects, such as fluctuations in motor response and the development of involuntary movements, classified as levodopa-induced dyskinesia. Currently, two surgical methods are used for the treatment of dyskinesia: pallidotomy with ablation of the internal globus pallidus and deep brain stimulation (DBS), the latter being the most used because neuromodulation is reversible and adjustable. However, the choice of method must be based on careful clinical analysis and socioeconomic conditions. This patient lives in a small city in the interior of the state of Paraíba, about 500km from the capital, and has difficulty in transport for clinical follow-up. She had disabling dyskinesia for more than 50% of the day. The treatment indicated was the ablative technique (pallidotomy) because it is the one that suits the patient's profile, as it is safe, low cost, effective and does not require adjustments such as neuromodulation. After surgery, dyskinesia totally regressed (UPDRS-part III pre: 65; 3 years later: 27). **Final Comments:** The therapeutic management of PD is based on a holistic view of the patient, which involves, in addition to their clinic, their socioeconomic conditions. Pallidotomy is a surgical procedure that offers excellent results, being indicated for patients who have difficulty in maintaining clinical follow-up due to inaccessibility to large centers.

Transtornos do Movimento

TL 1105381

TRAUMATIC PNEUMORRHACHIS: A CASE PRESENTATION IGOR FORTUNATO DA SILVA, FERNANDA GONÇALVES DE ANDRADE, FERNANDA AQUINO DE OLIVEIRA, HYASMIN MARIA OLIVEIRA CARVALHO

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Case Presentation: male, 25 years old, illicit drug user, victim of a beating, complaining of difficulty moving the left lower limb (LLL) after a stab wound in the right lateral dorsal region at the level of the thoracolumbar transition. He was conscious and oriented, incomplete deficit in the LLL with strength grade 4 in hip flexion, grade 2 in leg flexion and extension, grade 4 in plantar dorsiflexion and grade 5 in plantar flexion; Left patellar reflex absent and decreased Achilles deficit, partial sensory deficit in L1 to L4 dermatomes on the left, without motor or sensory deficits on the right. Computed tomography (CT) of the spine shows subarachnoid pneumorrhachis between T12 and L1, without fracture. Magnetic Resonance (MRI) shows an indolent lesion and contrast uptake with a trajectory starting in the skin on the right, right paravertebral obliquely in the midline at the level of the L1 direction, signs of rupture and direction of the T12-L1 ligamentum flavum and foci of hypersignal in the neural roots close to the left conus medullaris, compatible with traumatic radiculopathy. There were no signs of meningitis. In the outpatient follow-up after 1 month, the patient was able to walk without assistance, with partial recovery in some patients. **Discussion:** Pneumorrhachis was described for the first time in 1977, and it can have different causes (iatrogenic, after neurosurgery, spontaneous, infectious, etc.), with a traumatic cause being a rare condition. Its diagnosis is mainly made by CT. In the presence of neurological deficits and with a history of trauma, even without identification of spinal fractures, an MRI investigation should be followed to better determine the lesion. The neurological examination is always essential for the correct topographical diagnosis and for the appropriate request of complementary exams. The case in question had a difficult initial correlation between the right back injury and the left lower limb deficit. **Final Comments:** A careful examination of the back in search of injuries is part of polytrauma care. Even without obvious external signs, or unlikely mechanisms, there may be spinal injuries that are not initially identified. The topographic diagnosis of neurological injuries is often challenging.

Traumatismo cranioencefálico

TL 1105481

CHRONIC EPIDURAL HEMATOMA WITH ATYPICAL EVOLUTION POST TRAUMATIC BRAIN INJURY: CASE PRESENTATION

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Case Presentation: Patient, male, 27 years old, with hemiparesis and grade 4 motor strength – on the right, with a history of severe traumatic brain injury (TBI) and neurosurgical approach with left decompressive craniectomy and later cranioplasty, at age 19. A control cranial tomography revealed a passive epidural hematoma of resorption, which was chosen for a conservative approach. During the subsequent 7 years, there was no clinical worsening, but with gradual growth of the hematoma evidenced on CT. In 2022, CT showed a heterogeneous left frontoparietotemporal formation, predominantly hyperattenuating, measuring 8.7 x 8.6 x 6.3, with a midline deviation of 1.8 cm, dilatation of the ventricular system and CSF transudation, suggestive of Chronic Epidural Hematoma (CEH). A reapproach was decided, with preoperative MRI showing a lesion measuring 9.3 x 9.3 x 7.2 cm. During the procedure, the presence of fibrosis in the dura mater was noted, as well as a heterogeneous hematoma with no apparent origin of the bleeding, with an aspect of “hard clay” next to the “motor oil” aspect. In the postoperative period, the patient evolved with a bulging wound. Surgery and exteriorization of serous secretion, without exudate or foci of bleeding, with a diagnostic hypothesis of dural cerebrospinal fluid fistula. **Discussion:** The precise definition of CEH is not clearly defined, it is known, however, that it is more common in young men, associated with TBI and the source of bleeding is usually venous. Most reports state that only 15% of patients did not present clinical signs suggestive of CEH, as in this case. It is understood that a cerebrospinal fluid redistribution in the lateral ventricles could allow the creation of space for the hematoma to grow, not leading to clinical involvement, which may be the anatomopathological basis of this report. In addition, most cases have a mean time between CT and diagnosis. months, with few descriptions with more than years after the TBI, in this case the evolution of 8 years. In addition, the literature describes that CEH is isodense, corroborating the predominantly liquid presentation of CEH on surgical inspection, but with little association with the “hard clay” appearance. **Final Comments:** We present an CEH with a poorly documented chronological evolution and whose pathophysiology has not yet been elucidated. Its heterogeneous appearance, together with postoperative evolution, raises the diagnostic hypothesis of a dural CSF fistula.

Traumatismo cranioencefálico

TL 1106054

TRAUMATIC RETROCLIVAL TRAUMATIC SUBDURAL HEMATOMA DUE TO GUNSHOT WOUND TO THE SKULL: A CASE PRESENTATION AND LITERATURE REVIEW

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Case Presentation: A 48-year-old male patient was admitted to the emergency room with a gunshot wound to the skull. At baseline, he had 14 points on the Glasgow Coma Scale (GCS), left foot paresis, and two open gunshot wounds to the frontal bone. Computed tomography (CT) revealed projectile fragments entering the right frontal region, traveling in the left parietal and frontotemporal regions and lodged in the left frontoparietal region. A subdural hematoma was observed posterior to the clival ramp, with greater volume on the right, with a thickness of up to 0.6 cm. He underwent frontal craniotomy with debridement and duraplasty. Subdural HRC was treated conservatively. Postoperatively, he had a 15-point GCS and persistence of grade 1 paresis in the left foot. **Discussion:** Retroclival hematomas (RCH) are rare, representing 0.3% of acute extra-axial hematomas, and form a subgroup of extra-axial posterior fossa hemorrhage. They can be classified as subdural, blood up to the lower border of C3 and absence of blood anterior to the transverse ligament, or extradural, presence of blood loss at the attachment of the tectal membrane to the body of C2. The classification relates to neurological deficits, subsequent complications, and management of the patient's conditions. The most common clinical presentation is unilateral or bilateral abducens nerve palsy, as well as severe headache simulating subarachnoid hemorrhage and nerve involvement by direct compression or contusion. Most reports are epidural hematomas associated with traumatic brain injury in pediatric patients. We found only 31 reported cases of subdural RCH in adults, none of which were caused by a firearm. **Final Comments:** Traumatic posterior fossa hematomas are particularly rare conditions. The conservative approach with immobilization is sufficient in most cases.

Traumatismo cranioencefálico



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TL 1104711

THE INTERSECTION BETWEEN CEREBELLAR ATAXIA AND NEUROPATHY: A PROPOSED CLASSIFICATION AND A DIAGNOSTIC APPROACH

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Background: Neuropathy is a common associated feature of different types of genetic or sporadic cerebellar ataxias. The pattern of peripheral nerve involvement and its associated clinical features can be an invaluable aspect for narrowing the etiologic diagnosis in the investigation of cerebellar ataxias. **Objective:** In this review, we discuss the differential diagnosis of the intersection between peripheral nerve and cerebellar involvement, and classify them in accordance with the predominant features. Genetics, clinical features, neuroimaging, and neurophysiologic characteristics are discussed. Furthermore, a diagnostic approach for cerebellar ataxia with neuropathy is proposed according to the different clinical characteristics. **Methods:** This is an Educational and Descriptive review with the aim of medical education for the approach to the patients with cerebellar ataxia and neuropathy. **Conclusion:** The diagnostic approach to the patient with cerebellar ataxia with neuropathy requires a detailed medical history, phenotyping, characterization of disease progression and family history. Neuroimaging features and the neurophysiological findings play pivotal roles in defining the diagnosis. Establishing an organized classification method for the disorders based on the clinical features may be very helpful, and could be divided as those with predominant cerebellar features, predominant neuropathic feature, or conditions with both cerebellar ataxia and neuropathy. Second, determining the mode of inheritance is critical on cerebellar ataxias: autosomal dominant and recessive cerebellar ataxias, mitochondrial or sporadic types. Third, one must carefully assess neurophysiologic findings in order to better characterize the predominant pattern of involvement: damage location, mechanism of lesion (axonal or demyelinating), motor, sensory or sensory motor compromise, large or small fibers, and autonomic system abnormalities.

Ataxias

TL 1105070

SPINOCEREBELLAR ATAXIAS IN THE STATE OF ALAGOAS

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Background: Spinocerebellar ataxias (SCAs) are a group of autosomal dominant ataxias characterized by dysfunction of the cerebellum and its connections besides additional manifestations, such as pyramidal and extrapyramidal signs, peripheral neuropathy, visual impairment and epilepsy. SCAs have an average worldwide prevalence of 2.7 cases per 100,000 individuals and great ethnic and geographic variability. In Brazil, epidemiological studies are based on data obtained mostly at neurogenetic reference centers in south and southeast regions and some regions are still underrepresented. **Objective:** The present study aimed to identify and characterize SCAs in the state of Alagoas in order to contribute to the epidemiological and clinical profile of SCAs and to stimulate the development of strategies to detect people with SCA in regions without epidemiological data. **Methods:** From June 2018 to August 2021, we carried out active search strategies for people with a diagnosis or clinical suspicion of SCA and applied a clinical and molecular evaluation protocol that included clinical interview, heredogram construction, neurological exam, ataxia and functional scales and molecular analysis for SCAs type 1, 3, 6, 7, 8, 10, 12, 17 and DPRLA. **Results:** As a result, 73 people with SCA were identified, configuring a minimum estimated prevalence of 2.17 cases per 100,000 inhabitants, distributed as 75.3% patients with SCA3, 15.1% with SCA7, 6.8% with SCA1 and 2.7% with SCA2. SCA3 and SCA7 analysis showed age at onset and clinical features congruent with other studies; predominance of SCA3 subphenotype 2; gait alterations as the main initial manifestation and, in people with SCA7, accompanied by reduced visual capacity; inverse correlation between age at onset and number of expanded CAG; positive correlation between severity and disability scales and duration of illness; and that most people affected are not engaged in a rehabilitation program and there are large numbers of asymptomatic people at risk of developing SCA. **Conclusion:** Therefore, this study show that simple collaborative strategies can enhance the detection capacity of SCA families and demonstrate that Alagoas has a minimum estimated prevalence of SCA similar to the world average prevalence.

Ataxias

TL 1105166

SPINOCEREBELLAR ATAXIA TYPE 6 IS NOT A PURE CEREBELLAR DISEASE: DATA FROM SPINAL CORD IMAGING

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Background: Spinocerebellar ataxia type 6 (SCA6) is a rare autosomal dominantly inherited disorder caused by abnormal CAG trinucleotide repeat expansions in the CACNA1A gene (19p13.13). SCA6 is considered a prototype of pure cerebellar ataxia, with late onset and slowly progressive course, but extracerebellar findings have been described. The pathological hallmark of SCA6 is cerebellum and inferior olive atrophy, but degeneration has also been found on brainstem nuclei and spinal cord in autopsy studies. Previous MRI studies found cerebellar damage in SCA6 cohorts, but little is known regarding spinal cord imaging changes in these patients. **Objective:** To assess spinal cord involvement through MRI and its clinical correlates in a SCA6 cohort. **Methods:** We enrolled 17 SCA6 patients with genetic confirmation, but 4 patients were excluded due to compressive myelopathy. They underwent clinical evaluation, including SARA, INAS and ICARS scales, and MRI examination on a 3T Phillips scanner at the same day. Thirteen age and sex-matched healthy controls were also assessed. The Spinal Cord Toolbox (SCT) was employed to measure spinal cord area and eccentricity in C1-T2 levels and white matter tracts integrity by assessing diffusion parameters, which are fractional anisotropy (FA), axial diffusivity (AD), radial diffusivity (RD) and mean diffusivity (MD), in C2-C5 levels. Imaging data were compared between SCA6 and controls groups through a Mann-Whitney U test, employing age and sex as covariates. Bonferroni correction for multiple comparisons was performed. Correlations between imaging and clinical data were assessed through Spearman's rank correlation coefficients. **Results:** Mean age of the patients and disease duration were 68,1±7,0 and 15,3±6,9 years, respectively. Mean SARA score was 15,0±6,9. SCA6 group had increased AD in left tectospinal tract (p=0.023), left lateral vestibulospinal tract (p=0.027) and left ventral corticospinal tract (p=0.045), increased MD in right lateral vestibulospinal tract (p=0.032) and left tectospinal tract (p=0.019), and increased RD in right lateral vestibulospinal tract (p=0.032). These MRI parameters did not correlate with clinical data but, in an exploratory approach, left ventral corticospinal tract RD were directly related to INAS scale (p=0.011, r=0.699). **Conclusion:** These findings indicate that not only the cerebellum, but also the spinal cord is involved in SCA6. Quantitative spinal cord MRI might be a useful biomarker in SCA6.

Ataxias

TL 1105284

NGIROESTRIATAL DYSFUNCTION IS FREQUENT IN RFC-1 RELATED DISORDER

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Background: RFC1/CANVAS syndrome is a rare adult-onset hereditodegenerative disorder caused by biallelic AAAAG RFC1 expansions and characterized primarily by cerebellar ataxia, sensory neuronopathy (ganglionopathy), as well as bilateral vestibular hypofunction. Parkinsonism is now recognized as an additional feature in the disease, however no systematic evaluation of nigrostriatal dopaminergic function has been published so far. **Objective:** To describe striatal dopamine transporter (DAT) density in a sample of RFC1/CANVAS patients using dopaminergic transporter brain scintigraphy with 99mTc-TRODAT-1 (DAT imaging). **Methods:** This is an observational, single-center study conducted in a tertiary referral Hospital, which analyzed 10 patients with molecular confirmation of RFC1/CANVAS. Disease severity was assessed with the SARA scale. Each subject was carefully evaluated for the presence of parkinsonian features. DAT imaging was acquired and reconstructed in the transverse, coronal and sagittal planes 4 hours after venous injection of 99mTc-TRODAT-1. An experienced nuclear physician performed the visual analysis of all images. Quantification was performed drawing region of interest (ROI) in the striatum, putamen and caudate bilaterally. The specific uptake (mean uptake ratio) of each region was calculated using occipital region uptake as the reference. **Results:** Patients had a mean age of 63 ± 9.2 years, and there were 7 women. The mean SARA score was 16.9 ± 5.8 . Seven patients had abnormal DAT imaging results, characterized by reduced uptake in left putamen (100%), right putamen (85.7%), left caudate (71.4%) and right caudate (71.4%). Four patients (57.1%) had bilateral striatum abnormalities. Parkinsonism was noticed in 3/7 patients, all of which had abnormal DAT scans. Interestingly, four subjects had reduced DAT imaging uptake, but no clinical signs of parkinsonism. **Conclusion:** Nigrostriatal dysfunction is frequent in RFC1/CANVAS, suggesting that disease pathology is more widespread than previously thought. Additionally, the fact that patients without parkinsonism also had abnormal DAT imaging results might suggest that DAT imaging changes occur early along the disease course. These results have practical therapeutic relevance and suggest that dopaminergic agents might be useful at least for a subgroup of patients with RFC1-related disorder.

Ataxias

TL 1105785

ATROPHY OF CRANIAL NERVES V AND VIII IS A SPECIFIC NEURORADIOLOGICAL SIGN OF RFC1-RELATED DISORDER

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Background: The Cerebellar Ataxia, Neuropathy, and Vestibular Areflexia Syndrome (CANVAS) related to RFC1 is an important cause of late-onset progressive ataxia. In clinical practice, it may be difficult to differentiate it from other degenerative ataxias, such as spinocerebellar ataxias (SCA) and multiple system atrophy cerebellar type (MSA-C) due to its clinical interface. Thinning of cranial nerves V and VIII has been lately reported in MRI scans of patients with RFC1-related disorder, but its diagnostic specificity is not yet clear. **Objective:** To assess the usefulness of cranial nerves V and VIII imaging to differentiate RFC1-related disorder from SCA and MSA-C. **Methods:** Thirty-seven patients with late-onset ataxia were enrolled, including 9 individuals with RFC1-related disorder, 23 with SCAs (types 2,3 and 6) and 5 with MSA-C defined by clinical criteria. Nine healthy controls were also assessed. All subjects underwent MRI scans on a 3T Philips scanner and clinical evaluation on the same day. We acquired axial T2-weighted MRI with thin slices (balance sequence) for qualitative assessment of V and VIII cranial nerves. Images were reviewed by a neuroradiologist, blinded to patient and clinical data, to classify these nerves as atrophic or normal. We analyzed the frequencies of combined and isolated atrophy of nerves V and VIII for each subject. These frequencies were compared between groups using Fisher exact test. Level of significance was set at 0.05. **Results:** Mean age of patients with RFC1-related disorder, SCA and MSA were 66.7 ± 7.3 , 56.3 ± 16.6 and 64.4 ± 13.2 years, respectively. Atrophy of cranial nerves V (CNV) and VIII (CNVIII) were significantly more frequent in the RFC1 group when compared to SCA (CNV: $p < 0.001$; CNVIII: $p < 0.001$), MSA (CNV: $p < 0.001$; CNVIII: $p = 0.008$) and controls (CNV: $p < 0.001$; CNVIII: $p = 0.008$). In an exploratory sub-analysis assessing each SCA subtype, simultaneous atrophy of both nerves was also more frequent in the CANVAS group than in SCA2 ($p = 0.035$), SCA3 ($p = 0.015$) and SCA6 ($p = 0.009$) subgroups. **Conclusion:** MRI evaluation of cranial nerves V and VIII using a dedicated sequence is an easy-to-use tool that helps to distinguish RFC1-related disorder from other late-onset degenerative ataxias. This new neuroradiological sign should be sought in the routine evaluation of ataxias.

Ataxias

TL 1106076

NUTRITIONAL ASSESSMENT OF PATIENTS WITH HEREDITARY ATAXIAS IN NORTHEAST BRAZIL: A CASE-CONTROL STUDY

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Background: Hereditary Ataxias (HAs) comprise a wide spectrum of genetically determined neurodegenerative diseases with progressive ataxia as the main symptom. Few studies have evaluated nutritional profile in HA patients. **Objective:** To evaluate eating habits and nutritional status of a population of patients with HA in Northeastern Brazil. We also analyzed the association of these findings with clinical and genetic profiles of this population. **Methods:** We performed an observational case-control study of 2 reference centers in neurogenetic diseases in Fortaleza, a city located in Northeast Brazil. Body composition was assessed with bio-impedance analysis and dietary intake was estimated with a validated questionnaire (24-hour dietary recall). **Results:** The final sample consisted of 76 participants, 38 patients with hereditary ataxias and 38 controls. Mean body mass index (BMI) was lower in HA compared to controls ($p = 0.032$). Hereditary ataxia patients showed lower protein intake, higher frequency of dysphagia and higher incidence of nausea and diarrhea. The difference in average estimated caloric intake did not reach statistical significance. Disease severity measured by SARA scale was not associated with BMI, nor was ataxia subtype (autosomal dominant x non-autosomal dominant ataxias). **Conclusion:** Hereditary ataxia patients have lower BMI compared to healthy controls. There was no difference in this cohort between dominant or non-dominant ataxia regarding BMI. Weight loss may be a common finding among hereditary ataxias and may affect quality of life in these patients.

Ataxias

TL 1106564

THE IMPORTANCE TEVALUATION OF QUALITY OF LIFE WITH THE USE OF VIRTUAL REALITY IN PATIENTS WITH SPINOCEREBELLAR ATAXIA

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Background: Spinocerebellar ataxias (SCAs) are a heterogeneous group of neurodegenerative diseases, featuring the presence of progressive cerebellar ataxia, whose early clinical symptoms are balance and motor coordination decline and ocular disorders. **Objective:** To evaluate the living standard of patients with SCAs, by applying the Vestibular Disorders Activities of Daily Living Scale (VADL) and Activities-specific Balance Confidence Scale (ABC) questionnaires, before and after rehabilitation with virtual reality (ViR). **Methods:** An uncontrolled clinical trial study was conducted with 28 patients suffering from SCA (mean of 41.6 years, \pm of 16.9 years), underwent anamnesis, otolaryngologic examination, vestibular screening, and the application of protocols applied before and after rehabilitation with ViR. **Result:** The most reported otoneurological complaints in the anamnesis were: imbalance (85.7%), fall (28.5%), dizziness (17.8%), diplopia (10.7%), and tremor (7.1%); The vestibular exam was altered in 64.3% of the cases; The comparison **Results:** between the evaluation time (T1, T2 and T3) and the dimensions (functional, locomotion and instrumental) from the VADL did not show significant differences (1st assessment before the start of rehabilitation designated T1, after 10 rehabilitation sessions, considered T2 and, at the end of 20 rehabilitation sessions, called T3); The results from the ABC revealed that there was not significant differences when compared with the evaluation time (T1, T2 and T3); The result between the correlation of the VADL and ABC questionnaires showed significant **Results:** in all cases. The correlation between the ages and disease length with the VADL and ABC questionnaires was significant between the age and the instrumental dimension of the VADL questionnaire, in the T3 assessment. The correlation between the disease length and the VADL questionnaire was significant in all cases; The comparison of the ViR result (T1 to T2) showed significant difference for all of the applied games, except for the ski slalom. The comparison of the ViR result (T1 to T3) showed significant difference for all of the applied games. **Conclusion:** We can point out a direct improvement of the living standard, reflected by the reduction of falls, better balance and march, contributing to a higher self-confidence in patients in daily activities. We also point out the importance of ViR as the symptomatology in this kind of population improved.

Ataxias

TL 1106656

TRANSLATION, CROSS?CULTURAL ADAPTATION, AND VALIDATION TO BRAZILIAN PORTUGUESE OF THE CEREBELLAR COGNITIVE AFFECTIVE/ SCHMAHMANN SYNDROME SCALE

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Background: The Cerebellar Cognitive Affective Syndrome (CCAS) is characterized by deficits in executive functions, language processing, spatial orientation, and affect regulation in patients with cerebellar disease. The symptoms can occur isolated or along with motor and coordination symptoms. **Objective:** The aim of our study was to translate and culturally adapt the CCAS scale to Brazilian Portuguese and validate the scale in our population. **Methods:** We performed a cross-sectional study with patients with primary and secondary ataxia. The study included 111 individuals, aged between 20 and 80 years, of both genders, 20 without cognitive and/or affective complaints who participated in the pre-test phase, 40 with cerebellar disease (hereditary/neurodegenerative ataxia or acquired/secondary cerebellar ataxia), and 51 healthy controls with no evidence of cognitive impairment and no affective symptoms matched for sex, age, and educational level. The scale was translated, culturally adapted, and validated. Statistical analysis of the data was performed, with association tests, mean comparison, and ROC curve analysis. **Results:** In the association test, the items Semantic Fluency ($P = >0.999$) and Phonemic Fluency ($P = 0.386$) did not present a statistically significant difference when comparing the group of patients to the control group. The scale's internal reliability, assessed by Cronbach's alpha, was 0.752, demonstrating a good degree of reliability and consistency between the scale's subitems. Based on the analysis of the ROC Curve, optimal cutoff values were found for each subitem of the scale. **Conclusion:** The translated and adapted scale has good internal consistency, is reproducible, has good reliability, and has the potential to be a reliable tool for screening cognitive symptoms in patients with cerebellar disease.

Ataxias

TL 1104999

DIALYSIS-RELATED HEADACHE: PREVALENCE AND CLINICAL FEATURES IN HEMODIALYSIS PATIENTS AND AFTER KIDNEY TRANSPLANTATION

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Background: Headache is a common symptom, occurring in up to 70% of patients on hemodialysis. Dialysis headache is defined by the International Classification of Headache Disorders (ICHD-3) as a headache with no specific characteristics, occurring during and caused by hemodialysis, and which resolves spontaneously within 72 hours after the session has ended. There is no consensus on its pathophysiology or triggering factors. **Objective:** To evaluate the prevalence, clinical features and factors associated with dialysis headache in patients with chronic kidney disease on renal replacement therapy. **Methods:** Cross-sectional observational study with a quantitative approach. Participants were divided into two groups: (1) 25 patients on hemodialysis and (2) 25 patients on early post kidney transplantation. All participants were interviewed with a structured questionnaire, the Hospital Anxiety and Depression Scale, and the Epworth Sleepiness Scale. Blood pressure, weight, urea, glucose and electrolytes were evaluated before and after one hemodialysis session. Control groups were classified from the headache-free patients in each group. **Results:** In group (1), 8 patients (32%) were diagnosed with dialysis headache, predominantly pulsatile ($n=6$, 75%), accompanied by photophobia ($n=6$, 75%), phonophobia ($n=4$, 50%) and nausea or vomiting ($n=6$, 75%), with a mean pain score of 7.75 ± 1.58 . Individuals with headache had higher scores for anxiety (7.00 ± 3.93 vs. 3.82 ± 3.23 , $p=0.03$) and sleepiness (9.13 ± 3.94 vs. 4.76 ± 3.85 , $p=0.01$). Dialysis headache was associated with lower pre-dialysis serum calcium ($p=0.01$), higher pre-dialysis systolic ($p=0.02$) and diastolic ($p=0.02$) blood pressure. There was no correlation between headache and variations in serum urea levels. In group (2), 5 patients (20%) were diagnosed with dialysis headache, predominantly pulsatile ($n=4$, 80%), accompanied by nausea or vomiting ($n=4$, 80%), aggravated by routine physical activity ($n=3$, 60%), with a mean pain score of 8 ± 1.41 . Individuals with headache had higher sleepiness scores (9.20 ± 4.32 vs. 4.80 ± 4.51 , $p=0.029$) and younger age (38.93 ± 14.43 vs. 54.02 ± 8.31 , $p=0.03$). **Conclusion:** Headache is common among patients on hemodialysis, it has migraine-like features, and it is associated with increased anxiety, increased sleepiness, and higher pre-dialysis blood pressure levels.

Cefaleia

TL 1105053

TELEMEDICINE ON THE TREATMENT OF CHRONIC HEADACHE: A SYSTEMATIC REVIEW

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Background: Chronic headaches, such as migraines and tension-type headaches, are among the most common disorders in the world, but access to specialists for adequate follow-up is not yet feasible in all locations, especially in rural areas. Telemedicine/teleconsultation is a form of evaluation that can be used for several neurological disorders, including headaches, allowing a greater reach of health care. **Objective:** our goal was to evaluate the effectiveness of telemedicine/teleconsultation visits in the treatment of chronic headaches compared to a traditional visits. **Method:** A systematic review of literature, using PubMed, MEDLINE, Cochrane, Lilacs, and Scielo databases, yielded 262 articles, of which 9 studies were included for analysis. **Results:** a total of 1,834 patients were evaluated, mostly involving women and with migraines sufferers. They also included children, adolescents, and adults. Four studies were conducted during the coronavirus pandemic. All studies showed a high patient satisfaction rate with teleconsultation, whether adults or children with their families. It also showed a tendency to reduce the number of headache-days per month, the use of analgesics, as well as the frequency and duration of pain. **Conclusion:** telemedicine was found to be an effective and viable form of follow-up for patients with chronic headaches.

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TL 1105302

THE EFFICACY OF GREATER OCCIPITAL NERVE BLOCK FOR THE TREATMENT OF MIGRAINE: A SUBJECTIVE EVALUATION AND REPORT OF 31 PATIENTS

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Background: Migraine is the most common type of primary headache in outpatient clinics, even in the emergency room. With their debilitating crises that often end up invalidating the individual in their full work activity. An example of a treatment that has been extensively studied today is the occipital nerve block, which has shown to be quite promising. The greater occipital nerve (GON) is responsible for the cutaneous sensory innervation of the occipital and suboccipital regions. Recent research has shown that it also actively participates in brain nociceptive modulation, acting directly, together with the trigeminal nerve, on secondary nociceptive neurons located at the level of the brainstem. **Objectives:** Evaluate the efficacy of GON-block in the treatment of migraine. **Method:** The model of this study was a survey research, involving patients diagnosed with migraine. These patients were treated with GON-block. The instrument of data collection comprehends a questionnaire made through telephone calls, and was prepared with the aid of two sources: the medical records of the reference unit and relevant data from other research in the area. **Results:** The sample size presented a number (n) of 31 participants diagnosed with migraine, with a predominance of female patients (93.5%, n = 29). Concerning the age, the average was 38 years. In the survey, 93.5% (n = 29) reported pain relief after the procedure. Among these, they were asked to rate verbally their average pain relief on a scale ranging from 0 to 10. The average on the headache relief scale was 7.5, and 77.4% (n = 24) reported a relief greater than 6 points. The onset time for symptom relief was less than 48 hours for 45.1% (n = 14), with an average duration of 54 days. In the sample, 25.8% (n = 8) reported side effects, that include dizziness, holocranial headache and edema. Finally, 37 participants answered that they would do the procedure again. **Conclusion:** Greater occipital nerve block has proven to be an effective treatment method when properly indicated. However, further randomized controlled trials are needed to better assess the evidence of efficacy in the treatment of migraine.

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TL 1105305

USE OF MONOCLONAL ANTIBODIES IN PATIENTS WITH MIGRAINE: SUBJECTIVE ASSESSMENT OF USE AND IMPROVEMENT WITH THESE DRUGS INTRODUCTION MIGRAINE IS ONE OF THE MOST FREQUENT DISEASES IN NEUROLOGICAL CLINICAL PRACTICE, WITH ECONOMIC AND SOCIAL EFFECTS FOR PAT

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Background: Migraine is one of the most frequent diseases in neurological clinical practice, with economic and social effects for patients and the health system. The disease treatment is costly and those who suffer from migraine are affected in multiple domains. The prophylactic treatment uses antidepressants, anticonvulsants, triptans, among other drugs. They are safe drugs but have low efficacy, after all, 35% of patients - treated with Triptans - overgoing randomized clinical trials did not improve, and 40% of them experienced side effects. In an attempt to chance the outcome of the scenario, alternative therapies, such as immunological therapies, with use of Monoclonal Antibodies (mAb) such as Ajovy, Emigality and Pasurta, intend to improve the perspectives for patients and direct the investments of the health system in efficient treatments. **Objective:** To carry out a subjective evaluation with patients treated at the Proneuro Clinic, in the city of Cascavel-PR, who used mAb for the treatment of migraine, verifying if there was an improvement in the clinical condition with the new therapy. **Method:** This is a subjective assessment with the application of a questionnaire, carried out through calls, clarifying relevant information about the outcomes of immune therapy. **Results:** The sample consisted of 68 individuals diagnosed with migraine, treated by the same neurologist, who were instructed to use mAb. 34 did not answer the calls, 14 never underwent the treatment due to the high cost and 20 used mAb and answered the questionnaire. The results show that 70% of the collaborating patients resorted to use the immunological therapy because the previous treatments did not present good efficacy. 30% made the substitution due to side effects such as paresthesia, xerostomia and dizziness. Also, it was found that 70% of patients who used mAb had a reduction in the frequency of migraine attacks and 85% reported relief in intensity. However, 30% pointed out that the frequency remained the same and the intensity in 15% of them also did not change. **Conclusion:** The result with the mAb has a statistically significant effect when compared to conventional treatment. However, low adherence, due to the high cost, prevents the therapy from being applied globally so that the disease can have reasonable control in the majority of the population.

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TL 1105490

BOTULINUM TOXIN AS A PREVENTIVE TREATMENT FOR MIGRAINES

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Background: Migraine is a disease that affects about 12% of the world population. This headache can be characterized as intense and often debilitating, resulting in impairments in work capacity, in social life and generating large expenses for health services. As a result, prophylactic treatments for chronic migraines began to gain greater value and adherence among those affected by this disease. Among the wide range of possible treatments, botulinum toxin has been gaining prominence in the preventive treatment of chronic migraines for more than a decade, although its mechanism of action is gradually being understood. **Objective:** To systematically evaluate advances in the use of botulinum toxin as a preventive treatment for migraine, considering improvements in quality of life and considering its cost-benefits for the patient. **Methods:** This is a systematic literature review based on data extraction in indexes such as PUBMED and MEDLINE. Publications from 2018 to 2022 were considered, in English and with the following descriptors: Botulinum Toxin, Migraine and Treatments. **Results:** Since the approval of botulinum toxin as a preventive treatment for migraine, the results of the various studies carried out have demonstrated its strong efficacy, as it leads to a significant decrease in the number of hours in crises that the patient was affected in the month. In addition, patients who used this preventive treatment showed a positive result after about 2 months of medication use and quickly showed improvement in quality of life, in work and social activities. Finally, the studies point to a continuity in the clinical improvement after treatment, since the effects of the toxin remained over time. **Conclusion:** Preventive treatments for chronic migraine seek to reduce the number of hours and amount of attacks per month that each patient is affected. From this perspective, botulinum toxin gains prominence for guaranteeing both possibilities, in addition to offering a lower cost than the new monoclonal medications. This presented relationship guarantees the outstanding local botulinum toxin in the field of preventive medicine for these situations.

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TL 1105499

MONOCLONAL ANTI-CGRP ANTIBODIES AS A TREATMENT FOR MIGRAINE

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Background: Migraine is a highly debilitating pathology capable of causing a loss of ability to work and social life. However, only about 15% of patients who need prophylactic follow-up do so properly, the vast majority due to absence/efficacy or as a result of the secondary effects of the prescribed medications. In recent years, the development of a new family of drugs, anti-calcitonin gene-related peptide (CGRP) monoclonal antibodies, paves the way for more effective treatment of migraine migraines. This drug is based on blocking the CGRP pathway of action, which tends to have its levels increased during crises. **Objective:** To systematically evaluate advances in the treatment of migraine based on the use of monoclonal antibodies related to the anti-calcitonin gene. **Methods:** This is a systematic literature review based on data extraction in indexes such as PUBMED and MEDLINE. Publications between 2018 and 2022, in English and with the following descriptors: CGRP, monoclonal antibodies and Migraine, were considered. **Results:** CGRP is a neuropeptide synthesized at multiple sites in the central and peripheral nervous system and its role was noted from the observation of its elevation during moments of crises and also in the induction of crises in patients who underwent amino acid injection. The evaluation of CGRP inhibition has emerged as an effective method of treating migraine and studies have shown a significant reduction in the number of attacks in patients who used medication. Thus, the primary goals of the treatments with monoclonal antibodies were significantly achieved, since the patients pointed to a reduction in the number of attacks in the month. **Conclusion:** Treatment with anti-CGRP monoclonal antibodies appears as a positive aspect in the treatment of migraine attacks and of course there are still some barriers to be faced, such as the high cost and the lack of research on long-term treatment or relapses. However, this new line of research managed to guarantee significant results in the treatment with a reduction in the number of seizures per month in the vast majority of patients and thus allowed an improvement in the quality of their personal and social life.

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TL 1105548

THE INFLUENCE OF THE USE OF PERSONAL PROTECTIVE EQUIPMENT ON PREVIOUS HEADACHES OF HEALTHCARE PROFESSIONALS EXPOSED TO THE COVID-19 PANDEMIC

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Background: As a result of the COVID-19 pandemic, the use of personal protective equipment (PPE) has become indispensable for the safety. However, the impact of PPE use on previously diagnosed headaches of healthcare professionals is uncertain. **Objective:** To analyze the influence of the use of PPE on previous headaches of health professionals exposed to the COVID-19 pandemic. **Method:** This was a non-interventional, quantitative, observational, and cross-sectional study. Thus, the Snowball method was used for recruitment and an online questionnaire was applied to the participants. In addition, the Kolmogorov-Smirnov test was used to define data normality and Fisher's exact test served to correlate categorical variables. Headaches were classified according to diagnostic criteria present in the International Classification of Headache Disorders (ICHD-3). **Results:** The study showed that 20.6% of the participants had headaches diagnosed before, of which 70% had migraine headaches and 30% had tension-type headaches. Previous headaches, which are more common in women, had a significant difference between genders ($p < 0.001$). The difference between the types of headaches presented was not significant ($p = 0.5698$). Most professionals (67.5%) had changes in the pain pattern when using PPE during the pandemic, of which 45% had migraine and 22.5% had tension-type headaches, with no statistically significant difference between the change in the pattern and the classification ($p = 0.7158$). The increased frequency of crises was the main alteration mentioned by 60% of the professionals with previous headaches, although there was no statistical significance ($p = 0.9899$). Furthermore, 40% of the professionals with previous headaches reported an increase in the duration of the crises, without statistical significance ($p = 0.4898$). The analysis of the participants' perception, who reported an increase in the frequency of crises, regarding the relationship between the types of PPE and this change showed statistical significance ($p < 0.040$) for the use of masks. There was also statistical significance in the perception of the relationship between increased crisis duration and each type of PPE ($p = 0.0402$). **Conclusion:** A change was observed in most patients who reported previous headaches, in the pattern of crises, in terms of pain and frequency, with the use of PPE during the pandemic. Most patients considered the association between the types of PPE and these changes as very likely.

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TL 1105653

PREVALENCE OF EXTERNAL COMPRESSION HEADACHE CAUSED BY USE OF PERSONAL PROTECTIVE EQUIPMENT IN HEALTHCARE WORKERS EXPOSED TO THE COVID-19 PANDEMIC

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Background: External compression headache is classified as a primary headache resulting from compression of the soft tissues lining the skull. Pain is caused when peripheral nociceptors are stimulated due to some tissue lesion. In this perspective, with the use of protective equipment covering the skull in times of the COVID-19 pandemic, it is evident the possibility of the development of headache in health professionals, due to the prolonged use of these accessories. **Objective:** To determine the prevalence of external compression headache caused by the use of face shields and other personal protective equipment (PPE) in healthcare workers exposed to the pandemic of COVID-19. **Methods:** This was a non-intervention, quantitative approach, observational, cross-sectional study. 202 participants completed an online questionnaire, after approval by the local Research Ethics Committee. Of these, 8 were excluded because they used a cloth mask. ICHD3 criteria were used to confirm the presence of pre-existing headache and external compression headache. Kolmogorov-Smirnov's test and Fisher's exact test were applied using SPSS software. **Results:** 1949 healthcare professionals were included in the sample, which comprised 71.6% (139) women and 28.4% (55) men. 79.4% (154) of the professionals were between 22-40 years of age and 20.6% (40) were between 41-64 years of age. It was observed that 20.6% (40) had headaches diagnosed before the pandemic. Headaches prevalence associated with PPE use was high (81%). Regarding new headaches, 62% (49) could be classified as external compression headache, with 34 women. When calculating the prevalence of external compression headache in relation to the study sample, a value of 25.2% was obtained. To 15 of the 49 professionals (30.6%), headache was not associated with any symptoms. Furthermore, the frequency of painful episodes was 4 to 7 days per month for most of them (32.6%). **Conclusion:** Prevalence of external compression headache caused by PPE use was high in the study population, reaffirming the clinical characteristics of previous studies, and may have associated symptoms. There was no significant difference between the types of PPE and the prevalence of the occurrence of external compression headache, and a change in the preexisting headache pattern was also reported.

Cefaleia

TL 1105696

THE ASSOCIATION BETWEEN HEADACHE AND ANXIETY DURING THE COVID-19 PANDEMIC IN THE SOUTHEAST REGION OF BRAZIL

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Background: Headaches have scientifically established triggers, among the psychosocial factors are of great medical importance. So, comorbidities like psychosocial stress and anxiety can act primary cephalgia. Due to changes in the social dynamics during COVID-19 pandemic, debates in social media and studies about mental disorders have been received a great scientific interest. **Objective:** The aim of this study is to investigate an association between headaches and anxiety during the COVID 19 pandemic, analyzing new and pre-existing scenarios – and therefore aggravated – of cephalus and mental disorders. **Methods:** This is a cross-sectional, retrospective and descriptive study with a quantitative approach. The data was collected through an online form, on GoogleDocs, "Headache Prevalences during the Pandemic" between september/2020 to january/2021. The inclusion criteria are: over 18 years of age, signing the Free Informed Consent Term (FICT) available online. Exclusion criteria were participants who did not fully complete the questionnaire or who live outside Brazil. To carry out the analysis of the variables, sociodemographic data were considered – sex, age, ethnicity, and place of residence; as well as clinical data – previous diagnosis of headache, symptoms or previous diagnosis of anxiety, the impact of social isolation with the headache, among others. Statistical procedures are descriptive measures, frequency tables, mean and standard deviation. **Results:** In the present study, 59.3% of the volunteers (162, of which 147 were from the Southeast) reported headaches. The most prevalent was pulsatile headache, affecting 56.17% of the participants. Regarding location of pain, most participants reported bilateral pain (69.13%) and frontal headache was the most prevalent (51.85%). It was found that 70.7% (193) of the participants state that there was a worsening of anxiety symptoms during the COVID-19 pandemic and 36.3% (99) report that they simultaneously observed worsening of headaches. In addition, stress was listed as an important headache trigger throughout the study. **Conclusion:** The present study indicates the relation between anxiety and headaches, besides an increase of anxiety during the COVID-19 pandemic. Thus, the correlation of these data proves, considering its prevalence and great impact on patients' quality of life, the importance of the Unified Health System (SUS) to study and prioritize headaches and mental disorders.

Cefaleia

TL 1105961

RELATION BETWEEN TENSIONAL HEADACHE AND GENERALIZED ANXIETY DISORDER

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Background: Headaches are considered an important public health problem and there are several psychological factors linked to the manifestation of this symptom that is related, especially to anxiety. With that in mind, it became essential to study the connection involving these two factors in an attempt to explain this phenomenon and how to proceed with the treatment of this patient. **Objective:** The objective of this study is to relate the incidence of tension-type headache in patients with Generalized Anxiety Disorder, as well as its manifestation. **Methods:** This is a systematic review, conducted on Scielo and Google Scholar platforms. Were selected articles in Portuguese, published in the last twenty years with the descriptors "stress", "headache" and "anxiety". **Results:** Tension-type headache refers to a mild to moderate pain that is usually described as a feeling of a tight band around the head. It is also the most common kind of headache presented in clinics, expressed specially by pericranial muscular spasm and important myofascial pain. Several factors can cause a tension headache, especially stressful situations that predispose to anxious episodes, like work problems, study issues, and financial trouble. In this sense, it is clear that individuals who present classic anxiety symptoms, such as muscle tension, fatigue and palpitation, usually also have headache as a form of manifestation of an anxious state like a way of "alert" of an anxiety crisis, mainly in individuals with Generalized Anxiety Disorder. **Conclusion:** Therefore, it is a possible to affirm that there is a strong connection between episodes of tension-type headache in individuals who manifest anxious episodes frequently. Thus, neglecting this association can result in an ineffective treatment and it could also contribute to a decrease in the patient's life quality.

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TL 1106049

PREVALENCE AND IMPACT OF MIGRAINE AMONG HEALTH SCIENCES STUDENTS: A CROSS-SECTIONAL STUDY

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Background: It has been shown that migraine has a significant presence and negative impact on students' quality of life and academic achievement. Health Sciences Students are exposed to specific types of risk factors that may contribute to the prevalence of migraine. **Objective:** This study aims to evaluate the presence and persistence of migraine in students of health services and its impacts. **Methods:** This cross-sectional study was carried out at the University of Caxias do Sul, Brazil. A web-based survey between the second semester of 2020 and the first semester of 2021 was sent to students of health sciences' course. The survey contained questions regarding demographics, lifestyle, headache aspects and the ID-Migraine. The ID-Migraine™ was a questionnaire proposed in 2003 as a way to identify migraine in outpatient primary care and was found to be a valid and reliable screening instrument. It was the tool chosen to assess the prevalence of Migraine among the students. A multivariate model was used to investigate potential associations with migraine and disability reported. **Results:** The survey was responded by 504 students. The majority (90.1%; N=379) of students had headaches in the last three months. According to the ID-Migraine, 71.9% (N=350/454) of the students presented with migraine. To manage their headaches, 80.8% (N=367/454) of the respondents had resorted to the use of pharmacotherapy. Only 20.9% (N=77/367) of them had these medications prescribed by a physician. There was no significant association ($p < 0.05$) regarding both functional impairment and migraine with semester, hours of work per day, time spent at the university per week and time spent in physical exercises per day. Hours of study were significant ($p < 0.05$) on functional impairment. **Conclusion:** Migraine prevalence among health Science students is high compared to other studies. The population studied used self-medication to treat their pain. There was no difference in functional impairment and migraine when we consider the year of graduation, hours of work per day, time spent at the university per week and time spent in physical exercises per day. Hours of study were an important triggering factor for pain.

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TL 1106139

IMPROVING EMERGENCY DEPARTMENT HEADACHE TREATMENT: THE VIRTUAL HEADACHE TRAINING PROGRAM (VHTP)

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Background: Headache is among the most common causes of visits to the Emergency Department (ED), and migraine is the most common identifiable diagnosis in this context. The diagnosis of migraine is clinical, and neuroimaging should not be performed routinely unless an alarm sign is identified in the history or physical examination. Currently, the first-line treatment of acute migraine attacks should be performed with analgesics, non-steroid anti-inflammatory drugs, triptans, and antidopaminergic medications. Second-line treatment includes corticoids, magnesium, and nerve blocks. Opioids should be avoided for the treatment of migraine attacks². Despite these recommendations, we identified a high use of opioids and a high rate of neuroimaging exams in the emergency department of a tertiary Hospital in Brazil. **Objective:** To evaluate the impact of a virtual headache training program (VHTP) for healthcare providers at the ED of a tertiary Hospital in improving headache treatment. **Method:** We evaluated preliminary data including all visits to the ED with the final diagnosis of migraine (CID G43) and unspecified headache (CID R51). The intervention consisted of an asynchronous virtual training program of 90 minutes, aimed to review the clinical presentation of the primary headaches, the alarm signs for secondary headaches, and an evidence-based treatment protocol for primary headaches. Data were collected for 14 weeks before and 12 weeks after intervention. The primary endpoint was the reduction in the rate of opioids prescription. Secondary endpoints were the reduction in neuroimaging exams in the ED, reduction of inpatient admission rate, and duration of inpatient treatment. **Results:** we included 87 headache visits in the pre-VHTP group, and 96 in the post-VHTP group. The opioid prescription for headache patients was reduced in 75.1% ($p = 0.005$). There was no significant difference of the secondary outcomes. Neuroimaging was performed for 55.4% vs 41.8% ($p = 0.16$), mean length of ED visit was 249.3 vs 167.3 minutes ($p = 0.23$), inpatient admission rate was 5.5% vs 0.8% ($p = 0.08$) and length of inpatient admission was 2 vs 1 day ($p = 0.32$). **Conclusion:** The Virtual Headache Training Program successfully improved headache treatment, reducing the opioid prescription by 75.1%. Limitations: This is a single-center retrospective study with preliminary data from a small number of cases and a short period of observation.

Cefaleia

TL 1106222

MIGRAINE HEADACHE ASSOCIATION WITH CARDIOVASCULAR AND CEREBROVASCULAR DISEASES

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Background: The migraine (MI) is one of the most common neurologic disorder, represented by recurring headache episodes associated with motor and somatosensory transitory disturbances, and is an important cause of incapacitation in the world. In this scenario, several studies propose the correlation between MI, specially with aura, and cerebrovascular diseases (CVAs) and cardiovascular diseases (CVDs), due to pathophysiological aspects in common. However, the mechanisms of association between these diseases are not well enlightened, which difficulties a better approach for the patients. The study aims to contribute for a better understanding of the association between migraine and CVAs and CVDs. This article presents a narrative review of the literature based on the analysis of scientific articles published from 2012 to 2022, in PubMed and Science Direct, on MI and its association with cardiovascular and cerebrovascular diseases. Two criterias were applied, titles that did not mention the thematic association between Migraine and CVDs/CVAs and abstracts that did not address the theme migraine and cardiovascular and cerebrovascular diseases. After the application of the exclusion criterias, was obtained in total, 50 articles remained. MI is a very frequent neurological disorder, being reported as one of the most disabling diseases in the world, affecting the quality of life and social functioning of the patients. It is characterized by a pathological brain state that alters the patterns of neuronal activity, which can be accompanied by the presence of aura, generating pain associated with several motor and somatosensory manifestations that appear both prior to the moment of headache and after the end of the pain crisis. In addition to these clinical associations, MI can be related to an increased risk of developing cardiovascular and cerebrovascular diseases, due to its path physiology related to endothelial dysfunction, cortex spreading depression and increased coagulability markers and the inflammatory response, which can result in vasospasms and consequent hypoperfusion of some regions. This study discussed the relation between MI, especially with aura, and cerebrovascular and cardiovascular pathologies. Overall, it was evidenced that there is an association of mechanisms between these disorders. In this context, there is an urgent need for more specific research and dissemination of data on the association of Migraine and vascular heart and neurological diseases.

Cefaleia

TL 1106224

OVERVIEW OF HOSPITALIZATIONS DUE TO HEADACHE IN BRAZIL FROM 2016 TO 2022

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Background: Headache is a common cause of medical consultation, not only in primary care but also in specialist neurology outpatient clinics. Despite most of Covid-19 symptoms being respiratory, there was an increase in symptoms in the central and peripheral nervous system, including anosmia, ageusia and headache, possibly via direct viral injury, immunological mechanisms and the angiotensin-converting enzyme 2 (ACE2) receptor, and hypoxia. It is estimated that with the COVID-19 pandemic there has been a five-fold increase in the incidence of headache in the affected regions. There has also been described a possible new daily persistent headache in the aftermath of COVID-19. Therefore, it is fundamental to look at the epidemiological profile of the most affected groups. **Methods:** This consists of an ecological and descriptive study of the Hospitalizations in Brazil, according to age and gender, from January 2016 to April 2022, for which data collection was obtained via the Health Information (TAB- NET) and the SUS Hospital Information System (SIH / SUS). **Results:** During the past 7 years in Brazil there were 61.324 admissions for headache, while the region with the highest prevalence was the Southeast, with a total of 18.910 cases (30,84%), and subsequently the Northeastern region, in which there were 18.346 cases (29,91%). The region with the lower incidence was the Central-West, with only 3.138 cases (5,11%). The age group with most cases was from 20 to 49 years, with the highest number of cases from 30 to 39 years, in both genders (11.457 cases, of which 7.910 were female). There was a predominance of females, representing 40.073 cases (65,35%). From 2016 to 2021, there was an increase of 4,64% in the number of Hospitalizations, with a progressive increase until 2019, the year with the highest incidence. **Conclusion:** Hence, women from 20-39 were the most affected, and the region with most Hospitalizations was Southeast. In this case, it is fundamental to focus on the availability of headache treatments especially in primary care, while also making policies and approaches with individualized multidisciplinary planning, psychological resources and universalization of the access to such treatments. It is also important to make further studies to assess the real incidence of such cases, even before the need of Hospitalization.

Cefaleia

TL 1106442

THE INCREASING NUMBER OF HEADACHES IN THE CONTEXT OF INFOXICATION AND IN THE POST-PANDEMIC WORLD

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Background: At the current Digital Age, access to information is constant, whether through large communication devices or through smartphones. In the context of the Covid-19 pandemic, this problem has gotten worse, with not only the disclosure of necessary data, but also the spread of misinformation, causing emotional and mental overload. Given this, an increase in anxious and depressive symptoms was noted. In addition, there were neurological symptoms, among which the most common is headache, with emphasis on primary ones. Such headaches occur without the usual clinical or laboratory tests showing any etiology, as in, for example, migraine. In these cases, brain neurochemical disorders occur, involving neurotransmitter imbalance. In this bias, genetic and environmental factors act in these disorders. The present study will address the environmental factor contribution. **Objective:** To associate the impacts of disproportionate information overload with the increase in the number of headaches in the post-pandemic context. **Methods:** This is a literary review, in which research published in both English and Portuguese between the years 2019-2022 on Scielo, Google Scholar and PubMed were used. The following descriptors were used: "Headache", "Information" and "Covid-19", with 5 articles selected. **Results:** Based on these articles it can be seen that, with the increase in the use of electronic devices and with the excess of information available, incidence of headaches in the population in all age groups has been increasing. Within the spectrum of studying the cognitive function of the elderly, even before the covid-19 pandemic, anxiety and depression were already shown to be associated with headache. However, this situation was intensified during the intoxication resulted from Covid-19 and headaches in elderly people who have access to information proved to be an important health marker. In teenagers, excessive use of computers was considered a risk factor for the development of headache, especially in females. Furthermore, there was a high prevalence of primary headache among them, migraine being the most prevalent type, followed by tension-type headache. **Conclusion:** In this context, contributing factors are the excessive use of electronic devices and the information overload to which people were exposed during the Covid-19 pandemic, causing an immense emotional and mental damage and, finally, reflecting negatively on neurological health.

Cefaleia

TL 1106481

THE RELATIONSHIP BETWEEN TENSION-TYPE HEADACHE (TTH) AND ANXIETY DISORDER: A LITERATURE REVIEW

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Background: A headache is a pain with cephalic involvement, the most common neurological symptom and a very frequent medical complaint. Anxiety worsens the perception of pain, intensifies it, increases its duration, and, by an escalation mechanism, further increases the level of previous anxiety. In several clinical and epidemiological studies, a comorbidity especially focused on among patients with TTH is anxiety. **Objective:** To present relevant information about the relationship of anxiety symptoms among TTH sufferers. **Material and Methods:** This is a literature review that allows a broad approach to tension-type headache and its relationship with anxiety, providing knowledge and incorporation of the applicability definition of concepts, results: and studies. The collection began in May 2022, so that a survey of data and studies relevant to the research in question was carried out. The textual search was restricted to scientific publications from the Scientific Electronic Library Online (SCIELO) and PUBMED databases during the month of May of this year, using publications from the last 10 years, through the following descriptors: "tension-type headache" and "anxiety". Original articles indexed in the period between 2012 to 2022 were included. **Results:** Seven articles were found in the initial search, and after analyzing the abstracts, 4 papers remained for this review. Among the primary headaches, tension-type headache stands out, characterized by light or moderate compressive pain sensations and its relation with anxiety disorder. It is subclassified according to its duration into infrequent episodic, constant, and chronic. Its pathophysiology is complex and involves peripheral and central pain mechanisms, and it must be differentiated from other types of headache. And through the analyzed studies we concluded that the patient diagnosed with anxiety has a higher frequency of occurrence of headaches when compared to those without mental disorders. The characteristics of headache, frequency, intensity, duration of crises were more aggravated in patients with anxiety. This disorder is preventable, with changes in habits in order to reduce or eliminate triggering factors. **Conclusion:** Tension headache and anxiety, although recurrent, is underdiagnosed, usually due to lack of knowledge by general practitioners, a fact that may result in the adoption of inappropriate treatment plans and cause impairment in the quality of life of patients.

Cefaleia

TL 1106652

"NOT OTHERWISE SPECIFIED HEADACHE" IN EMERGENCY DEPARTMENT: AN ANALYSIS OF 149,603 VISITS TO NINE UPA'S 24H IN FORTALEZA

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Background: Headache is one of the most common neurological symptoms and is almost a universal experience that requires medical attention on many occasions. Patients often seek medical advice for headaches in primary or secondary health care systems. In these scenarios, establishing the correct diagnosis of headache and referring further treatment to a specialist is essential. Hundreds of low-complexity Emergency Care Units (called UPAs 24h) were opened throughout the country in the last decade. Working 24/7, many UPA 24 h are one of the preferred places by patients to treat acute headaches. **Objective:** This study aims to evaluate the frequency of the "Not otherwise specified headache diagnoses" in patients who seek UPA's 24-h for headache. **Methods:** We evaluated the ED charts of 149,603 visits motivated by headache to nine UPAs 24h in Fortaleza, Ceará, from January 2017 to April 2022. In addition to the demographic data, the care flow chart, the MTS classification, the times of each step of care (from risk classification to discharge or death), and the final diagnosis coded by the attendant physician using the International Classification of Diseases (ICD-10). Data were analyzed using statistical software, and a $p < 0.05$ was considered for significance. **Results:** The 149,603 consultations, which represented 3.5% of the total, were made by men and women (72 and 28% respectively) with 38.2 (± 14.6) years of age, on average. Regarding the diagnosis, only 25,121 (16.8%) patients received the diagnosis of migraine, and 4,671 (3.1%) were diagnosed with "other cephalic algic syndromes". The remaining 119,811 (80.1%) had the not otherwise specified headache discharge diagnosis in their charts. **Conclusion:** The UPA's 24 h represents an important care channel for patients with acute headaches; however, most patients in this setting do not receive the correct diagnosis, which undoubtedly contributes to inadequate treatment and prognosis. A training program for the health professionals who work there is urgently needed to improve the care of thousands of headache patients.

Cefaleia

TL 1106654

OCCURRENCE OF SILENT LACUNAR INFARCTS DETECTED BY VOLUMETRIC ANALYSES OF MAGNETIC RESONANCE IMAGING IN WOMEN WITH MIGRAINE

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Background: The development of neuroimaging techniques aroused interest in structural brain changes associated with migraine. Silent lacunar infarcts (SLI) are related to cerebral microvascular disease and frequently found in neuroimaging studies in migraine. **Objective:** To describe the occurrence of ILS in different groups of migraineurs and to characterize them in terms of location. **Methods:** Sixty patients were selected, without cerebrovascular risk factors, from the headache clinic of a tertiary neurology center. The volunteers were equally allocated into four groups according to the third International Classification of Headache Disorders: Episodic migraine without aura (MoA), Episodic migraine with aura (MA), Chronic migraine (CM) and control group (CG). All participants were submitted in a 3T MRI scan with volumetric protocols. The acquired images were analyzed by two independent neuroradiologists. **Results:** ILS were detected in only three volunteers, two of them from the MoA group and one from the MA group; no ILS were identified in the MC and CG. Only one lesion was identified in each patient. Two SLI were located in the frontal lobe and the other one in the occipital lobe. No infratentorial SLI were identified in our study. **Conclusion:** the occurrence of ILS was low in the sample, and presented in the anterior circulation. Prospective studies are needed to determine whether migraine is related to a higher risk of SLI formation.

Cefaleia

TL 1106655

ANALYSIS OF CLINICAL AND VOLUMETRIC PARAMETERS OF WHITE MATTER LESIONS IDENTIFIED BY MAGNETIC RESONANCE IMAGING IN WOMEN WITH MIGRAINE

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Background: Migraine is a very prevalent primary headache characterized by attacks of pulsatile, unilateral pain, associated with nausea, photo and phonophobia. It is believed that repeated headache episodes throughout life may cause structural changes in the central nervous system, especially in those patients with a high frequency of attacks and with aura subtype. White matter lesions (WML) are the most reported structural changes in neuroimaging studies. **Objective:** This work uses volumetry and voxel-based morphometry (VBM) techniques to analyze WML in migraineurs. **Methods:** Sixty volunteers from a tertiary headache center were selected and divided equally into four groups: episodic migraine without aura (MoA), episodic migraine with aura (MA), chronic migraine (CM) and controls (CG). The participants performed 3.0T magnetic resonance imaging (MRI). WML variables were compared between groups and about the clinical characteristics of the sample. **Results:** There were no differences in WML variables between groups. There was a positive weak correlation between age and the number and total volume of WMLs, which persisted in the comparison categorized by size and location by brain lobe, except for the occipital lobe. Disease duration was positively correlated with the number and total volume of WML, but when the variable was controlled by age; the correlation was significant only for the insular lobe. Aura frequency was relevant for the frontal and temporal lobe WMLs. There was no statistically significant correlation with attack frequency, pain intensity and use of prophylactic medication. **Conclusion:** The type of migraine does not influence the formation of LSBs. Number and volume of WML were correlated with insula, and the presence of frontal and temporal lobe WML were correlated with aura frequency.

Cefaleia

TL 1105292

THE PREVALENCE OF VESTIBULAR SYNDROMES IN A BRAZILIAN TERTIARY CENTER

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Background: Vestibular symptoms are common complaints and sometimes challenging to discriminate among different syndromes and etiologies. A great number of these patients are referred to the neurologist, who must be able to determine whether there is a central or peripheral cause for the problem, as the management and therapy are very distinct in each situation. **Objectives:** The goal of this study was to quantify the most frequent vestibular syndromes in a tertiary dizziness reference department. **Methods:** Data of 1649 patients between 1999 and 2022 were evaluated retrospectively in our institution. These data were analyzed and the diagnoses of these most frequent vestibular syndromes were established by anamnesis, physical examination, and vestibular tests and images when necessary. **Results:** The most common syndromes were: benign paroxysmal positioning vertigo (BPPV) (24.2%), central vertigo (12.7%), migraine (8.6%), perceptual postural persistent dizziness (PPPD) (7.8%), Menière disease (6.5%), clinical non-neurological causes (7.4%), vestibular neuritis (4.9%), other causes (26.7%), and undetermined (1.2%). **Discussion:** Our findings are in similar to the literature, and show that the most frequent causes of vertigo can be treated and have good prognosis. A small difference in number of some diagnosis may have different reasons such as publications new diagnostic criteria for vestibular migraine, and changes the referral system in our institution. **Conclusion:** The knowledge of the epidemiology in a reference dizziness department is important for the best planning and conducting those patients. The early diagnosis and most adequate treatment are critical since these syndromes may have a good prognosis when correctly handled.

Distúrbio Vestibulares e do Equilíbrio

TL 1106590

BALANCE REHABILITATION WITH VIRTUAL REALITY IN PATIENTS WITH HEREDITARY SPASTIC PARAPLEGIA: A PRELIMINARY STUDY

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Background: Hereditary spastic paraplegia is a heterogeneous group of hereditary degenerative diseases involving the disorder of a single gene and is characterized by the progressive retrograde degeneration of fibers in the spinal cord. Among the symptoms, vestibulopathy is one of the causes of discomfort and a decrease in quality of life. **Objectives:** Determine the benefits of vestibular rehabilitation involving virtual reality by comparing pre intervention and post intervention assessments in individuals with hereditary spastic paraplegia. **Methods:** Randomized controlled clinical trial from the Rebec platform, were evaluated 40 patients. The interventions included vestibular rehabilitation with virtual reality using the Wii® console, Wii-Remote and Wii Balance Board and pre- and postintervention assessments. Group I included twenty volunteers who performed balance games. Group II included twenty volunteers who performed balance games and muscle strength games. The sessions were performed twice a week for 10 weeks (total: 20 sessions). **Results:** This study provides an assessment of the effectiveness of a virtual reality vestibular rehabilitation program in decreasing the progression of the imbalance (above 50%) in the hereditary spastic paraplegia, and this treatment can be personalized and affordable. It is observed that the improvement was more evident in the group that performed the muscle strength exercises concomitantly (56.4%). **Conclusion:** The study contributed to the reduction the progressive effect of hereditary spastic paraplegia and serve as an alternative treatment option that is accessible and inexpensive.

Distúrbio Vestibulares e do Equilíbrio

TL 1104942

ASSESSMENT OF THE PRESENCE OF INDICATORS OF ATRIAL HEART DISEASE IN PATIENTS WITH ISCHEMIC STROKE WITHOUT PREVIOUSLY DIAGNOSED ATRIAL FIBRILLATION

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Background: Ischemic Stroke may have paroxysmal atrial fibrillation as a potential risk factor, which existed before the ischemic event, but remained undiagnosed even with the etiological investigation. **Objective:** To study whether the presence of markers of atrial heart disease in patients with stroke has a statistical correlation with the identification of AF at follow-up. **Methods:** Retrospective longitudinal study with evaluation of data in electronic medical records of patients Hospitalized for stroke from 2014 to 2021. A total of 872 patients with complete data in medical records and without known AF were analyzed. The markers of atriopathies studied were LA size, reduced ejection fraction, presence of intraventricular septal fibrosis, interatrial septal aneurysm or atrial appendage thrombus identified on echocardiography. Using Holter, we analyzed the presence of arrhythmias, as well as the presence of extrasystoles. We also tabulated data such as age, image topography, main comorbidities and failure to use antithrombotics in the population studied. **Results:** We identified that LA greater than or equal to 42 mm, age greater than or equal to 70 years, the presence of septal aneurysm and NHSS greater than or equal to 6 at admission are variables with statistical significance associated with the risk of developing atrial fibrillation in the follow-up. With these variables, it was possible to structure a risk score for the diagnosis of AF in the follow-up, in which the patient who scores equal to or more than 2 points has a risk of AF in the follow-up 5 times greater. **Conclusion:** Based on the present study, we concluded that in the sample of stroke patients from the Instituto de Neurologia de Curitiba, without known AF before the event, the presence of markers of atrial heart disease showed a positive correlation with the detection of AF at follow-up. In addition, it was possible to apply an AF risk score which could be validated in larger samples in future studies.

Doença Cerebrovascular

TL 1105123

CHARACTERIZATION AND FUNCTIONALITY OF INDIVIDUALS POST STROKE OF THE NORTHERN PLATEAU OF SANTA CATARINA: ASSISTED IN STROKE UNIT

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Background: Stroke is known as the most disabling chronic disease, affecting not only the individual, but also his family and society as a whole, mainly caused by the significant reduction in the individual's level of independence. Therefore, the objective was to characterize post-stroke individuals Hospitalized in U-stroke, regional reference for the North Plateau of state of SC in terms of epidemiological, clinical and functional profile. Prospective, observational and longitudinal study carried out between Nov/2019 to Dec/2020. With individuals 718 years old; post-stroke confirmed by CT or MRI. Individuals with other diseases and conditions that could lead to motor deficits or functional changes other than the stroke were excluded. Sociodemographic data, ICTUS and therapeutic window were analyzed. The TOAST scale, NIHSS, Functional Independence Measure (FIM) and Modified Rankin Scale (ERm) were also applied at discharge. ERm was also measured at 30 and 60 days after stroke, using the Brazilian and validated version of the interview via telephone contact. Sample composed of 44 patients; 45 to 92 years old; 79.5% elderly (>61 years), 16% 46-60 years, 4.5% 31-45 years and no young adults (12-30 years). 56.8% were male and 88.6% self-declared white. 96% had modifiable risk factors; 79% hypertensive (only 73% undergoing treatment); 22.7% with Atrial Fibrillation (only 4.5% using anticoagulants); 48% with a previous history of stroke/transitory stroke without sequelae; 84% considered sedentary. Only 9% were eligible for thrombolytic therapy. In tasks of daily living according to the total FIM: they were predominantly dependent (complete, 54%; and moderate, 39%). Considering the motor FIM classification, the levels of functionality were 70% complete dependent and 23% moderate. As for the functional evolution by Rankin, at the time of discharge, 82% of the individuals were dependent. After thirty days, 57% were able to walk independently and seven died. After sixty days, 27% had negligible disability; 29% moderate disability; 23% reported severe disability and one died. The findings point to important correlation between the prevalence of modifiable risk factors and functional disability even after Hospital discharge. This scenario reflects on the population of the Planalto Norte Catarinense and the necessary health care measures, thus enabling the design of strategic measures that aim to meet the characteristics of the population.

Doença Cerebrovascular

TL 1105134

FUNCTIONAL BIOMARKERS OF TRUNK AND LIMB CONTROL AND QUALITY OF LIFE IN POST-STROKE PATIENTS

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Background: Stroke has a great impact on the lives of many individuals. The identification of post-stroke biomarkers is important to assume functional independence and the impact of the disease on the patient, family and society. The present study aimed to characterize the functional capacity of post-stroke patients, evaluated by the application of functional scales for the trunk (Trust Control Test, TCT) and upper limb (Frenchay Arm Scale), in addition to scale for the assessment of quality of life after stroke using the Specific Quality of Life Scale for stroke (EQVE-AVE). This study was carried out with patients treated in stroke unit that is reference in the Planalto Norte in the state of Santa Catarina, including total of 20 patients, evaluated in period of six months (out of a total of 48 months of study). Therefore, the mean age range was 67 ± 17.6 years, 55% were female, and the mean NIHSS for the first 24 hours was 9.7. It was evidenced as a predominant risk factor for SAH (90%), sedentary lifestyle (90%) and DM (45%). As for functional capacity by TCT, the **Results:** show great variability, obtaining an average of 51.9 ± 36.03 . In this test, the examinee is proposed to perform 4 maneuvers dependent on the skeleton, muscle and axial nerves, responsible for stability and postural integrity – the main disabilities present were: ability to balance while sitting on the edge of the bed with the feet off the floor; activity of rolling to the compromised side and rolling to the healthy side. In the Frenchay Arm Scale, the mean score was 2.4 ± 2.27 points (from 0 to 5 points), which reflects in a heterogeneous sample regarding the capacity of proximal control of the upper limb and manual dexterity. On the analysis of quality of life, through EQVE-AVE, an average value of 125.3 ± 48.8 points were obtained, which reflects their significant impairment – on this, the most affected domains were Mood, Self-care and Social Role; the least affected were work, energy and family roles. Thus, the present study points to a positive correlation on functional findings and post-discharge reflexes, mainly in emotional and social aspects, not showing significant loss in relation to work (possibly because the majority are already out of the job market). Interestingly, energy and disposition were also less affected. Thus, the important correlation between the loss of trunk and limb functionality and quality of life in relation to mood and self-care is highlighted here.

Doença Cerebrovascular

TL 1105168

BILATERAL THALAMIC INFARCTION PRESENTED AS A LOSS OF CONSCIOUSNESS

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Case Presentation: A 62-year-old female presented in the emergency department with sudden loss of consciousness. At admission, the patient was conscious but confused. She had clinical history of hypertension. On examination, she was somnolent, and the rest of the physical examination was unremarkable. Her electrocardiogram showed a sinus rhythm. Echocardiogram reported concentric remodeling of the left ventricle. A non-contrast CT scan revealed a hypodensity in the medial portion of both thalami, corroborated by MRI. Angio-CT demonstrated stenosis in the distal V4 segment of the left vertebral artery. **Discussion:** The thalamus is part of the diencephalon and is the relay center of the brain, retransmitting information between the cerebral cortex and subcortical structures. The inferolateral, polar, paramedian, and posterior choroidal arteries are the main arteries that supply blood to the thalamus. There is considerable variation in the territory supplied by each artery. Gerard Percheron described four variants of paramedian artery that supply the mediodorsal and intralaminar thalamic nuclei and occasionally the upper brainstem. One of the variants is the artery of Percheron. It arises from P1 segment of the posterior cerebral artery as a single artery and supplies blood to the bilateral paramedian thalamic nuclei and occasionally the rostral mesencephalon. Occlusion of the artery of Percheron can lead to a bilateral paramedian thalamic infarct. It can present with a variety of clinical manifestations such as an altered mental status, increased somnolence, memory impairment, a vertical gaze palsy, aphasia, hemiplegia, movement disorder, cerebellar ataxia and even coma. **Conclusion:** Due to the large number of possible differential diagnoses, vigilance impairment poses a common diagnostic challenge. The occlusion of the artery of Percheron, an anatomical variation whereby both paramedian arteries arise from a common P1, often leads to a bilateral paramedian thalamic infarction. Thalamic infarcts account for 11% of vertebrobasilar infarcts, of which 4-18% are due to the artery of Percheron occlusion and only 0.6% of cerebral infarctions are bilateral thalamic. In a case with no obvious cause for sudden loss of consciousness, occlusion of the artery of Percheron should be considered as one of the differentials. The severity of vigilance impairment is significantly more pronounced in bilateral paramedian infarction than in unilateral lesions.

Doença Cerebrovascular

TL 1105187

IMPACT OF COVID-19 ON ACUTE STROKE CARE

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Background: The COVID-19 pandemic had impact on stroke care around the world, but middle and low-income countries may have struggled more to adapt to the rapid changes in workflow because of its more fragile health care systems. At Hospital das Clínicas/University of São Paulo, major changes were made in the stroke workflow after the onset of the pandemic. **Objectives:** To assess changes in metrics of acute ischemic stroke care at a large-volume Hospital, before and after the pandemic. **Methods:** In this prospective, observational study we compared key performance measures collected from the University of São Paulo Stroke Data Base, one year before and one year after the report of the first cases of COVID-19 in São Paulo. **Results:** We analyzed 1.584 stroke admissions: 797 from March 16, 2019 to March 15, 2020 and 787 from March 16, 2020 to March 15, 2021. There were no statistically significant differences between patients admitted during the two periods regarding age, sex or NIH Stroke Scale (NIHSS) scores. Before the pandemic, 132 procedures of intravenous thrombolysis (IVT) and 29 mechanical thrombectomies (MT) were performed and afterwards, 110 IVT and 19 MT. There was no statistically significant difference in the monthly average between the one-year periods for stroke admissions, IVT or MT. After the pandemic, there was an increase in icus-to-admission and door-to-imaging (DTI) times compared to before, but there were no statistically significant differences regarding door-to-needle time (DTN) or door-to-puncture (DTP) times. **Conclusion:** In the largest medical center from Latin America, despite significant increases in delays in the interval from onset of symptoms of stroke to admission, as well as from admission to performance of CT scans, median DTN times did not significantly change and remained below 45 minutes. Median DTP times did not significantly change, possibly due to the limited power, given the relatively low number of thrombectomy procedures performed. Training of the stroke team and maintenance of high volumes of patients may have contributed to the lack of significant difference in key performance metrics.

Doença Cerebrovascular

TL 1105192

NEUROPSYCHIATRIC DISORDERS IN POST-STROKE PATIENTS

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Background: Stroke patients have a high prevalence of neuropsychiatric disorders. Post-stroke depression, for instance, can affect around a third of these patients, in addition to the occurrence of anxiety disorders, post-traumatic stress disorder, bipolar disorder and others. However, psychiatric diagnosis and treatment is still limited. **Objectives:** We aimed to identify the prevalence of neuropsychiatric disorders in patients previously diagnosed with stroke and compare commonly used depression and anxiety assessment scales to a structured interview, in order to assess their reliability in this population. **Methods:** We evaluated ischemic stroke patients. Sociodemographic, health data and information on brain lesion location were collected and the Mini-International Neuropsychiatric Interview (M.I.N.I.) was applied for diagnosis of neuropsychiatric disorders. The Beck (BDI-II and BAI) and Hamilton (HAM-D, HAM-A) depression and anxiety scales were then applied, at same day, for comparison and analysis. **Results:** Of 81 included participants, 45.7% were diagnosed with at least one neuropsychiatric disorder. Post-stroke depression had the highest prevalence (35.8%), followed by anxiety disorders (34.6%), post-traumatic stress disorder (7.4%), obsessive-compulsive disorder (6.2%), and other (4.93%). Only 14.8% of participants had a psychiatric diagnosis at the moment of the interview. Brain lesion location was not statistically relevant. The depression rating scales BDI-II and HAM-D had an accuracy of 97% and 96%, respectively, when compared to M.I.N.I. as the gold standard. The anxiety rating scales, BAI and HAM-A, accuracy of 91% and 92%, respectively. **Conclusion:** The prevalence of neuropsychiatric disorders in post stroke patients is high, being underdiagnosed and undertreated. The Beck and Hamilton depression and anxiety scales can be used in this population with good accuracy and reliability, improving screening, diagnosis and follow-up of these patients.

Doença Cerebrovascular

TL 1105196

EPIDEMIOLOGICAL AND CLINICAL CHARACTERISTICS OF ANEURYSMAL SUBARACHNOID HEMORRHAGE PATIENTS IN A BRAZILIAN TERTIARY NEUROSURGICAL CRITICAL CARE UNIT

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Background: Subarachnoid hemorrhage (SAH) is a rare and life-threatening cerebrovascular disease. Ruptured aneurysms are the most common cause of atraumatic SAH. It is one of the main medical emergencies associated with high rates of morbidity and mortality and years lived with disability. In Brazil epidemiology of aneurysmal SAH remains unknown. **Objectives:** To evaluate epidemiological and clinical data and outcomes such as mortality, time of mechanical ventilation of aneurysmal subarachnoid hemorrhage patients interned in a tertiary neurointensive critical care unit. **Methods:** This is a cross-sectional study. Patients with SAH admitted to the neurointensive critical care unit (neuroICU) in a tertiary care public Hospital were eligible. The inclusion criteria were a minimum stay in the intensive care unit (ICU) of 24 h, due to subarachnoid hemorrhage from a nontraumatic spontaneously ruptured cerebral aneurysm, and Hospital admission within 24 h after the onset of symptoms. **Results:** Our population was mainly composed by women (67.2%). The most reported symptoms were syncope (26%), isolated headache (23.2%), headache and syncope (16.3%). Twenty-eight percent presented Hunt Hess Scale grade II and 62.1% presented Fisher scale grade 4. Sixty-eight percent required mechanical ventilation, 48.6% presented vasospasm, 5.1% presented rebleeding, 36.7% acute hypertensive hydrocephalus, 40.7% died. Females presented higher mean of age than males (women: 57.21±12.20 years; men: 51.34±12.54 years; p=0.03) and also had the longest waiting time for an intensive care unit bed (women: 6.12±6.71 days; 4.21±3.56 days; p=0.01). When stratified by sex there was no statistical significance among outcomes such as rebleeding, vasospasm, delayed cerebral ischemia, acute hypertensive hydrocephalus, death. When stratified by type of discharge, death was statistically associated with vasospasm (OR 2.138 p=0.01) and acute hypertensive hydrocephalus (OR 2.136 p=0.01). **Conclusion:** This study can address relevant data about epidemiological and clinical factors involving subarachnoid hemorrhage patients. As reported in literature the majority of patients were female. Percentages of complications are similar from the previous reported. It is noteworthy that women spent more time waiting for a specialized bed.

Doença Cerebrovascular

TL 1105218

HEADACHE AT THE CHRONIC STAGE OF CEREBRAL VENOUS THROMBOSIS

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Background: Headache is the most frequent symptom of cerebral venous thrombosis (CVT) but there is limited information about the frequency and phenotype of headache, weeks to months after CVT (post-CVT headache, PCH). **Objective:** To assess the frequency, characteristics and predictors of PCH. **Methods:** In this cross-sectional study, the frequency and characteristics of PCH were assessed in CVT survivors. Patients were interviewed between 6 months and 5 years after the CVT diagnosis. Clinical and imaging characteristics at the time of CVT diagnosis, as well as history of headache prior to CVT were compared in subjects with (GroupPCH) and without PCH (Groupcontrol). **Results:** Subjects (n=100; 82% women) were assessed, on average, at 1.1 ± 1.6 years of follow-up. PCH was present in 59% of the patients, had phenotypes of tension-type-like headache in 31/59 (52.6%) and of migraine-like headache, in 16/59 (27.1%). History of primary headache prior to CVT was significantly more common (OR: 6.4; 95% CI: 1.7-36.3) in GroupPCH (33.9%) than in Groupcontrol (7.3%). **Conclusion:** PCH was present in more than half of the patients. History of prior headache may be a risk factor for PCH. Prospective studies are required to confirm these findings and determine mechanisms, as well as interventions for prevention and treatment of PCH.

Doença Cerebrovascular

TL 1105252

EPIDEMIOLOGICAL ASPECTS OF PATIENTS AFFLICTED BY STROKE IN REFERRAL SERVICES OF THE PAJEÚ MICROREGION IN THE STATE OF PERNAMBUCO

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Background: The profile of patients afflicted by stroke differs between countries and geographical regions. An area's demographic and socioeconomic aspects have a direct impact on stroke determinant risk factors, whose identification is necessary for the development of primary and secondary prevention strategies. In Brazil, the availability of epidemiological data is low, which reflects the reality of regions with higher sociodemographic indexes. In Pernambuco, most of all in the "Sertão do Pajeú" region, there aren't any available studies with this outline. **Objective:** Trace the epidemiological profile of patients with stroke admitted into the emergency services of "Serra Talhada/PE", reference for 17 cities of the Pajeú microregion. **Method:** Descriptive, transversal study. The instruments used were the Individual Registration File of SUS and Evaluation File elaborated according to the protocol of the Brazilian Society of Cerebrovascular Diseases. **Results:** The sample consisted of 114 participants with male predominance (n=64; 56,1%), black self-declaration (n=86; 75,5%) and higher distribution in the 61-80 age group (n=57; 50%). Most from Pernambuco (n=109; 95,6%), with 56,1% (n=64) from Serra Talhada/PE. Around 48% (n=55) were farmers; 36,8% (n=42) had no schooling and 42,1% (n=48) had incomplete elementary school. The main comorbidities observed were hypertension (n=93; 81,6%) and diabetes (n=42; 36,8%); despite that, 74,5% (n=85) did not follow the diet. As for the life habits, approximately 30% (n=34) smoke; 12% (n=14) consume alcoholic beverages 3 times a week and 87% (n=99) are sedentary. The alteration on the consciousness level was light at 41,2% (n=47) and the neurological compromise was severe at 41,2% (n=47). The predominant stroke type was ischemic (n=101; 88,6%) and the etiology undetermined at 45,6% (n=52). **Conclusion:** The study made it possible to trace the epidemiological profile of the patients attended at the Hospitals of reference of the "Sertão do Pajeú", identifying the predominance of the elderly, males and black people, reaffirming some of the main risk factors for stroke, such as hypertension, a sedentary lifestyle and bad dietary control of comorbidities. The knowledge of stroke determinant characteristics in this group shows the importance of the development of prevention strategies directed at the region's profile for control over the main modifiable risk factors.

Doença Cerebrovascular

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UNUSUAL PRESENTATIONS OF SUDDEN FOCAL NEUROLOGICAL DEFICIT: "STROKE CHAMELEONS"

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Background: Stroke is a medical emergency, present with sudden onset focal neurologic disorder and it may lead to severe disability or death in worse case scenarios. There are false-negatives when the clinical sign suggests other disorder conditions. These are known as "stroke chameleons". **Objective:** Trace the unusual presentations of sudden focal neurological deficit caused by stroke capable of dismissing the diagnostic suspicion and the appropriate treatment. **Method:** This study is a literature review. The chosen database was PUBMED, with keywords "stroke chameleon" and "stroke". Inclusion criteria were reviews, systematic reviews, clinical trials and meta-analysis with free full text available, published between 2017 and 2022. **Results:** The common presenting symptoms include dizziness/vertigo without trigger symptoms, nausea/vomiting, confusion, decreased level of consciousness, altered mental status, generalized weakness, and severe hypertension. In addition, it is presented with wrist drop syndrome purely motor or sensorimotor with hypotonia, decreased myotatic reflexes and normal superficial reflexes during the acute phase of stroke in precentral gyrus. Furthermore, transient global amnesia, hyperkinetic movement disorders defined as abnormal, repetitive, involuntary movements rarely occur in acute stroke presentations. Among these, are the Alien limb syndrome, consisting of involuntary movements and foreign-sense of limb, the Anton's syndrome, the denial of blindness and confabulation of visual sensory information, and the Gerstmann syndrome consists of a tetrad of symptoms: agraphia, acalculia, finger agnosia, and right-left confusion. A few stroke chameleon cases are diagnosed as psychiatric diseases. **Conclusion:** It appears that detailed history taking, comprehensive neurological examination, and appropriate imaging studies are important to identify the clinic manifestation and, when acute onset symptoms are present, stroke should be considered.

Doença Cerebrovascular

TL 1105270

EIGHT YEARS OF EXPERIENCE WITH STROKE PROTOCOL IN A HOSPITAL IN THE COUNTRYSIDE OF MINAS GERAIS

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Background: Stroke is the second most frequent cause of death in Brazil, and the main cause of disability. The use of care protocol guides the development of best practices in health work processes, and can directly impact the indicators related to stroke. **Objective:** To express data and evaluate the evolution of quality indicators related to stroke over 8 years after the adoption of care protocols by a Hospital in the countryside of Minas Gerais, which serves approximately 400,000 inhabitants. **Method:** Descriptive, cross-sectional, documentary study that used secondary data collected from the medical records of patients diagnosed with stroke during the years 2014 and 2021. **Results:** In the analysis, the chi-square test was used, at the nominal level of 5% of significance, which indicated a significant difference in all data presented. In 8 years, 2,608 cases of stroke were managed, of which 2,083 (79.87%) were of ischemic type and 525 (20.13%) were hemorrhagic; of the ischemic strokes, 142 were thrombolysed and another 165 were admitted in a timely manner for thrombolysis, but this was not performed due to contraindications to the use of thrombolytics or failure in care. The number of thrombolysis performed in this period of time were, respectively: 4, 18, 15, 23, 13, 18, 24, and 27. Of the patients who arrived at the Hospital less than 4.5 hours after the onset of symptoms, the percentage that had cranial computerized tomography (CCT) performed in less than 25 minutes between 2018 and 2021 were, respectively: 55.9, 69.4, 62.2, and 76.9. Of all the stroke cases, 601 (23.04%) evolved to in-Hospital death, the others were discharged. Stroke mortality rates between 2014 and 2021 in percentages were, respectively: 23.4, 24.4, 23.5, 18.9, 21.5, 21.4, 26.0, and 24.2. **Conclusion:** The adoption of the protocol makes it possible to observe progress in quality of service indicators, such as CCT execution time and number of thrombolysis performed. There is also a progressive drop in mortality between 2014 and 2019, but this rise again, reaching peaks in the years of the Covid-19 pandemic, probably mortality increased in this period due to the absence of 1st and 2nd prevention, which made strokes more frequent and more severe during this period. Therefore, it is stated that the use of protocols organizes the service and directly impacts stroke mortality. Remembering that the real improvement in the indicators comes in health policies in all areas of health.

Doença Cerebrovascular

TL 1105271

PREDICTORS OF HEMORRHAGIC TRANSFORMATION IN ISCHEMIC STROKE PATIENTS: EXPERIENCE OF A BRAZILIAN TERTIARY HOSPITAL

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Background: Hemorrhagic transformation (HT) is one of the most common stroke-related complications. Predictors of this complication are still unclear in the literature and understanding them may be a resource of unique clinical value to predict this dreaded condition. **Objective:** Identify predictive factors for HT in patients with ischemic stroke submitted or not to reperfusion therapies (RTs). **Methods:** We analyzed retrospectively data from all patients admitted to a South-brazilian Stroke Center, from 2017 to 2019. **Results:** 821 patients were included in the analyzed sample. The prevalence of any HT was 9.7%. Thrombolysis with thrombectomy (n=12) was independently associated with TH (OR 4.7, 95% CI 1.4-16.1, p=0.013). Thrombolysis with tPA was not associated with TH (p=0.1), in contrast to performing thrombectomy alone (n=12/805, 1.5%). In general, TH was not related to a worse clinical outcome (mRS3-6) in adjusted analysis. **Conclusion:** The value of low ASPECTS score, high admission NIHSS, and cardioembolic etiology were possible predictors for HT in patients who did not undergo rt-PA thrombolysis.

Doença Cerebrovascular

TL 1105336

STROKE ETIOLOGIES IN PATIENTS WITH COVID-19

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Background: Patients with severe COVID-19 may have an increased risk of venous and arterial thrombotic events, including ischemic stroke (IS). It has been hypothesized that the infection by SARS-CoV-2 alone may be a risk factor for IS, particularly in young subjects. We designed an observational retrospective cohort to compare stroke etiologies in patients with IS, with (ISCOVID+) or without (ISCOVID-) SARS-CoV-2 infection, defined by molecular detection of SARS-CoV-2 before the stroke, or within 15 days after stroke or detection of serum antibodies before or within 30 days after the IS. The primary outcome was the stroke etiology according to the Causative Classification System for Ischemic Stroke (CCS) algorithm. In addition, two subgroups were determined for the category of Other uncommon Causes (OC): iatrogenic causes (OC-IC) and Other uncommon Causes excluding iatrogenic (OC-EI). A total of 212 patients were included (160 in ISCOVID+ and 42 in ISCOVID-). Cardio-aortic embolism (CE) was the most common etiology in the two groups. Two patients in the ISCOVID+ group but none in the ISCOVID-group had unexplained thrombosis of the common carotid artery. There were independent associations between younger age (RR 0.91, 95% CI 0.87 – 0.95) as well as COVID-19 infection (RR 4.08; CI 95% 1.03 – 16.14) and IS due to OC-EI. Classic stroke etiologies accounted for most of the cases of IS and concomitant COVID-19 infection, but large-artery thrombosis was more common in infected than in non-infected patients.

Doença Cerebrovascular

TL 1105365

ANALYSIS OF THE FUNCTIONAL CHARACTERISTICS OF THE PATENT FORAMEN OVAL AND DIAGNOSTIC ACCURACY USING CONTRASTED TRANSCRANIAL DOPPLER

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Background: Patent foramen ovale (PFO) is an interatrial embryonic remnant that results in right-to-left circulation (RLC) communication, therefore associated with paradoxical embolism events. It is associated with cases of stroke of undetermined source in young people, although it is present in about 30% of the general population and its incidental finding in complementary exams is common. The causal relationship has been explored in many studies, and the search for high-risk markers is the subject of numerous meta-analyses. Contrast-enhanced transcranial Doppler (cTCD) is considered the screening test of choice because it is a low-cost, non-invasive diagnostic tool with high sensitivity and specificity. **Methods:** The database of the Neurosonology Laboratory of the Hospital de Clínicas of the Federal University of Paraná was used, with a review of information from 901 patients from January 2015 to May 2019. Of the total, 701 patients who underwent cTCD were included and 551 who had a complementary investigation with transthoracic echocardiography (TTE) or contrast transesophageal echocardiography (c-TEE). The functional pattern by the cTCD followed criteria previously published in the Latin American Consensus. **Results:** Our study showed a significant difference in the “curtain” pattern at rest in the cerebrovascular disease group when compared to migraine with aura and incidental finding groups. In the evaluation of the subgroup that performed the cTCD and c-TEE, we found a high sensitivity and specificity of the cTCD when compared with the gold standard exam. **Conclusion:** The “curtain” rest pattern resulted in the greatest association of functional risk for the event related to cerebrovascular disease and can be considered a high-risk characteristic. Comparison of diagnostic tests for RLC resulted in a sensitivity of 98.6% and a specificity of 91.3%, with a PPV of 96% and a NPV of 96.9%.

Doença Cerebrovascular

TL 1105369

CLINICAL AND EPIDEMIOLOGICAL PROFILE OF MEDICAL CARE IN ACUTE STROKE AT THE EMERGENCY ROOM AT AMECOR HOSPITAL (CUIABÁ-MT)

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Background: Stroke is divided into two categories: ischemic (iCVA) and hemorrhagic (hCVA). The iCVA is caused by the interruption of blood flow to a specific brain area, having as main causes thrombosis, embolism and systemic hypoperfusion. The hCVA is the presence of intracranial bleeding, due to parenchymal or subarachnoid hemorrhage. Ischemic CVA is the most common, however, hemorrhagic CVA is associated with a higher mortality rate. Both can cause complications and need for different approaches in the acute phase. **Objectives:** Describe the clinical-epidemiological profile of patients treated for acute stroke in a reference service in Cuiabá-MT. **Methods:** This is a descriptive observational study with retrospective data collection from patients diagnosed with acute stroke. **Results:** In the year of 2021, 48 patients presented suspected clinical features of acute stroke, and the stroke protocol was open; 35% of these patients were aged between 61 and 80 years, 54% were male and 46% were female. Of the 46 stroke protocols opened, 24% were confirmed as iCVA and 8% as hCVA, and 56% were discarded cases. Stroke protocols were more opened in July (16.6%) and August (16.6%). In relation to the time from ictus until arrival at the referral service, 62% were admitted between 0 and 4.5 hours (thrombolysis window), 21% between 4.5 and 12 hours, 12% between 12 and 24 hours and 5% after 24 hours. Of the cases confirmed as iCVA that entered in the window for thrombolysis, 20% had indication for thrombolytic therapy, being successfully performed in approximately 3 hours after the ictus. The median door-to-imaging time was 1h and 23m, while the median door-to-needle time was 01h and 24m. The mean Hospital stay time was 5 days. There was no registered deaths in patients with iCAV. **Conclusion:** Thrombolytic therapy when properly indicated provides effective results and the analysis of this series demonstrates that adequate information is needed for greater agility in the care of the acute stroke phase in the reference unit.

Doença Cerebrovascular

TL 1105380

LACUNAR STROKE AS A PREDICTOR OF MORE FAVORABLE OUTCOMES: THE RELEVANCE OF BAMFORD CLASSIFICATION WHEN EVALUATING ISCHEMIC BRAIN EVENTS AND PROGNOSTIC PREDICTION

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Background: The occlusion of perforating arteries is often the cause of a ischemic stroke and it appears as a syndrome called lacunar syndrome (LACS). That description is made based on Bamford classification and its use can help handling a patient and in their prognostic evaluation. **Objective:** To investigate possible relations between lacunar and non-lacunar syndromes of a ischemic stroke, the severity of neurological deficit, and the early functional result in patients admitted in the emergency room. **Method:** This is a descriptive, analytic and transversal study. The variables that were analyzed were obtained by the use of the National Institutes of Health Stroke Scale (NIHSS) instruments, the Bamford classification, the modified Rankin Scale (mRS) and the Barthel Index. **Results:** The sample had 103 patients, most of them being men (n = 57; 55.3%) between ages 51-80 (n = 69; 67%). The LACS was seen in 26.2% (n = 27) of the patients and it showed a significant correlation (? = 0.46; p < 0.001) with better functional results (mRS). There was a greater sampling distribution in light incapacity levels (22.2%) and insignificant incapacity (29.6%) when compared to non-lacunar syndromes, which had a higher percentage in moderate to severe incapacity level (29%) and severe incapacity level (29%). The analysis of the independence of a patient during their daily activities showed a significant correlation between the LACS and the Barthel Index (? = 0.42; p < 0.001). It was possible to highlight higher percentage numbers on total dependence (17.8%) and on severe dependence (49.3%) in patients with non-lacunar syndromes. No case of total dependence was found on patients with LACS. Among those with severe neurological damage (NIHSS ? 16), 95% of them (n = 37) showed non-lacunar syndromes. The analysis of the correlation between LACS and NIHSS showed a significant number (? = -0.32; P < 0.001), so that the bigger the severity of the deficit the smaller was the distribution of patients with that stroke subtype. **Conclusion:** Lacunar Syndrome has a correlation with functional and independent results higher than non-lacunar syndrome does. Moreover, the neurological damage in patients with LACS tends to be lighter. That result reveals the importance of the right classification of the ischemic stroke as well as allows the establishment of parameters in order to better guide a patient's rehabilitation.

Doença Cerebrovascular

TL 1105388

THE RELATION BETWEEN THE SEVERITY OF THE NEUROLOGICAL DEFICIENCY AND THE FUNCTIONAL DEPENDENCY DEGREE IN PATIENTS DURING THE ACUTE PHASE OF A STROKE

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Background: The ischemic stroke is the second most cause of people with some sort of physical impairment. The severity of the initial neurological deficiency is a relevant predictor of a post-stroke functional prognosis. **Objective:** To analyse the correlation between the neurological deficiency and the functional dependency in victims of a stroke during the periods of: admission to the Hospital, Hospital discharge and 3 months post Hospital discharge. **Method:** Longitudinal study done in the XI Gerência Regional de Saúde de Pernambuco between September/2021 and April/2022. The instruments of research were the National Institutes of Health Stroke Scale (NIHSS), which was used on admission (T0) and Hospital discharge (T1); and the Barthel Modified index, which was used on Hospital discharge (T1) and 3 months after Hospital discharge (T2). **Results:** 43 patients participated, most of them had a ischemic stroke (88%), men (53%), age between 51-80 (63%) and blacks (72%). Medical record of hypertension (86%), sedentary lifestyle (88%) and a non-regulated diet (65%). The median of the NIHSS T0 was 13 [4-22.5]. It shows a moderate neurological deficiency. The median of the NIHSS T1 was 6 [1-18], which shows a light neurological deficiency. There was a significant statistic difference ($p=0.001$) between the scores of the NIHSS T0 and T1. The evaluation of the functionality showed a severe level of dependency with a median of 20 [10;5-43] on Bathel (T1) to the detriment of the severity of the neurological deficiency reduction. After three months at home, there was a slight Barthel (T2) reduction to 17 [0-43.5], however, the severe dependency level didn't change. We could also see a significant correlation between the NIHSS scores (T0) and Barthel's (T1) ($r = -0.77$; $p < 0.001$); and between NIHSS (T0) and Barthel (T2) ($r = -0.65$; $p < 0.001$). Plus, there was a significant correlation ($r = 0.79$; $p < 0.001$) between the scores of Barthel T1 and T2. **Conclusion:** More severe levels of neurological deficiency verified by the NIHSS during the acute phase of a stroke grant a worse expectation of a functional prognosis in a three month period. Therefore, the relevance of therapeutic interventions directed towards the reduction of the severity of the neurological condition and the intensification of Hospital rehabilitation should be reiterated.

Doença Cerebrovascular

TL 1105422

DIAGNOSTIC CONSIDERATIONS IN CERVICAL AND INTRACRANIAL ARTERIAL DISSECTIONS THROUGH A CASE SERIES STUDY

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Background: Cervical and intracranial vessels dissections are often misdiagnosed given their symptoms heterogeneity. Headache, partial Horner Syndrome and focal deficits are important clinical features. **Objective:** Report epidemiological and clinical features of 6 patients diagnosed with arterial dissections in a neurology emergency department. **Method:** We have retrospectively studied six cases of cervical and intracranial arterial dissections and extracted information regarding age, symptoms, vaccine status, covid-19 infection and radiological aspects of the dissections. **Resultados:** Medium age of onset was 43 years-old. Five patients had headaches, in which two cases were occipital/cervical pain, one orgasm-associated headache, one headache plus transient expression aphasia and one new-onset bilateral frontal headache. New headache pattern was the main red flag for further investigation. Unresponsiveness to usual analgesics led to the diagnosis in a patient with migraine. Direct trauma caused an extensive carotid bulb dissection complicated with distal embolization and ipsilateral M1 occlusion in a patient. The mean time to diagnosis was 4.8 days. Diagnosis was achieved with angiotomography or magnetic resonance angiography. Anterior circulation was stricken in 2 cases, with a carotid bulb dissection related to trauma and a bilateral carotid spontaneous dissection. Extracranial vertebral dissections were seen in 3 cases, plus 1 case of intracranial V4 dissection. Four patients had had respiratory viral infections or vaccine shots within 30 days of symptoms onset. Five patients were discharged with no disability. No deaths occurred. **Discussion:** Cervical arterial dissections are present in up to 20% of strokes in young patients. Intimal damage or vasa vasorum rupture are proposed mechanisms. Hypertension, migraines, recent infections and direct trauma are established risk factors for arterial dissections, which usually occur after carotid bifurcations. **Conclusion:** This case series highlights important pitfalls in diagnosing arterial dissections: (1) unresponsive and new occipital or cervical headaches prompt further investigation with contrasted images. (2) Infections and vaccines may cause transient endothelial inflammation, weakening the arterial wall, with further risk of dissection. (3) Neurologic focal deficits arising from both anterior and posterior circulations can be subtle and should be carefully searched in patients presenting with high-risk headaches.

Doença Cerebrovascular

TL 1105435

AN EPIDEMIOLOGICAL ANALYSIS OF THE ENDOVASCULAR VS MICROSURGERY TREATMENT OF INTRACRANIAL ANEURYSMS BETWEEN THE BRAZILIAN REGIONS

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Background: The advances of the technology for the endovascular treatment for intracranial has brought a change in the approach for this pathology. Therefore, the choice for course of treatment in these lesions have been a topic of recent discussions. **Objective:** Compare, through a prevalence analysis, the endovascular and the microsurgical treatment of intracranial aneurysms among the Brazilian regions in the last decade (2012-2021), whilst comparing cost and mortality rate (MR). **Methods:** Epidemiological, retrospective, descriptive study, carried out with data obtained from the Hospital Information System (SIH/SUS) and from the Institute for Applied Economic Research (IPEA). From these data, the average dollar cost per procedure was calculated. We also compared both treatments for intracranial aneurysms over and under 15 mm. **Results:** In the period analyzed, 32,010 intracranial aneurysms were treated. When treating aneurysms >15mm the embolization technique was, throughout the regions, the most utilized, had the lowest MR but was also the most expensive. The biggest average cost gap between approaches was in the northeast region (108%) and the smallest in the north with 59%. The north region had the most difference in MR (8,49%) and the central-west region had the least (1,68%). The choice of approach had its biggest contrast in the northeast region with 76%, and smallest in the north region (11%). Regarding the course of treatment for intracranial aneurysms <15 mm, the north region was the only one to have a higher MR in endovascular treatment, and to showcase a higher average cost with surgery but was also the region with the highest percentage of this approach (89%). The north region had the smallest average cost variance (6%), with the southeast having the biggest (74%), and had the biggest difference in course of treatment (78%) while the central-west region has the smallest (19%) both favoring the surgery route. The MR showed its largest variation in the southeast region (6,62%) with its smallest in the central-west with 1,21%. **Conclusion:** We conclude that although its higher cost, the endovascular treatment for aneurysms >15 mm is already more utilized in all the regions. While the approach for aneurysms <15 mm still varies between the regions, with both the north and central-west regions having the lowest percentage for this treatment. Furthermore, we affirm that further studies are needed to evaluate these regional differences in Brazil more deeply.

Doença Cerebrovascular

TL 1105464

CEREBRAL AND CERVICAL ARTERY DISSECTION: 6 YEARS CLINICAL REVIEW

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Background: Arterial dissections are rare, despite being a common cause of stroke in the young; they occur upon lesion of the arterial wall, allowing formation of intramural hematoma. Aneurysm or hematoma formation and vessel dilatation may cause local and result in stenosis or occlusion, which can lead to cerebral ischemia. The optimal treatment of dissection remains a challenge due to limitations of rapid definitive diagnosis, low incidence, low recurrence rate, and marked variation in patient characteristics. **Objective:** Evaluate the profiles and outcomes of patients with cerebral or cervical artery dissections, as well as the main symptoms and characteristics of the affected vessel, in patients treated in a Brazilian tertiary care Hospital. **Methods:** Retrospective, single-center observational study evaluating patients between January 2016 and December 2021. Data were collected from medical records; patient characteristics were analyzed (sex and age, initial symptoms, risk factors, history of trauma, previous comorbidities). Imaging methods data include location of affected vessel and treatment. **Results:** 87 patients were screened with intra or extracranial arterial dissection, with female predominance of 60.9% and mean age of 40.9 years. 28 cases occurred in the internal carotid artery, 17 on the left, with 53 in vertebral arteries, 22 on the left; a higher incidence of cases in the V2 and V3 segments (19 and 26, respectively) was seen; 31 cases were multisegmentar. The main symptoms were headache and neck pain, in 57 and 37 cases respectively; there were also clinical presentation with nausea (12 cases), dizziness (12), stroke (19), Horner's syndrome (9) and visual alterations (9). Only 36.7% of the cases had a history of trauma, 19.5% were using hormonal therapy, 8% had some coagulopathy after etiological investigation. All cases were initially investigated with AngioCT or AngioMRI and 55% underwent angiography to confirm the diagnosis. Patients were treated with warfarin; 4 patients were thrombolysed and 3 underwent angioplasty. **Conclusion:** Arterial dissections are an important cause of stroke in young patients and do not always evolve with a typical clinical manifestation, so they should be suspected with minor symptoms, such as headache and neck pain, in order to be treated early and prevent stroke. Treatment with anticoagulation seems to have good efficacy, but it is still the subject of several studies, given the advance in new therapies.

Doença Cerebrovascular

TL 1105498

VALIDATION OF AN ARTIFICIAL INTELLIGENCE IN THE PREDICTION OF STROKE RECURRENCE: PRELIMINARY RESULTS

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Background: Stroke remains the main cause of death in Ceará (Brazil). The high number of deaths in the state reflects the large incidence of primary and recurrent strokes. With the advances in digital health, technologies such as Artificial Intelligence (AI) can help the development of prediction studies. Machine Learning, an area of AI, is defined as the ability of a machine to learn "by itself" through database analysis and perform predictions, such as the recurrence of a stroke in a timely manner with minimal error. **Objective:** To validate an artificial intelligence that helps predict the recurrence of stroke. **Method:** The database used for training the predictive algorithm consolidated the records of 571 individuals residing in Ceará. It was built from three databases, with 127 records acquired with the Neurofunctional Physiotherapy League of the Federal University of Ceará database; 66 records by a survey from the Fortaleza General Hospital; and 378 records, from a population database, by an internet form survey. The variables included in the model were: occurrence of stroke, sex, age, marital status, hypertension, heart disease, type of work, local of residence, diabetes mellitus, tobacco use, alcohol use, and depression/anxiety. **Results:** These are preliminary results of an ongoing study. Of the 571, 237 people had a primary stroke and 51 of those had a recurrence. The algorithm with the best predictive performance was the Light Boosting Gradient Model (LBGM) with the highest accuracy of 90.0% and an AUC (Area Under the Curve) of 96.5%. The higher the accuracy and the AUC, the more positive model's accuracy. Among the variables, those with high influence scores on the stroke recurrence were: 1st stroke (80.1), heart disease (76.2), age > 50 years (48.5), Hypertension (46.9), BMI > 24 (18.4), Diabetes mellitus (12.5), Alcohol use (8.5), Depression/Anxiety (5.18), urban residents (4.77), married (3.03), retired (2.81), male (0.61) and smoking (0.26). Individuals after the first stroke frequently stopped smoking, this shows that the change in this habit makes it less explicative in the model. **Conclusion:** The AI algorithm is valid and has accuracy in stroke recurrence prediction models, with LBGM, a modern machine learning algorithm, being the most accurate. According to the analysis, the first stroke makes the individual susceptible to a second stroke, and the change in modifiable factors may be related to a lower influence of recurrence of stroke.

Doença Cerebrovascular

TL 1105500

INVESTIGATION OF PLASMA BIOMARKERS IN CASES OF CAVERNOMA-RELATED EPILEPSY

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Cerebral cavernous malformations (CCM) are a type vascular disease found in the central nervous system. CCM are triggered by mutations in different genes, called CCM1, CCM2 and CCM3, whose loss of function modifies the endothelial cytoskeleton and results in lesion. Although most cases are asymptomatic, the increased vascular permeability and fragility at the site can trigger cavernoma-related epilepsy (CRE). Currently, there are not clinically established or user-friendly biomarkers to determine the likelihood of this complication. Furthermore, most available studies have used symptomatic hemorrhages in cavernous angioma (CASH) as the only primary endpoint, with CRE being considered a minor comorbidity for CCM patients. In this context, from a cohort of 128 patients with CCM, we tried to identify possible plasma biomarkers for CRE. The disease was classified as familial or sporadic according to the distribution of lesions in SWI sequence, with familial being synonymous to the presentation of multiple lesions in this method. From this, the division between the groups was made using clinical history and complementary exams. CRE was defined with documented epileptic syndrome arising in CCM topography. We selected and obtained plasma samples from 20 patients (8 asymptomatic and 12 with CRE diagnosis). Vascular Endothelial Growth Factor (VEGF) and Thrombomodulin proteins, linked to vasculogenesis and coagulation, respectively, were selected for analysis from a pre-designed panel of molecules of interest. Both proteins were measured in our samples by ELISA assays. Patients diagnosed with asymptomatic CCM had a mean level of 49.45 pg/mL for VEGF and 482.88 pg/mL for thrombomodulin, while patients diagnosed with CRE had 24.45 pg/mL and 441.57 pg/mL for VEGF and thrombomodulin, respectively. A decrease in VEGF was observed in patients with CRE when compared to asymptomatic patients, with statistical significance ($P < 0.05$). Disruption of VEGF expression has been widely studied in CCM disease and models. Low levels of this molecule are known to alter blood-brain barrier permeability and induce progression of CCM lesions. Interestingly, decreased plasma VEGF has been described as a predictor of disease severity. This pilot study opens a path for VEGF as potential target in future studies on CCM biomarkers and justifies the understanding of CRE as a distinguishable clinical entity, while larger samples for a more meaningful analysis are still needed.

Doença Cerebrovascular

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HOSPITALIZATION AND MAIN MODIFIABLE RISK FACTORS OF STROKE IN THE STATE OF CEARÁ (BRAZIL): VIGITEL DATA

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Background: Stroke is the leading cause of death in Ceará (Brazil) as well as continued with high rates of Hospitalization. Since 90% of stroke cases can be avoided by preventing stroke risk factors, understanding the health and risk profile of this population is necessary. **Objectives:** To describe the rate of Hospitalization of stroke and its modifiable risk factors in the state of Ceará (Brazil) between the years 2020 and 2021. **Methods:** The number of Hospitalizations for stroke in the state of Ceará to calculate its incidence rate was provided by the website of DATASUS-tabnet and the Brazilian Institute of Geography and Statistics (IBGE). In addition, specific data regarding the main modifiable risk factors for stroke (Systemic Arterial Hypertension, Diabetes mellitus, alcohol consumption, smoking, physical inactivity and overweight) were obtained from the surveillance system of risk and protective factors for chronic diseases by telephone survey (VIGITEL). **Results:** The incidence rate of Hospitalization per 100,000 inhabitants was 76.12 in 2021, a higher number than in 2020 (71.93). In the same period, there was an increase in the frequencies of risk factors for stroke: Systemic Arterial Hypertension [2020 (21.4%); 2021 (22.8%)], Diabetes Mellitus [2020 (7.3%); 2021 (9.0%)], and regular alcohol consumption [2020 (17.6%); 2021 (18.3%)]. Active smoking was the only factor that decreased its frequency, from 7.6% to a total of 7.2% of smokers. Despite this, levels of exercise and quality diet, protective factors for stroke, showed lower values in 2021, with less than half of individuals following the recommended level of adequate physical activity of 150 minutes per week [2020 (45.9%); 2021 (35.3%)]. Approximately 65.7% of these individuals were totally sedentary. In addition, most of the population keeping overweight or obese [2020 (82.2%); 2021 (82.9%)]. **Conclusion:** The high number of Hospitalizations for stroke, and the incidence of risk factors suggest the need to implement effective actions to control stroke in Ceará. The still elevated numbers of common modifiable risk factors for stroke and the unwanted decline of protective factors prompt the emergence of new public health approaches to stroke prevention.

Doença Cerebrovascular

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EPIDEMIOLOGY OF CEREBROVASCULAR ACCIDENT IN PATIENTS WITHOUT COMORBIDITIES: A DESCRIPTIVE ANALYSIS

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Cerebrovascular accident (CVA) is a sudden neurological deficit caused by ischemia or hemorrhage of the central nervous system, with a predominance in females, aged over 50 years, with the middle cerebral artery being the most affected. Among the risk factors, the highest is systemic arterial hypertension. In addition, smoking, accumulation of abdominal fat, a diet rich in fats and carbohydrates, dyslipidemia, diabetes, heart disease, and previous vascular events are included. Stroke patients may present with some signs such as aphasia, dizziness, headache with no known cause, decreased or loss of consciousness, nausea or vomiting, and motor deficits. The aim of the study is to analyze the epidemiology of stroke in patients who did not have associated comorbidities. The study consists of a descriptive quantitative analysis of patients treated at the Neurosurgery Service of Hospital Santa Isabel, in Blumenau, Santa Catarina, between the periods of 2016 and 2020. From a group of 27 patients, 7 patients were selected who did not have comorbidities associated with the vascular condition. Of the 7 patients selected, the variables selected for frequency analysis were sex, age, symptoms, stroke characteristics, procedure, and comorbidities (SAH, diabetes, dyslipidemia, smoking, previous stroke, or previous TIA). Of the patients analyzed, 71.4% were female and 28.6% were male, and of these 57.1% were between the 5th and 6th decade of life, 14.3% in the 8th decade of life, 14.3% in the fourth decade of life, and 14.3% in the 2nd decade of life. Regarding the symptoms presented, 71.4% had some motor deficit, 57.1% of the patients had headaches, 57.1% had aphasia, 57.1% had altered level of consciousness, 14.3% had dizziness and 14 had 3% nausea. As for the characteristics of the lesion, 85.6% of the patients had a stroke after endovascular procedures and 14.3% of the patients had a spontaneous stroke. 71.4% of patients had lesions in the middle cerebral artery, 14.3% in the anterior cerebral artery, and 14.3% in the cavernous segment of the internal carotid artery. Of these injuries, 71.4% occurred on the left side and 28.6% on the right side. In 71.4% of the patients, the mechanism of approach was chemical fibrinolysis and in 28.6% it was mechanical thrombectomy. None of the patients had previous treatment or comorbidities. Thus, it is clear that the patients, even without presenting risk factors, met the epidemiological data that characterize stroke in the literature.

Doença Cerebrovascular

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REDE CUIDAVC APP: DEVELOPMENT OF A MOBILE APPLICATION FOR STROKE PREVENTION

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Background: The fast-growing stroke burden in the world suggests that current healthcare is failing to reduce modifiable risk factors in the population. Mobile apps became part of our everyday life may offer an opportunity to improve healthy behaviors and potentially be useful for stroke prevention. **Objective:** To describe the development of the Rede CuidAVC App as new digital technology to improve long-term health behaviors and trace the stroke survivors' needs according to their residence location. **Methods:** The App development took four stages to be concluded: 1. conceptual modeling; 2. navigation project; 3. abstract design of the interface and 4. Implementation. The design was based on criteria for Primary Stroke/CVD Prevention Digital Tools: Scientific evidence; Target population and purpose of the tool; Scalability; Interface, usability, and privacy protection; Interactivity and Engagement. **Results:** The first stage of Conceptual modeling includes what is recommended as evidence-based preventive recommendations; Target population and purpose of the tool with general information about stroke; and Interactivity and engagement with alerts and reminders with at least one motivational technique. At this stage, the seven (7) main risk factors were defined according to the American Heart Association and included a self-reported questionnaire approaching the main modifiable risk factors. Based on the user responses, recommendations for changes in lifestyle habits and how to manage the risk factors will be sent to users. The App also complies with scalability by being designed to run on a computer, smartphones, and internet browsers. The second and third stages, Navigation project and Abstract interface design, applies to the Interface, usability, and privacy recommendation. In those stages, the concern was to provide a clean, simple, and easy-to-use interface. For this, the applications were divided into screens: Home; Address registration; Cardiovascular Health profile and make an appointment (for Stroke individuals users with motor disorders) for an evaluation with the neurological team of physiotherapists of the Federal University of Ceará. The fourth stage is still in progress with validation and implementation studies to be carried out with health professionals and users. **Conclusion:** The Rede CuidAVC App has the potential to reduce modifiable risk factors and better manage access to post-stroke rehabilitation in the state.

Doença Cerebrovascular

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WHAT CAUSES THE INCREASED FREQUENCY OF ISCHEMIC CEREBROVASCULAR ACCIDENT CASES IN PATIENTS INFECTED WITH SARS-COV-2 VIRUS? A SYSTEMATIC REVIEW

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Background: The hypercoagulant characteristic of Covid-19 is a factor of high impact in the lives of affected patients. The syndrome hypercoagulation promotes cases of ischemic stroke, which occurs when there is blockage or rupture of a cerebral artery, even after the passage of high waves of contamination, however, specific studies of the characteristic are still insufficient, making treatment and prevention of the problem inefficient. **Objective:** To analyze the causes of the prevalence of ischemic stroke cases in patients contaminated by the SARS-CoV-2 virus. **Method:** The results established by incidence studies published from the year 2017 to 2022 with the keywords: "SARS-CoV-2", "Covid-19", "Ischemia" and "Stroke" in the MEDLINE and PUBMED databases were selected and analyzed for this systematic review, excluding publications that portrayed patients who were contaminated with the SARS-CoV-2 virus only after presenting ischemic conditions and publications that portrayed cases of ischemia that were not classified as cerebral ischemia. Their characteristics and results were analyzed to determine the probable reasons for the increase in cases of ischemic stroke in infected persons. **Results:** Studies have stated that cases of ischemic stroke in patients infected with the SARS-CoV-2 virus are more common in patients with severe infections and in patients with preexisting cases of ischemic risks. Research also shows that cases of stroke accompanied by Covid-19 are associated with cases of endotheliopathy, systemic inflammatory responses, and a high Von Willebrand Factor. **Conclusion:** The increase in ischemic stroke cases is shown to be related to the hypercoagulant characteristic of the SARS-CoV-2 infection, a characteristic influenced by points such as a high Von Willebrand Factor. Despite the presentation of a number of probable motivations, there is no study to guarantee that new factors may not have a major influence on the increase in cases. New studies are also needed to evaluate if there is a predominance of cerebral ischemia or a homogeneous distribution among other types of ischemia due to the influence of the virus and its characteristics.

Doença Cerebrovascular

TL 1105630

PROFILE OF PATIENTS ADMITTED TO A STROKE UNIT OF A TERTIARY HOSPITAL AND OUTCOMES AFTER A 3-MONTH POST-DISCHARGE FOLLOW-UP

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Background: Stroke is a major public health issue that leads to elevated mortality and morbidity. Specialized Hospital care and standardized protocols are crucial in minimizing disability and intra-hospital complications. Stroke Units (SU) provide the best multi-professional care, leading to reduced Hospital stay length and adverse outcomes for these patients. **Objective:** To describe the profile of inpatients of a SU in a Tertiary Hospital and functional outcome after a period of 3-month follow-up post-discharge. **Methods:** This is a single-center, retrospective study of inpatients admitted to a SU in a Tertiary Center during 2020, which received standardized multi-professional health care. Fifty-nine individuals with hemorrhagic or ischemic stroke were enrolled. The following data were assessed: chemical thrombolysis with Alteplase (rtPA), thrombolytic door-to-needle time (DNT), the prevalence of major adverse outcomes due to rtPA, NIH Stroke Scale (NIHSS) at Hospital admission, NIHSS at SU admission, NIHSS at discharge length of Hospital stay, the prevalence of adverse outcomes during the Hospital stay, functional status through modified Rankin Scale (mRS) at Hospital discharge and after 3 months of discharge, through a remote assessment by telephone call. **Results:** Mean age was 65.7±18 years, and 52% were women. Six patients (10.1%) were diagnosed with hemorrhagic stroke and 53 (89.9%) with ischemic stroke, which included 6 individuals with wake-up stroke. Among the ischemic stroke group, 19 (35.8%) received rtPA, with a mean DNT of 47 ±18 minutes, and none had hemorrhagic transformation or anaphylaxis. Median NIHSS was 7 at Hospital admission, 3 at SU admission, and 2 at discharge. Regarding adverse outcomes during the Hospital stay, 1 patient (1.6%) deceased, 8 (13.5%) had pneumonia, none had DVP or pressure ulcers. The mean length of Hospital stay was 7.5. Mean mRS at Hospital discharge was 02. After a 3-month period post-discharge, 44 patients were reassessed and 30 (68.2%) individuals had mRS 0-2, 8 (18.2%) had mRS 3-5 and 6 (13.6%) had mRS 6 by any cause. **Discussion:** SU and standardized multi-professional health care might benefit stroke patients and reduce the length of Hospital stay and adverse outcomes. Our data regarding a Brazilian SU stroke are in alignment with previous international data.

Doença Cerebrovascular

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DEATHS FROM STROKE IN THE STATE OF CEARÁ IN THE PANDEMIC (2020-2022) AND PRE-PANDEMIC (2017-2019) PERIOD: AN ECOLOGICAL TIME SERIES STUDY

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Background: The stroke is a neurological emergency in which there is an obstruction in the oxygenation to the brain. It consists of an alteration in the blood flow in the nervous tissue that can be caused by ischemia, when there is an obstruction of blood vessels, or hemorrhage, when there is a rupture of the vessel. This interruption in the blood flow causes death of nerve cells and damage functions in the body, according to the injured brain region. **Objective:** To analyze the variation in the number of deaths from stroke in the state of Ceará in the pandemic period (2020-2022) and pre-pandemic period (2017-2019). **Methods:** This is an ecological time-series study that analyzed the number of deaths from stroke in the state of Ceará from March 2020 to March 2022 and compared with the pre-pandemic period, January 2017 to January 2019. The data were collected through the Tabnet Platform (DATASUS) and analyzed according to age group, sex, metropolitan region, year and month of occurrence and color/race. **Results:** In metropolitan regions, there was an increase in the incidence of cases, except for the Cariri region, which had a decrease of 13.0%. The analysis according to the gender variable revealed an increase in the total number of cases, however, in the race/color variable, only mixed race expressed an increase in the number of cases. When analyzing the age group, people aged 10 to 29 years and 80 years or older had a lower mortality rate in the pandemic period, while those aged 30 to 79 years had a higher mortality rate. When analyzing the data by month, it was found that from January to July was presented a drop in the number of cases, and the numbers from July to December continued to increase. **Conclusion:** The public health dynamics during the Covid-19 pandemic reflected on the prevalence and prognosis of various diseases, as well as stroke. In the state of Ceará, there was a 5.9% increase in cases in the pandemic period. The proportionally greater increase found, 44.0% in brown people in the color/race variable, may be related to the fact that these people are the majority of those who are in situations of social vulnerability. The results of the study require further research to investigate the real impact of the pandemic on stroke mortality, since it may have various influences.

Doença Cerebrovascular

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CARDIAC MAGNETIC RESONANCE IMAGING ABNORMALITIES ARE ASSOCIATED WITH STROKE RECURRENCE IN PATIENTS WITH EMBOLIC STROKE OF UNDETERMINED SOURCE: A PROSPECTIVE COHORT STUDY

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Background: An unknown fraction of embolic strokes of undetermined source (ESUS) are thought to be cardioembolic. Previously published baseline data from this cohort showed that cardiac magnetic resonance imaging (CMRI) abnormalities are present in over one-quarter of ESUS patients, especially in patients with coronary artery disease or Chagas disease. However, it is not known if these CMRI abnormalities can affect the incidence of stroke recurrence. **Objectives:** To identify the association between an abnormal CMRI and stroke recurrence. **Methods:** Prospective cohort study. Participants were 18 years old, with previous ESUS according to the Causative Classification of Stroke. CMRI was performed at baseline, between 2012-2016. An abnormal CMRI was defined as the presence of wall fibrosis, wall edema, intracardiac thrombus or left ventricle aneurysm. Follow-up occurred every three months, using the Questionnaire for Verifying Stroke-Free Status and neurologic evaluation with neuroimaging to identify stroke recurrence. Survival analysis was performed for the primary outcome. **Results:** Follow-up was obtained from 30 (61%) out of 49 normal CMRI and 13 (77%) out of 17 abnormal CMRI patients. Baseline clinical characteristics were similar for both groups except for a higher frequency of Chagas disease among patients with abnormal CMRI (46% versus 17%). Mean age was 52 ± 15 and $23 (53,3\%)$ were female. No patient had coronary artery disease. Mean follow-up time was 75 ± 21 months for abnormal CMRI and 81 ± 24 months for normal CMRI. Stroke recurrence incidence was 39.8 (IC95% 8-116) per 1000 patient-years if CMRI was abnormal and 5 (IC95% 0-28) per 1000 patient-years if CMRI was normal (log-rank test: $p=0.04$). Regarding health status during follow-up, anticoagulation was started in 5 (38%) patients with abnormal CMRI and 1 patient (3%) with normal CMRI ($p=0.03$). We noticed no difference in emergency department visits or atrial fibrillation diagnosis during follow-up. **Conclusion:** An abnormal CMRI is associated with higher stroke recurrence in ESUS patients, suggesting that this method may be useful to further stratify stroke risk in patients with ESUS.

Doença Cerebrovascular

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OVERVIEW OF VASCULAR NEUROSURGERY PROCEDURES IN BRAZIL BEFORE AND AFTER THE COVID-19 PANDEMIC: AN ECOLOGICAL TIME-SERIES STUDY

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Background: Vascular neurosurgery comprises a wide range of procedures for the treatment of various health problems, which include brain arteriovenous malformation, ischemic and hemorrhagic strokes, and cerebral aneurysms. The Covid-19 pandemic created a global health crisis and had an overall impact on health care, delaying the scheduling of elective treatments and reducing the demand for emergency care. It is not known, however, if the pandemic has affected more complex procedures, such as vascular neurosurgery. **Objective:** to analyze the frequency and variation of vascular neurosurgery procedures in Brazil, from 2012 to 2021. **Methods:** This is an ecological time-series study that used secondary data from the SUS's Hospital Information System. Vascular neurosurgery procedures registered from 2012 to 2021 in Brazil were analyzed. The study's main outcome was the evolution of the surgery, and the data were analyzed according to the region of residence. The mortality rate was calculated according to the total number of procedures performed, as a percentage. The prevalence of events was estimated according to occurrence per 1,000,000 inhabitants. **Results:** 22,053 vascular neurosurgeries were performed in the period, of which 2,361 resulted in deaths. The mean (\pm SD) of procedures performed in 2020 and 2021 (pandemic period) was 41% lower than between 2012 and 2019 ($1,260 \pm 170$; $2,170 \pm 904$, respectively). The analysis of the year-on-year percentage change reveals that the largest changes were recorded between 2014 and 2015 (-26%) and 2020 and 2021 (-17%). The total number of procedures oscillated by -65% in the period. The mortality rate was around 9%, with the year 2021 having the highest mortality in the period (14%). The South region had the highest mean (\pm SD) number of surgeries/1,000,000 inhab. (13 ± 7) and the Northeast region the smallest (8 ± 4). **Conclusion:** the number of vascular neurosurgeries procedures performed in Brazil decreased from 2012 to 2021 and has shown a sharp drop in the years 2015 and 2021. The comparison between the period before and after the Covid-19 pandemic revealed that, in the latter, the average of procedures was lesser. The mortality rate was higher in 2021. More studies should be carried out to confirm the hypothesis that the Covid-19 pandemic changed the care dynamics in the vascular neurosurgery scenario and influenced the prognosis of the procedures performed.

Doença Cerebrovascular

TL 1105672

INCREASED FOVEAL AVASCULAR ZONE IN CHAGAS DISEASE REINFORCES VASCULAR-MEDIATED HYPOTHESIS: A CROSS-SECTIONAL STUDY

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Background: Chagas Disease (CD) is associated with cerebral atrophy, silent cerebral microembolism, stroke, and cognitive dysfunction in patients with heart failure (HF), all independently of HF severity. Two theories were proposed to explain these associations: microvascular damage and/or neurodegeneration. The vascular and nerve fiber layers of the retina are assessable non-invasively using Optical Coherence Tomography (OCT) and Optical Coherence Tomography Angiography (OCT-A), which may be useful to understand if the pathophysiology of CD is vascular- or neurodegenerative-mediated. **Objective:** To investigate the association between CD and abnormalities detected in OCT and OCT-A. **Methods:** Cross-sectional study from a prospective cohort of individuals aged ≥ 18 years, fulfilling Framingham criteria for HF. CD was defined by a positive ELISA or immunofluorescence test performed in duplicate. Exclusion criteria were diabetes mellitus, pregnancy, glaucoma, previously diagnosed retinal diseases, or stroke. We performed OCT and OCT-A in patients between 2020-2022 and excluded tests with exam quality $<50\%$. We conducted a univariate analysis considering the mean of both eyes. **Results:** We included 32 patients with CD and 20 without CD. Baseline clinical characteristics were similar for both groups except for a higher frequency of females (88% vs 50%) and higher ejection fraction (mean 65.5 ± 9.2 vs mean 41.0 ± 19.5) among patients with CD. In OCT, the foveal thickness was lower in CD patients (mean $230 \mu\text{m} \pm 18.1$) compared to those without CD (mean $241 \mu\text{m} \pm 24.7$), but not statistically significant ($p=0.13$). No difference was noticed in the thickness of retinal nerve fiber or ganglion cell layers. Regarding OCT-A, the superficial plexus foveal vessel density was lower in CD patients (mean $12.9\% \pm 5.10$) compared to those without CD (mean $16.6\% \pm 4.23$), with $p=0.02$. The deep plexus foveal vessel density in CD (mean $26.7\% \pm 7.10$) was lower than in patients without CD (mean $30.8\% \pm 6.21$) but not with statistical significance ($p=0.05$). There was no difference in vessel density considering the whole image, perifoveal or papillary regions. **Conclusion:** Chagas disease was associated with decreased foveal vascular density, but no changes in nerve fiber or ganglion cell layers of the retina. This finding is common to microvascular retinopathies, suggesting that CD may damage microcirculation beyond neuroimaging-detected damage with tests such as magnetic resonance imaging.

Doença Cerebrovascular

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HOSPITAL ADMISSIONS FOR STROKE AND STROKE IN ADULTS BEFORE AND DURING THE COVID-19 PANDEMIC. AN ANALYSIS OF DATASUS

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Background: Over the course of the COVID-19 pandemic, numerous evidences have demonstrated a close relationship between severe acute respiratory syndrome 2 (SARS-CoV-2) and the development of serious neurological manifestations, such as stroke. Given that Brazil was one of the countries most affected by the pandemic, an analysis of the variation in the number of Hospitalizations for stroke before and during the pandemic period is essential. **Objective:** To carry out an investigative analysis of changes in the epidemiological profile regarding the number of Hospitalizations for stroke in Brazil during the pandemic in view of the pre-pandemic period. **Methods:** Epidemiological, retrospective, cross-sectional and quantitative study, which collected data from the Department of Informatics of the Unified Health System (DATASUS) regarding the occurrence of Hospitalizations for stroke in Brazil in adults aged between 20 and 39 years, in a period of 24 months before the SARS-CoV-2 pandemic compared to 24 months after its onset. The study included Hospitalizations for strokes not specified as ischemic (istroke) or hemorrhagic (hstroke) (ICD-10 I64), transient ischemic strokes (ICD-10 G45) and cerebrovascular syndromes (ICD-10 G46). variables studied were: number of Hospitalizations, age group, sex and color/race. Statistical analysis was performed using the Excel program, using the Collective Health measures present in descriptive statistics. **Results:** 18,733 Hospitalizations were observed during the pre-pandemic period and 17,394 during the pandemic period. In the male population, there was a variation from 8,367 to 7,956 and in the female population, from 10,366 to 9,438. In adults aged 20 to 29 years, the variation was from 5,508 to 5,111 and in the age group from 30 to 39 years, it was from 13,225 to 12,283. The only variable that showed an increase was in the indigenous population, with an increase of 5.55 %, however the number of reported cases was small, from 18 cases before the pandemic to 19 cases during the pandemic period. **Conclusion:** In general, as in the group analyses, there were no changes regarding the number of Hospitalizations comparing the pre-pandemic period with the first two years of the pandemic by SARS-COV-2 in SUS through DATASUS.

Doença Cerebrovascular

TL 1105725

ASSESSMENT OF HOSPITALIZATIONS FOR TIA IN THE UNIFIED HEALTH SYSTEM FROM 2017 TO 2022

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Background: Transient ischemic attack (TIA) is an important risk factor for ischemic stroke. **Objective:** To determine the frequency of Hospitalizations for TIA in the different federative regions of Brazil in a period of 05 years. **Methods:** Through the database of the Ministry of Health, the Hospital Information System of the SUS (SIH/SUS), the records of transient ischemic strokes (ICD-10 G45) of patients Hospitalized between January 2017 and January were selected. 2022, according to SUS Hospital Morbidity and place of residence in Brazil. Being stratified in the federative regions: North, Northeast, South, Southeast and Midwest. The analysis was performed per 100,000 inhabitants. **Results:** 10,1447 cases were identified, 38.2% in the Southeast region, 26.7% in the South region, 20.8% in the Northeast region, 8.6% in the North region and 5.4% in the Midwest region. When analyzed per 100,000 inhabitants, the southern region had the highest prevalence, 0.089/100,000 inhabitants, followed by the North region (0.046/100,000 inhabitants), the Southeast region (0.043/100,000 inhabitants), the Northeast region (0.037/100,000 inhabitants) and the Midwest region. (0.033/100,000 inhabitants). **Conclusion:** according to the analysis of DATASUS, the rates of Hospitalization for TIA in the SUS occur with higher prevalence in the southern region compared to other regions of the country.

Doença Cerebrovascular

TL 1105852

ANALYSIS OF DOOR-TO-CT AND DOOR-TO-NEEDLE TIME IN PATIENTS WITH SUSPECTED STROKE AT THE CLINICAL HOSPITAL OF THE FEDERAL UNIVERSITY OF MINAS GERAIS, FROM 2018 TO 2021

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Background: Stroke is one of the most important causes of morbimortality around the world. Trombolysis can improve patients outcomes, but time is crucial: When the patient gets to the emergency room, a brain computed tomography (CT) must be performed in 25 minutes and the intravenous (IV) trombolysis (first-line therapy) must be initiated no more than 4.5 hours after the symptoms onset and within one hour (60min) after admission. **Objective:** To evaluate the latency to perform brain CT (door-to-CT time, DTC) and to initiate IV trombolysis (door-to-needle time, DTN) in patients with suspected stroke at the Clinical Hospital of the Federal University of Minas Gerais, and to identify possible flaws in the management of these patients. **Methods:** We analyzed medical records of patients admitted with suspected stroke, from 2018 to 2021. We also invited professionals involved in stroke care (neurologists and nurses, both in the emergency room and in the intensive care unit – ICU) to fill forms containing questions about the initial management of stroke. **Results:** A total of 947 patients were selected. 63 were submitted to IV trombolysis. The average DTC time and DTN time were 22min and 81min, 17min and 63min, 12min and 56min, 19min and 62min in 2018, 2019, 2020 e 2021, respectively. In addition, 30 professionals answered the questions, among these, 12 neurologists and ICU physicians, 16 nurses and 2 others professionals (clinical pharmacy and radiology technician). 20% answered correctly about maximum DTC time, 46,7% about maximum DTN time and 70% about maximum time after symptoms onset to initiate IV trombolysis. Also, 17 professionals have reported problems with high demand in the emergency room and 22 with team communication. **Conclusion:** Despite achieving the goal of performing a CT within 25min or less, the time to initiate the IV trombolysis after patient admission had not been reached. Furthermore, the percentages of correct answers indicate the need of professionals education update and to optimize the Hospital protocols in order to minimize complaints about team communication and professionals overload.

Doença Cerebrovascular

TL 1105875

RECANALIZATION THERAPIES IN ACUTE ISCHEMIC STROKE: THE EXPERIENCE OF A BRAZILIAN UNIVERSITY HOSPITAL

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Background: Stroke is the second leading cause of death and disability in the world. Ischemic stroke (IS) corresponds to 85% of cases and its treatment reduces sequelae and mortality. However, recanalization therapies for stroke are available only to a small portion of the population and there are few data in the Brazilian literature on this topic. **Objectives:** To evaluate the clinical characteristics of patients undergoing recanalization therapy treated at a public University Hospital in Ribeirão Preto; to identify the predictors of in-Hospital mortality and symptomatic hemorrhagic transformation (HTs) and good functional outcome using the Modified Rankin Scale (mRS) 0-2 at 90 days after therapy was instituted. **Methods:** A review of the medical records of patients with acute stroke who underwent endovenous thrombolysis and/or endovascular treatment between the years 2001 to 2017 was performed. **Results:** There were 869 patients treated, most of them male (57%), mean age of 66.5 ± 13.1 years, median NIHSS 15 and 65.7% with proximal occlusion. The rate of intravenous thrombolysis was 87.5% and endovascular treatment was 28.7%. Mean admission-needle time was 57 minutes [35-70] and ictus-needle time was 204 minutes [150-248]. HTs occurred in 5.9% of patients, in-Hospital death in 16.8% and the median mRS after 90 days was 3. The predictors of good functional outcome (0-2) were age and NIHSS; for Hospital mortality, in addition to these, diabetes mellitus, proximal occlusion and HTs; for HTs, NIHSS, proximal occlusion and blood glucose. **Conclusion:** The profile of patients with acute ischemic stroke is predominantly male, in the sixth decade of life, with several comorbidities, admitted within 2.5 hours of symptom onset, high blood pressure and blood glucose levels, in addition to severe and severe symptoms. high rate of proximal occlusion. The HT rate was similar to those reported in international and national studies. The shorter treatment time had an impact on the functional outcome, supported by previous evidence. The present study presents the largest series of patients treated with thrombolysis and/or thrombectomy in Brazil, throughout a historical serie, showing the safety of recanalization therapies and less chance of sequelae, with 40% of patients having a good functional outcome.

Doença Cerebrovascular

TL 1106028

NEURORADIOLOGICAL MARKERS OF VASCULAR COGNITIVE IMPAIRMENT AFTER STROKE

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Background: Cognitive vascular impairment (CCV) is a frequent, but overlooked, possible consequence of stroke. The most relevant markers in Magnetic Resonance Imaging (MRI) seem to be, among others, strategic location, severity of white matter changes, as well as the degree of atrophy of the medial temporal lobe. **Objective:** To assess the relationship between stroke and CCV using markers from MRI. **Methods:** Systematic review of observational studies published between 2005 and 2020. The search was carried out in the PubMed and SciELO databases with the keywords "stroke", "MRI", "Vascular cognitive impairment". The PRISMA check-list was used to guide this review. **Results:** 8 studies were selected. "Event location" was the marker in MRI of the skull most frequently considered (7 studies). It proved to be a statistically significant marker ($p < 0.05$) for the prediction of CCV in 6 out of 7 studies. 75% of the studies included in this review evaluated the relationship between the presence of "hyperintensity in the white matter" at MRI and CCV. However, this marker was shown to be statistically significant in 50% of these studies. **Conclusion:** A review that brought together the assessment of a wide range of possible neuroradiological predictors of CVD after stroke had not been carried out so far. Yet it would be particularly useful to evaluate the markers in a more homogeneous way in a study with a larger sample size.

Doença Cerebrovascular

TL 1106040

SHORT-TERM EFFICACY OUTCOMES OF TENECTEPLASE VERSUS ALTEPLASE FOR ACUTE ISCHEMIC STROKE: META-ANALYSIS OF 5 RANDOMIZED TRIALS

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Background: Tenecteplase (TNK) has been shown to be non-inferior to Alteplase (ALT) for long-term efficacy and safety outcomes. Whether this also applies to short-term efficacy outcomes such as early clinical improvement and recanalization is unknown. **Objective:** Compare TNK and ALT regarding the short-term efficacy outcomes early neurological improvement and recanalization. **Methods:** The Preferred Reporting Items for Systematic Reviews and Meta-Analyses PRISMA was used to conduct a meta-analysis, adapted to noninferiority analysis. The primary outcome was early (24-72 hours) neurological improvement, defined as either National Institutes of Health Stroke Scale (NIHSS) score 0 or reduction of at least 8 points compared to baseline. Recanalization was a secondary outcome. The noninferiority margin was set at 6.5%. **Results:** Search strategy yielded 5 randomized clinical trials (1585 patients: 828 TNK, 757 ALT). Across all trials, mean age was 70.8, 58.8% were men, mean baseline NIHSS was 7 and mean onset-to-treatment time was 148 minutes. Patients in intervention group received TNK at doses of 0.1mg/kg (6.8%), 0.25mg/kg (24.6%), and 0.4mg/kg (68.6%), while all ALT patients received 0.9mg/kg. In random-effects meta-analysis, TNK was non-inferior to ALT for the primary outcome early major neurological improvement (risk difference 8% in favor of TNK, 95% CI 1-15%). Recanalization was also non-inferior for the TNK compared to the ALT group (risk difference 9% in favor of TNK, 95% CI -6% to 23%). Fixed-effects models yielded similarly non-inferior results and signaled for a possible TNK superiority for both early neurological improvement and recanalization. **Conclusion:** TNK is non-inferior to ALT at the short-term efficacy outcomes early neurological improvement and recanalization.

Doença Cerebrovascular

TL 1106065

PROFILE OF PATIENTS HOSPITALIZED FOR VENOUS THROMBOSIS CEREBRAL IN NORTHEASTERN BRAZIL

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Background: Cerebral venous thrombosis (CVT) is a rare disorder and presents a high risk of morbidity and mortality. The signs and symptoms can be diverse, which makes this condition a diagnostic challenge. **Objective:** to characterize the clinical and epidemiological profile of patients diagnosed with cerebral venous thrombosis at a University Hospital in Teresina. **Methods:** This is a retrospective, cross-sectional and descriptive study, carried out through the evaluation of electronic medical records of patients admitted to the Neurology Service of a University Hospital in Teresina/PI, from May to July 2021. **Results:** The sample consisted of 13 medical records. Most CVT patients were female (69.2%), with a mean age of 39.2 years. Headache (92%) was the most common clinical manifestation. The use of oral contraceptives was the risk factor most associated with CVT, with 46%. The mean length of stay observed was 18.1 days. **Conclusion:** Despite being a potentially serious disease, no death was observed in the study population, which may be linked to adequate diagnosis and timely treatment, capable of reflecting on the successful outcome of patients with CVT.

Doença Cerebrovascular

TL 1106141

CEREBRAL VENOUS THROMBOSIS SECONDARY TO THE MUTATION IN THE PROTHROMBIN GENE IN THE CONTEXT OF VACCINATION AGAINST COVID-19: CASE PRESENTATION

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Case Presentation: A 15-year-old woman came to the consultation with a progressive headache for 6 weeks, after 30 days of vaccination against the coronavirus with the Ad26.COV2 vaccine. S. She Denied use of contraceptives. She evolved with drowsiness, neck stiffness, abducens nerve paresis, low visual acuity (BAV), vomiting, ataxia and mild asymmetric tetraparesis. Optical Coherence Tomography (OCT) showed bilateral papilledema. Brain Magnetic Resonance (MRI) and CSF examination without alterations. Spinal manometry confirmed Intracranial Hypertension (ICH) (75cmH2O). A relief lumbar puncture was performed and acetazolamide was started. Arteriography showed Venous Thrombosis of the sigmoid sinus and left transverse. The investigation of alteration of factor V Leiden, antithrombin III, Protein C and S, antiphospholipid antibodies were normal. A positive result was obtained for the mutation of the prothrombin gene 20210A in heterozygosity. Full anticoagulation was performed with 5 mg warfarin (INR between 2 and 3) and bridging with enoxaparin, associated with topiramate 25 mg/day. The patient remains under outpatient follow-up, with no signs of disease recurrence. **Discussion:** Cerebral venous thrombosis (CVT) is defined as the presence of a thrombus within a venous sinus, superficial intracranial vein or deep intracranial vein. CVT has a higher incidence in young women, with additional risk factors, such as pregnancy, puerperium and use of oral contraceptives². Headache is the most common symptom, but focal neurologic deficits, seizure episodes, and encephalopathies are also evidenced. Digital subtraction angiography is indicated for inconclusive cases. The coagulogram may indicate the presence of conditions that aid in the pathogenesis of CVT and the search for thrombophilic status and mutations is useful. Lumbar puncture is important to rule out meningitis³. In addition, other conditions such as previous vaccination for COVID-19 should be investigated, as vaccines based on adenovirus vectors are related to thrombotic episodes, through a mechanism called Vaccine-Induced Thrombocytopenic Thrombosis (VITT). **Conclusion:** It is noteworthy that CVT is an uncommon and reversible condition if properly diagnosed and treated. Its association with mutations in the prothrombin gene 20210A is well established and in relation to vaccination it is important to recognize that the risk of CVT by COVID-19 in the general population outweighs the risk of vaccines.

Doença Cerebrovascular

TL 1106156

AN UPDATED NON SYSTEMATIC REVIEW ON CEREBRAL AMYLOID ANGIOPATHY (CAA)

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Background: Cerebral amyloid angiopathy (CAA) is described as the accumulation of amyloid fibrils on the walls of blood vessels of the parenchyma of the central nervous system (CNS) and leptomeninges. Due to abnormal production or impaired clearance of the amyloid beta protein, structural changes in the vessels are lead, causing ischemic lesions and intracerebral hemorrhages (ICH) owing to small vessels fragility. **Objectives:** To describe the pathophysiology, criteria diagnosis and progression of CAA. **Methods:** Non systematic review in the Pubmed, Lilacs and Scielo databases, selecting articles in english, published from 2020 to 2022. **Results:** The pathophysiological progression of CAA remains uncertain, but has a strong contribution of a genetic component (APOE 2 and 4), related to changes in the astrocytarian reactivity and deficit of solubility factors. In addition, the impairment of perivascular spaces on the blood-brain barrier (BBB) or lymphatic system, accumulation of amyloid beta proteins, in the integrity of BBB by the loss of proteins on tight joints, leads to inflammatory effects by oxidative stress, which alters the permeability of the BBB and promotes toxicity and changes in vessel structure. Currently, the management of CAA remains focused in the prevention of recurrent ICH episodes, and the protocols for diagnosis still lack padronization - there is no consensus in the meanings of CAA's neuropathology. Also, a definitive diagnosis can only be confirmed via postmortem examination of the brain. Thus, to identify possible CAA patients, the Boston criteria was developed as a combination of clinical, pathological, and radiographic criteria that uses MRI findings, classifying the results in "possible" and "likely". According this criteria, the analyzed aspects of the MRI are strictly lobar cerebral microhemorrhages (small cerebral hemorrhages restricted to the cortical and subcortical regions of the brain), and superficial cortical siderosis (deposition of blood degradation products in the cortical sulci over the convexity of the cerebral hemispheres). The use of this criteria, in synthesis, can improve the chances of obtaining a diagnosis during the patient's lifetime. **Conclusion:** The poor understanding of the disease mechanisms reflects the lack of therapies able to limit the disease progression. The combination between experimental results and clinical data from large observational studies are mandatory to its understanding.

Doença Cerebrovascular

TL 1106208

CHAGAS' DISEASE AND ISCHEMIC STROKE: A STUDY OF ASSOCIATED FACTORS

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Background: Chagas' disease is an independent risk factor for stroke. The management and prevention of these cases are based on incomplete information or extrapolations of knowledge about other stroke etiologies. The occurrence of strokes without cardiac involvement in Chagas' disease has also been documented. **Objective:** The present study aims to verify the risk factors (clinical and laboratory) for the occurrence of stroke in patients with Chagas' disease. **Methods:** This is a cross-sectional study that included patients seen at the Chagas Disease outpatient clinic in the Cardiovascular Emergency Room of Pernambuco and the Neurovascular Outpatient Clinic at Hospital São Paulo - Federal University of São Paulo. Demographic, clinical and complementary cardiological exams (echocardiogram, electrocardiography, Holter-electrocardiography), in addition to those related to the investigation of stroke and its mechanisms were analyzed. It was defined as significant $p < 0.05$. The local research ethics committee of each participating center approved this study. **Results:** The sample consisted of 678 patients, 72 (10.6%) had a confirmed stroke. The mean age of the patients was 62 years old ($SD = \pm 12.3$) ranging from 14 to 94 years old, mostly women 67.6% and born in the state of Pernambuco (82%). The variables that showed a statistically significant association with stroke were: sex, congestive heart failure, alcoholism, not-sinus electrocardiogram, left bundle branch block, right bundle branch block, anterosuperior divisional block, atrial fibrillation, left ventricular ejection fraction, and use of oral anti-coagulants ($p < 0.05$). Variables with $p > 0.20$, except for drugs that are a consequence and not a cause of stroke, were included in the multivariate analysis, with heart failure, atrial fibrillation, right bundle branch block and anterosuperior divisional block being the risk factors that remained significant. **Conclusion:** Although risk factors for atheromatosis such as diabetes, dyslipidemia, obesity, smoking and alcoholism are present in patients with stroke and Chagas disease, the variables related to heart disease are the most related to the occurrence of stroke.

Doença Cerebrovascular

TL 1106215

MORTALITY AND FUNCTIONAL STATUS FIVE YEARS AFTER SURVIVING A STROKE: A PROSPECTIVE COHORT STUDY OF 1.005 PATIENTS IN FORTALEZA, NORTHEAST, BRAZIL

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Background: Stroke is among the leading causes of death and the second most common cause for disability-adjusted life years globally. Over the past decades, mortality related to stroke has decreased globally. The same, however, is not observed regarding impacts on personal, family, social and productive life aspects due to the ensuing disabilities, which remain high. Knowing the survival and disability rates after a stroke helps evaluate the performance of the line of care from primary prevention to pre-Hospital and Hospital care in the acute setting to rehabilitation and secondary prevention. In addition, it permits having a better understanding of the evolution of the disease and the effects of therapeutic interventions. **Objective:** To study the mortality and functional status five years after surviving a stroke in Fortaleza, Northeast Brazil. **Methods:** This prospective cohort study was carried out in Fortaleza to analyze patients' survival and functional state five years after a stroke. A translated and culturally adapted Brazilian Portuguese version of the Modified Rankin Scale (mRS) validated to be used over the telephone was adopted to interview 2,214 patients who survived having a stroke between February 3 and December 31, 2014. **Results:** From the 3,052 patients investigated by the State Stroke Registry, 838 (27.5%) died while Hospitalized. Of these 2,214 survivors, 1,209 patients were excluded (27 refused to participate, 217 did not answer the phone calls, and 961 changed their telephone number). The remaining 1,005 had a mean age of 66.8 ± 14.8 years. After five years, 347 (34.5%) patients had died; 124 (12.3%) did not present any deficit or disability; 116 (11.5%) presented some deficit but no disability, and 88 (8.7%) presented deficits that were considered severe disabilities and were bedridden. **Conclusion:** Our data show that five years after surviving a stroke, approximately one-third of patients are independent, one-third rely on assistance for their everyday lives, and one-third are deceased. These numbers are aligned with other studies. Age above 50 years, previous stroke, and atrial fibrillation were strong poor prognostic predictors, though obesity and dyslipidemia were paradoxically associated with better prognoses.

Doença Cerebrovascular

TL 1106220

PALLIATIVE CARE IN PATIENTS HOSPITALIZED FOR STROKE: AN 8-MONTH TIME FRAME DURING THE SARS-COV-2 PANDEMIC

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Background: Since March 2020, Brazil has been experiencing the waves of the pandemic caused by the SARS-COV-2 virus. Stroke is an important cause of mortality and morbidity in the country and in the world, and therefore it is imperative to consider Palliative Care in cerebrovascular diseases. During the pandemic, it was reported that patients were afraid to look for emergencies with symptoms of stroke for fear of contracting COVID-19. **Objective:** To describe Hospitalizations for stroke from October 2021 to May 2022; describe the cases of COVID-19 in these patients during Hospitalization; describe the outcome of these Hospitalized patients; describe epidemiological variables of patients (age and sex). **Method:** Cross-sectional, observational, quantitative study in patients admitted through the emergency department of a private Hospital from October 2021 to May 2022. Inclusion criteria were patients Hospitalized for ischemic, hemorrhagic stroke and Cerebral Venous Thrombosis (CVT). Patients Hospitalized for other cerebrovascular syndromes, as well as those who had a stroke after Hospitalization, were excluded. Data were collected from the patients' medical records and analyzed using Excel and SPSS programs. The study is part of the research project approved by the Hospital's CEP -CAE 48835021.6.0000.5043. **Results:** 75 patients were studied, 60 of which were diagnosed with Ischemic Stroke, 14 with Hemorrhagic Stroke and 1 with CVT. The mean age of the patients was 75.8 years and 60% were female. 86.66% were discharged from Hospital, 3 remained Hospitalized until the end of the clipping and 7 of them died. Of all patients, 6.66% were diagnosed with COVID-19 during Hospitalization. 11 patients were indicated for palliative care, and of these patients, 54.54% died, 18.18% were discharged from the Hospital and 27.27% were Hospitalized by the end of data collection. **Conclusion:** Most patients were Hospitalized for ischemic stroke, aged over 70 years and were discharged from Hospital. Of the 75 patients, 5 cases were found with a positive COVID-19 test and of the 7 deaths, 6 entered the Hospital's palliative care line. The study is relevant in assessing the importance of the stroke itself, even in the face of the pandemic. The authors also emphasize the importance of Palliative Care for stroke patients. There is a need for further analysis on the subject, in order to approach the patient and his family in the most appropriate and comprehensive way possible.

Doença Cerebrovascular

TL 1106249

MACHINE LEARNING APPROACH IN PREDICTION OF NASOENTRAL TUBE IN ACUTE ISCHEMIC STROKE

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Background: Dysphagia occurs in about 20 to 50% of acute stroke patients, and it may persist longer than six months after stroke. Nasoenteral tube feeding (NTF) must be judiciously prescribed to avoid dysphagia complications and, at the same time, prevent its unnecessary usage, which is not free of adverse events. **Objective:** Our main aim was to state independent predictive factors associated with dysphagia and nasoenteral tube feeding. Besides, we aimed to develop one prediction model for nasoenteral tube feeding through a machine learning modeling approach. **Methods:** This is a prospective cohort study. All consecutive ischemic acute stroke patients were included. All patients had at least one phonoaudiological evaluation for dysphagia screening. Data analyzed included age, sex, Glasgow coma scale, NIHSS, Aspects score, Seattle comorbid index, Rankin scale, previous and at discharge, TOAST classification for stroke subtypes, presence of major stroke risk factors and data from CT scan of all patients. **Results:** This research studied 1101 acute stroke patients. Twenty-eight percent of stroke patients went through nasoenteral tube feeding. They were older ($p < 0.001$), had a more severe stroke ($p < 0.001$), and presented consciousness disturbance and dysarthria more frequently ($p < 0.001$ for both measures). These findings predicted independently nasoenteral tube feeding in acute stroke. The decision tree model disclosed a sensitivity of 75% and specificity of 87%, with 84% accuracy for predicting nasoenteral tube feeding. On the other hand, the artificial neural network predicted 83% accurately, disclosing a sensitivity of 70% and specificity of 86%. An application for clinical decision support was developed based on the decision tree model, which disclosed better sensitivity and accuracy for nasoenteral tube feeding prediction. **Conclusion:** Dysphagia occurred in one third of cases. Older age, stroke severity, dysarthria, reduced conscious level at onset independently predict the need for nasoenteral tube. The tree decision model is a accurate tool for predicting NTF in acute ischemic stroke patients.

Doença Cerebrovascular

TL 1106261

TELECARE IN NURSING CONSULTATION TO STROKE PATIENTS USING WARFARIN: EXPERIENCE REPORT

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With The COVID-19 pandemic, health services had to make adjustments in their routines to harm patient care. Thus, medical and nursing telecare was incorporated as a strategy to combat the pandemic. This study aimed to report the experience of telecare in nursing consultation to stroke patients using warfarin. This was a descriptive study, the type of experience report. The scenario was the neurology outpatient clinic of a tertiary public Hospital, located in the city of Fortaleza, Ceará. Results a nurse and a neurologist implemented telecare for patients using warfarin in September 2020, through synchronous consultation, on Tuesdays, in the afternoon. The average number of attendance/day was 25. The neurology outpatient clinic was equipped with devices, which allowed real time video calls with users, and the health professionals were trained to use telecare system. The patients were previously scheduled, by phone call, and on the scheduled day, they started the consultation via WhatsApp, waiting in a virtual waiting room for care with the nurse. During the nursing consultation, the main activities developed were: patient/family welcoming; health status assessment/complaints/complications; monitoring of prothrombin time, whose examination file was sent by the patient; and registration in the follow-up form; adjustment of the dose of warfarin, according to the outpatient anticoagulation protocol adopted in the service; return consultation schedule; educational guidance on care related to the use of warfarin and registration of the telecare in the patient's medical record. In case of clinical complications, the patient was referred to teleconsultation with a neurologist via system. Telecare proved to be a useful tool that continued to be requested by patients, even with the return of face-to-face care. Patients who preferred this modality of care were those who with physical or financial problems or residents in other cities. The nursing teleconsultation allowed the simultaneous interactivity of the nurse with the patient using warfarin, contributing to the continuity of care and minimize health problems resulting from the treatment.

Doença Cerebrovascular

TL 1106267

IMPACT OF DELIRIUM DURATION ON STROKE OUTCOMES

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Background: Delirium is a common complication after stroke and its occurrence is associated with worse outcomes in this population. Longer delirium duration seems to be associated with higher cognitive impairment in critical illness. However, literature is scarce in studies investigating its association with death and functional dependence in stroke patients. **Objective:** We aimed to evaluate the impact of delirium duration on stroke prognosis. **Methods:** Prospective cohort of stroke patients with symptom onset within 72 hours before research admission. Delirium was diagnosed by Confusion Assessment Method in an Intensive Care Unit and coma was defined by Richmond Agitation-Sedation Scale of minus 4 (responsive only to physical stimulus) or minus 5 (unresponsive to physical stimulus). Delirium duration measurement was adjusted by considering as the main predictor the number of days alive free of delirium or coma (DFDC) after admission. We calculated delirium/coma-free days (DCFDs) as follows: DCFDs = 30 - days of delirium - days of coma - days deceased. We did not require that delirium/coma-free days occur consecutively. Our main outcome was functional dependence or death (mRS>2), evaluated by modified Rankin Scale, at 90 days post-ictus. **Results:** Three hundred sixty three patients were enrolled. Delirium was diagnosed in 85 (23,4) patients, who presented a median number of 25 (19,5-29,0) days alive without delirium or coma. Patients who developed delirium and died or became functionally dependent in long term had fewer DFDC [median (IQR): 25 (17,2-28) vs 29 (26-29) days, $p < 0,001$]. Multivariate analysis showed DFDC (odds ratio, 0,80 [IC 95%, 0,75-0,86]) as an independent predictor of mRS > 2 in 90 days, as well as age, sex, Charlson Comorbidity Index, National Institutes of Health Stroke Scale on admission, thrombolytic treatment and infection occurrence. **Conclusion:** A first episode of delirium after stroke is usually not the only one, being followed by other days of delirium or coma. Longer duration of delirium is associated with higher risk of death and worse functional outcome in this population. Delirium duration seems to be a parameter of significant clinical relevance, being a valid outcome to be adopted in clinical trials of delirium prevention or mitigation. To our knowledge, it is the first study to investigate prospectively the effect of delirium duration on stroke functional outcome.

Doença Cerebrovascular

TL 1106272

MILD CAROTID STENOSIS FREQUENCY IN PATIENTS WITH EMBOLIC ISCHEMIC STROKE OF UNDETERMINED SOURCE (ESUS)

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Background: Etiological classifications define carotid stenosis ≥50% as a source of ischemic stroke. However, some studies showed a higher frequency of mild carotid stenosis (<50%) ipsilateral to Embolic Ischemic Stroke of Undetermined Source (ESUS), as well as high-risk features, such as intraplaque hemorrhage, thrombus and ulceration. **Objectives:** To investigate the frequency of mild carotid stenosis and to assess whether it occurs more frequently ipsilateral than contralateral to cerebral infarction in ESUS. **Methods:** Cross-sectional study based on medical records of adult (≥18 years) ESUS patients from a stroke outpatient clinic. ESUS was defined as imaging-confirmed non-lacunar stroke; absence of ≥50% stenosis on arteries that supply the area of ischemia; and no cardioembolic or other rare specific sources of stroke. The frequencies of mild carotid stenosis and side of cerebral infarction were compared using Fisher's exact test. **Results:** A total of 55 participants with ESUS were enrolled. The mean age was 61±11.3 years and 28 (50.9%) were women. Mild carotid disease was present in 27 individuals (49.1%) and this group was significantly older (65.4±9.3 vs 56.8±12.2, p<0.001). Among these, 21 (77.8%) had bilateral mild carotid stenosis and unilateral cerebral infarction and two (7.4%) had bilateral stenosis and bilateral infarcts. Of patients with unilateral stenosis, two (7.4%) had contralateral and one (3.7%) had ipsilateral infarct. One (3.7%) participant presented with unilateral stenosis and bilateral cerebral infarction. There was no difference of mild carotid stenosis frequency when comparing ipsilateral, contralateral or bilateral infarct groups (p=0.456). Considering carotids as individual observations, we evaluated 110 carotids, 50 of them presenting mild stenosis, with 27 (54.0%) ipsilateral to stroke and 23 (46.0%) contralateral to cerebral infarction, with no significant association (p=0.848). **Conclusion:** In our sample, assessment of stenosis laterality is not sufficient to suggest an association with stroke, given the large volume of patients with bilateral stenosis. The evaluation of plaque high-risk features by means of exams such as carotid plaque MRI is a resource to be studied.

Doença Cerebrovascular

TL 1106293

CEREBRAL OXIDATIVE STRESS IN SYSTEMIC INFECTION AFTER ISCHEMIC STROKE

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Background: Ischemic Stroke (AVCI) represents 87% of all stroke, and in Brazil already reached 222.773 thousand individuals. AVCI has a greater effect in women than in men, then women have more events and are less likely to recover. The pathophysiological mechanisms of AVCI determine even such as neuroinflammation and oxidative stress, leading the individual to a systemic immune suppression, being sepsis one of them. The role of oxidative stress associated with inflammatory conditions is well characterized, event that can be observed in brain areas such as the prefrontal cortex and hippocampus. **Objective:** To evaluate the relationship between AVCI and sepsis through oxidative parameters in female rats submitted to the animal model of AVCI and sepsis. **Methods:** Female Wistar rats aged 60 days were randomized into groups: SHAM+SHAM, SHAM+MCAO, SHAM+CLP, MCAO+CLP, and submitted on day 0 to Middle Cerebral Artery Occlusion (MCAO) and on day 7 submitted to Cecal Ligation and Perforation (CLP) technique, suffering painless death after 24 hours and the hippocampus and prefrontal cortex were removed for biochemical analysis. **Result:** We observed that sepsis exacerbated reactive oxygen and nitrogen species in animals that suffered AVCI as well as increased sensitivity of lipid peroxidation in sepsis. The brain damage caused by AVCI induces protein oxidation and the association of two conditions was effective to potentiate a decrease of antioxidant enzyme catalase decrease. **Conclusion:** Sepsis can potentiate brain oxidative stress in female rats undergoing AVCI.

Doença Cerebrovascular

TL 1106307

CAUSES FOR NOT PERFORMING INTRAVENOUS THROMBOLYSIS FOR ISCHEMIC STROKE PATIENTS IN A HOSPITAL WITH A CERTIFIED STROKE PROGRAM

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Background: Thrombolytic treatment (TT) reduces the morbidity and mortality of ischemic stroke when performed within 4.5 hours after stroke symptoms. Therefore, it is crucial to expand the performance of this treatment. **Objective:** To understand the reasons for not performing TT in ischemic stroke patients. **Method:** We retrospectively evaluated the database of the stroke program of a private Hospital in the city of SP over one year. All data from patients admitted to the program during this period were analyzed. Patients that were transferred from other services were excluded from this analysis. The percentage of patients undergoing TT and the exclusion reasons were analyzed, based on the criteria of non-eligibility for this treatment. **Results:** Data from 272 patients were analyzed. Of these, 137 (59%) were included in the protocol (ischemic and hemorrhage stroke, TIA, subarachnoid hemorrhage). Ninety-five patients (69%) were confirmed as having ischemic stroke (69%). Of these, 40 patients were excluded because they had been transferred from other services. Of the 55 patients included in the analysis, 8 (14%) were submitted to TT. Of the 47 (86%) who did not receive TT, the causes were: ischemic stroke occurring outside the therapeutic window (75%), large ischemic stroke area on arrival or previous stroke that occurred less than 3 months ago (2%), postoperative period of recent major surgery (5%), use of oral anticoagulants (7%), and ischemic stroke with multiple comorbidities (12%). **Conclusion:** We found that the most important challenge to increase the percentage of cases treated with TT is to reduce the arrival time of patients to the health service. Educational actions for the population are crucial in this regard.

Doença Cerebrovascular

TL 1106320

CEREBRAL VENOUS THROMBOSIS IN TIMES OF PANDEMIC SARS-COV-2: NEUROCOVID STUDY CASE SERIES

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Background: Cerebral venous thrombosis (CVT) is a rare cause of stroke associated with significant morbidity and mortality. COVID-19 began in 2019, when an outbreak of atypical pneumonia occurred in the city of Wuhan (China), and the etiologic agent was identified as a novel human pathogen from the family Coronaviridae, called Severe Acute Respiratory Syndrome Coronavirus type 2 (SARS-CoV-2). As the pandemic progressed, cases of stroke in young patients without cardiovascular risk factors were reported and new cases of CVT associated with both the infection and vaccination were also described. **Objective:** To describe the clinical, epidemiological, and laboratory characteristics of patients with cerebral venous thrombosis admitted to a public referral Hospital. **Methods:** This is a case series study nested within the multicenter case-control entitled NeuroCOVID: Association of SARS-CoV-2 with the occurrence, prognosis and pathogenesis of cerebrovascular diseases, funded by the Inova Fiocruz Program (Fundação Oswaldo Cruz - COVID-19 Emergency Fund: VPPCB-005 FIO-20-2-22). **Results:** Over a 1-year period of data collection, 19 patients included had a clinical and radiological diagnosis of CVT. It is evident that 84.2% of the sample was composed of women and 42.1% self-declared as brown. Only 26.3% and 5.3% reported having hypertension and diabetes mellitus, respectively. Approximately 62.5% of women used contraceptives. Only 2 patients reported a diagnosis of COVID-19 in the 30 days preceding Hospitalization. Headache was the most reported symptom (89.5%), followed by seizures (26.3%). Approximately 63.2% had venous infarction and 52.6% had an associated hemorrhagic event. Thirty-six percent received covid19 vaccination with pfizer; 21.1% received astrazeneca. The minimum, median, and maximum times between vaccination and the occurrence of CVT were, in days: 22, 75, and 159, respectively. Two patients (10.5%) were RT-PCR positive for COVID19 on admission; 63.2% were IgA positive for COVID19 and 73.7% were IgG positive for COVID19. The minimum, median, and maximum number of platelets were: 158.000, 221.350 and 382.500/mm³. **Conclusion:** Our study highlights the clinical and laboratory characteristics of patients with CVT in pandemic COVID19. The main limitation is that this is a small case series from a single center.

Doença Cerebrovascular

TL 1106348

LESION MAPPING IN PATIENTS WITH STROKE SUBMITTED TO TRANSCRANIAL DIRECT CURRENT STIMULATION FOR UNILATERAL SPATIAL NEGLECT

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Background: Transcranial direct current stimulation (tDCS) may be useful in the management of patients with stroke and unilateral spatial neglect. In a previous investigation we described a better recovery of patients submitted to anodal tDCS. **Objective:** Use neuroimaging to access structural predictors of the outcomes those patients submitted to tDCS. **Methods:** Patients with stroke and unilateral spatial neglect were randomized in 3 groups (anodal tDCS [group 1] cathodal tDCS [group 2] and control [group 3]). High resolution computerized tomography scans in the sub-acute phase of the stroke were obtained. Lesions were semi-automatically segmented and normalized. Extension of the lesions based of regions of interest (ROIs) were extracted using the Atlas of Anatomical Labeling. Only gray matter of right hemisphere was evaluated, and the total area analyzed was 642.745mm³. Statistical analysis was conducted using chi-square test. **Results:** For group 1 (n=8, mean age 66 years), extension of damage obtained was 194.241mm³ (30% of all ROIs) centered in the amygdala; for group 2 (n=8, mean age 61), 79.100mm³ (12% of all ROIs) centered in the amygdala and for group 3 (n=7, mean age 68) 49.789mm³ (8% of all ROIs) centered in the insula. Group 1 was statistically different when compared to group 2 and 3 (p=0.001 and p<0.001). There was no difference between group 2 and 3 (p=0.9). **Conclusion:** A larger area of damage was observed in patients with stroke submitted to anodal tDCS. Despite the increased area of damage compared to the other groups these individuals presented a better outcome indicating that this approach may be useful in these patients.

Doença Cerebrovascular

TL 1106360

TELENEUROLOGY CARE IN ACUTE CEREBROVASCULAR DISEASE

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Background: Stroke treatment is extremely effective, but its benefit is time-dependent, making it challenging to achieve the goals established by the guidelines. The neurologist participation is essential for the best clinical decision and teleneurology has been used to increase the access of these cases to specialized management. **Objective:** Demonstrate the results of a private teleneurology network. **Methods:** We evaluated patients admitted to the database from January 2021 to April 2022 from a private network of Hospitals located in 2 Brazilian states (SP and RJ), totaling 15 Hospitals that use teleneurology. Several interventions were carried out to improve the indicators in previous years, such as: continuous training, monthly meetings, managed protocol. The KPI evaluated were: Median door-to-image time (mPI); median door-to-needle (mPA); venous thrombolysis rate (txTV); mortality rate (tx mort) and median length of stay (mTI). **Results:** A total of 2081 patients were admitted during the study period, with mPI=24min; mPA=68min; txTV=15%; txmort=7%; mTI=4 days. **Conclusion:** About 4000 patients are included in the database annually, and approximately 1800 patients with a stroke diagnosis confirmed. Telemedicine provides access to vascular neurology expertise for Hospitals without it, and its use has increased rapidly in recent years. Access to such technology demonstrates an improvement in the KPIs of the acute phase of stroke. The results founded are congruent with previous studies, showing that teleneurology in a private network is an important support and measures of continuous improvement of the process are fundamental.

Doença Cerebrovascular

TL 1106434

PROFILE ASSESSMENT OF MOYAMOYA SYNDROME IN PATIENTS FROM THE NORTHEAST: CASE SERIES

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Background: MoyaMoya syndrome is an uncommon cerebral vasculopathy worldwide and rare in South America, in which reduced blood flow in the internal carotid artery promotes compensatory angiogenesis. Known risk factors associated are systemic arterial hypertension, type 2 diabetes and Down Syndrome. **Objectives:** Describe clinical-epidemiologic profile of 7 patients with MoyaMoya Syndrome **Methods:** Report of seven cases of MoyaMoya Syndrome in a tertiary neurology department **Results:** In our series, all patients were female, in an age range of 18-49 years. All of them had sudden onset of focal neurological symptoms, which prompted the investigation. Three patients had ischemic stroke, with varied Bamford Classification of anterior circulation syndrome. Four patients had hemorrhagic events, one with association of Subarachnoid Acute Hemorrhage (SAH) and Subdural Hemorrhage, one with Tetraventricular Hemorrhage and two Frontal Lobar Intraparenchymal Hemorrhage. The angiographic study of the patients showed Suzuki-Takaku stages of 3 and 4. Two patients had neurosurgery indication, but one refused and another died in anesthesiological procedure. Two patients had brain death because of hemorrhage. The other patients remain in neurologic department clinic follow-up. **Conclusion:** Moya Moya Syndrome is rare in our population, with high potential to provoke cerebrovascular events, with disabilities and death. There is low evidence of pharmacologic treatment, being surgery the main strategy of approach in these cases. Therefore, we recommend furthering studies of pathogenesis, prognosis and treatment of this disease, to minimize complications.

Doença Cerebrovascular

TL 1106485

POST-STROKE DYSPHAGIA IN PATIENTS UNDERGOING THROMBOLYTIC TREATMENT: A RETROSPECTIVE COHORT

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Background: For the treatment of ischemic stroke in its acute phase, the use of intravenous thrombolysis with Alteplase (rt-PA) is currently recommended, as it is capable of reducing the functional deficits and the severity of the lesion. However, despite the advances in care and reduction of functional deficits in stroke patients, dysphagia is a common dysfunction. Nevertheless, there are few studies related to thrombolytic treatment and its effects on dysphagia, with divergent results regarding the benefits of this drug on the severity and incidence of dysphagia. **Objective:** To analyze dysphagia at Hospital discharge after stroke in patients undergoing and not undergoing thrombolytic treatment. **Method:** A retrospective cohort was carried out in a stroke referral Hospital in Salvador - BA, in which patients were divided into two groups: submitted and not submitted to thrombolytic treatment. This research was submitted to and approved by the Research Ethics Committee. The patient's socio-demographic and clinical characteristics were analyzed, and the incidence and severity of dysphagia between the groups were compared. The Dysphagia Outcome and Severity Scale (DOSS) was used to analyze dysphagia. Patients with some degree of dysphagia (DOSS \geq 5) or no dysphagia (DOSS $>$ 5) were considered. Data were analyzed using descriptive statistics and compared by parametric or nonparametric tests, when appropriate. Moreover, a univariate analysis was performed for the outcome dysphagia at Hospital discharge and the variables that were significant ($p < 0.05$), were taken to logistic regression to assess whether the thrombolytic is an independent protective factor. **Results:** A total of 120 thrombolized and 117 nonthrombolized patients were included in the final analysis. Dysphagia at Hospital discharge affected 23 (19.2%) thrombolized and 41 (35%) nonthrombolized patients ($p = 0.006$). There was no difference regarding the severity of dysphagia between the groups ($p = 0.158$). For multivariate analysis, thrombolysis was an independent protective factor for dysphagia at Hospital discharge [OR: 0.29 (CI: 0.08-0.95); $p = 0.042$]. **Conclusion:** The incidence of dysphagia at Hospital discharge was significantly higher in nonthrombolized patients, with no effect on dysphagia severity. Furthermore, thrombolysis was an independent protective factor for dysphagia at Hospital discharge.

Doença Cerebrovascular

TL 1106488

HAS THE IMPACT OF CEREBROVASCULAR DISEASES CHANGED DURING THE COVID-19 PANDEMIC?

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Background: Stroke, both ischemic and hemorrhagic, is an important cause of morbidity and mortality worldwide. Due to the SARS-COV-2 pandemic, patients were afraid to seek medical care for fear of contracting COVID while health professionals were, amid the search for beds to care for all patients with respiratory symptoms, alerts for the association of COVID with stroke, in addition to strict precautions so that patients do not contract COVID after Hospitalization. **Objective:** To describe the number of patients Hospitalized for ischemic stroke, hemorrhagic stroke and cerebral venous thrombosis (CVT) and their respective outcomes from March 2019 to February 2022. To describe the number of patients who were diagnosed with COVID-19 in the period of March from 2020 to February 2022. To assess whether there was a difference in the incidence of cerebrovascular diseases during the COVID-19 pandemic in a private Hospital in Fortaleza. **Methods:** The study design was cross-sectional, observational, descriptive, evaluating patients Hospitalized for ischemic stroke, hemorrhagic stroke and CVT from March 2019 to February 2022 who had a diagnosis of COVID after their Hospitalization. Patients who did not have a stroke confirmed by imaging or clinical findings were excluded from the study. Data were collected from the patients' medical records. The study is an arm of the approved research project at the Hospital's CEP. **Results:** From March 2019 to February 2020, the year in which the pandemic had not started, 85 patients were Hospitalized, mean age 75 years, 53% female, 74% with stroke, 26% stroke, 14% death. From March 2020 to February 2021, 95 patients were Hospitalized, mean age 72 years, 55% female, 77% stroke, 17% stroke, 6% CVT, 15.7% death, 3.1% COVID+. From March 2021 to February 2022, 90 patients were Hospitalized, mean age 72 years, 55% female, 79% stroke, 6% stroke, 5% CVT, 14.4% death, 10% COVID+. **Conclusion:** The number of Hospitalizations for stroke was not impacted in the 24 months of the pandemic. However, an increase in the incidence of CVT related to COVID+ patients was observed in most cases, which corroborates the association found in the literature. Regarding contamination by COVID after admission, there is no evidence that patients Hospitalized for stroke contracted it.

Doença Cerebrovascular

TL 1106492

RECOGNIZING AND REACTING TO A HYPOTHETICAL STROKE SCENARIO IN BRAZIL'S NORTHEAST

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Background: Recognizing the alarming signs of stroke is the first step to starting a proper treatment reaction chain. **Objectives:** We aimed to assess the frequency of awareness of the laypeople on recognition and reaction to stroke. **Methods:** Cross-sectional survey-based study, which data was collected in 2020, in twelve cities of Brazil's Northeast. The participants were exposed to a stroke case study and then responded to an open-ended semi-structured questionnaire to evaluate their stroke knowledge. **Results:** A total of 1,475 subjects were enrolled in this study (52.6% women, 36.2±15.3 years old). 1,220/1,475 (82.7%) recognized the situation as a stroke; 1,148/1,475 (77.8%) would react to it by taking the patient to the emergency room. Of those who recognized the stroke, 19.9% (243/1,220) did not take an appropriate reaction to it, meaning a recognition-reaction gap. **Conclusion:** The frequency of stroke recognition and reaction to it were acceptable, but could be improved. The recognition-reaction gap needs to be addressed by specific campaigns focusing on time-sensitivity aspects for successful stroke outcomes.

Doença Cerebrovascular

TL 1106496

THE COMPLICATIONS OF PATIENTS HOSPITALIZED IN STROKE UNIT CARE OF THE HOSPITAL IN PERNAMBUCO

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Background: Stroke is a condition characterized by reduced or absent blood flow to a certain area of the brain. According to its etiology, it can be classified as ischemic, when caused by the occlusion of an intracranial or extracranial artery. Stroke can also be classified as hemorrhagic, when caused by the rupture of intracranial vessels due to conditions such as arteriovenous malformations or aneurysms. Among the types of stroke, the one with the highest incidence is ischemic, which globally reached the mark of 7.2 million cases in 2019, which represented 62% of the total. In second place is intracranial hemorrhage responsible for 3.4 million cases of stroke or 28% of the total, followed by subarachnoid hemorrhage with 1.2 million cases in 2019 and approximately 10% of the total. Despite the severity of the disease, there have been many advances in the matter of treatment, especially for ischemic stroke, for which, in eligible patients, intravenous thrombolysis or mechanical thrombectomy can be performed. **Objective:** To describe the complications of patients from the stroke unit care of "Hospital da Restauração" in the months of April and May 2022, based on the service quality indicator questionnaires. **Methods:** An observational, retrospective and cross-sectional study was carried out on patients admitted to the stroke unit care of Hospital da Restauração in Pernambuco in the months of April and March 2022. **Results:** The study observed that of the 66 patients who were Hospitalized during the period, 4 had pressure ulcers with lesion stages ranging from 2 to 3. In addition, 15 patients used antibiotics to treat infection in the period, with most used antibiotics ceftriaxone, which was used in 6 cases and clindamycin, used in 2 cases. Finally, another complication identified was the use of nutritional devices (11 cases), mainly a nasogastric tube, which was used by 9 patients, and a urinary catheter (1 patient). **Conclusion:** The study showed that approximately ¼ of the patients had some complication post stroke. Some measures can be taken to avoid them, for example, the follow-up with speech therapists, aiming to reduce the use of nasogastric tube, use of protocols to avoid bronchoaspiration, reducing the use of antibiotics and change of decubitus to avoid pressure ulcers.

Doença Cerebrovascular

TL 1106520

THE STROKE AND ISCHEMIC HEART DISEASE DISCONNECTION: THE NUMBERS FROM CEARÁ, BRAZIL

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Background: Stroke and ischemic heart disease (IHD) are Brazil's leading causes of death. However, mortality rates have declined for both diseases since the 70s. This has been more pronounced in regions with higher socioeconomic status (mainly in the southeast and south), probably due to better control of risk factors, access to health care services, and treatment of hypertension and diabetes through national government programs. There are very few epidemiological studies on stroke and IHD mortality trends in regions with lower socioeconomic status, such as the Northeast. **Objective:** This study aimed to investigate the trend of mortality rates from stroke and IHD in Ceará, a state with low socioeconomic status in Northeast Brazil, from 2009 to 2019. **Methods:** The mortality data were obtained from the Brazilian Mortality Information System made available by the National Health Ministry. The mortality rate estimates, expressed as the number of deaths per 100 000 inhabitants, were calculated for each year—population estimates derived from the Brazilian Institute of Geography and Statistics (IBGE). The proportion of deaths caused by stroke and ischemic heart disease was also calculated, and statistical significance was tested. **Results:** A consistent decline in stroke mortality was observed from 2009 to 2019 (10.0%, 9.9%, 9.5%, 9.2%, 8.9%, 8.3%, 8.5%, 8.7%, 8.2%, 7.5% and 7.5%; $p < 0.001$) but the coronary heart disease mortality increased consistently over the years studied (8.0%, 7.7%, 8.1%, 8.5%, 8.0%, 8.1%, 8.4%, 8.8%, 8.5%, 9.6% and 9.5%; $p < 0.001$). **Conclusion:** Stroke is no longer the leading cause of death among non-communicable diseases in Ceará, overtaken by ischemic heart disease, which has shown a consistent increase in recent years. Population-based epidemiological studies are necessary to clarify why mortality rates exhibit such a different tendency in diseases that share most risk factors.

Doença Cerebrovascular

TL 1106523

DIFFERENT NAMES FOR STROKE IN BRAZIL'S NORTHEAST

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Background: There are different names for stroke in Brazil, but it is not clear if those names mean the same idea of disease. **Objectives:** To assess the different words for stroke in Brazil and both intended response to it and symptoms associated. **Methods:** Cross-sectional study enrolling healthy individuals from urban areas in twelve cities in all nine states of Northeast Brazil for an open-ended survey. We presented a typical clinical case of a stroke (an elderly who had sudden onset of hemiparesis and slurred speech) and asked “what is happening?”. **Results:** From 1,475 interviewed individuals, 1,220 (82.7%) recognized the scenario as a stroke, and they used 4 words to identify the situation as a stroke, which were “AVC”, “derrame”, “trombose” and “aneurisma”. There were significant differences among them concerning demographic, economic, educational and geographical aspects. The group who stated “AVC” was younger, had more school years and lived in capitals. “Derrame” was cited often in Bahia and in the countryside cities; this group was made mostly of Blacks and Mullatos. “Trombose” was mentioned only in 5 states (Paraíba, Ceará, Maranhão, Piauí and Rio Grande do Norte), other states did not cite this name at all. **Conclusion:** There are at least four words for stroke in Portuguese (“AVC”, “derrame”, “trombose” and “aneurisma”). Stroke campaigns should apply those alternative names to improve the comprehension of the general population.

Doença Cerebrovascular

TL 1106558

NURSING CONSULTATION TO STROKE PATIENTS USING WARFARIN IN A NEUROLOGY OUTPATIENT CLINIC

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The outpatient nursing consultation consists of a strategy of resolute and quality care, supported by law and exclusive to the nurse. In the care of patients, using warfarin is show to be a tool of great value, because they need close follow-up, given the specificities of the treatment, such as drug interaction, food and the need for correction in the dose of the anticoagulant. This study aimed to report the experience of the nursing consultation to stroke patients using warfarin. This was a descriptive study, the type of experience report. The scenario was the outpatient clinic of a tertiary public Hospital, located in the city of Fortaleza, Ceará. The nursing consultation aimed at this target audience began in 2020 and took place on Tuesdays, in the afternoon. The service was performed by a nurse, in two moments. Initially, a face-to-face care occurred and then telemedicine care intended for patients with difficulty in physical mobility or residents in other cities. The main activities developed by the nurse were: patient/family welcoming; health status assessment/complaints/complications; monitoring of prothrombin time and registration in the follow-up form; adjustment of the dose of warfarin, according to the outpatient anticoagulation protocol adopted in the neurology outpatient clinic and return consultation schedule; educational guidance on care related to the use of warfarin, by providing an information leaflet; referral to the neurologist in case of clinical complications and registration in the patient's medical record. The nursing consultation proved to be beneficial to patients using warfarin, as it allows systematized and individualized actions, which contributes to minimize health problems resulting from the treatment.

Doença Cerebrovascular

TL 1106573

EPIDEMIOLOGICAL AND CLINICAL INDICATORS OF A TYPE III EMERGENCY CARE CENTER FOR STROKE IN JUIZ DE FORA (MINAS GERAIS)

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Background: Stroke is one of the leading causes of morbidity and mortality in Brazil and in the world. Thrombolytic therapy and stroke units were important advances to patients suffering from stroke. The Stroke Unit of the reference stroke center in Juiz de Fora has a multidisciplinary team to assist stroke patients 24 hours a day, and it serves the entire macro-region of Juiz de Fora, in Minas Gerais state, exclusively within the scope of “Sistema Único de Saúde” (SUS). **Objective:** To study epidemiological and clinical indicators of a stroke care center in Juiz de Fora (Minas Gerais), Brazil. **Method:** Medical records of patients admitted to the emergency department and referred to the Hospital from September 2020 to March 2022 were retrospectively reviewed. **Results:** A total of 732 individuals (371 [50.8%] male, mean age 67 years) with a predictive diagnosis of stroke were referred to the Hospital from September 2020 to March 2022 and their medical charts were reviewed. With regards to the comorbidities, 560 (76.6%) had arterial hypertension and 212 (29.0%) were diabetics. Regarding lifestyle habits, 385 (52.7%) patients were smokers and 428 (58.5%) were alcohol consumers. Mean symptom onset occurred 691 minutes (11.5 hours) before admission. Mean NIHSS score value was 8.0 points. Thrombolysis was performed in only 57 (7.9%) of patients. Cranial CT scans were normal in 264 (36.2%) of patients. 423 (57.8%) scans showed ischemic stroke while 45 (6%) showed hemorrhagic stroke. The average time the patients stayed in the Hospital was 11 days. At discharge, the average NIHSS was 4.0 points and the mean RANKIN scale score was 3.0 points. 543 (74.2%) were discharged with platelet antiaggregation, 633 (86.5%) with statin prescription. Among Hospitalized patients, 610 (83.4%), received deep vein thrombosis prophylaxis in the first 48hrs. At Hospital discharge, 579 (79.1%) patients received prescription of a prophylactic and rehabilitation therapy plan. The total mortality rate in 10 months of the study period was 71 (9.7%) and the mean number of deaths was 7.1 deaths, ranging from 4 (0.55%) deaths in September 2021 to 10 (1.36%) deaths in July 2021. **Conclusion:** Thrombolysis eligibility rate was rather similar to other stroke reference centers in Brazil. We also observed a long time between the onset of symptoms and arrival at the service, which was the main barrier to performing thrombolysis, due to loss of the therapeutic window.

Doença Cerebrovascular

TL 1106592

PANORAMA OF HOSPITALIZATIONS AND DEATHS ATTRIBUTABLE TO STROKE IN BRAZIL FROM 2016 TO 2022

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Background: Cerebrovascular diseases are the second cause of death in the world, and stroke a leading cause of long-term disability. In the United States (US) in 3% of males and 2% it led to disability, and caused 6.6 million deaths in 2019. Early treatment is critical, but there are several factors that lead to disparities, while in the US whites have higher proportional rates of access to treatments, worldwide the differences are of overall access to quality health care. **Methods:** An ecological and descriptive study of the Hospitalizations due to Stroke in Brazil, from January 2016 to April 2022, for which data collection was obtained via the Health Information (TAB- NET) and the SUS Hospital Information System (SIH/SUS). **Results:** During the past 7 years in Brazil there were 127.956 stroke admissions. The region with the highest incidence was the Southeast, with a total of 48.861 cases (38,18%), followed by the Southern region with 35.453 cases. The region with the lower incidence was the Central-West, with only 7.119 cases (5,56%). There was a predominance of males, representing 66.612 cases (52,05%). The age group with most cases was from 70 to 79 years, with 33.272 cases (26%). There were a total of 10.020 deaths due to stroke, with the highest incidence in patients with 80 years or more (4.023 cases, 40,14%). From 2016 to 2021, there was a decrease of 10,97% in the number of Hospitalizations, with a fluctuation of incidence. While 2019 was the year with the highest incidence of Hospitalizations, the year with most deaths due to stroke was 2016, with 1.755 cases, followed by a decrease up until 2020, with the lowest cases 1.469, and a new increase in 2021 with 1.555 cases. When it came to racial differences, patients with the highest Hospitalization rates for Stroke were whites, with 38.270 cases (41,9%), followed by browns, blacks, yellows, with the lowest incidence among indigenous people, and deaths followed the proportions of Hospitalizations, with the highest among whites 4.057 and lowest among indigenous people, 6. **Conclusion:** Hence, white men from 70-79 were the most affected, having the highest amount of Hospitalizations and deaths, and the region with most Hospitalizations was Southeast. It is crucial to focus on primary prevention, especially in primary care, along with educational programs towards the population, so that they may seek medical attention sooner, while also making policies and approaches to improve access to medical care.

Doença Cerebrovascular

TL 1106619

IMPACT OF IN-HOSPITAL ACUTE KIDNEY INJURY ON DEATH OR FUNCTIONAL DEPENDENCY AFTER STROKE

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Background: Renal dysfunction and acute kidney injury (AKI) have been identified as a risk factor to worse outcomes in the general population of intensive care units. According to previous studies, AKI has been a frequent complication after an acute cerebrovascular event, with an overall prevalence around 11.6%. **Objective:** Our aim is to evaluate the impact of AKI during Hospitalization on stroke patients. **Methods:** Prospective cohort of stroke patients with symptom onset within 72 hours before research admission. AKI was diagnosed by the occurrence of an increase in the serum creatinine level by approximately 0.4 mg/dl in less than 72 hours during Hospitalization for ischemic or hemorrhagic stroke. Our main outcome was functional dependence or death, evaluated by modified Rankin Scale (mRS), at 90 days post-ictus. **Results:** Two hundred and fifty one patients were enrolled. AKI was diagnosed in 78 (37.1) patients. The frequency of death or functional dependence (mRS > 2) was lower among those who were not diagnosed with acute kidney injury [U = 4.523; 38 (48.7) vs 40 (51.3) cases, p=0.033]. However, multivariate analysis failed to show the occurrence of AKI as independent predictor of mRS > 2 in 90 days (odds ratio, 1.19 [IC 95%, 1.13-1.25]), which was observed to variables as age, sex, National Institutes of Health Stroke Scale on admission, type of stroke, pre-ictus mRS score and infection occurrence. **Conclusion:** Our study suggests that, although in-Hospital acute kidney injury may be associated with worse outcomes in stroke patients, we were not able to describe it as an independent predictor. That conclusion indicates there may be unknown variables, which our data was not able to detect, that can influence this association.

Doença Cerebrovascular

TL 1105287

AMYOTROPHIC LATERAL SCLEROSIS ATTENDED IN A REFERENCE CENTER IN THE CITY OF RIO DE JANEIRO

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Background: Amyotrophic lateral sclerosis (ALS) is, to date, one of the incurable progressive neurological diseases with the worst clinical prognosis. The Neuromuscular Diseases Service of the Pedro Ernesto University Hospital (HUPE/UERJ) is a reference center that receives patients referred from the primary and secondary care levels of the SUS, which provides the meeting of rarer and more complex neurological cases, such as ALS. **Objective:** To carry out a retrospective epidemiological study on confirmed cases of ALS treated at the Neuromuscular Diseases Outpatient Clinic at HUPE/ UERJ between 2004 and 2022 to identify the locoregional profile of the patients. **Methods:** Review of care records of patients with suspected or confirmed cases of ALS and treated at the Neuromuscular Diseases Outpatient Clinic of HUPE/ UERJ between 12/17/2004 (first record found) and 05/31/2022, identifying and organizing information related to gender, age, etiology and clinical phenotype of ALS cataloged cases. **Results:** Ninety-five suspected cases of ALS were identified in the 17 years and 5-month interval considered, of which, after reviewing the consultations, 16 cases were discarded, making a total of 79 ALS records. In this series, 45 were men (57%), and 34 were women (43%). The vast majority of cases were idiopathic (76 cases, 96% of the total), while only 3 cases showed genetic/family inheritance (4%). The most affected age group was 50-69 years-old (46 cases, 58%), with a mean age of 55 years-old, and the most common phenotype was classic ALS (65 cases, 82%), although were found 4 bulbar ALS, 4 cases of progressive muscular atrophy, 2 cases of primary lateral sclerosis, 2 cases of monomelic atrophy, 1 flail-arm syndrome and 1 Mills variant. **Conclusion:** The data collected on a locoregional scale, despite the small number of patients considered (because it is a rare disease), are in line with the epidemiological characteristics of ALS already demonstrated in other studies of national and international scope, showing a predominance of men, sporadic cases as opposed to hereditary cases, and average age at onset of the typical condition from the sixth decade of life.

Doenças do Neurônio Motor - Esclerose Lateral Amiotrófica

TL 1105456

MOTOR CORTEX HYPOINTENSITY IN SUSCETIBILITY WEIGHTED IMAGING: A RADIOLOGIC BIOMARKER OF AMYOTROPHIC LATERAL SCLEROSIS?

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Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease of the central nervous system characterized by progressive loss of upper (UMN) and lower motor neurons that results in loss of voluntary movement control. The diagnosis of definite ALS is based on clinical or electrophysiological evidence of lesions in the two aforementioned motor segments. The presence of hypointensity in the motor cortex in the magnetic susceptibility sequence (SWI) may indicate injury to the UMN. The objective was to verify the presence of hypointensity in the motor cortex in SWI of brain magnetic resonance imaging (MRI) in patients diagnosed with ALS, its correlation with clinical characteristics and also the association with corticospinal tract (CST) radiologic findings in the magnetization sequence transfer (MT) of the same patients. This is a descriptive study that evaluated data from 20 patients with ALS and 20 control subjects. The exams were performed at the same imaging center and analyzed by the same radiologist with experience in neuroimaging. Clinical data were collected from medical records. Statistical analysis was performed using Bioestat 5.0 software. The statistical significance of the variables was verified using the T test and the linear regression test, with alpha levels of 0.01 and 0.05, respectively. Clinical variables did not statistically influence the presence of hypointensity in the motor cortex of ALS patients, however in control group there was a statistical significance of older age in the presence of hypointensity. When comparing the presence of hypointensity in ALS patients and healthy individuals, this finding occurred especially in ALS subjects. There was also a statistical influence of the CST hyperintensity of the MT on the presence of hypointensity in the motor cortex in SWI. It was concluded that the presence of hypointensity in the motor cortex of ALS patients is not influenced by clinical variables despite being more frequent in ALS patients than in healthy individuals. Age can influence the presence of hypointensity in healthy individuals, but in the ALS group there was no such relationship, which may represent neurodegeneration caused by the disease itself. Some patients with hypointensity at SWI in the cortex also presented hyperintensity in the CST at MT, finding that suggest a possible to use the SWI as another tool for detecting lesions in the UMN and thus help in ALS diagnosis.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1105601

ANALYSIS OF TELEMONITORING AND EARLY INDICATION OF HOME NONINVASIVE MECHANICAL VENTILATION IN PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS FOLLOWED UP IN A TERTIARY CARE HOSPITAL

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Background: Amyotrophic lateral sclerosis (ALS) is a rare neurodegenerative disease that invariably results in significant ventilatory impairment. In this sense, respiratory function monitoring is essential, including the early indication of noninvasive ventilation (NIV), although there is still no consensus on the ideal moment of its indication or how to promote satisfactory adherence and adaptation during its use. **Objective:** To describe the effects of NIV telemonitoring in patients with ALS followed up in a tertiary Hospital in Brazil. **Methods:** NIV was indicated for ALS patients with forced vital capacity (FVC) lower than 70% (late indication) or with FVC higher than 70% (early indication). The following variables were observed during 9 months of telemonitoring: hours of use, leak, apnea hypopnea index (AHI), respiratory rate, minute volume, and trigger. **Results:** 22 patients with a definitive diagnosis of ALS were included, with a predominance of males (68%; 15/22) with a mean age of 60.8 (± 9.25) years. 40.9% (9/22) underwent endoscopic gastrostomy. 22.2% (5/22) had FVC over 70. 22.2% (5/22) had a fall of more than 10% between sitting and lying position, none had satO₂ less than 93.8%. The ALS Functional Rating Scale (ALS-FRS-S) had a mean of 22.38 at inclusion and 24.5 at month 9. In the first month of telemonitoring, we observed: 3.4 (± 2.0) hours of daily compliance, AHI of 6.7 (± 2.2) events/hour, leak of 3.15 (± 14.4), RR of 25.5 (± 5.7). 56.25% had poor adherence, defined as usage time of less than 4 hours. Comparing the first and ninth month of telemonitoring, we observed a reduction in the following parameters: AHI by 11%, leakage by 74%, RR by 21%, MV by 25% and firing by 38%. In this period, there was an increase of 117% in the time of daily use. **Conclusion:** The increasing use of new technologies has made it possible to offer better quality in the performance of health care activities. In this sense, telemonitoring is a tool capable of providing information about diagnosis, prescription and treatment that contributes to an increasingly earlier indication for NIV. In addition, it makes it possible to improve care, allowing the identification of problems and complications not observed in periodic evaluations in determining appropriate solutions and interventions.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1106062

A GEOGRAPHICAL STUDY ON AMYOTROPHIC LATERAL SCLEROSIS IN RIO GRANDE DO NORTE, BRAZIL, FROM 2005 TO 2018

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Background: Amyotrophic lateral sclerosis (ALS) is a rare and fatal neurodegenerative disorder with variable incidence and prevalence worldwide. However, clinical-epidemiological studies on ALS are scarce in Brazil. **Objective:** This study investigated whether ALS incidence had uniform spatial distribution in a population-based cluster analysis in Rio Grande do Norte state (Brazil), from 2005 to 2018. **Methods:** New cases of ALS were identified in a database of the ALS multidisciplinary care centre of the Onofre Lopes University Hospital in Natal (Rio Grande do Norte, Brazil). Approaches were based on incidence (empirical Bayes estimator and Moran's I analysis) and cluster analyses (Moran scatter plot and spatial correlogram). **Results:** A total of 177 patients (59% males) participated in the study; mean age of ALS onset was 57 years. Mean annual incidence of ALS was 0.3769 per 100,000 inhabitants (95% confidence interval of 0.0889), higher in males than in females (0.4516 per 100,000 vs. 0.3044 per 100,000). According to spatial statistics, patients were homogeneously distributed throughout the studied area. **Conclusion:** A low estimate was observed compared with other populations. Results did not indicate areas of increased risk or significant spatial geographic dependence, suggesting a random ALS incidence in Rio Grande do Norte.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1106132

AMYOTROPHIC LATERAL SCLEROSIS TYPE 8 HAS LESS COGNITIVE DEFICITS AND DISTINCT BEHAVIOURAL PROFILE THAN THE SPORADIC FORM: A COMPARATIVE STUDY

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Objective: To compare the cognitive performance and the behavioural profile of patients with sporadic Amyotrophic Lateral Sclerosis (sALS) and patients with ALS type 8 (ALS8), a familial form of ALS caused by mutations on VAPB gene. **Methods:** We included 29 symptomatic ALS8 patients (17 men; median age = 49 years-old), 20 sALS (12 men; median age = 55 years-old) and 30 healthy controls (16 men; median age = 50 years-old), matched for sex, age, and educational level. Participants underwent a neuropsychological assessment focused on executive functions, visual memory, and facial emotion recognition. Behavioural and psychiatric symptoms were investigated with the Hospital Anxiety and Depression Scale and the Cambridge Behavioural Inventory. **Results:** Clinical groups (sALS and ALS8) had lower global cognitive efficiency than controls, and also impaired performance in cognitive flexibility and inhibitory control. Verbal fluency deficits were not found in ALS8 patients, but they seem to have impairments in working memory subcomponents. On the psychiatric and behavioural aspects, apathy was shown to be frequent in both clinical groups. ALS8 patients presented more eating changes and more stereotypical behaviours. **Conclusion:** Overall, sALS and ALS8 had similar deficits in most cognitive domains, but ALS8 patients exhibit milder degree of executive dysfunction than sALS, besides distinctive pattern of behavioural changes. ALS8 has a specific cognitive-behavioural profile, which should be considered in the clinical care of patients.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1106185

UNVEILING CONFLICTING OPINIONS ABOUT AUTONOMY, END-OF-LIFE DECISIONS AND MEDICAL MANAGEMENT OF AMYOTROPHIC LATERAL SCLEROSIS (ALS): RESULTS FROM A SURVEY IN 3 CITIES FROM THE STATE OF CEARÁ

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Background: ALS is an irreversible neurodegenerative disorder. End-of-life decisions vary according to cultural perspectives about the limits of medical treatment. In Brazil, assisted suicide and euthanasia are illegal and the boundaries of patient's autonomy for decisions are not precise. **Objective:** To evaluate the opinions of the educated general population about different medical decisions related to management of ALS patients. **Method:** Presentational written opinion survey conducted with unidentified volunteers (minimum education: complete High school) after signing informed consent with demographic data collection, pre-test teaching about the subject and 4 case simulations. **Results:** 510 volunteers, age 28.9±12.6 years (range 18-89), 50.7% female, 18.5% married. 45.4% were for-, 35.7% against euthanasia. 71.8% were for-, 11.5% against orthoethanasia. 15.2% were for-, 63.8% against dys-ethanasia. In situation 1 (ALS patient requested the medical team not to undergo gastrostomy that would increase life expectancy from 3 to 36 months): 17.1% considered this suicide, 64.1% orthoethanasia, 7.1% euthanasia done by family and 2.6% murder by either physician/family. If the ALS patient was restricted to a wheelchair (situation 2), only 5.9% considered refusal suicide but 5.5 and 5.7 considered murder by family or physician, 5.9% medical malpractice. However, if the same patient suffered from severe depression (under treatment) and required gastrostomy and tracheostomy but declined (situation 3): 47.2% considered this suicide, 53.8% orthoethanasia, 18.4% euthanasia, 9.1 and 5.8% murder by family or physician, 20.7% failure to provide adequate medical assistance, 12.9% medical malpractice. In situation 4, a 40-year-old patient who progressed to "vegetative state" after cardiac arrest had previously informed that she would refuse CPR/aggressive treatments if severely disabled. Following her wishes, her family refused to allow hemodialysis. The patient died. 6.9% considered this suicide, 74.3% considered orthoethanasia, 19.3% euthanasia, 14.1% considered failure to provide adequate medical assistance. **Conclusion:** In this educated general population from 3 cities of Ceará (Fortaleza, Cratubar, Sobral), after prior teaching, there is great confusion about physician's role, patient's autonomy/rights, euthanasia and end-of-life decisions. A large % of respondents considered accepting patient's autonomy the same as allowance of suicide, euthanasia, murder or malpractice.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1106204

IS COGNITIVE RESERVE A PREDICTOR OF THE CLINICAL OUTCOME OF AMYOTROPHIC LATERAL SCLEROSIS? A LONGITUDINAL STUDY

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Background: Amyotrophic Lateral Sclerosis (ALS) is a neurodegenerative disease with motor neuron involvement and non-motor symptoms, such as cognitive and behavioral deficits. It is unclear whether cognitive reserve has a protective factor in the rate of functional decline in patients with sporadic ALS (sALS). **Objective:** To investigate the effect of cognitive reserve on the clinical outcome of sALS patients. **Methods:** This longitudinal study included patients with the diagnosis of probable or definite ALS according to Awaji criteria, between March/2018 and March/2020. Patients underwent regular clinical assessments, including the ALS Functional Rating Scale-Revised (ALSFRRS-R). Patients were evaluated at the baseline, 6 months, 12 months and 18 months after inclusion. We considered years of education (schooling) as a proxy of cognitive reserve. Correlation and linear regression analysis were performed to test the association between schooling and ALSFRS-R decline. **Results:** We included 55 individuals (31 male) with sALS (mean age=55.3 years, mean schooling=7.16 years and median disease duration = 2.1 years). Eighteen patients died during the follow-up. The mean (±SD, number of patients) for ALSFRS-R scores were, respectively: 31.22 (±10.7, 55), 28.2 (±11, 48), 24.8 (±10.4, 41) and 22.2 (±10.1, 37), at baseline, 6, 12 and 18 months. Multivariate analyses including ALSFRS-R variation, schooling, disease duration, age and spinal/bulbar onset, found that disease duration was positively correlated with ALSFRS-R decline (B=0.73, p<0.05). The other variables did not show association with disease progression. **Conclusion:** In this series, schooling was not a predictor of clinical decline in sALS. More studies, with larger samples, are warranted to investigate the influence of cognitive reserve in the clinical outcome of ALS patients.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1106263

CLINICAL DECISION SUPPORT SYSTEM APPLIED TO AMYOTROPHIC LATERAL SCLEROSIS PROGNOSIS

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Background: Amyotrophic Lateral Sclerosis (ALS) is a rare and incurable disease that affects the neurons of the human motor system. The communication between the brain and muscles is gradually interrupted, leading patients to paralysis and death. The average life expectancy is 3-5 years after symptoms onset, and the worldwide incidence is about 1.9 cases per 100,000 individuals per year. ALS is clinically heterogeneous, with different symptoms and disease progression among its patients, making it challenging to perform prognoses (e.g., survival time and disease progression). Research using Machine Learning (ML) algorithms has been applied to improve the prognosis of diseases. ML could extract information from the training data, transform it into knowledge, and use it to solve different categories of problems. Thus, it is crucial to collect patient data to perform relevant studies and create ML solutions to help physicians in their daily work, e.g., developing a Clinical Decision Support System (CDSS). **Objectives:** The primary objective is to develop a CDSS to assist Brazilian health workers in their tasks related to ALS prognosis. The proposed CDSS should provide helpful information about prognosis prediction, including the survival time, the disease stage, and the moment when respiratory or nutritional support will be needed. Consequently, we intend to create a Brazilian ALS Prognosis Database to support future research in this field. **Methods:** This study is clinical, prospective, observational, and longitudinal. Data will be collected from patients followed by Brazilian Research Centers, which include demographics, clinical, laboratory, functional, and respiratory biomarkers. The data collected will be processed by ML algorithms to learn and create models to provide the target predictions defined. **Results:** A CDSS will be developed to allow the health workers to register information and obtain the prognosis predictions about their patients. The system will comprise the Electronic Medical Record and the Prognosis Dashboard modules. **Conclusion:** This research aims to develop a CDSS to help physicians in their ALS clinical practice. This system will allow health workers to register and monitor their ALS patients. We expect that the proposed CDSS can provide valuable information to the health workers about prognosis predictions and that it could represent a valuable tool for knowledge dissemination among all interested health workers and researchers in this area.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1106384

PROPOSAL OF AN INTEROPERABLE ELECTRONIC HEALTH RECORD FOR THE MULTIDISCIPLINARY FOLLOW-UP OF PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS

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Amyotrophic Lateral Sclerosis (ALS) is characterized as a rare neurodegenerative disease with an unknown cure. Due to its irreversible progression and complexity, it is essential that patients diagnosed with ALS are followed up by a multidisciplinary team. By involving different professionals in the health care of a patient with ALS, it is essential to consider the use of a platform capable of collecting and sharing information by each specialty, building a concise and cohesive clinical history to assist the multidisciplinary team in decision-making. It is in this perspective that electronic health records arise. This tool stores processable information about patients' health, allowing the sharing of this information to the multidisciplinary team safely and efficiently. Due to the evolution of technological resources, these tools have more functionalities and, consequently, more significant importance in the health area. In addition to containing information that allows communication between the entire medical team, the electronic health record can provide continuity of clinical care, gathering important information that, when analyzed, can contribute to the diagnosis and control of the evolution of diseases and adequate treatment. Given this context, the present work proposes developing a platform for collecting and storing structured data related to patients with ALS. The main objective is to present a blockchain-based architecture for developing a specific electronic health record for ALS patients. Given the need for information exchange, this architecture will be interoperable, following the Health Level 7 (HL7) standards with the Fast Healthcare Interoperability Resources (FHIR) specification, in accordance with the standards of the National Health Data Network (RNDs, its Portuguese acronym) and adherent to the General Data Protection Law. It is expected that this architecture will provide greater integration between the professionals who make up the multidisciplinary team, guaranteeing security, privacy, and confidentiality in managing information in the electronic health record for ALS. Once developed, this electronic health record will store essential data that can be used to plan health interventions, support decision-making processes, and follow up in more detail on the evolution of ALS in the patients.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1106426

THE PROTOCOL AND THE IMPLEMENTATION STEPS OF THE BRAZILIAN AMYOTROPHIC LATERAL SCLEROSIS REGISTRY

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Background: ALS is a rare disease. Recent studies have reported the incidence of ALS between 0.6 and 3.8 per 100,000 person-years, with possible geographic variation. In Brazil, its incidence and prevalence and the distribution of cases in the national territory are unknown. In recent years, the public health importance of rare diseases has been recognized. The use of a population-based registry, allowing the collection of information on all individuals diagnosed with ALS, allows a reliable estimate of the incidence and prevalence of the disease, in addition to other data that can be analyzed. **Objectives:** Our mission is to describe the study protocol and the steps to implement the ALS registry in Brazil. **Methods:** The Brazilian ALS registry began in 2022, with a retrospective and prospective collection of cases, as a multicenter observational study of a population-based cohort. A web platform was developed by the Laboratory of Technological Innovation in Health (LAIS) to collect data on new and monitored cases of ALS in different regions of Brazil. Demographic variables such as age at onset, ethnicity, sex, and family history of ALS and FTD will be collected. This data can be added by the physician (neurologist or neurophysiologist) or by the patient (self-registry). Six months later, the physician will be invited to update the patients' clinical situation. All participating centers have authorization from the corresponding research ethics committee. **Results:** The ALS registry was successfully implemented. Different centers throughout the Brazilian territory have already contributed to the registration platform and data from ALS patients have already been collected. Os dados serão analisados no final de 2022. **Conclusion:** We reached consensus on a central dataset and data sharing processes established with various partners to address the need for high quality information on the epidemiology of ALS in Brazil. In this way, we can plan health planning interventions, support decision-making processes regarding the diagnosis and evolution of the disease.

Doenças do Neurônio Motor – Esclerose Lateral Amiotrófica

TL 1105009

THE COMMUNICATION OF THE DIAGNOSIS OF SPINAL MUSCULAR ATROPHY IN ENDOGAMOUS X NON-ENDOGAMOUS REGIONS

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Background: The communication of the diagnosis of a rare, genetic and neurodegenerative disease may leave sequelae in the lives of those involved. **Objective:** To compare the communication of the diagnosis of spinal muscular atrophy (SMA) in endogamous x non-endogamous regions. **Methods:** A cross-sectional study obtained by means of a questionnaire containing the Impact of Event Scale-Revised, sent to volunteers with SMA or their families. **Results:** The sample consisted of 100 volunteers, 83% female, and aged 36.65 ($\pm 10,19$) years old and homogeneous for the variables of sex, age, color, education, religion and SMA subtype (1,2,3 and 4) between groups. The endogamous regions, despite being economically less favored, showed more satisfaction and inclusion in medical care and health services, with less trauma and signs of post-traumatic stress disorder related to the time of SMA diagnosis. **Conclusion:** the positive impact of AME communication in endogamous regions resulted in less emotional damage, favoring free and informed decision-making.

Doenças Neuromusculares

TL 1105031

MYOTONIC DYSTROPHY TYPE 1: A DISEASE WITH MULTIPLES OPHTHALMOLOGIC FINDINGS

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Background: Several ocular abnormalities have been documented to occur in patients with myotonic dystrophy type 1. **Objective:** To evaluate the frequency of these abnormalities and to alert the importance of multidisciplinary for better patient management. **Methods:** All participants underwent a thorough ophthalmologic examination, including visual acuity assessment, slit-lamp biomicroscopy, ocular motility, dynamic refraction, and fundus examination in addition to optical coherence tomography, a complementary exam that is fundamental for retinal evaluation, since many of these patients may have low visual acuity unrelated only to the presence of cataracts. **Results:** We evaluated 68 eyes of 34 patients aged 3 to 71 years (mean 42.7 ± 16.9), 17 of which were men. IOP was reduced in 20 eyes (29,4%). We found cataract or positivity for surgery in 31 eyes (45,6%) and ptosis in 40 (58,8%). Myopia in 36 eyes (52,9 %), hyperopia in 16 eyes (23,5%), ocular motility limitations in 14 eyes (20,6 %), blepharitis in 12 eyes (17,6 %), epiretinal membrane in 24 eyes (35,3 %) and pigmentary retinopathy in 13 eyes (19,1 %). **Conclusion:** Cataract and ptosis are frequent in DM1 and associated to motor dysfunction. An expressive reduction of intraocular pressure was observed in our patients as well. A finding described in the literature that was not found in our patients was lamellar macular holes and foveal schisis. However, we observed an important and relatively common alteration that was an irregularity and pigmentary changes in the choriocapillary/RPE complex which could only be analyzed and visualized with the advent of the spectral domain optical coherence tomography technology. We report an increased prevalence of epiretinal membrane in the myotonic dystrophy type 1 group. Therefore, it is important to emphasize the importance of the inclusion of different specialties in multisystem diseases like DM1, once that brings benefits to both patients, in their diagnosis and follow-up, as about the quality of life and prognosis, once again ophthalmological changes above found have treatment.

Doenças Neuromusculares

TL 1105048

NEUROIMAGING, GENETIC AND CLINICAL CORRELATIONS IN LAMA2- CONGENITAL MUSCULAR DYSTROPHY

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Background: LAMA2-Congenital muscular dystrophy (CMD) is the most common cause of CMD. LAMA2 gene codifies a protein named laminin alpha-2 (merosin), which binds to alpha-dystroglycan and is expressed in skeletal muscle fibers, and also in the brain. **Objective:** Characterize motor, genetic and neuroimaging changes, and in a large cohort of CMD-LAMA2. **Methods:** Patients were divided according to motor severity, and, for each group, comparisons were made among motor, genetic and central nervous system findings. **Results:** The study cohort consisted of 56 patients. (1) Three patients (5,3%) had the severe congenital form and were not able to sit without support. (2) Forty-three (76,8%) patients had the classic congenital form and were able to sit unassisted. (3) Five patients (8,9%) had the mild congenital form and were able to walk; among them, two patients who had acquired independent gait at four and five years of age became wheelchair dependent at age of eight years. (4) Five patients (8,9%) had the limb girdle muscular dystrophy form and acquired gait within the expected age. 40 different mutations were found, 26 variants were novel. All three patients with the severe congenital form had null variants in both alleles. 36 (83,7 %) patients with the classic congenital form presented null variants in both alleles and 7 (16,3 %) had one null variant with one missense or inframe variant. Among 5 patients with mild congenital form, 3 had one missense variant plus one null or CNV variant, and two had two null variants. The patients with 2 null variants were those who lost ambulation. Among patients able to walk, there was a higher frequency of missense variants than null variants ($p < 0.05$). Among the three patients with severe congenital form, one had associated cortical malformation. Cortical malformations were also found in 8 (18,6%) patients with the classic congenital form. Among patients who were able to acquire gait none had cortical malformation. All patients who presented cortical malformations had null variants in both alleles. Cortical malformations were: occipital polymicrogyria, temporal polymicrogyria, occipital lissencephaly-pachygyria and white matter changes. They were more frequently seen in patients with mutations located in the LG domains, region that binds Laminin-211 to alpha dystroglycan ($p < 0.05$). **Conclusion:** The most severe motor presentations are related to cortical malformations and null variants, and to variants present in the LG domain of LAMA2 gene.

Doenças Neuromusculares

TL 1105049

PSEUDOMETABOLIC SYMPTOMS IN CONGENITAL AND DYSTROPHIC MYOPATHIES

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Background: Metabolic myopathies comprise a diverse group of muscle disorders that result in impaired energy production due to defects in glycogen, lipid, or mitochondrial metabolism. Most of the patients have exercise intolerance, pain, muscle cramp, myoglobinuria, variation in CK levels, with or without progressive muscle weakness. Structural myopathies can also present with symptoms triggered by exercise, and thus mimic the metabolic myopathies, being referred to as the pseudo-metabolic myopathies. To date, there is only few articles about pseudometabolic symptoms in structural myopathies, and the actual frequency and genetic correlations is not yet well established in large cohorts. **Objective:** identify and genetically characterize patients with pseudometabolic symptoms and to identify the frequency of this symptoms amongst patients with RYR1-myopathies **Methods:** Observational study conducted from 2020 to April 2022 at Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo. São Paulo SP, Brazil. Among 1310 patients evaluated, 30 (2,3%) had symptoms resembling metabolic myopathies. Inclusion criteria was the presence of at least one of the following four typical metabolic symptoms, being the most important complaint: (1) exercise intolerance, (2) muscle pain, (3) muscle cramps, (4) myoglobinuria, with or without muscle weakness. Exclusion criteria: patients whose most important complaint/symptom was weakness, patients with inconclusive genetic test or genetic test confirming a metabolic myopathy. **Results:** From 30 selected patients, twelve (40%) had genetic confirmation of structural myopathy. Four patients had RYR1-myopathy (33,3%), three patients had Dystrophinopathy (25%), two siblings had Gamma-sarcoglycanopathy (16,6%), and the other three patients had FKRP, ANO5 and COL6-related myopathy, respectively. All patients had exercise intolerance and muscle pain, five (41,7%) patients had muscle cramps and four (33,3%) had myoglobinuria **Conclusion:** Our cohort highlight the need to consider structural myopathies in the differential diagnosis of patients who present with complaints suggestive of a metabolic myopathy, particularly when metabolic studies have been unrevealing. It also highlight the need to consider not only genes of muscular dystrophy, but genes of structural myopathy, specially RYR1-related myopathy.

Doenças Neuromusculares

TL 1105176

PHENOTYPICAL PATTERNS OF CONGENITAL MYASTHENIC SYNDROMES

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Congenital myasthenic syndromes (CMS) comprise a rare group of genetic diseases with phenotype that can be difficult to be distinguished between their selves. In previous study we found that a pure ocular syndrome is not a usual phenotype of CMS, and that usually the phenotype is predominantly limb weakness or predominantly axial weakness (impairment of bulbar, neck and/or respiratory muscles). This study verifies which genes are related to each pattern of CMS phenotype in a large CMS cohort with genetically confirmed cases. The clinical and molecular characteristics of a large cohort of 92 patients with suspected CMS were analyzed. Data from clinical evaluations and ancillary exams were collected and blood samples were submitted to new generation sequencing. After molecular results, 66 patients were considered confirmed cases of CMS. These confirmed cases were stratified into three categories: predominantly limb weakness; predominantly axial weakness and generalized weakness. CHRNE was the defective gene in 36 patients, and was followed by DOK7 (8), RAPSN (5), COL13A1 (4), COLQ (4), GFPT1 (2), GMPPB (2), CHRNA1 (1 receptor deficiency, 1 slow channel) SCN4A (n=1), CHAT (n=1), MUSK (1). Comparison between gene related cases was done with Fisher's test. Twenty-nine cases were considered as predominantly limb weakness: 19 CHRNE (52% of CHRNE cases), four DOK7 (50%), two RAPSN (40%), two GFPT1 (100%), one GMPPB (50%) and one COLQ (25%). Predominantly axial weakness was considered to be the phenotypical pattern in 14 cases: four DOK7 (50% of DOK7 cases), four COL13A1 (100%), two RAPSN (40%), two COLQ (50%), one MUSK and one SCN4A. 23 cases were considered as generalized weakness: 17 CHRNE (48% of CHRNE cases), one RAPSN (20%), one COLQ (25%), 1 GMPPB (50%), 1 CHRNA1 (receptor deficiency), 1 slow channel, 1 CHAT. CHRNE cases were significantly not related to predominant axial weakness ($p=0.0001$), and DOK7 cases were not related to generalized weakness ($p=0.0430$). COL13A1 cases were related to predominant axial weakness ($p=0.0014$). In a cohort larger than our previous, we still find pure ocular symptoms not a phenotype typical of CMS, once no CMS cases had this phenotype. Predominant axial weakness seems to be the less frequent of the three patterns verified, and is related to COL13A1 cases, but it is not expected as a CHRNE phenotype. Similarly, generalized weakness is probably a rare presentation of DOK7 CMS.

Doenças Neuromusculares

TL 1105178

MOLECULAR ANALYSIS OF PATIENTS WITH CLINICAL SUSPICION OF ACUTE HEPATIC PORPHYRIA FROM A TERTIARY CENTER IN BRAZIL

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Background: Acute hepatic porphyrias (AHP) consist in acute intermittent porphyria (AIP), hereditary coproporphyria (HCP), variegate porphyria (VP) and porphyria due to delta-aminolevulinic acid deficiency dehydratase (ALAD). Data from USA indicate that the most common is AIP, followed by VP and HCP. Epidemiological data from Brazil are scarce due to lack of available specific biochemical exams and genetic studies. **Objectives:** To describe epidemiological and genetic profile of patients with AHP in a tertiary center in Brazil. **Methods:** We included patients based on clinical history: recurrent abdominal pain, altered colored urine and peripheral or central nervous system manifestation. We reviewed medical records and evaluated all patients. We performed a next generation sequencing panel using a commercial kit. **Results:** We identified 34 suspected patients from 26 different families. Age varied from 18 to 80, median age of 43 years. Majority of patients were female (25/34). 21 patients identified as being white and 13 patients as brown. A total of 26 patients underwent genetic testing. AIP was diagnosed in seven patients from four families, VP was diagnosed in nine patients from five families. Two patients had inconclusive tests and eight patients had a negative panel. Regarding AIP, each family had a different pathogenic mutation: 973C>T (p.Arg325*), c518G>A (p.Arg173Gln); deletion of exon 1; and a splice donor variant (c.912+2T>C). For VP, four families shared the same missense variant in exon 6, c503G>A (p.Arg168His), and one patient had a deletion in exon 5-6. Five out of nine patients with VP identified as brown, and four out of seven with AIP, as white. We also identified seven cases of pathogenic intronic variant for erythropoietic porphyria (c.315-48T>C), in six of them associated with AHP. **Conclusion:** Our data suggest that VP is more common in our population than previously reported. This may be related to our country's history and colonization. VP originated in South Africa, whereas AIP originated in Scandinavia and Northern Europe. More studies are necessary to confirm this epidemiological pattern.

Doenças Neuromusculares

TL 1105206

MUSCLE ULTRASOUND CHANGES CORRELATE WITH RESPIRATORY MUSCLES INVOLVEMENT AND FUNCTIONAL IMPAIRMENT IN SPINAL MUSCULAR ATROPHY

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Background: Although new therapies modified the course of Spinal Muscle Atrophy (SMA), these treatments are costly and clinical trials have been substantially based on clinical assessments. Reliable techniques to evaluate disease progression and therapy response could support more robust clinical trials. Here we investigated ultrasound patterns of muscle involvement in different types of SMA and their correlation with functional status to determine the validity of this non-expensive, and easy-to-use assessment. **Objective:** We examined 41 patients with SMA (types 1 to 4) and 46 age-, and gender-matched healthy controls using B-mode ultrasound (whole muscle method) for grey scale analysis (GSA), area (biceps brachii and rectus femoris) and diaphragm thickening ratio. **Methods:** Functional scales were applied to patients only. We analyzed ultrasound abnormalities in specific clinical subtypes and correlated findings with functional status. Also, we analyzed if it's possible to predict respiratory involvement using GSA. **Results:** Compared to controls, patients had reduced muscle area and increased mean GSA with correlation with the SMA severity, and the GSA of their intercostal muscles differed more in relation to the controls, as compared to differences in the diaphragm histograms. Biceps brachii GSA was correlated with diaphragm and intercostals GSA ($p=0.019$ and 0.008), and linear regression showed that it could predict respiratory muscle involvement ($R=0.456$ and 0.489 respectively). **Discussion:** Muscles are affected on ultrasound in SMA patients with correlation with SMA severity. Biceps brachii GSA can possibly be used to predict respiratory involvement in SMA patients because it is correlated with diaphragm and intercostal muscle involvement.

Doenças Neuromusculares

TL 1105230

CLINICAL-EPIDEMIOLOGICAL PROFILE AND FACTORS ASSOCIATED WITH PROGNOSIS IN PATIENTS WITH MYASTHENIA GRAVIS IN A REFERENCE HOSPITAL IN NORTHEASTERN BRAZIL

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Background: Myasthenia gravis (MG) is an acquired chronic autoimmune, neuromuscular disorder due to autoantibodies formed against the relevant post-synaptic membrane proteins at the neuromuscular junction synapsis. According to the Myasthenia Gravis Foundation of American Clinical Classification (MGFA), the severity varies from mild weakness limited to the ocular muscles to generalized limb or axial muscles and bulbar weakness. This classification is designed to identify subgroups of patients who share distinct clinical features or severity of disease that may indicate different prognoses or responses to therapy. **Objective:** This study aims to identify potential clinical predictors for the prognosis of MG, comparing groups with mild-moderate disease (MGFA I to III) and with severe disease (MGFA IV and V). **Methods:** Within 88 patients followed at the Neuroimmunology Service from January 2006 to December 2021, this retrospective study included 79 patients whose medical records were available. The diagnosis of MG was based on a combination of clinical, and laboratory criteria, and electromyography. **Results:** Regarding gender, 47 (59.5%) were female, corresponding to an overall female-male ratio of 1.46: 1. In the clinical presentation, generalized weakness predominated (48.1%), followed by exclusively ocular symptoms, (40.5%), and less frequently by the involvement of the bulbar muscles (11.4%). As for the myasthenic crisis, 24 (20.4%) patients had at least one episode during follow-up, and for 9 of them, this was the initial clinical presentation. When comparing the group with MGFA classification ranging from I to III, to the group in classes IV to V, it was shown that greater severity was associated with the bulbar form of MG presentation, and more frequent positivity of anti-AChR antibodies, plus the greater frequency of thymoma and thymic hyperplasia. All these associations reached statistical significance. **Conclusion:** In summary, this study showed that initial presentation of the bulbar form, positivity for Anti-AChR, and association with thymic alterations are prognostic factors for MG.

Doenças Neuromusculares

TL 1105239

GENOTYPIC AND PHENOTYPIC SPECTRUM OF FIG4 VARIANTS IN CHARCOT MARIE TOOTH DISEASE

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Charcot Marie Tooth disease (CMT) type 4J is a rare inherited peripheral neuropathy, affecting less the 1% of patients with CMT. It is caused by biallelic disease causing variants in FIG4 gene, with patients presenting a variable phenotype and onset ranging from early childhood to adulthood. According to the literature, most individuals typically have both proximal and distal asymmetric muscle weakness of the upper and lower limbs. In addition to the CMT phenotype, pathogenic variants in FIG4 gene have also been related to motor neuron disease, bilateral temporooccipital polymicrogyria, and Yunis-Varon syndrome. Here, we describe the clinical, genetic, neurophysiological and other ancillary data from 3 unrelated individuals with CMT4J evaluated at two referral centers for neuromuscular diseases. All patients have early-onset disease, with symptoms beginning before age 5 years, and ENMG exam with a demyelinating pattern. One of the patients started to use a wheelchair as a mobility aid at the age of 43 and the other 2 patients maintain an independent gait. Two patients who underwent brain MRI did not have significant radiological findings. One of the patients had an abnormal pulmonary function test with FVC: 50% and FEV1: 54%, indicating a restrictive pulmonary pattern. Spinal radiography evaluation showed mild scoliosis in two individuals. Genetic test revealed the variant c.122T>C (p.Ile41Thr) in a homozygous state in the FIG4 gene in all three patients. Despite being a rare variant of CMT, patients with disease causing variants in FIG4 usually have more severe forms, including potential respiratory involvement. Accurate genetic diagnosis is very important for appropriate genetic and reproductive counseling of patients.

Doenças Neuromusculares

TL 1105312

CHARACTERIZATION OF THE RESPIRATORY PROFILE OF PATIENTS WITH SPINAL MUSCULAR ATROPHY IN A TERTIARY CENTER

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Background: Spinal muscular atrophy (SMA) is a disease of motor neurons due to mutations in the SMN1 gene. Respiratory involvement is the main cause of death in SMA and respiratory care should be introduced early in disease course. Knowledge about the respiratory clinical profile is necessary to ensure the appropriate ventilatory management in SMA patients. **Objective:** To present the respiratory clinical profile of SMA patients followed up at a tertiary center. **Methods:** Descriptive and retrospective study carried out from July 2021 to June 2022. Data was collected from medical records of respiratory physiotherapy assessments, such as use of invasive and non-invasive ventilation support, forced vital capacity (FVC), peak cough flow (PCF), oxygen saturation, heart rate, chest deformity and scoliosis. **Results:** A total of 45 SMA patients were included, 27 (60%) male and 18 (40%) female. Mean age was 12.9±9.5 years, mean height was 1.39±0.26 m, and mean weight was 38.2±22.43 kg. Most patients (49%) were SMA type 2, 36% were type 3 and 14% were type 1. Only 11% had recurrent respiratory infections, 13% had symptoms of alveolar hypoventilation, 0.04% had a paradoxical pattern and 13% complained of dyspnea. Three (6%) patients had a history of weight loss and 13 (29%) had dysphagia. Regarding the use of ventilation support, 14 (31%) use non-invasive and 4 (8%) use invasive. The mean FVC was 1.81±1.45 l, with 24% of patients with severe restrictive ventilatory impairment, 6% with moderate restrictive impairment, 11% with mild restriction and 37% of the patients had no decrease in FVC. The mean PCF was 250.27±139.98 l/min. Mean oxygen saturation was 97±0.70%, heart rate was 94±15 beats per minute, respiratory rate was 20±5 breaths per minute and 55% of the patients had chest deformities. **Conclusion:** Through an evaluation of ventilatory clinical parameters, it was possible to classify and identify the degree respiratory involvement SMA patients. It is remarkable that severe ventilatory restriction is the most frequent finding. Such information will be useful for planning the respiratory management and care of these patients.

Doenças Neuromusculares

TL 1105325

PERIOPERATIVE NURSING CARE MEDIATING THE CARE OF MYASTHENIC PATIENTS SUBMITTED TO TIMECTOMY: AN INTEGRATIVE REVIEW

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The pathology of Myasthenia gravis (MG) is of idiopathic cause, resulting in alteration in the neuromuscular junction by the presence of Anti-Acetylcholine receptor (AChRs), preventing the physiological action of this chemical mediator resulting in several clinical forms of this disease: neonatal MG, congenital MG, MG of drug origin and autoimmune MG. **Objectives:** This study aimed to highlight the importance of perioperative nursing care as a mediator in the qualification of care to be provided in myasthenic patients submitted to thymectomy. **Methods:** This is an integrative review bibliographic study that resulted in an active search in the VHL Regional Portal and Scielo Research Portal with selected titles between 2008 and 2021. Thirty publications that met the proposed objectives were selected and grouped, categorized, synthesized, and described in thematic axes according to the similarity of the subjects treated. **Results:** In the analyzed articles, it was observed that the patient who needs myasthenia presents generalized weakness and, in this condition, besides the ocular muscles, there is an impairment of the muscles of the limbs and the trunk, that may affect the muscles of the oropharynx and muscles involved with breathing, making the use of the Perioperative Nursing Care Systematization (PNCS) a valuable tool in the dynamization of nursing care through the three phases of the surgical experience: the immediate and mediate preoperative, intra- or transoperative and immediate postoperative. **Conclusion:** It is concluded that the knowledge about MG aligned to PNCS will corroborate with the delineation of actions involving the care planning to be directed to the patient, from clinical judgment supporting evidence of signs and symptoms.

Doenças Neuromusculares

TL 1105443

CAREGIVER OVERLOAD IN THE CONTEXT OF RARE NEUROMUSCULAR DISEASES

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Background: Rare neuromuscular diseases are chronic degenerative, presenting, in some cases, a motor and/or bulbar deficit. The patient may evolve with physical and psychological impairment, thus becoming dependent on a caregiver, which impacts the life of the patient and his companion. In view of this, the caregiver may present an overload in the face of the new demands that arose with the illness of their loved one. **Objectives:** The objective of this study is to identify the presence of burden in caregivers of patients with rare neuromuscular diseases. **Methods:** Cross-sectional and descriptive study, with selected rare neuromuscular diseases. As inclusion criteria, having rare neuromuscular disease and over 18 years of age. We excluded patients whose medical conditions made it impossible to understand or perform the tests (such as acute confusional state, aphasia, reduced level of consciousness, dementia, and sensory deficits). The following clinical instruments were applied to caregivers of patients: form with sociodemographic factors; ZARIT scale (caregiver burden assessment) and FACIT (Functional Assessment of Chronic Illness Therapy-Spiritual Well-Being/FACIT-Sp). The functional independence measurement (FIM) and activities of daily living (BARTHEL) scales were applied to the patients, in addition to the PHQ-9 (Patient Health Questionnaire-9) to screen for major depressive episodes (MDE). **Results:** 43 caregivers were included in the study; 28 patients (65.11%) were female and age was 42 ±18.4 years. ZARIT Scale was 14.0 (9.2-22.5), being a mild classification to caregiver overload, and the total score of FACIT-Sp was 33 (28.0-37.0). These results are correlated with caregivers of a group of patients who have 77 (55.2-111.5) of measure of functional independence, with modified dependency up to 25% and PHQ-9: 9 (5.0-13.0), with mild depression. However, this sample shows severe dependence for activities of daily living which showed in Barthel Scale with 42 (25.0-78.7). **Conclusion:** The assessment of caregiver burden was mild in the studied sample of caregivers of our patients. This sample, despite presenting with severe dependence for activities of daily living, shows functional dependence modification of up to 25% and low probability of depression, besides the FACIT Spiritual Well-Being of the caregivers. It is possible that happens psychological biases when caregivers, especially family members, are questioned about their position as caregivers.

Doenças Neuromusculares

TL 1105468

EVALUATION OF DYSPHAGIA, SARCOPENIA AND PULMONARY FUNCTION IN PATIENTS WITH RARE NEUROMUSCULAR DISEASES

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Background: Neuromuscular diseases (NMDs) affect the Central Nervous System (CNS) and the Peripheral Nervous System (PNS). They can be chronic, progressive and degenerative. Dysphagia is characterized by changes in one or more phases of swallowing biomechanics, preventing adequate safe, efficient and comfortable oral intake. It can cause important functional damage to the individual, such as pneumonias, malnutrition, dehydration and mortality. Another frequent manifestation is sarcopenia, which is defined as the natural and progressive loss of muscle mass and strength, whether peripheral or respiratory. **Objective:** The objective of this study is to evaluate these manifestations in a group of patients from an outpatient clinic for rare neuromuscular diseases, followed by a multidisciplinary team, using the "Eating Assessment Tool – EAT-10, in the classification of the risk of dysphagia. **Methods:** The following instruments were applied: sociodemographic and clinical data; Eating Assessment Tool (EAT-10) scale; Speech and voice assessment; Assessment of strength and resistance of lips and tongue measured by Iowa Oral Performance Instrument (IOPI); Nutritional assessment, including calf circumference and handgrip strength measurement; Maximum inspiratory and expiratory pressure and peak expiratory flow values. **Results:** Seventy-two patients were analyzed, 38.88% men and 61.11% women. Fifty – nine patients with ALS, twenty-three with SMA. According to EAT-10, it was classified patients into risk or not risk for dysphagia. In the risk group, age was 62.0 ± 15.39 years. BMI, calf circumference and hand grip were respectively 25.10 ± 11 Kg/m², 36.0 ± 3.3 cm and 9.8 ± 10.22 kPa. The P_{lmax} and the P_{Emax} were 40.00 ± 29.62 cmH₂O and 31.00 ± 31.82 cmH₂O. Peak flow value and the maximum time of sustained phonation (TMF) were 160.0 ± 103.3 L/min and 6.46 ± 9.66 s. The maximum peak of tongue tip and dorsum were, respectively, 17.50 ± 13.56 kPa and 17.00 ± 12.20 kPa. Finally, the endurance strength result was 9.00 ± 7.19 s. Values ??from the group without risk of dysphagia were higher, but no significant differences when the groups were compared. **Conclusion:** The importance of a multidisciplinary team in the follow-up of patients with neuromuscular diseases in order to promote comprehensive care is highlighted. This study presented partial results, requiring an expanded effort to better assess the outcomes of rare conditions that affect the quality of life of these individuals.

Doenças Neuromusculares

TL 1105473

PREVALENCE OF DYSPHAGIA, DYSPHAGIA AND ITS CORRELATION TO TONGUE STRENGTH AND ENDURANCE IN A CASE SERIES OF PATIENTS WITH POMPE DISEASE

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Background: Pompe's disease is defined by the deficiency of acid alpha-glucosidase (GAA) of autosomal-dominant hereditary inheritance that leads to the gradual and systemic accumulation of glycogen inside the lysosomes and generates a picture of progressive muscle weakness, cardiac disorders, insufficiency breathing and a host of other symptoms. Based on the clinical analysis of patients affected by the disease, the involvement of the orofacial anatomy and its musculature was observed, leading to the condition of tongue strength deficit that can have an outcome of dysphagia and dysarthria. **Objective:** report the prevalence of dysarthria, dysphagia and its correlation to tongue strength and endurance in a Serie of patients with Pompe disease. **Methods:** Seven patients were evaluated at a neuromuscular rare disease outclinic from a university Hospital of Rio de Janeiro, from March 2020 to December 2021. Tongue strength and endurance was evaluated using the Iowa Oral Performance Instrument (IOPI). Speech intelligibility was assessed from spontaneous speech. Dysarthria was characterized as mild, moderate or severe according to the screening of the 5 motor bases of speech. Dysphagia was evaluated using the water swallowing test. Tongue strength and endurance was evaluated using the Iowa oral performance instrument (IOPI). **Results:** The average age was 48.42 ± 14.92 years. 42.9% were female (3) and 57.1 male (4). The prevalence of dysarthria was 42.9% (3), being characterized as mild in all patients. Speech intelligibility was presented in 42.9% (3) of the sample. The prevalence of dysphagia was 28.6 (2). The average strength and resistance of the tongue was low, when compared to the reference values ??for age (28 ± 11.07 kPa; 10.71 ± 8.03 kPa respectively). The means of tongue strength and resistance were significantly lower in the groups of patients with dysarthria, dysphagia and impaired speech intelligibility. **Conclusion:** Low tongue strength and resistance are related to the increased prevalence of dysarthria, dysphagia and impairment on speech intelligibility, which demonstrates the importance of including these measurements in the screening of patients with pompe's disease.

Doenças Neuromusculares

TL 1105485

ASSESSMENT OF EXECUTIVE FUNCTIONS IN PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS

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Background: Rare neuromuscular diseases are chronic degenerative, presenting, in some cases, a motor and/or bulbar deficit. Some studies mention a significant presence of executive dysfunctions in patients with Amyotrophic Lateral Sclerosis (ALS). The Five Digit Test (FDT) aims to measure mental processing speed and the ability to direct and switch their attentional control, demanding executive function skills such as response inhibition and cognitive flexibility. **Objectives:** The objective of this study is to describe executive functions in patients with ALS with a multilingual and non-reading test like FDT. **Methods:** Cross-sectional and descriptive study, in which patients treated at an outpatient clinic for rare neuromuscular diseases were selected. As inclusion criteria, having rare neuromuscular disease and over 18 years of age. We excluded patients whose medical conditions made it impossible to understand or perform the tests (such as acute confusional state, aphasia, reduced level of consciousness, dementia, and sensory deficits). The FDT test was applied to assess executive functions in patients with ALS. All percentiles below the 25th are considered "changed", given that scores below the 25th percentile indicate mild difficulties in executive functioning and processing speed, even without necessarily having clinical significance, and scores below the 5th percentile are more indicative of prominent deficits. **Results:** A total of 16 patients were included in the study; 10 patients (62.5%) were male and mean (SD) age was 56 (11.04) years. More than half of patients showed changes to FDT decoding and retrieving (75% and 68.75% respectively). However, the control processes are 56.25% not changed in Response Inhibition and 62.5% not changed in cognitive flexibility. **Conclusion:** Patients with ALS can manifest executive dysfunction severe enough to classify them as cognitively impaired. However, in this sample, despite the automatic processes (reading and counting) being altered, the inhibition and cognitive flexibility, which are control processes and depend on executive function, responses remained preserved. Furthermore, more studies are needed to better understand and monitor executive functions in patients with ALS, as well as discuss an assessment more adapted to the progression of the disease.

Doenças Neuromusculares

TL 1105491

THE AVALGLUCOSIDASE ALFA PHASE 3 COMET TRIAL IN LATE-ONSET POMPE DISEASE PATIENTS: EFFICACY AND SAFETY RESULTS AFTER 97 WEEKS

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Avalglucosidase alfa, a recombinant human GAA enzyme replacement therapy with increased mannose-6-phosphate content for increased cellular uptake, is approved in the United States for late-onset Pompe disease (LOPD) patients ≥1 year of age and in Japan for all Pompe disease patients (Nexvzyme(TM), Sanofi Genzyme, Cambridge, MA). During the 49-week, double-blinded primary-analysis period (PAP) of the Phase 3 COMET trial (NCT02782741) comparing avalglucosidase alfa with alglucosidase alfa, avalglucosidase alfa resulted in greater improvements in forced vital capacity (FVC), 6-Minute Walk Test (6MWT), and other outcomes and a more favorable safety profile than alglucosidase alfa in treatment-naïve LOPD participants. During the open-label extension treatment period (ETP), 51/51 participants receiving avalglucosidase alfa during the PAP continued this treatment and 44/49 receiving alglucosidase alfa during the PAP switched to avalglucosidase alfa. Changes (LS mean [SE]) from Baseline at Week 97 are reported. Changes in FVC %predicted were +2.65 (1.05) for avalglucosidase alfa PAP participants and +0.36 (1.12) for alglucosidase alfa PAP participants. Changes in 6MWT distance (meters) were +18.60 (12.01) for avalglucosidase alfa PAP participants versus +4.56 (12.44) for alglucosidase alfa PAP participants. Similar trends occurred in other Week-97 outcomes. Treatment-emergent adverse events (AEs) during the ETP occurred in 49 (96.1%) and 42 (95.5%) participants from the avalglucosidase alfa and alglucosidase alfa PAP groups, respectively. Five participants discontinued during the ETP by Week 97 due to AEs (ocular hyperemia, erythema, urticaria, respiratory distress, acute myocardial infarction, pancreatic adenocarcinoma). Twenty-two participants had treatment-emergent serious AEs during the ETP. Patients who switched from alglucosidase alfa to avalglucosidase alfa presented no safety- or immunogenicity-related concerns. Overall results demonstrate sustained treatment effect for improvements observed with avalglucosidase alfa during the PAP and stabilization of treatment effect after switching from alglucosidase alfa to avalglucosidase alfa over 97 weeks, supporting long-term maintenance of outcomes and persistence of avalglucosidase alfa's effect.

Doenças Neuromusculares

TL 1105504

AVALGLUCOSIDASE ALFA IMPROVES HEALTH-RELATED QUALITY OF LIFE (HRQOL) IN PATIENTS WITH LATE-ONSET POMPE DISEASE (LOPD) VS. ALGLUCOSIDASE ALFA: PATIENT-REPORTED OUTCOME MEASURES (PROMS) FROM THE PHASE 3 COMET TRIAL

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The phase 3 COMET trial (NCT02782741) assessed PROMs in treatment-naïve adult LOPD patients receiving avalglucosidase alfa (AVAL, n=51) or alglucosidase alfa (ALGLU, n=49). PROMs comprised generic measures (12-item Short-Form Health Survey [SF-12], Patient Global Impression of Change [PGIC]), EuroQoL-5D-5L [EQ-5D-5L]), and LOPD-specific measures (Pompe Disease Symptom Scale [PDSS], Pompe Disease Impact Scale [PDIS], and Rasch-Built Pompe-Specific Activity [R-PAct]) that cover various domains: breathing, mobility, fatigue, disease-related symptoms, upper extremity weakness, pain, morning headache, daily activities, physical functioning, and mental health. Change from baseline to Week 49 was analyzed using mixed models for repeated measures. AVAL was numerically better than ALGLU across all domains except for pain/discomfort (EQ-5D-5L) and mood (PDIS). Between-group difference was nominally significant (p-value <0.05) for PGIC questions: daily activities (least squares mean difference vs. ALGLU [95% confidence interval [CI]]: 0.57 [0.08, 1.06]), disease-related symptoms (0.78 [0.31, 1.25]), mobility (0.65 [0.18, 1.12]); and EQ-5D mobility (-0.31 [-0.62, 0.00]). Post-hoc responder analyses numerically favored AVAL vs. ALGLU for all PROMs. Proportion of responders was nominally significantly greater (p<0.05) for all PGIC questions (daily activities [62.0% vs. 37.0%], disease-related symptoms [74.0% vs. 49.0%], ability to breathe [66.0% vs. 41.0%] and mobility [66.0% vs. 29.0%]), EQ-5D-5L (usual activities [54.0% vs. 30.6%]), and PDSS (shortness of breath [26.0% vs. 4.1%], overall fatigue [22.0% vs. 2.0%], fatigue/pain [18.0% vs. 4.1%], and morning headache [22.0% vs. 4.1%]). Cumulative distribution functions showed a clear separation in favor of AVAL for full range of thresholds in PDSS (shortness of breath, overall fatigue, and fatigue/pain) and PDIS (difficulty in performing daily activities). These LOPD-specific PROMs provide further support for greater therapeutic benefit of AVAL over ALGLU, including the most relevant LOPD domains - mobility, usual activities, and breathing. This dataset confirms meaningful impact of AVAL on LOPD patient's quality of life.

Doenças Neuromusculares

TL 1105564

DIAGNOSTIC YIELD OF WHOLE-EXOME SEQUENCING AND GENETIC PANEL IN ADULT BRAZILIAN PATIENTS WITH MYOPATHY

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Background: Myopathies in adults have multiple etiologies, but a significant proportion of cases are due to genetic variants in more than 200 known genes. Whole-exome sequencing (WES) and genetic panels (GP) have been employed as a diagnostic test in this setting with variable success rates. Factors such as age, ethnic background and phenotypic characterization of patients seem to influence in both accuracy for myopathies. Herein, we investigated the diagnostic yield of WES and GP in a cohort of adult patients with myopathy coming from a population with mixed ethnic background (Brazil). **Objective:** to determine the accuracy of WES and commercial GP in a population of patients from a reference neuromuscular center with suspected genetic myopathy. **Methods:** 106 patients attending a reference neuromuscular center were recruited. Diagnosis for all of them remained elusive after physical examination, muscle biopsy, electrophysiology and lab tests. WES was performed following standard procedures for 30 patients, while GP was performed for 76. We identified variants labeled as pathogenic or likely pathogenic following ACMG guidelines. Then, diagnostic yield of WES and GP were determined for the whole cohort and for subgroups defined according to the major phenotypes. **Results:** Mean age of patients was 38 years and there were 43% men. Definite genetic diagnosis was found in 12 from 30 patients in WES group (40% accuracy), while 36 from 76 in the genetic panel group (47%), leading to a diagnostic yield of 45% in this cohort. Pathogenic variants were found at TRAPPC11, SCN4A, ACADS, COL6A2, SGCB, CAPN3, SGCB, POLG, TWNK, RYR1, TRIM32, TTN, EMD, SLC25A4, TPM3, DOK7, DMD, ANO5, CHRNE, GAA, ADSS1, CLCN1, PYGM, PNPLA, HAD1, NEB, DPAGT1, MYH7, FKTN. **Conclusion:** WES and GP are useful diagnostic test for adult patients with undefined myopathy. In this mixed ethnic background cohort, diagnostic yield was 45% and variants in multiple genes were identified.

Doenças Neuromusculares

TL 1105650

FHL1-RELATED MYOPATHY IS ASSOCIATED TO INFLAMMATORY FINDINGS IN SKELETAL MUSCLE HISTOLOGY

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Background: FHL1 (Four-and-a-Half-LIM domains 1) is a protein involved in signaling processes, differentiation and maintenance of structural cellular elements. Mutations in FHL1 can lead to a myopathy with myofibrillar derangement and intracytoplasmic bodies, known as reducing body myopathy (RBM). RBM is characterized by progressive weakness in childhood and the presence of cytoplasmic aggregates that reduce nitro-blue-tetrazolium (NBT) in muscle fibers. In addition to RBM, other phenotypes are described, such as Emery-Dreifuss, X-linked scapuloperoneal myopathy and hypertrophic cardiomyopathy. We recently described a case of RBM presenting as inflammatory myopathy. However, there is no data systematically investigating inflammatory findings in muscle biopsy in FHL1-related myopathy. **Objective:** To evaluate inflammatory findings in skeletal muscle histology of patients with FHL1-related myopathy. **Methods:** We retrospectively identified patients with FHL1-related myopathy in a tertiary neuromuscular center. We reviewed medical records to clinical description. We evaluate histopathological findings of muscle biopsy of the patients using the following stains: hematoxylin and eosin, modified Gömöri's trichrome, NADH-tetrazolium reductase, succinate dehydrogenase, cytochrome-c-oxidase, acid phosphatase, NBT and immunohistochemistry using commercial antibodies against desmin, CD4, CD8, CD68 and major histocompatibility complex type I (MHC-I). **Results:** Five patients with FHL1-related myopathy were clinically evaluated. Two patients (one male and one female) presented the classical RBM phenotype, and three females presented an X-linked scapuloperoneal myopathy. All patients presented increased serum CK over 10x the UNL. Histopathological analysis from all patients showed intracytoplasmic desmin-positive aggregates, NBT-reducing bodies, necrotic fibers and intense increased acid phosphatase activity. Moderate endomysial CD4+, CD68+ cell infiltrates, in addition to milder CD8+ infiltrates, with mild to moderate increase in MHC-I staining was observed in all patients. **Conclusion:** FHL1-related myopathy is associated with intense inflammatory findings in skeletal muscle histology. Some cases can be misdiagnosed as inflammatory myopathy. Curiously, anti-FHL1 antibodies was previously described in 25% of patients with idiopathic inflammatory myopathy, which indicate that FHL1 can be an epitope able to induce an inflammatory response.

Doenças Neuromusculares

TL 1105706

PROFILE OF RESPIRATORY CHAIN ENZYME ACTIVITY IN PATIENTS WITH EXERCISE INTOLERANCE AND MILD CHANGES AT MUSCLE BIOPSY

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Background: Exercise intolerance is a nonspecific term used to describe disproportionate fatigue, myalgia and/or cramping as a consequence of muscular exertion. Although it is a common complaint, a specific etiologic diagnosis is rarely achieved, even after comprehensive investigation. **Objective:** In the present study we aim to define the profile of respiratory chain enzyme activity in skeletal muscle of patients with exercise intolerance and mild and nonspecific changes at muscle biopsy. **Methods:** One hundred and three adult patients with long lasting unexplained exercise intolerance were included in the study. They were followed at the outpatient clinic of our University Hospital and were subjected to muscle biopsy in the course of diagnostic investigation. Inclusion criteria: 18 years old or above; chronic (2 years or more) myalgia and/or fatigue in relation to exercise; non-specific findings in the muscle biopsy. Exclusion criteria: insufficient muscle sample size for enzyme assays. Respiratory chain activities of complexes I, II, III, II+III and IV were measured in whole muscle homogenates. Citrate synthase activity was measured to estimate the mitochondrial mass, which was used to correct enzyme activity. **Results:** Activity of respiratory chain enzymes was unremarkable in 70% of patients. However, 16% of patients presented a complex I deficiency (mean 0.38 and SD 0.29 micromol/min/g tissue; when corrected for citrate synthase activity). Another 9% of patients presented isolated deficiency of complexes II (mean 0.48 and SD 0.34), III (mean 0.72 and SD 0.25) or IV (mean 1.12 and SD 0.3). In 5% of patients, there was a combined deficiency of complexes I, III and IV. **Conclusion:** Although skeletal muscle biopsy with routine histochemical studies frequently show mild and nonspecific changes in patients with exercise intolerance, respiratory chain enzyme assays may contribute to a better understanding of the pathophysiology of exercise intolerance in a significant portion of those patients.

Doenças Neuromusculares

TL 1105713

HISTOPATHOLOGICAL FINDINGS OF RYANODINE RECEPTOR 1 (RYR1)-RELATED MYOPATHIES IN A COHORT FROM A TERTIARY NEUROMUSCULAR CENTER

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Background: Ryanodine receptor 1 (RYR1)-related myopathy presents with a variety of phenotypes characterized by early onset of symptoms such as hypotonia, delayed motor development, proximal muscle weakness, ptosis and/or ophthalmoparesis, facial weakness, thoracic deformities, and spinal involvement. Among the congenital myopathies, RYR1-related myopathy is the most frequently diagnosed which includes a heterogeneous spectrum of histopathological and clinical subtypes followed by variable disease severity. **Objectives:** To describe the epidemiological and histopathological findings on muscle biopsy of a case series with RYR1-related myopathies in a tertiary referral center. **Methods:** We retrospectively selected patients who had a clinical history of proximal myopathy or congenital myopathy and a positive genetic test for the RYR1 gene. We evaluated clinical features and histological aspects. The evaluation of muscle biopsies included grading the fiber variability, nuclei internalization, presence of type 1 or type 2 atrophy, intermyofibrillar disorganization, presence of cores and presence of necrosis. **Results:** Thirty-one patients with RYR1-related myopathy were included. Age varied from 4 to 62 years old (median age of 17). Most patients (61.3%) were male. The most common phenotype was classical congenital myopathy (25/31), representing 80%, followed by later-onset congenital myopathy with 9.6%. One patient presented with rhabdomyolysis phenotype and one patient with a distal myopathy. Ptosis was reported by 22.5% of the patients and a history of malignant hyperthermia in one patient. Regarding histological findings, central core pattern appeared in five cases. The centronuclear pattern occurred in six patients, as well as dystrophic pattern was seen in other six ones. Importantly, one patient showed abundant necrosis on muscle biopsy, in addition to the presence of intracytoplasmic cores. **Discussion:** We present a large cohort of patients with genetically confirmed RYR1-related myopathy. Histopathological findings varied, as did phenotypes. Most patients had dystrophic or centronuclear patterns. An interesting finding was the presence of abundant fiber necrosis in one patient, which is not a characteristic usually described in these patients. The broad spectrum of RYR1-related myopathy is not only clinical but also histological.

Doenças Neuromusculares

TL 1105739

THE RELATION BETWEEN MYASTHENIA GRAVIS AND COGNITION: A REVIEW OF LITERATURE

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Background: Myasthenia Gravis (MG) is an autoimmune chronic disease that involves neuromuscular junction generating muscle weakness and fatigability. In addition to peripheral muscle involvement, cognitive impairment has been hypothesized for a long time. Controversial results about the impact of MG in cognitive functions are present in varied studies. **Objective:** To evaluate the literature regarding relation between MG and cognitive dysfunction. **Methods:** An extensive literature review was carried out based on analysis and selection of articles and publications found on MEDLINE, SCIELO, LILACS and grey literature. We included in this review studies referring to congenital myasthenia to access myasthenic syndromes. **Results:** In this search, 415 records were found, corresponding to 101 different studies selected. About 20 studies compared cognitive impairment between patients with myasthenia and healthy control. The majority of those studies demonstrated correlation between myasthenia and cognitive impairment. The main domains affected were language, visuospatial function, information processing and memory. **Conclusion:** Despite the fact that MG is traditionally seen as a purely motor disease, there are evidences suggesting it is related to cognitive dysfunctions in multiples domains. Beside that, it is associated with more neuropsychiatrist symptoms and state of fatigue. Further studies are needed to evaluate the impact of cognitive impairment in myasthenic patients and the pathophysiological mechanisms.

Doenças Neuromusculares

TL 1105743

PEDIATRIC NEUROMUSCULAR DISORDERS IN A TERTIARY HOSPITAL AT NORTHEASTERN BRAZIL

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Background: Neuromuscular disorders (NMD) are diagnosed across the lifespan and create many challenges especially with infants, children, and adolescents. NMD typically cause muscle weakness, and usually, the symptoms slowly get worse over time. **Objective:** To evaluate the epidemiological, clinical, and diagnostic profile of pediatric patients with neuromuscular diseases. **Method:** A retrospective cross-sectional study with consultation of outpatient records of child and adolescent population symptomatic for neuromuscular diseases treated at a specialized referral center at a university Hospital in northeastern Brazil. The statistical study was descriptive of the variables analyzed. **Results:** We evaluated 77 patients (60% male), age at onset of symptoms 1 day of life-15 years. Affected age group: <2 years=26 (34%, 65% men; and <6months 65.4%); 3-6years=25 (32.4%, 60%men); 7-10 years= 19 (24.6%, 52.6% men); and >11 years=7 (9%, 44.4% men). Diagnoses were grouped into: Myopathies= 45 patients, 58.4% (Mitochondrial 22.2%; Congenital 18%; Metabolic 6.6%; Inflammatory 6.6%; Duchenne Dystrophy (DMD) 15.6%; Sarcoglycanopathies 11.1%; LAMA2- 11.1%; Emery-Dreifuss 4.4%; Becker 2.2%; Collagen 6- 2.2%); Motor Neuron Diseases= 14 patients, 18.2% (Spinal muscular atrophy - SMA-1 50%; SMA-2 7.2%; SMA-3 28.5%; others 14.3%); Neuropathies=11.7% (Charcot Marie Tooth- CMT 77.8%); Myoneural Junction Disease= 7.8% (Myasthenia Gravis (MG) 83.3%; Myasthenia congenita 16.7%); Plexopathy= 3 (3.8%, Traumatic 66.7%; Parsonage Turner 33.3%). Muscle involvement occurred in 11.7% eye (66.7% MG), 28.5% face (32% SMA), 31.2% swallowing (29% SMA), 19.5% neck (46.7% SMA), 72.7% upper limbs, 84.4% lower limbs. Respiratory involvement 26% (SMA 40%; LAMA2 20%). Impairment in walking 53.2% (SMA 29.2%; Dystrophies 19.5%; CMT 14.6%). Intellectual Deficit 10.4% (DMD 50%). Heart disease 6.5% (DMD 40%). **Conclusion:** Our study showed that neuromuscular diseases begin more frequently in boys, infants (especially, younger than 6 months) and preschoolers. More than half of patients have Myopathies (mainly, mitochondrial). The most affected muscle groups were those of 4 members, mainly the lower ones. However, SMA patients were frequently affected by the face, swallowing, neck, breathing and walking, being a greater gravity. Thus, understanding our pediatric population with neuromuscular diseases, accurate diagnosis, correct therapeutic and better prognosis can be performed.

Doenças Neuromusculares

TL 1105842

HOSPITALIZATIONS FOR TREATMENT OF ACUTE DEMYELINATING POLYRADICULONEURITIS AND THE PANDEMIC: A QUANTITATIVE STUDY

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Background: Acute immune-mediated polyneuropathies, also called Guillain-Barré syndrome (GBS), is one of the main causes of acquired weakness. The pathogenesis occurs when an immune reaction, in response to a previous infection, cross-reacts with shared epitopes in the peripheral nerve. This process generates a clinical condition characterized by symmetric and progressive limb weakness, sensory symptoms and dysautonomia. Some cases require immediate Hospitalization, since among the main complications is bulbar weakness, which can lead to respiratory failure. Several cases of GBS associated with COVID-19 have been reported in the literature, although no direct causal relationship has been established. **Objective:** This article aims to verify if there was an increase in the number of GBS diagnoses in Brazil during the pandemic, looking for a possible causal correlation with COVID-19. **Methods:** This is a descriptive quantitative study, conducted in the database of the Ministério da Saúde, DATASUS, in the Hospital Procedures section (SIH/SUS) with data on Hospitalizations for treatment of acute demyelinating polyradiculoneuritis referring to the period from January 2017 to December 2021. The values were categorized by year of attendance. Data analysis was performed in Microsoft Excel version 2019 program. **Results:** A reduction in the number of Hospitalizations for treatment of acute polyradiculoneuritis was observed during the years 2020 to 2021, characterized by a total of 3740, in relation to the amount found from March 2018 to February 2020, 4362 Hospitalizations, with the year 2020 having the lowest Hospitalization rate, 21.7%, compared to the total of the period analyzed. About the reduction of expected cases compared to the real data, it is hypothesized that there was a decrease in the number of beds available for treatment of GBS cases due to the resettlement for severe cases of covid. Also, one can speculate about an underreporting of this pathology and, finally, the safety protocols during the period avoided transmission routes for etiological agents of GBS, possibly decreasing its incidence. **Conclusion:** The data presented point to a lower incidence of GBS in the period. Future studies will be necessary to better understand the relationship between COVID-19 and acute polyradiculoneuritis, which was found to be unassociated by the present study. The lack of specific data for GBS case diagnosis in the DATASUS system was a limitation of the study.

Doenças Neuromusculares

TL 1105879

EPIDEMIOLOGICAL PROFILE OF 494 PATIENTS WITH NEUROMUSCULAR DISEASE IN A BRAZILIAN UNIVERSITY CENTER

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Background: It is of utmost importance to seek cross border collaboration and common ground to facilitate disease's diagnosis and management. Nonetheless, little effort has been made regarding epidemiological profile of neuromuscular disorders. **Objective:** This report analyses the epidemiological profile of 494 patients that presented for consultation in our university Hospital, a tertiary care center for neuromuscular patients in Brazil. **Methods:** Data was collected from April 2015 to April 2022. The patients were stratified according to topography – (1) nerve; (2) muscle; (3) motor neuron; (4) neuromuscular junction – and etiology. **Results:** The most prevalent disorder was neuropathy (314 cases, 63,5%), followed by neuromuscular junction disorders (73 cases, 14,1%), myopathy (53 cases, 11,8%) and motor neuron disease (40 cases, 8%). Among patients with neuropathy, 276 (87,8%) cases were related to acquired etiologies and 38 (12,25%) to hereditary causes. In the subgroup of acquired neuropathies, major etiologies were inflammatory in 97 cases (35,2%), metabolic in 48 (15,3%), infectious in 35 (11,1%), toxic in 4 (1,1%) and there were 17 cases with more than one etiology hypothesized, 34 cases with other etiologies (nutritional, post-surgical, compressive or traumatic) and 39 (14,13%) cases with no etiology defined (currently under investigation). **Conclusion:** The majority of neuromuscular disease in our cohort was represented by nerve disorders and the inflammatory etiology was the most frequent. Since this study is limited to only one center, and little effort in specialized literature has been made to set common referral patterns, selection bias and information bias may affect the interpretation of the data. Nonetheless, this study contributes for further analysis including topographic and etiological epidemiology of neuromuscular diseases and calls out for national as well as cross-border collaboration to proceed a standardized approach to neuromuscular diagnosis.

Doenças Neuromusculares

TL 1106082

SENSORY NEURONOPATHY IN MULTIPLE ACYL-COENZYME A DEHYDROGENASE DEFICIENCY (MADD)

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Background: Multiple acyl-CoA dehydrogenase deficiency (MADD) is a recessive disorder due to ETFA, ETFB, and ETFDH pathogenic variants. Typical late-onset presentation includes exercise intolerance, cyclical vomiting, and progressive muscle weakness. Only a few reports of sensory neuronopathy were associated with MADD, with variable improvement after riboflavin supplementation. **Objectives:** We aim to describe MADD-associated sensory ganglionopathy from a cohort of MADD patients in a tertiary neuromuscular center. **Methods:** This retrospective study presents clinical, molecular and response characteristics after treatment. **Results:** We identified five cases with myopathy and genetically confirmed MADD. Three patients manifested with sensory neuronopathy. All these cases were female, aged 34, 67 and 74. Physical examination was remarkable for severe sensory ataxia and global weakness. Two patients were unable to walk, complained of fluctuating hoarseness, and had a dropped head on examination. Electrodiagnostic studies were consistent with a ganglionopathy in all cases, but two patients also had a superimposed myopathy. Serum CK levels were high in only one patient. Muscle MRI of the three patients had fat substitution or edema. Acylcarnitine profiles were collected in two patients, with only one test consistent with MADD. Muscle biopsies of all patients showed lipid droplets and mild mitochondrial dysfunction. A next-generation sequencing panel for myopathies led to different heterozygous ETFDH pathogenic variants in the patients. Riboflavin, coenzyme Q10, and L-carnitine were supplemented in all patients with a significant response in motor function, and variable response in sensory symptoms. One patient regained the capacity to walk. **Conclusion:** MADD, a curable myopathy, is often missed in its typical presentation. An atypical presentation as presented here is even more ignored, and probably missed. Despite extensive investigations, many sensory ganglionopathies remain idiopathic, therefore, we propose MADD should be in differential diagnosis of sensory neuronopathy, especially with the phenotype here described. All three patients had a heterozygous variant, and we could not conclude whether there is a dominant inheritance related to this atypical manifestation, or the second variant was not found. In author's experience, the motor function has a better response than sensory symptoms, but further studies and phenotype characterization are needed.

Doenças Neuromusculares

TL 1106120

ANALYSIS OF THE GENETIC AND EPIDEMIOLOGICAL PROFILE OF PATIENTS WITH LIMB-GIRDLE MUSCULAR DYSTROPHY IN REFERENCE CENTERS FROM PERNAMBUCO

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Background: Limb-girdle muscular dystrophies (LGMD) are the fourth leading cause of genetically generated muscle weakness. LGMDs form a heterogeneous group of diseases, which includes hereditary pathologies that preferentially affect the proximal muscles of the pelvic and scapular girdle. There are currently 29 subtypes, the vast majority of which are autosomal recessive. As it is a rare pathology, prevalence data are limited. There is a lack of studies that evaluate the epidemiological and genetic profile of LGMDs in the Brazilian northeast region. **Objective:** Evaluate the epidemiological and genetic aspects of patients with suspected limb-girdle muscular dystrophy who underwent a molecular panel at two referral centers in Pernambuco. **Methods:** This is a cross-sectional observational study. Data were collected from the medical records of patients treated in two centers from Pernambuco. Demographic data such as age, gender and origin were analyzed, as well as the results of the molecular panels performed by the patients. **Results:** Information was collected from the medical records of 37 patients, 17 had their diagnosis confirmed, the majority being sarcoglycanopathies (70.6%), especially alpha-sarcoglycanopathy with 35.5% of all LGMD subtypes found. The distribution by age and sex was similar in the group of 17 patients diagnosed with LGMD, with a higher prevalence of alpha-sarcoglycanopathy among those younger than 18 years (66.7%), with a higher number of women in this subtype (88.3%) being also observed. The origin of 14 of the 17 diagnosed patients was found, the majority (four) were from Recife-PE. All patients underwent a molecular panel, a total of 52 different mutations were observed, being 53.8% pathogenic variants, 17.3% probably pathogenic variants and 28.8% variants of uncertain significance (VUS). Regarding the pathogenic and probably pathogenic variants, most were found in the SGCA gene and in CAPN3. The most common mutation was c.229C>T in the SGCA gene (61.5% of the variants in this gene). Among the VUS, most corresponded to DYSF gene mutations with 33.3%, followed by FKRP and CAPN3 with 20% each. **Conclusion:** Despite presenting a small population sample, the study presented relevant data on the genetic and epidemiological profile of LGMDs in Pernambuco. The results presented can promote studies, in addition to strengthening general population and professional health knowledge.

Doenças Neuromusculares

TL 1106133

THE EVALUATION OF A SECOND CYCLE OF INTRAVENOUS IMMUNOGLOBULIN FOR PATIENTS WITH GUILLAIN-BARRÉ SYNDROME ON MECHANICAL VENTILATION

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Background: Approximately 25% of patients with Guillain-Barré syndrome (GBS) do not respond to the first cycle of intravenous immunoglobulin (IVIg) infusion and are offered a second cycle. There are a few studies on the effectiveness of this second infusion. **Objectives:** To evaluate the effectiveness of the second cycle of IVIg in individuals with GBS who required mechanical ventilation in the acute phase. **Methods:** We performed a retrospective review of the database of patients with GBS who required mechanical ventilation, between the years 1994 to 2018, aged > 6 years, with Brighton level 1 or 2 and who received 1 or 2 cycles of IVIg, in the state of Rio Grande do Norte. Demographic, clinical and outcome data up to 6 months were compared. **Results:** 74 individuals from a sample of 350 (21.4%) were selected. 59 (79.7%) received 1 cycle IVIg and 15 (20.3%) received 2 cycles IVIg. There was no difference between age (34 x 39 years; median; $p=0.746$) and sex ($p=0.5501$). There was also no difference between nadir (8 x 6 days; $p=0.3979$), facial paralysis ($p=1.000$), sensitive alterations ($p=0.2207$), presence of dysautonomia ($p=0.1932$), presence of anti-GM1 ($p=1.000$) and demyelinating and axonal electrophysiological subtypes ($p=0.1241$). The 1-cycle IVIg group had less diarrhea ($p=0.0002$) and more proximal weakness ($p=0.0051$). The 2-cycles IVIg group had more global weakness ($p=0.0141$) and tracheostomy ($p=0.0077$). The ability to walk at 6 months was similar (48X13 cases; $p=1.000$). However, the median time to walk was shorter in the 1-cycle IVIg group (31 x 90 days; $p=0.0093$), as was the time to gain 1 point on the Hughes disability scores (14 x 40, $p=0.0004$), the time on mechanical ventilation (13 x 34; $p=0.0001$) and the days of Hospitalization (43 x 59.5; $p=0.0014$). **Conclusion:** This observational study did not observe better results after a second cycle of IVIg in GBS under mechanical ventilation. The absence of a history of diarrhea and the presence of proximal weakness were significantly present in the 1-cycle IVIg group, suggesting a good prognostic marker.

Doenças Neuromusculares

TL 1106175

PHENOTYPE-GENOTYPE CORRELATION IN DESMINOPATHY: VARIANT LOCATION IN DES GENE DETERMINES THE SEVERITY OF PHENOTYPE AND THE MORPHOLOGY OF INTRACITOPLASMIC AGGREGATES

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Background: Desmin (DES) is the main intermediate filament in the muscle, arranged in a tridimensional structure that connects myofibrils to each other as well as to the nuclei, the sarcolemma and organelles. Pathogenic variants in DES cause desminopathy, a disorder affecting skeletal muscles and the heart. It is characterized by a spectrum of phenotypes and arrangements of intracytoplasmic aggregates. **Objectives:** We aimed to analyze the clinical features and the morphology and distribution of desmin aggregates in the skeletal muscle biopsies of patients with desminopathy, and to correlate these findings with genetic characteristics. **Methods:** This is a retrospective study that included patients with molecularly confirmed desminopathy from two national referral centers (Brazil and Spain). **Results:** Thirty patients from 20 unrelated families were included. The mean age of disease onset was 25 years old. Cardiopathy was present in 86.7% of them, and in 14 patients, the cardiac symptoms preceded musculoskeletal weakness. From 21 available biopsies, two distinct patterns of desmin aggregates were identified: subsarcolemmal well-demarcated aggregates (in eight patients) and aggregates with diffuse and poorly delimited borders (in 13 patients). Pathogenic variants in the 1B segment and the tail domain presented cardiopathy and the proximal distribution of weakness at the onset of symptoms more frequently when compared with patients with variants in other segments of the desmin. All patients with mutations in the 1B segment presented the well-demarcated desmin aggregate pattern, and no patients with variants in other segments showed this histological arrangement. **Conclusion:** We suggest a phenotype-genotype correlation that associates the presence of variants in the 1B segment and the tail protein domain with a more prominent cardiac presentation and a proximal-onset weakness distribution. Additionally, we found a morphotype-genotype correlation, with variants in the 1B segment causing well-demarcated subsarcolemmal desmin aggregate formation. These findings may be useful for identifying more severe patients, thus optimizing clinical care in this subgroup.

Doenças Neuromusculares

TL 1106189

CLINICAL AND MOLECULAR SPECTRUM OF MYOFIBRILLAR AND DISTAL MYOPATHIES

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Background: Distal myopathies (DMs) are a group of muscle diseases featuring a predominance of weakness in the distal extremities, while myofibrillar myopathies (MFMs) are skeletal and cardiac muscle diseases with Z-disk protein aggregates and myofibril disarray. Both conditions may present with predominant distal weakness and demonstrate a vacuolar histology, confounding clinical classification. **Objectives:** To characterize patients with DMs and MFMs, comparing both groups and classifying their genetic subtypes. **Methods:** This observational study identified patients at a tertiary neuromuscular center with MFM and DM diagnoses. Patients underwent a next-generation sequencing panel, followed by whole-exome sequencing. **Results:** We included 54 patients from 42 families: 34 patients (23 families) with MFMs, and 20 patients (19 families) with DMs. Among the MFM families, variants in DES were the main cause ($n=8$), followed by TTN ($n=4$), FHL1 ($n=3$), FLNC ($n=2$), BAG3 ($n=2$), MYOT ($n=1$) and HNRNPA2B1 ($n=1$). Among the DM families, DYSF ($n=6$) and GNE ($n=4$) were the most frequent disease-causative genes, followed by MYH7 ($n=2$), NEB ($n=2$), TTN ($n=1$), ANO5 ($n=1$), RYR1 ($n=1$), and DNM2 ($n=1$). The severity of motor impairment occurred in a similar proportion between distal myopathy and myofibrillar myopathy, being more pronounced in patients with variants in the BAG3, FHL1, MYH7 and GNE. Myofibrillar myopathies had a higher proportion and severity of cardiorespiratory impairment. DES myopathy was related to more frequent cardiopathy. BAG3 myopathy was an early and severe subgroup with axial contractures, cardiac and respiratory impairment. Child FHL1 patients presented with inflammatory histology, and adult female presented a scapuloperoneal phenotype. Miyoshi phenotype was seen in dysferlinopathy and anoctaminopathy. GNE myopathy, in turn, presented with involvement of the anterior leg muscles. Some genes related to congenital myopathies were also causes of DMs (TTN, RYR1, DNM2) and were confounded with hereditary neuropathy during the first years of disease, although we only noted concomitant peripheral nerve involvement in patients with BAG3 myopathy. **Conclusion:** DMs and MFMs comprise heterogeneous groups, but their phenotypic characteristics allow relative categorization into subgroups. Recognizing these myopathies is important due to their potential severity, early cardiac complications, and respiratory failure, which can lead to preventable death.

Doenças Neuromusculares

TL 1106195

USEFULNESS OF ULTRASOUND IN DETECTING MUSCLE INVOLVEMENT PATTERN IN POMPE DISEASE

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Background: Pompe disease is a treatable autosomal recessive disorder caused by deficiency of acid alfa-glucosidase. Two classical phenotypes are described: the infantile and the late-onset forms. Among late-onset Pompe disease, most patients present with a limb-girdle weakness pattern, although axial involvement is common. Muscle ultrasound may evaluate clinical and subclinical involvement of muscles, identifying some particular distribution pattern, and acting as adjunct to the physical exam. **Objective:** To describe the pattern of muscle involvement by ultrasound, including the tongue, paravertebral and diaphragm muscles in patients with Pompe disease. **Methods:** Eight patients with molecular diagnosis of Pompe disease were clinically assessed and underwent muscle ultrasound using the setting for muscle evaluation with a fixed gain (Sonosite Edge, Fujifilm) with a 7-15MHz transducer. The followings segments were assessed: tongue, upper and lower limb (mostly for proximal muscles), abdomen, diaphragm and paravertebral muscles. For the diaphragm we measured the thickness during expiration and forced inspiration. All ultrasound images were obtained and scored based on Heckmatt's rating scale. **Results:** Five from eight patients were male. The mean age was 42 years old, with 10 years of disease duration. Only one patient complaint of dysphagia and five reported dyspnea. The most involved and weakest muscles were abdominal (mean MRC 2), gluteus maximus (mean MRC 2) and gluteus medius (mean MRC 3), and adductors (mean MRC 2.5). On ultrasound the most affected muscles were abdomen, vastus intermedius, adductors and lumbar paravertebral muscles, scaled as Heckmatt 3 in virtually all patients. We also observed a pattern of brachialis more involved than biceps brachii, and vastus intermedius more affected than rectus femoris in all patients. Five patients had imaging involvement of the tongue without any complaint of dysphagia. Considering diaphragm, seven patients had high echointensity, and all patients with Heckmatt 3 had a thickness ratio (considering inspiration and expiration thickness) lower than 30%, which was considerably lower than in Heckmatt 2 patients (which had a thickness ratio between 30 and 60%). **Conclusion:** Muscle ultrasound was able to detect a recurrent pattern of muscle compromise in all patients. Subclinical involvement of the tongue and diaphragm may help early disease recognition and predict potential respiratory complication.

Doenças Neuromusculares

TL 1106231

COMPARING HORMONAL EFFECT IN SYMPTOMS IN MYASTHENIA GRAVIS AND CONGENITAL MYASTHENIC SYNDROMES

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Background: Fluctuations due to hormonal alterations are known features of myasthenic syndromes. In autoimmune myasthenia gravis a reasonable explanation of this phenomenon is hormonal influence in autoimmune response, as usually seen in other autoimmune disorders. Congenital myasthenic patients also experience these fluctuations, but their syndrome is not associated with autoimmune mechanisms. Therefore, the influence of symptoms might be due to other mechanisms of neuromuscular junction impairment. **Objective:** Evaluate whether there are differences in menstrual period, pregnancy, or menopause in patients with congenital myasthenic syndromes and autoimmune myasthenia gravis. **Methods:** data of patients were all obtained by phone call and outpatients visits, and then compared between two groups by Fisher's test. **Results:** A total of 34 participants with autoimmune myasthenia gravis (22 seropositivity for anti-acetylcholine antibody was present in 22 patients, and two for anti-Musk) and 27 participants with congenital myasthenic syndromes (15 CHRNE, 2 COL13A1, 2 GFPT1, 2 GMPPB, 1 RAPSN, 1 COLQ, 2 DOK7, 1 CHRNA1, 1 MUSK) were interviewed. Mean age was 42.8, MG-ADL median score was 4.7 and pregnancy rate 1.5, in the autoimmune group. In the congenital group the mean age was 34.6, MG-ADL median score 5.1 and pregnancy rate 1.5. There were no statistical differences of worsening rates during menstrual period and anticonception medications or improvement after menopause. Pregnancy in patients with congenital myasthenia resulted in improvement in 55% of patients compared to 70% of worsening symptoms in myasthenia gravis. Congenital myasthenic patients were significantly more likely to have symptoms improved during pregnancy compared with autoimmune patients ($p = 0.018$). **Conclusion:** Most results showed that no statistical differences could be observed in patients with congenital myasthenia or myasthenia gravis. An exception was the improvement in symptoms during pregnancy in congenital myasthenic women. This finding suggests that hormonal changes related to pregnancy may favor neuromuscular junction function but may also have an unfavorable effect on immunological attack to neuromuscular junction. These conflicting events would have a neutral, or even deleterious resulting effect on symptoms of autoimmune myasthenic patients. On the other hand, congenital patients, who do not present immunological disorder, would be subject only to the beneficial effects.

Doenças Neuromusculares

TL 1106270

EFFECT OF THE COVID-19 PANDEMIC ON PATIENTS WITH INHERITED NEUROMUSCULAR DISORDERS

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Background: The COVID-19 pandemic has brought substantial challenges to current practices in treating hereditary neuromuscular disorders (hNMDs). However, this infection has not been the only concern for these patients. Social distancing has compromised multidisciplinary assistance and physical activity and has brought about several mental health issues. We presented a follow-up on 363 patients with hNMDs at a Brazilian tertiary center during the peak of the COVID-19 pandemic. **Objective:** We aimed to show the frequency and severity of SARS-CoV-2 infection among hNMD patients and to demonstrate the effects of the pandemic on life habits, disease progression, and multidisciplinary supportive care status. **Methods:** Three hundred and sixty-three patients (58% male and 42% female) were followed for three months through three teleconsultations during the peak of the COVID-19 pandemic in Brazil. **Results:** There were decreases in the numbers of patients who underwent physical, respiratory, and speech therapies. For several patients, their appetite (33%) and sleep habits (25%) changed. Physical exercises and therapies were interrupted for most of the patients. They reported new-onset/worsening of fatigue (17%), pain (17%), contractions (14%) and scoliosis (7%). Irritability and sleep, weight and appetite changes, and especially diminished appetite and weight loss, were more frequent in the group that reported disease worsening. There was a low COVID-19 contamination rate (0.8%), and all infected patients had a mild presentation. **Conclusion:** The isolation by itself was protective from a COVID-19 infection perspective. However, this isolation might also trigger a complex scenario with life habit changes that are associated with an unfavorable course for the NMD.

Doenças Neuromusculares

TL 1106271

SEROPATHOLOGICAL CORRELATION IN PATIENTS WITH INFLAMMATORY MYOPATHY: A COHORT STUDY

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Background: Muscle biopsy have been considered the gold standard pattern for the diagnosis of idiopathic inflammatory myopathies (IIM). Myositis-specific antibodies (MSA) and myositis-associated antibodies (MAA) have been shown to play an important role in diagnosis and prognosis. **Methods:** Describe a cohort of IIM, correlating histopathological and immunohistochemical with serological findings (MSA and MAA). **Results:** Out of 44 patients, 40 tested for MSA or MAA and 38 were underwent muscle biopsy, as the following: 15 (39%) immune-mediated necrotizing myopathy (IMNM), 8 (21%) dermatomyositis (DM), 4 (10.5%) perimysial myopathy (PMM), 3 (7.9%) immune-mediated myopathy (IMM), 2 (5.2%) lipid-storage myopathy (LSM), 5 (13%) other myopathies and 1 (2.6%) inclusion body myositis. A total of 25 cases (62.5%) were seropositive for the following antibodies: 6 (24%) anti-SRP, 5 (20%) anti-HMGCR, 3 (12%) anti-Ro52, 2 (8%) anti-Jo1, 2 (8%) anti-Mi2, 2 (8%) anti-Ku, 1 (4%) anti-Jo1 + anti-Ro52, 1 (4%) anti-SRP + anti-Ro52, 1 (4%) anti-SAE1 + anti-Ro52 and 1 (4%) anti-SAE1 + anti-Ro52 + anti-Ku. Among the IMNM, 5 (33.3%) were antiSRP positive (+), 3 (20%) anti-HMGCR+, 1 (6.6%) anti-Ku+, 1 (6.6%) anti-Ro52+ and 1 (6.6%) anti-SRP+ and anti-Ro52+, (sensitivity: 60%). The DM subtypes 1 (12.5%) tested positive for anti-Mi2, 1 (12.5%) anti-SRP, 1 (12.5%) anti-NXP2, 1 (12.5%) anti-SAE1 and anti-Ro52 and 1 (12.5%) anti-SAE1 and anti-Ku + anti-Ro52. Regarding the 4 cases of PMM, 2 (50%) were positive for anti-Jo1 and 1 (25%) for anti-Mi2. Among IMM cases, 1 (33.3%) was positive for anti-Ku and 1 (33.3%) for anti-Jo1 and anti-Ro52. In LSM, 1 (50%) tested positive for anti-HMGCR. **Conclusion:** The positive test for MAA and MSA occurred in 62.5% of our sample, demonstrating sensitivity above that reported in the literature (50%). Anti-HMGCR and anti-SRP antibodies were highly specific and sensitive for IMNM, while anti-Jo1 showed a correlation with PMM. On the other hand, anti-Ro52 proved to be the less specific. Lipid accumulation in anti-HMGCR myopathy was probably a secondary finding. The positive case for anti-Mi2 in 1 case of PMM points out to the possibility of DM diagnosis in this subgroup. The presence of MAA in the nonspecific IMM group highlights the value of testing these autoantibodies in cases only suggestive of an immune process. Overall, the future will decide how important these antibodies are to replace or not the muscle biopsy in IIM.

Doenças Neuromusculares

TL 1106289

ROLE OF MUSCLE BIOPSY IN GENETIC ERA

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Background: Innovations in genetic testing have revolutionized the diagnosis of hereditary myopathies. However, the detection of variants of uncertain significance makes the interpretation of some cases difficult. In addition, hereditary myopathies can mimic acquired diseases that have specific treatment, such as inflammatory myopathies. Thus, muscle biopsy may have a defining role in the diagnosis of myopathies. Here we present three cases in which the muscle biopsy allowed a diagnostic conclusion by complementing or guiding the genetic evaluation. Case 1: female infant with neonatal hypotonia and delayed motor development. NGS panel revealed heterozygous VUS in the RYR1 gene. The biopsy showed signs of neurogenic disease. New genetic testing revealed homozygous pathogenic variant in the SMN1 gene, concluding the diagnosis of spinal muscular atrophy. Case 2: 4 year old boy with neonatal hypotonia, muscle weakness, dysphagia and respiratory distress. Exome sequencing found no pathogenic variants that would justify the clinical picture. Muscle biopsy showed myotubular myopathy. A microarray detected a deletion in MTM1 gene, which causes X-linked myotubular myopathy. Case 3: 12 year old male, with frequent falls, gait alteration and progressive muscle weakness. Genetic investigation of symptomatic relatives did not clarify the diagnosis. In muscle biopsy, several reducing bodies were identified. Genetic testing detected a deletion in the FHL1 gene, which causes, in addition to reducing body myopathy, Emery-Dreifuss muscular dystrophy type 6, X-linked myopathy with postural muscle atrophy and X-linked scapulohumeral myopathy. **Discussion:** The case 1 shows limitations of the exome as a tool for genetic diagnosis, as the reading depth can be variable and sometimes insufficient. In case 2, the exome did not detect the pathogenic variant because it is a copy number variation; the biopsy directed the microarray to the region of the X chromosome where the suspected gene is located, confirming the diagnosis. In case 3, the final diagnosis was guided by the histopathological finding, which suggested the causative gene; this gene, however, can cause different myopathies, and it is only possible to define the type through muscle biopsy. **Conclusion:** In genetic era, muscle biopsy may play a role in diagnosing hereditary myopathies when genetic testing is inconclusive.

Doenças Neuromusculares

TL 1106300

A UNIVERSITY NEUROMUSCULAR CENTER EXPERIENCE IN CHRONIC AUTOIMMUNE DEMYELINATING POLYNEUROPATHY DIAGNOSTIC WORKUP

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Background: Chronic autoimmune neuropathies are a heterogeneous spectrum of disorders with distinct pathophysiology, phenotypes and treatment. This group accounts for both economic and quality of life burden. Misdiagnosis remains common, especially for chronic autoimmune demyelinating polyneuropathy (CIDP) variants, which is why diagnostic criteria have recently been reviewed. Widespread interest in immune neuropathies relies on the fact that they are treatable conditions. **Objective:** An analysis of our university Hospital cohort of patients presenting with CIDP variants, aiming to contribute to diagnostic workup debate. **Methods:** A cohort of 46 suspected CIDP patients was identified in our neuromuscular outpatient clinic, after thorough chart review from 2018 to June/2022. Patients underwent analysis based on EFNS/PNS diagnostic criteria. **Results:** Out of 46 suspected acquired demyelinating neuropathy patients, 9 fulfilled criteria for multifocal motor neuropathy and 2 were diagnosed with metabolic neuropathy, and were all excluded from further analysis. Out of 35 remaining patients, 14 are female and 21 male. 12 patients presented with relapsing forms, 3 progressive, 11 secondary progressive, 8 clinical remission and 1 still unidentified. According to 2021 EFNS/PNS electrodiagnostic criteria, 20 patients were classified as definite CIDP and 7 as possible CIDP. 8 did not fulfill criteria. Twenty three patients presented with typical CIDP, 2 with multifocal CIDP, 1 with focal CIDP and 1 with sensory CIDP. Median time for diagnosis after initial symptoms was 53 months. **Conclusion:** Accurate diagnosis and full understanding of different clinical presentations guide therapeutic management and prognosis. They range from multifocal to diffuse, from sensory to motor, from benign to severe and from relapsing to progressive forms. Out of our cohort of clinically suspected and treatment responsive patients, 77.14% fulfilled criteria for possible diagnosis of CIDP, which reinforces importance of supportive criteria when diagnosis is uncertain. Sensibility may be reduced for focal and sensory forms, besides severe cases with axonal loss. Most frequent clinical course was relapsing. 85.2% of patients showed typical presentations. CIDP diagnosis remains a challenge, which is evidenced by the prolonged time lapse between initial symptoms and definite diagnosis.

Doenças Neuromusculares

TL 1106352

STRUCTURAL DAMAGE IN DYSTROPHINOPATHIES: A MULTIMODAL NEUROIMAGING STUDY

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Background: Dystrophinopathy is a X-linked recessive progressive neuromuscular disorder characterized by muscle weakness. Dystrophinopathies are also characterized by central nervous system damage, but pattern and extent of such damage are not yet clear. For this reason, we designed a cross-sectional multimodal MRI-based study to address this point. **Methods:** Mean age of patients and disease duration were 12 and 8, respectively. We enrolled 50 patients (40 duchenne and 10 becker) and 40 age-sex matched controls. All subjects underwent MRI in a 3T device to assess gray (GM) and white matter (WM). To evaluate the cerebral and cerebellar cortices, we used measures from FreeSurfer. DTI-multitlas was used to investigate microstructural abnormalities in cerebral WM. All analyses were corrected for multiple comparisons. **Results:** Group analyses showed WM microstructural abnormalities at bilateral fornix, being characterized by increased axial, medial and radial diffusivities, with large effect sizes (>0.8) for all significant diffusion parameters. We also found a negative correlation between axial diffusivity and Motor Functional Measure scale ($r=-0.338$, $p=0.022$) of such structure. Regarding volumetric analysis, patients showed volumetric reduction of both left thalamus and cerebellar cortex. **Conclusion:** Patients with dystrophinopathies are characterized by structural damage at fornix, thalamus and cerebellar cortex. Such structural signature helps us to understand some phenotypic traits in this disease. Our results also shed some light in the dystrophinopathy pathogenesis and highlight potential neuroimaging marker for therapeutic trials.

Doenças Neuromusculares

TL 1106380

GENETIC PANEL TESTING'S DIAGNOSTIC ACCURACY FOR HEREDITARY NEUROMUSCULAR DISEASES IN HOSPITAL UNIVERSITÁRIO ANTÔNIO PEDRO

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Background: Neuromuscular disorders (ND) represent an important fraction of neurological diseases. They affect the peripheral nervous system, which comprehends a vast spectrum of syndromes and etiologies. In this regard, hereditary NDs are characterized by an early age manifestation, family history and progressive loss. Furthermore, the diagnosis is challenging and of exclusion, naturally requiring a detailed investigation and complementary exams, such as and beyond genetic sequencing and electromyography. Owing to the diagnostic's difficulty, the genetics tests are value resources when it comes to identifying the disease, allowing a better understanding and orientation of the patient's symptoms and outcomes. This ongoing study aims to specify the most correlated factors with the test's positivity. In addition, a decisive algorithm for the genetic approach will be established, along with the patient's sample characterization. To that end, this is an observational retrospective and descriptive current study related to genetic test's accuracy. Patients included will have hereditary ND's suspicion, age over 18 years and other etiologies excluded. In total, 54 patients matched the previous criteria and preliminary **Results:** show that 51% are women and 29,6% are between 53-62 years. The hereditary ND was identified in 33% of the patients, yet a Charcot-Marie-Tooth phenotype was most prevalent with mutation of the PMP22, DNM2 and SH3TC2 genes. **Conclusion:** Therefore, genetic testing is an important instrument for hereditary ND's diagnosis and disease specification. In consequence, the patient experiences an improvement in the orientation and outcome's anticipation, besides an elementary comfort in knowing one's disorder. However, genetic panel testing doesn't ensure precise diagnosis and, as a result, a wide range of patients continues without an answer. For this reason, studying those tests accuracy is mandatory, highlighting hotspots in need of improvement.

Doenças Neuromusculares

TL 1106408

ELECTROPHYSIOLOGICAL FINDINGS IN A SAMPLE OF GENETICALLY CONFIRMED BRAZILIAN CITIZENS WITH CANVAS

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Cerebellar ataxia, neuropathy and vestibular areflexia syndrome (CANVAS) is a late-onset, slowly progressive neurological disorder. Despite recently described, its etiopathogenesis is already in the process of elucidation due to the identification of a biallelic intronic AAGGG repeat expansion in the RFC1 gene as the genetic cause of the disease. Meanwhile, electrophysiological findings are still to be completely described as patients receive their definite diagnosis. Based on these findings, this study evaluated nerve conduction study and electromyography findings of genetically confirmed CANVAS patients from a single Brazilian center. In order to genetically identify this expansion, flanking polymerase chain reaction (PCR) and repeat-primed PCRs (RP-PCR) for the pathogenic allele expansion configuration in RFC1 were performed in patients with at least one clinical feature of the Syndrome. If flanking PCR did not show any product, the DNA sample would undergo RP-PCR. Biallelic AAGGG expansion was found in nineteen patients of the sample group (18%). Subsequently, confirmed CANVAS patients were submitted to electrophysiological studies (nerve conduction study and electromyography). Fifteen out of the nineteen confirmed cases were submitted to electrophysiological evaluation. Only one patient had a normal electrophysiological study and clinically presented with cerebellar ataxia related symptoms alone. Six patients (40%) had motor involvement associated with neuropathy. Notably, eight patients (53%) had exclusively sensory involvement and primarily axonal deterioration for most cases. Additionally, 6 patients were submitted to the blink test during the exam and 3 (50%) had the R2 reflex impaired. With regards to autonomic evaluation, four patients were evaluated for cutaneous response for electric, respiratory and sound stimuli. One of these patients showed diminished response to the electric stimulus. Ultimately this study aims to contribute to the understanding of the electrophysiological findings of the CANVAS patients in the Brazilian population.

Doenças Neuromusculares

TL 1106411

CLINICAL MANIFESTATION OF NEBULIN-ASSOCIATED NEMALINE MYOPATHY

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Background: Nemaline myopathy (NM) is a genetically heterogeneous inherited myopathy related with at least twelve genes while pathogenic variants in NEB gene are the most common genetic cause. The clinical spectrum of NM caused by NEB pathogenic variants (NM-NEB) is very broad ranging from mild to severe presentations manifesting with generalized weakness, respiratory and bulbar involvement. Currently there is not enough data regarding the progression disease. **Objective:** Here we present a mutational and phenotypic spectrum of 33 NM patients caused by NEB variants (NM-NEB) classified according to age groups and the use of ventilatory support. **Methods:** Clinical and genetic data from NM-NEB patients followed in one specialized center were collected through regular consultations. Thirty-three patients were evaluated regarding motor, bulbar and respiratory function. **Results:** 32% of the NM-NEB patients used a G-tube, 65% were able to walk unsupported and 45% did not used ventilatory support (VS). Scoliosis and dysphagia were more common among patients that used VS. Comparing the patients grouped by age we found that proportionally, older patients had more scoliosis and respiratory dysfunction than younger groups suggesting the progression of the disease in these domains. In addition to that, we showed that ventilatory support use was associated with scoliosis and dysphagia. **Discussion:** NM-NEB is a very debilitating disease. There is an association between scoliosis and respiratory dysfunction while patients using VS have more often scoliosis than the no-VS group. Grouping patients by age suggested disease stability in motor and swallow function but a progression in respiratory dysfunction and skeletal deformities. All these observations are relevant in the management care of NM-patients.

Doenças Neuromusculares

TL 1106416

CORRELATION BETWEEN AGE, RESPIRATORY AND MOTOR FUNCTION IN CHILDREN WITH LAMA2 CONGENITAL MUSCULAR DYSTROPHY

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Background: LAMA2 Natural history studies have been increasing in the last years due to the new era of emerging gene therapy. In this type of congenital muscular dystrophy, the age is associated with poor respiratory and motor function. A better understanding of how age relates to the course of respiratory and motor aspects can contribute to multidisciplinary management and treatment for this children. This study aimed to investigate the relationship between age, respiratory and motor function in LAMA2 children. **Methods:** Eighteen children (11.4±5.8 years; 11 male/7 female; 7 with nocturnal noninvasive ventilation) had the forced vital capacity percentage (FVC) and cough peak flow (CPF) measured by spirometer and peak flow meter, respectively, and the motor function evaluated by three domains and total score of Motor Function Measure (MFM) – 32. The Shapiro-Wilk test demonstrated that the data were non-normally distributed. The relationships between respiratory and motor functions and also between age and respiratory and motor functions were tested by Spearman correlation test. The correlation coefficients were classified as strong ($r \geq 0.70$), moderate ($0.40 \leq r < 0.70$), or weak ($r < 0.40$). **Results:** FVC strongly correlated to MFM-D2 (axial and proximal function) and moderately correlated to MFM-D1 (standing and transfers function) and to MFM-total. No correlation was found between CPF and motor function items, but CPF was moderately correlated to age. Age strongly correlated to MFM-D2 and moderately correlated to MFM-D1 and to MFM-total. **Conclusion:** A good pulmonary function (FVC) was positively correlated to high motor function to perform axial/proximal, standing and transfers activities. Interestingly, CPF was not correlated to motor function, but was negatively correlated to age. Additionally, age was negatively correlated to axial/proximal and total motor function in LAMA2 children

Doenças Neuromusculares

TL 1106424

TRIPLE-FURROW TONGUE ATROPHY IN NEMALINE MYOPATHY: A NOVEL FINDING THAT CHANGES MANAGEMENT CARE

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Background: Inherited Nemaline myopathy (NM) is a clinically and genetically heterogeneous myopathy. Orofacial and bulbar muscles are commonly involved in NM patients. **Objective:** Previous data showed that one third of patients needed feeding tube and that dysphagia might lead to recurrent pulmonary. However there is a lack of data characterizing such a common comorbidity. **Methods:** We evaluated patients with NM molecularly and/or histologically diagnosed and stratified them according to swallow function using the Neuromuscular Disease Swallowing Status Scale (NdSSS). **Results:** 48 patients were seen, 33 were caused by NEB, 10 by ACTA1, 2 by TPM2 variants and 3 had no defined genetic cause. All patients presented high arched palate and most of them had facial weakness, less pronounced in the two patients with childhood onset. Neonatal dysphagia and failure to thrive was referred by 68% of the patients, half of them improved in the next few years of life. At the evaluation, around 30% used feeding device, but 20% additional, despite eating by mouth, needed to adapt their food and take supplements. Comparing ACTA1 with NEB patients, dysphagia was more common among the ACTA1 group. We described a novel finding which is tongue atrophy in a triple furrow pattern. The tongue atrophy was present 50% of the accessed patients, but it was more frequent in the dysphagic patients (83%). **Discussion:** Dysphagia is a very common disability caused by NM myopathy and is present in around 50% of the patients, being more frequent in patients with ACTA1 variants. Tongue atrophy is a novel finding which is associated with dysphagia and can help us to stratify patients needing a more detailed bulbar assessment.

Doenças Neuromusculares

TL 1106436

MOLECULAR INVESTIGATION USING NGS FOR INHERITED MYOPATHIES : EXPERIENCE OF A BRAZILIAN TERTIARY CENTER

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Background: Inherited myopathies represent a large and variable group of disorders. The age of onset ranges from neonatal presentations to late-adult forms. Most of the conditions can be caused by multiple genes, making the unique gene testing unfeasible. Advances in next generation sequencing techniques and the participation of industry in providing no-cost panels has made this genetic evaluation a new reality in Brazil. **Objectives:** Our aim is to present the experience of the use of these technologies in a tertiary center and present the gene distribution among the main groups of inherited myopathies: congenital muscular dystrophy (CMD), congenital myopathy (CM), congenital myasthenia (CMS), mitochondrial myopathies (MiM), metabolic myopathies (MeM), myofibrillar and distal myopathies (M_DM), limb-girdle muscular dystrophy (LGMD). For analysis purposes we included dystrophinopathy patients with the LGMD. **Results:** We evaluated 427 patients and performed panel for 249 patients and whole exome sequencing for 178 patients presenting diagnostic rates of 69% for the exomes and 66% for the panels. We identified a genetic cause for the condition in 68% of patients with a hipotesis of canalopathy, 90% of the CMD, 76% of the MC, 72% of the CMS, 16% of the MiM, 58% of the MeM, 46% of the M_DM and 71% of the LGMD. **Discussion:** We showed the performance of NGS technologies among many highly curated patients for a particular phenotype. We had higher diagnostic rates for several groups of disorders such as CM, CMD, CMS and LGMD than previously reported in the literature. We linked this finding to a continuous vigilance among new genes in the literature and to the importance of the clinical phenotype stratification for the positive results. In the opposite direction, we had lower diagnostic rates for MiM than previously reported, but mtDNA variants were not yet checked. **Conclusion:** NGS has facilitated the diagnosis in the inherited myopathies but requires a careful assessment of its limitations. A detailed clinical assessment is very important to provide high diagnostic rates.

Doenças Neuromusculares

TL 1106445

MITOCHONDRIAL MYOPATHY CAUSED BY TK2 DEFICIENCY

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Background: The nuclear gene TK2 encodes the mitochondrial thymidine kinase, an enzyme involved in the phosphorylation of deoxycytidine and deoxythymidine nucleosides. Biallelic TK2 mutations lead to mitochondrial DNA instability and are associated with a wide clinical spectrum of disease (TK2d). Current classification includes infantile (? 1 year), childhood- (1-12 years), and late-onset (?12 years) forms. These forms differ for progression and life expectancy. Deoxynucleosides reposition were shown to be effective to disease controls in a compassionate use. **Objectives:** Here we report the clinical findings and mutational spectrum of a series of Brazilian cases presenting infantile and childhood-onset TK2d. This report characterized patients with TK2-deficiency enhancing the importance of a clinical recognition of patients with a severe, potentially lethal, and potentially treatable condition. **Methods:** Patients with TK2d were evaluated in a Tertiary Center. We presented clinical and molecular findings from 8 patients (6 families) harboring pathogenic variants in TK2 gene. **Results:** Four patients presented the early-onset infantile form. Of those, three passed away still in their first decade of life. P5 was 2 years old in the last evaluation, he had normal motor development until the age 11 months, when a weakness started. After 5 months of the beginning of symptoms he had lost his gait, head support and started on night-time non-invasive ventilatory support at age of 18. At 23 months he was hypotonic, had generalized weakness and was requiring ventilation also for a couple of hours during the day. Five patients presented the childhood form (mean of age average of 32 +/- 8 years old). Despite the slow disease progression, all of them noticed first symptoms in their first decade of life, they had difficulties gaining weight and frequent episodes of diarrhea. They had severe proximal and axial weakness and started on NIVS around 20 years old. Four of them used NIVS night-time only and one used for 15 hours/day. **Conclusion:** With the advent of a specific treatment and given the severity of TK2d, clinicians need to know how to recognize this disorder. The mortality rates are high mainly among the infantile presentation; however, childhood-onset are also a debilitating condition. Ptosis and hypoacusis were not found in our series. But all patients had proximal and axial weakness, global hypotrophy, dysphagia and respiratory involvement.

Doenças Neuromusculares

TL 1106458

PHYSICAL EXERCISE IN PATIENTS WITH DEMYELINATING DISEASES

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Background: People with demyelinating diseases, such as Multiple Sclerosis (MS) and Neuromyelitis Optica (NMO), despite the evidence of benefits, have reduced the practice of physical exercises, given the fear of worsening the symptoms. This increases physical disability and worsens stability, in addition to reducing muscle strength, mobility and quality of life. In this context, some studies have demonstrated beneficial results of aerobic training on brain gray matter volume and on the white matter tract unit, in addition to greater functional connectivity of the hippocampus and cortex. **Objectives:** To analyze the practice of physical exercise by patients with demyelinating diseases in the State of Paraíba. **Methods:** A retrospective, cross-sectional study carried out by reviewing the medical records of 115 patients, selected in a non-probabilistic manner and for convenience, with demyelinating diseases in a Multiple Sclerosis center, using the variables sex, age, Body Mass Index (BMI), diagnosis, frequency and category of physical exercise. **Results:** Among the study participants, a distribution of 101 (87%) patients with MS and 14 (13%) with NMO was observed. There was a predominance of females (70%). In terms of age group, 3% belonged to the age group under 16 years old, 9% from 16 to 25 years old, 34% from 26 to 35 years old, 31% from 36 to 45 years old; 10% from 46 to 55 years old; 10% from 56 to 65 years old and 3% from those over 65 years old. Regarding BMI, 3% had a thin BMI, 49% were eutrophic, 32% were overweight and 16% were obese. Before the diagnosis of MS or NMO, 43% rarely practiced physical exercise and 27% at least 5 days a week. After diagnosis, it was observed that 24% of the patients increased the frequency of physical exercises, 29% decreased it and 47% maintained the frequency. Before the diagnosis, 12% practiced Tension and Aerobic Exercises (TAE); 42% Aerobic Exercise (AE); 18% Tension Exercises (TE); 28% did not practice activities. After diagnosis, 20% practice TAE; 41% AE; 18% TE; 21% do not practice activities. **Conclusion:** A predominance of patients between 26 and 45 years (65%) and the presence of 48% of participants with an overweight or obese BMI is observed. Regarding the practice of physical exercise, it is noted that 76% decreased or maintained the frequency, in addition, an increase of 8% was observed in TAE practitioners and a decrease of 7% in individuals who do not practice physical exercise.

Doenças Neuromusculares

TL 1106469

LATER DIAGNOSIS OF DUCHENNE MUSCULAR DYSTROPHY IN BRAZILIAN PATIENTS: WHERE COULD WE IMPROVE?

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Background: Duchenne muscular dystrophy (DMD) is caused by a mutation in the dystrophin gene and is the most common form of childhood-onset muscular dystrophy affecting approximately 1 in 3500 newborn boys. The disease is progressive and most patients exhibit signs of muscle weakness before 6 years. Despite all the advances in management and treatment of DMD over the last decades, the mean age at diagnosis of DMD has been reported to be around the age of 4.5-5 years in several countries with a delay of about 2 years between the first symptoms are noted, and the diagnosis. **Objective:** This retrospective study had objective to investigate the age at diagnosis of disease in a group of Brazilian patients followed in a tertiary center. **Results:** We identified 122 Brazilian boys with phenotype/genotype compatible with DMD in the last 7 years (2014-2021). The most common mutational event was represented by intragenic deletions, accounting for 58% (71/122) of all mutations. Duplications accounted for 14% and 20% of patients had a point mutation (including 12/122 with nonsense mutation). In 7 boys (5%) was found an intronic mutation and in 2 the muscle biopsy confirmed absence or near absence of dystrophin in the muscle. The mean age at onset of the disease was 3.3 years. Frequent falls were the more frequent symptoms (55/122) at initial clinical presentation. The mean age at diagnosis was 6.9 years. The age of diagnosis was defined as the age of molecular test was realized, confirming the disease. Steroid therapy was initiated in 120/122 patients (prednisolone 36/120 or deflazacort 84/120). The mean age at start treatment with steroid was 7.3 years. Thirty-seven (30%) lost the capacity to walk until the last evaluation. The mean age of lost was 10 years. In all patients, CK levels were markedly increased in the early stages of the disease. **Conclusion:** In this group of Brazilian patients with DMD, an important delay in diagnosis was observed, which led to a delay in the beginning of steroid therapy, when compared to the onset of initial symptom. This late onset of therapy is probably related to an earlier age of loss of capacity to walk observed in this group. Despite the availability of access to molecular testing, we still observed difficult in recognizing the disease, which may be improved with wider dosage of serum CK in patients with motor/global development delay and weakness.

Doenças Neuromusculares

TL 1106542

LONG-TERM FOLLOW-UP OF SMA TYPE 1 TREATMENT WITH NUSINERSEN: A SINGLE-CENTER EXPERIENCE

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Background: Spinal Muscular Atrophy (SMA) is a genetic motor neuron disease caused by mutations in the SMN1 (Survival Motor Neuron) gene, which leads to hypotonia, muscle weakness and respiratory involvement. Its most severe form, SMA type 1, starts before 6 months of life and has a high mortality due to ventilatory failure. Nusinersen, the first approved treatment for SMA, is an antisense oligonucleotide for intrathecal use, which leads to greater survival and gain in motor acquisitions. Studies on the safety and efficacy of long-term treatment are still scarce. **Objective:** To present long-term results (4 years of follow-up) in SMA type 1 patients under treatment with Nusinersen. **Methods:** We followed a total of 24 patients, all with SMA type 1 (20 patients with 2 copies of SMN2). The patients were evaluated by the functional scale CHOP-INTEND (The Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders) and in relation to gain of motor milestones (head control, sitting with or without support, standing and walking), in addition to assessment of survival and use of permanent ventilation (PV) and feeding route. **Results:** Twelve patients were female, only 11 patients (45.8%) started treatment before 12 months of illness. 22 patients (91.6%) were already using gastrostomy at the beginning of treatment. After 4 years of follow-up, 22 (91.6%) patients were alive, two deaths occurred: one after gene therapy and the other after respiratory failure. Two patients received gene therapy but continued to use Nusinersen (combined therapy). Eight patients gained some motor milestone, all of them started treatment before 12 months disease. The greatest gains in CHOP-INTEND occurred up to 24 months of treatment, and after this period, the scores tended to stabilize, without further gains. 19 patients (79.1%) were already using PV (>16h/day) at the beginning of treatment and 15 patients were using PV after 4 years of treatment. Even in those patients who were on PV, there was a reduction in the duration of ventilation use and an improvement in the management of airway secretion. **Conclusion:** Nusinersen showed continuous benefit over 4 years of treatment, bringing motor improvement mainly within the first 2 years of treatment and maintaining motor function acquired at 4 years. Only patients who started treatment before 12 months of illness gained some motor milestone. No new adverse events were reported in this long-term follow-up.

Doenças Neuromusculares

TL 1106551

SAFETY AND EFFICACY OF GENE THERAPY FOR PATIENTS WITH SPINAL MUSCULAR ATROPHY: A REAL-LIFE STUDY IN A BRAZILIAN COHORT

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Background: Spinal Muscular Atrophy (SMA) is a genetic motor neuron disease caused by mutations in the SMN1 (Survival Motor Neuron) gene, which leads to hypotonia and muscle weakness with high mortality related to respiratory involvement. Gene therapy (GT) (onasemnogene ABERPVOVEC) for SMA, through an adeno-associated viral vector 9 (AAV9) was recently approved in our country, but its safety and efficacy outside the context of clinical trials is still poorly understood. **Objective:** To present early results regarding safety and efficacy in SMA patients treated with GT. **Methods:** We followed a total of 33 patients treated with GT for SMA from 6 months to 1 year of treatment. The patients were evaluated by the functional scales CHOP-INTEND (The Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders) and in relation to gain of motor milestones. In addition, assessment of survival and use of continuous ventilation (CV) was performed and also data regarding transaminase elevation, liver function, hematological data, elevation of troponin and duration of corticosteroid use. **Results:** 33 patients were included, 26 SMA type 1 and 7 SMA type 2. The mean age at dosing was 18.5 months (14.0 - 23.2), with a mean weight of 9.9 kg (8.3 kg) - 16.3) and 28 patients (87.5%) were using nusinersen previously. After 1 year of treatment 32 patients (96.9%) were alive, 7 patients (21.2%) remained on CV (>16h/day) versus 11 (33.3%) patients at dosing. Regarding the gain in the CHOP-INTEND score, the mean baseline score was 30.50 (19.50, 40.75) to 46 (40.00, 52.00) at 6 months and to 56 (50.00, 58.00) points at 12 months. Regarding motor milestones, from those with SMA type 1, nine patients (42.9%) sat and four patients (19%) stood with support, and three patients acquired gait with support among SMA type 2. In terms of safety, the highest transaminase peak occurred in weeks 3 and 6 after infusion. Only 10 patients (30.3%) had transaminase levels similar to baseline at week 8. 15 patients (45.4%) had thrombocytopenia in the first week and one patient met criteria for thrombotic microangiopathy. The mean time of prednisolone use was 105 days (60.0 - 122.2). **Conclusion:** GT is effective in real life but with the potential for serious adverse events. There is a need for strict monitoring of transaminases, platelets and troponin and the occurrence of liver damage beyond 2 months of drug use, especially in patients with an older profile than in clinical studies.

Doenças Neuromusculares

TL 1104633

NEUROPLASTICITY INVOLVED IN THE REVERSAL OF NEUROPATHIC PAIN INDUCED BY MOTOR CORTX STIMULATION

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Background: The motor cortex stimulation (MCS) have been widely used in clinical medicine as a tool for pain control, used in patients with different pain syndromes, mainly of a chronic nature, which do not respond satisfactorily to any type of conventional analgesia. Preliminary studies, obtained by our group, have demonstrated that MCS reverts the neuropathic pain in rats, inhibiting neurons of the ventral posterior lateral and medial thalamus nuclei and dorsal horn spinal cord, and activating neurons of the anterior cingulate cortex and central and basolateral nuclei of the amygdala. Despite this evidence, the mechanisms involved in that nociceptive modulation are not yet understood. **Objective:** The aim of this research project was to evaluate the neuroplasticity induced by MCS in a persistent peripheral neuropathic pain model in rats, investigating the participation of glial cells and of cannabinoids involved with the activation and inhibition of the nociceptive response, respectively. **Methods:** involved induction of neuropathic pain by chronic constriction injury of the sciatic nerve, implantation of transdural electrodes over the primary motor cortex and evaluation of the nociceptive response in tests of mechanical hyperalgesia and allodynia. Immunoblotting and immunohistochemical assays were performed to detected glial fibrillary acidic protein (GFAP, an astrocyte marker), OX-42 (a microglial marker) and cannabinoid receptor type 1 (CB1) in spinal cord obtained from sham rats or animals with neuropathic pain, stimulated or not. In animals with MCS reversed neuropathic pain, a decrease of microglial activation was seen, as well as increased CB1 immunolabel in the spinal cord, compared to unstimulated animals with neuropathic pain. The data obtained suggest that MCS inhibits spinal microglia, which contribute to the induction of nociceptive processes. Also, the cortical stimulation induces the activation of CB1, which is important for the modulation of pain sensitivity. **Conclusion:** These results will contribute to a better understanding of the role of the motor cortex in the modulation of neuropathic pain and to an improvement of therapeutic strategies to treat persistent pain.

Dor

TL 1105627

BOTULINUM NEUROTOXIN TYPE A FOR TINNITUS MANAGEMENT IN PATIENTS WITH TEMPOROMANDIBULAR DISORDERS: A DOUBLE-BLIND RANDOMIZED CONTROLLED TRIAL

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Background: Tinnitus is a common otological symptom eventually leading to the development of serious physical and emotional disorders. Although uncertainties remains about pathophysiological mechanisms, the high prevalence of such complaint and the relationship to temporomandibular disorders make this and important topic on clinical medicine. Treatment is challenging and currently there are no approved drugs. **Objective:** To evaluate efficacy and safety of BoNT-A injection in the masticatory muscles to treat tinnitus intensity compared to 2% lidocaine and 0.9% saline solution in patients with TMD and tinnitus. **Methods:** A double-blind, placebo-controlled, randomized controlled trial (RCT) involved 96 patients who were diagnosed with TMD and tinnitus. The Fonseca Anamnestic Index (FAI) was assessed to characterize the severity of TMD symptoms in patients. The protocol included evaluation using the Visual Analog Scale (VAS), Tinnitus Handicap Inventory (THI), and Survey of Pain Attitudes—brief version (SOPA-brief). Patients in all the 3 groups received injections in bilateral masseters and temporalis muscles, according to the American Pain Society criteria. Insulin needles and a 1-mL syringe (Ultra Fine Syringe 1 mL 6 × 0.25; UNIQMED) were used for injection. Group A received 100 IU of onabotulinumtoxin A (Botox) diluted in 2 mL of 0.9% saline solution; group B received 2 mL of 2% lidocaine (without vasoconstrictor); and group C received 2 mL of 0.9% saline solution. The dose was equally divided and administered in the right and left sides. **Results:** Myofascial pain was found in 92 patients (92.7% of TMD patients). Myofascial pain with limited opening was found in 71 patients (71.7%), disc displacement with reduction in 51 patients (51.5%), and osteoarthritis or osteoarthrosis in only 17 patients (17.2%). Evaluation using THI after 30 days of the treatment showed a significant improvement in tinnitus symptoms in the treated group [F (2.94) = 6.498; p < 0.001]. The post hoc least significant difference test showed that the average THI score measured on the 30th day of the treatment in BONT-A group differed from that of the lidocaine group (27.9 ± 4.6 vs 22.5 ± 4.3; p < 0.001) and the saline solution group (27.9 ± 4.6 vs 23.4 ± 3.8; p < 0.001). **Conclusion:** This study demonstrated that BoNT-A is effective in treating tinnitus in TMD patients. However, further studies are necessary to understand better the longterm effects of BoNT-A on tinnitus.

Dor

TL 1105784

FIBROMYALGIA IMPACT WORSENERD DURING THE COVID-19 PANDEMIC

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Background: Pain is an unpleasant experience modulated by sensorial, emotional, cognitive and social components. Fibromyalgia is a chronic pain syndrome whose manifestation is influenced by these factors that were aggravated in the context of fear, uncertainty and restrictions of COVID-19 pandemic. This study aimed to evaluate if the impact of fibromyalgia worsened during this period. **Methods:** This is a longitudinal retrospective analysis of the database of Fibromyalgia Impact Questionnaire (FIQR) scores collected in the visits in the Fibromyalgia Outpatient Clinic of Hospital de Clínicas da Universidade Federal do Paraná (HC-UFPR) in Curitiba, Brazil. Two analytical approaches were done. The analysis of monthly evolution of FIQR medians indicates the pattern of disease manifestation during pandemic in the months of pandemic and compared to the months of three previous years. The hypothesis testing if there was an intrinsic variation of the parameter was done in an analysis of four periods between March, 16 and March, 15 of the following year. **Results:** In descriptive analysis of monthly variation of FIQR median, registers of 326 patients were analysed, and a worsening of this parameter was observed after March, 2020, when the pandemic was officially declared in Curitiba. This trend was also observed in November, 2020 when COVID-19 epidemiologic indices worsened again in Paraná State. 22 patients were included in hypothesis testing if there was a FIQR score variation in pandemic compared to previous periods, a p-value <0.05 was observed only in comparison to 19-20 (p=0.026). In comparison to 17-18 and 18-19, the p-values obtained were 0.101 e 0.065, respectively. However, the confidence interval analysis demonstrated a trend of relative worsening of FIQR scores in pandemic comparing to all previous periods: 17-18 (CI 95% = [-1.55; + 13.48]), 18-19 (CI 95% = [-0.48; +12.36]) and 19-20 (CI 95% = [+0.97; +16.25]). **Conclusion:** This retrospective analysis of FIQR scores registers collected in HC-UFPR can indicate a trend of worsening of fibromyalgia impact in life of patients during the pandemic compared to previous periods.

Dor

TL 1105165

STROKE ATTENTION JOURNEY: RESEARCH, TEACHING AND EXTENSION IN THE NORTH PLATEAU OF SANTA CATARINA

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Background: Stroke is the second most deadly disease in the world, however, with 80% of the causes considered preventable because they are modifiable factors and the time between the recognition of the signs and medical care is crucial for the chance of saving lives. Having this, in a continuous way and permanent education, already in its second version (1st in 2019 and 2nd in 2021), the Stroke Attention Journey in the North Plateau of Santa Catarina has been carried out. Characterized by a partnership between a Hospital institution, educational institution and private institutions providing health services. **Objective:** The objective of the actions linked to this journey is medical and health education, promoting the achievement of recognition at different levels of care focused on stroke risk factors and stroke patient care. For this, technical and professional lectures, courses and community actions were held in favor of knowledge about the risk factors for stroke, what is stroke, signs of a stroke, diagnosis and possible treatments and prognosis, as well as, how the post-stroke patient rehabilitation. The actions were aimed at different age groups, from early childhood education to graduate studies. Professionals and academics from different areas made a great contribution, covering the modality of research, teaching and extension. For this, in raising awareness of the proposal, data from regional surveys were used, which show that patients with suspected and later confirmed stroke were mostly transferred to U-AVC by ambulances (65.9%), and only 25% is performed by SAMU. In addition, those who arrived outside the therapeutic window were justified due to the failure to identify the stroke by family members (34%) and delay during the regulation/transfer from the municipalities to the U-AVC (18%). In this scope, more than 5700 people were reached with a frequency computed directly, estimating a greater indirect reach. There are numerous reports and data that show the permeability of the proposal in the different target communities. Continuing health education is the focus of different world organizations in favor of health promotion. This action shows the multi and inter-professional interaction, aimed at the different spheres of education and community. It is demonstrated here not only the scope of this action, but also the **Objectives:** behind it, as they bring direct benefits to the community and to the regional health market.

Educação Médica

TL 1105527

NEUROANATOMIC DISSECTION OF CORPSES AS A STUDY MATERIAL IN A MEDICINE COURSE

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Background: The study of neuroanatomy is extremely important in the medical course, both for academics who wish to pursue neurology and for the training of general practitioners. Teaching-learning **Methods:** that focus on the development of practical skills of the academic, such as the dissection of human brains for the study of neuroanatomy, facilitate the acquisition of detailed knowledge of human anatomy. **Objective:** The aim of this work is to describe and demonstrate the dissections performed on parts of human brains intended for the neuroanatomical study of the Medicine course. Literature data referring to neuroanatomical dissection were reviewed and parts of human brains from the Anatomy Laboratory of the Universidade Regional de Blumenau were photographed. Dissection constitutes a selection of fundamental techniques for the preparation of the piece that will be the subject of study and is an efficient method for the learning of human anatomy by medical students. At the Laboratory of Human Anatomy of the Regional University of Blumenau, medical students have the opportunity to have contact with parts of the nervous system of human cadavers, which are dissected and prepared for visualization of the anatomical structures, at different levels of depth and relationship with other structures, also presenting serial cuts, which are similar to cuts performed in imaging exams, thus demonstrating the connections between them and the basis of neuroscience to be used when future professionals. The dissection process allows the student to transform the organ into a puzzle that has several ways to assemble and consequently absorb the content to be learned. Unlike the study only in synthetic, plaster or plastic models, neuroanatomical parts in cadavers allow a three-dimensional view of the nervous system, fundamental for the formation of the basic neuroanatomy of the future doctor. With the dissection and preparation of natural pieces, it is also possible for the academic to recognize anatomical variations, a source of practical learning, encouraging the student to actively participate in the search for knowing such variations and relating them to possible pathological processes. Thus, the use of human cadavers in the study of neuroanatomy in medical universities proves to be a differential for academic training, providing an adequate and efficient method for learning the nervous system.

Educação Médica

TL 1105533

PRACTICAL CLASSES IN NEUROANATOMY: THE USE OF STUDY SCRIPT

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Background: Neuroanatomy is the study of the structures of the central and peripheral nervous systems. Its study constitutes a mandatory subject in many health courses, introducing concepts and structural knowledge essential for the understanding of areas of activity. Due to the complexity and extensive program content, anatomy teachers need pedagogical resources that facilitate the understanding of the academics in their practical classes. A resource that helps and guides students in their learning process, inside and outside the laboratory, are the scripts in practical classes, leading students to build their own knowledge. **Objective:** The aim of the study is to describe the use of study guides in the Laboratory of Human Anatomy as a method of practical study of neuroanatomy by the medical course of the Regional University of Blumenau - FURB. The scripts of FURB academics and the curriculum in which it is inserted were used, according to the description of the **Methods:** of the laboratory professors. At the Laboratory and Human Anatomy of FURB - Universidade Regional de Blumenau, practical neuroanatomy classes are carried out using a script of practical classes, which includes a list of the nomenclature of the structures to be studied by academics and images that point to the location from them. This guide is previously made available to academics so that they can seek information about the structures and become familiar with the anatomical terms. In the first practical classes, the structures contained in the scripts are demonstrated in the anatomical pieces, initially in synthetic models, by means of numbered pins, which correspond to the structures listed in the anatomical pieces. This first contact serves for academics to observe the location and relationship between the structures. In the following classes, the professors present the structures to the academics in the human natural parts, making a relationship between the anatomical structure and its concept, function, relationship and location, integrating theoretical and practical content. Thus, it was demonstrated that the human anatomy scripts in practical classes are effective tools in the learning process in Neuroanatomy, directing the student to understand and fix knowledge and facilitating the teaching-learning process.

Educação Médica

TL 1105567

DESCRIBING HORNER SYNDROME: AN ILLUSTRATED GUIDE

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Background: Horner Syndrome (HS) is classically characterized by a clinical triad: miosis/anisocoria, ptosis and facial anhidrosis. Its pathophysiology is related to the interruption of the oculosympathetic pathway; however, the mechanisms that lead to the onset of the syndrome are diverse, and some are even potentially fatal, highlighting the importance of its early identification. **Objective:** The aim of this study was to elucidate the main anatomical, etiopathogenic and clinical aspects of this neurological condition, based on original illustrations. **Method:** In order to synthesize the central aspects of the anatomy, clinical and pathophysiological aspects of Horner syndrome, original illustrations of own authorship were developed. In addition, neuroradiological examinations and photographs of patients with the syndrome were included. **Results:** Although uncommon, it is important to know how to recognize, diagnose and properly investigate patients with HS. Understanding the anatomy involved in the sympathetic innervation of the eye is fundamental to both understanding the pathophysiology of the syndrome and screening for its possible causes. The oculosympathetic pathway involves three neurons: first order (central), second order (preganglionic) and third order (postganglionic). The etiologies of HS include infections, neoplasms, vasculopathies and even iatrogenic lesions. Besides the clinical picture composed of the clinical triad, pharmacological and neuroradiological tests are often indicated for the diagnostic investigation of HS. **Conclusion:** We present an illustrated guide with targeted information on the anatomy, clinical picture and most important complementary tests in the context of HS, in order to provide a didactic guide on this syndrome for neurologists and the general medical community.

Educação Médica

TL 1105600

ANALYSIS OF THE COMMUNICATION QUALITY OF THE DIAGNOSIS OF SPINAL MUSCULAR ATROPHY IN THE VIEW OF PATIENTS AND FAMILY MEMBERS

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Background: The communication of the diagnosis of a genetic and neurodegenerative disease, such as Spinal Muscular Atrophy (SMA), requires a transmission centered on the patient and or on the family, giving autonomy to those involved and strengthening the millenary doctor-patient binomial. **Objective:** To analyze, from the perspective of patients and family members, the quality of disclosure of the SMA diagnosis. **Methods:** Semi-structured interview, via telecall, whose analysis was carried out by systematic condensation of the text, summarized in four thematic axes. **Results:** Fifty-seven volunteers from all regions of Brazil reported positive and negative experiences about clarifying the diagnosis, communicating the prognosis, affective memory related to the event, in addition to advice to doctors. **Discussion:** The individualized, clear, honest and welcoming transmission, emphasizing positive aspects, in the presence of family members and with the possibility of continued monitoring, were important to meet the communication demands. Among the barriers found, the lack of empathy, the estimation of life time and the absence of guidance and follow-up stand out. **Conclusion:** Failures in the transmission of the SMA diagnosis, in the perception of patients and families, alert about the need for improvement and adequacy of communication techniques and protocols, for the application of ethical principles in medical practice.

Educação Médica

TL 1106192

EXTENSION PROJECTS AS A STRATEGY TO STROKE PREVENTION

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Background: In medical education, students are usually invited to participate, throughout their training, in activities that bring them closer to the community, in exercise of developing empathy. In the academic environment, extension projects offer students the opportunity to be in closer contact with a particular medical condition of great relevance to the community. In these projects, students can get in touch with people outside academic environment and disseminate essential information that is often little known. In this context, the Interest Group of Neurosciences of the State University of Ceará (Neuruece) applies an extension project named "health education in stroke in primary care and in social networks". **Objective:** Perform educational strategies relates to stroke prevention in patients and/or population with very high risk for stroke in the context of Primary Health Care. **Method:** This project aims to enlighten the population about the risk factors for stroke, the importance of detection of its symptoms as well as immediate intervention to avoid sequelae. The target audience consists of hypertensive individuals at high or very high risk for stroke followed up in Primary Health Care Units of Fortaleza (CE), as well as its family members, companions and community health agents. In addition, educational posts on aspects involving stroke are elaborated and posted, both for the target audience and for the general public that has access to the profiles of Neuruece's social networks. **Results:** The health promotion action in the Primary Health Care Units reached 104 people who participated in training on the main characteristics of stroke, such as: risk factors, main symptoms, how to identify a stroke and when to call emergency services. In addition, 2 materials were posted on the following topics: "what is a stroke?" and "main signs and symptoms of stroke", which gathered an average of 98 likes and 93 shares. In total, 1118 accounts from different cities were reached (Fortaleza (CE), São Paulo (SP), Maracanaú (CE) and Eusébio (CE)); 87 visits to the @neuruece profile and 34 new followers gained, currently totaling 2550. **Conclusion:** The impacts of these educational strategies on the participants can prevent new cases of stroke or reduce complications resulting from the sequelae of people already affected by the disease. Neuruece's social networks may impact positively the current knowledge of the population about stroke.

Educação Médica

TL 1106238

CREATING SHORT VIDEOS AS AN ACTIVE LEARNING TOOL FOR NEUROANATOMICAL STUDIES

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Background: Despite the traditional predisposition to favor textual instruction, research has been carried out since the 1980s suggesting the importance of image-based education. Among these lines, Levin, Anglin and Carney proposed the five functions of textual illustrations: Decorative; Representational; Transformational, facilitating the memorization of crucial information; Organizational; and Interpretation, in order to contribute to the understanding of the information. Seeing the importance of new media and exposure to images for learning, as well as first-hand contact with the subject, short videos were chosen to be made as a means to learn Neuroanatomy, a crucial subject for medical education, also as an active learning tool. **Methods:** A Bibliographic Narrative Review was carried out on the different ways of learning, memory functioning, use of illustrations and active teaching methods, as well as a text-book approach for the contact with the subject (Neuroanatomy), followed by note-taking on the subject. Structured scripts were created for the voiceover and the images, in order to build a narrative from individual photograms, featuring an animation. The scripts were directed to the construction of short scenes, favoring memorization (transformational), the organization of information in a coherent way (organizational), leading up to understanding (interpretation). The photographs were taken using a semi-professional camera, Canon T5i and a tripod, as well as a Smartphone Apple SE. Editing was performed using the Final Cut Pro program. **Results:** Two (02) short animation films of 03 to 06 minutes were made, with animations alongside the voiceover explanation for easy understanding of the contents regarding the function and naming of the cranial nerves, as well as another video explaining encephalic gyri and sulci. **Conclusion:** Experiments carried out on the performance of students with different teaching methods showed that students with access to multimedia had the greatest success, being a playful and at the same time efficient way to expand the student's contact with the subject and then to consolidate the knowledge, especially if the Student is involved during the preparation of the study material itself.

Educação Médica

TL 1106385

"FORMANDO SINAPSES": INSTAGRAM AS A TOOL FOR TEACHING NEUROSCIENCE, AN EXPERIENCE REPORT

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Background: Digital technologies have been consolidating themselves as important educational forms in the teaching and learning process. Among these technologies, social networks have become important tools for the dissemination of educational content and for the democratization of knowledge, with Instagram being one of the main platforms used for this. Thus, it is important to explore this social media as a potential neuroscience teaching network. **Objective:** To report the use of Instagram as a tool for teaching neurosciences. **Method:** This is an experience report on the actions carried out by medical students in the university extension project "Formando Sinapses", from February 2022 to June 2022. 8 educational publications were carried out in the thematic axes: "Neuroinformação" (posts containing information about important matters for Neurology), "Neuroartes" (posts relating artistic expressions to Neurology), "Dicionário de Neuro" (explanatory post on neurological terms), "Neuroimagem" (explanatory post on neuroradiology signs) and "Tópicos em Neurologia" (explanatory video on essential topics in Neurology). The publications were aimed at the public with the aim of disseminating information on neurosciences. The posts were made using PowerPoint and Photopea and the videos were made using the cell phone camera. All publications were posted on the Instagram profile @neuruece and the feedbacks were accounted and analyzed for using a spreadsheet created in Excel. **Results:** The 8 posts published had the following distribution: 4 "Neuroinformação", 1 "Neuroartes", 1 "Dicionário de Neuro", 1 "Neuroimagem" and 1 "Tópicos em Neurologia". The analysis by Excel accounted for an average of 76 likes, 366 shares and 374 views on the video. In total, 3471 accounts from different cities were reached (Fortaleza (CE), São Paulo (SP), Sobral (CE), Caucaia (CE) and Maracanaú (CE)); 1018 visits to the @neuruece profile and 47 new followers were gained, currently totaling 2562 followers. **Conclusion:** University extensions projects may contribute to production and dissemination of scientific knowledge to society. The use of Instagram as an educational platform made it possible for more people to be reached with the knowledge disseminated. We believe that the project "Formando Sinapses" significantly increase the diffusion of neuroscience knowledge to our students.

Educação Médica

TL 1106437

MARKETING, SOCIAL MEDIA AND NEUROLOGY: RECOGNITION OF ETHICAL ASPECTS BY NEUROLOGISTS

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Background: The control of information in the face of a universe surrounded by easy access to the media is a challenge found in all areas. In Medicine, it is evident that the role of disseminating scientific content, dictated as medical advertising, must exclusively follow ethical principles of primarily educational orientation, guaranteeing the dissociability of merely commercial practices from the production of content with an instructive content. **Objective:** To analyze the posts made by neurologists and quantify the nonconformities carried out in opposition to the guidelines of the Federal Council of Medicina (FCM). **Method:** This is a cross-sectional and quantitative study carried out on the social network Instagram, through the search for an anonymous profile with a disabled locator. The descriptors used were: neuro, neurology, headache, dementia, parkinson, migraine, stroke, epilepsy and dystonia; postings from January to May 2022 in each neurologist profile were analyzed, among the first 50 posts found in the "most relevant" and "recent" tags. The variables were related to ethical infractions (before and after photos, self-promotion, sensationalism, praise from third parties, gifts and sweepstakes, prizes with no scientific value, photos with patients, announcement of exclusive technique), recommendations (educational posts, disclosure of the registry expertise, contact disclosure) and not recommendations (work selfies, personal posts). **Results:** 63 public profiles were found through the selected descriptors, showing that about 50.8% contained at least one infraction, with selfies being pointed out as the most prevalent in 73% of the total cases, followed by self-promotion and company advertisements, private institutions, and only 14.3% had no infractions. Regarding non-recommendations, personal posts prevail in 69.8% of the profiles. In addition, when dealing with recommendations, only 42.9% of physicians strictly follow what has been proposed. **Conclusion:** The study pointed out the great disparity between the norms presented by the FCM and the infractions presented in the surveyed profiles. Furthermore, by understanding the basic premise of caring for the individual, current Medicine presents the continuous need for the dissemination of evidence-based knowledge and the review of norms, highlighting the limits of medical advertising in the face of the vulnerability in which the general population is inserted.

Educação Médica

TL 1106499

DEVELOPMENT OF AN ONLINE COURSE AS A TOOL TO TEACH DEMENTIA MANAGEMENT FOR PRIMARY HEALTH CARE DOCTORS

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Background: Due to the progressive aging of the population and the exponential increase in the number of elder people, it became urgent that health professionals are able to manage the main diseases that affect this population, especially dementia in its most varied etiologies. **Objectives:** This work aimed to build a virtual educational technology in the form of a basic course in the management of elderly patients with dementia for Primary Health Care physicians, using an active methodological proposal. **Methods:** This is a methodological study. The construction of the tool was divided into 3 phases. In December 2021, the first phase of the study began with a review of the literature in health databases. The next phase consisted in the elaboration of the course project with the pedagogical planning. In the third phase, the educational technology for the dementia management course was developed with an active methodological proposal using strategies such as storytelling, role play, interactive activities (multiple choice questions, clinical cases, images, videos, quizzes, memory game) and short expository video classes. The material prepared was then inserted and made available in a virtual environment with publication of the material on the Moodle platform at the electronic address: "cursodeemencia.net". **Conclusion:** It is believed that the course developed is an adequate strategy for the improvement of health professionals in primary care, whose potential to change the health situation of the population is still underused and it is expected that it will result in the improvement of care provided to the elderly population under your care.

Educação Médica

TL 1106606

SIMULATION TRAINING HELPS IN LEARNING THE NEW BRAZILIAN BRAIN DEATH PROTOCOL?

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Background: In 2017, the Federal Medical Council released the new Brain Death (BD) protocol with many modifications compared to the last one. According to the protocol, the training course should be theoretical-practical, however the resolution hasn't detailed how was this practical activities. Simulation-based teaching has been increased use in medical education and could be part of practical program. **Objectives:** to verify if the BD simulation training during the medical course modifies the learning about the new BD protocol. **Methods:** This is a cross-sectional analytical study, performed at the University of Fortaleza, Brazil, from March to December of 2018. Forty-five, fourth-year medical students composed the sample. The students were divided into three groups, Group A – students who participated in theoretical class and simulated station; Group B – students who participated only in theoretical class. Group C – students who did not engaged in any activity. **Results:** Group A has scored a mean of 8.13 ± 1.01 , group B has scored 6.49 ± 1.52 , and group C has scored 5.54 ± 1.83 ($p = 0.000$). A significant difference between the groups in the prerequisite ($p = 0.004$) and the clinical examination step ($p = 0.003$) was found, with the biggest mistakes in Groups B and C. **Conclusion:** the BD simulated stations developed contributed significantly to the learning of the new Brazilian BD protocol, for the fourth-year medical students.

Educação Médica

TL 1106651

THE USAGE OF OSCE IN NEUROLOGY AS A METHOD TO EVALUATE CLINICAL KNOWLEDGE AND TECHNIQUE IN MEDICAL STUDENTS

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Background: oral and written exams allow for a traditional evaluation of clinical knowledge and skills in medicine, failing to gauge both technique and emotional skills in the students. The objective Structured Clinical Examination, known as OSCE, is vastly used to supplement written exams, allowing students the chance to practice medical skills (such as taking patient's history, going through the physical exam and formulating diagnosis) in a controlled space. In the Federal University of Alagoas' Medical School (FAMED/UFAL), the OSCE is implemented during the 6th semester with the sole purpose of evaluating student's technique and clinical thinking in the subject of Neurology. **Objective:** To report the neurologists' assistant students' and 6th semester students' experience with the OSCE's development and implementation in FAMED/UFAL. **Method:** The OSCE happened in FAMED/UFAL's Abilities Laboratory in the rotation format; four stations were displayed with clinical cases addressing typical topics in neurology that were discussed during classes. Accessory devices were used to heighten the students' immersion, such as videos, audio and actors. 1 minute was reserved for clinical cases' initial reading and 4 minutes for the cases' resolution, making each rotation lasting 20 minutes. **Results:** both assistant students and 6th semester students gave positive feedback to the neurology department about the OSCE. Evaluated students shared great interest in the possibility of conjoining theoretical and practical skills and the capacity to reinforce the knowledge passed on by the professors. The practice of both physical exam and clinical thinking were the highlights for the evaluated students, with most of them considering themselves ready to face neurological cases in real life. **Conclusion:** underestimated by most subjects in medical schools of Brazil, OSCE allows for a better implementation and evaluation of clinical and emotional skills, theoretical knowledge and practical technique in a controlled space, better preparing students for their future as professionals. Therefore, expansion of the OSCE method is needed for other clinical subjects in Brazilian medical schools, as a way to improve learning, reinforce skills that can not be gauged during written and oral exams and better prepare students for their future as doctors.

Educação Médica

TL 1105018

TEMPORAL LOBE EPILEPSY WITH MESIAL HIPPOCAMPAL SCLEROSIS (TLE-HS): LITERATURE REVIEW

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Background: Temporal Lobe Epilepsy (TLE) is the most common focal epilepsy in adults. Hippocampal sclerosis is associated with TLE and is present in 70-80% of cases. This is an epileptic syndrome commonly associated with drug-resistant epilepsies, and represents one of the most frequent indications for surgery for epilepsy. **Objective:** To understand the current scenario of TLE-HS. **Methods:** This is an integrative literature review with descriptive character, of articles indexed in the Medical Literature Analysis And Retrieval System Online MEDLINE/Pubmed, Latin American and Caribbean Literature in Health Sciences LILACS, and Scientific databases Electronic Library Online (SciELO), researched in the period between October 2021 and March 2022. **Results:** Initially, 136 articles were found in the databases, which after reading, 32 articles were selected in the research that corresponded to the proposed objective. **Conclusion:** Attention to the treatment of mesial temporal lobe epilepsy associated with hippocampal sclerosis remains of special interest due to its high prevalence and frequent resistance to antiepileptic drugs. Thus, surgery should be considered in these patients. The preoperative evaluation is responsible for determining the brain area where the crises originate and the precaution of a safe resection without postoperative sequelae. It is a multidisciplinary process that involves the performance of structural tests, functional tests, video-EEG monitoring and psychosocial assessment. The main data to consider are magnetic resonance, interpreted in the context of clinical data and EEG. Surgical treatment for drug-resistant mesial temporal lobe epilepsy with hippocampal sclerosis (ELT-HS) is the most effective alternative, although the literature reports seizure recurrence in 30% of patients treated surgically. Among the standard techniques for patients refractory to clinical treatment, the anterior temporal lobectomy (ATL) stands out. The analyzed studies demonstrate to be the most effective treatment for the control of crises. This technique consists of removing the anterior portion of the temporal lobe, removing structures such as the amygdala, hippocampus and temporal neocortex. The ATL surgical technique is a consolidated and effective procedure, with minimal mortality after the surgical procedure. The surgical treatment of TLE-HS has been shown to be effective for the complete resolution of crises in most patients.

Epilepsia

TL 1105072

THE INCIDENCE OF NEPHROLITHIASIS IN PATIENTS ON TOPIRAMATE THERAPY: A LITERATURE REVIEW

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Epilepsy is a chronic disease related to the nervous system, in which the brain oscillates between moments of stability and instability, in the latter presenting epileptic seizures. In order for the individual to control the incidence of seizures, there are antiepileptic drugs (AEDs), among them Topiramate (TPM), which acts by blocking voltage-dependent sodium channels, improves chloride influx, weakly inhibits carbonic anhydrase, potentiates the GABA receptor, and blocks the AMPA receptor. Among the adverse effects manifested in patients taking long-term PMT, there is the incidence of nephrolithiasis. The objective of this study is to describe the occurrence of kidney stones in individuals, especially epileptic patients, who are treated with the drug TPM, based on research in PubMed and SciELO databases. The research showed that, despite having a low incidence rate, some patients on long-term therapy with the drug may develop renal lithiasis. However, in most of the reported cases, discontinuation of the drug improved the symptoms and the stones disappeared. It was also possible to notice a certain incredulity described by older authors about this adverse effect, in which many did not consider relevant the relationship between renal lithiasis and the use of TPM. In contrast, more current studies emphasized the relationship as something significant. One study from 2002 does not report the incidence of nephrolithiasis, but, the duration of treatment of the individuals who participated in the study ranged from 20 to 1143 days, a time that may be insufficient to determine the risk of kidney stones, since this is an effect provided by long-term therapy. Finally, one study tried to associate the use of the drug with a ketogenic diet, which proved to be contraindicated because the ketogenic diet provides a significant reduction in HCO₃⁻ levels, increasing metabolic acidosis, a predisposing factor for the occurrence of kidney stones. It can be concluded that nephrolithiasis may have a great impact on the patient's quality of life, and may result in life-threatening complications such as sepsis and renal failure. Knowing the predisposing factors for its occurrence in each individual is of utmost importance when prescribing drugs that inhibit carbonic anhydrase, such as Topiramate, in order to provide quality of life and longevity to them, especially to patients with epilepsy.

Epilepsia

TL 1105103

EVALUATION OF ADHERENCE TO TREATMENT OF PATIENTS WITH EPILEPSY

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Background: Epileptic seizure is characterized by the transient occurrence of signs and/or symptoms about the excessive and synchronous neuronal activity in the brain. The factors that affect the quality of life of chronic disease patients is medication. Drugs will only be effective if taken regularly. **Objectives:** Compare adherence to treatment of patients with epilepsy and patients with other chronic diseases. **Methods:** Cross-sectional observational controlled study, with application of a questionnaire to a group of 100 patients diagnosed with epilepsy and 50 controls, who were patients with chronic diseases use medication. **Results:** By the controlled partial correlation, there was a positive and strong correlation between the scores of the Morisky tests and the scores of the BMQ Regimen and BMQ Memories test regarding age, disease duration and scholarship in years. Morisky's test and BMQ Regimen score ($r = 0.317$; $p < 0.05$) and BMQ Recall score ($r = 0.383$; $p < 0.05$) correlated strongly and positively when controlling for the number of focal and bilateral seizures. The effect about having or not having epilepsy using the Mann-Whitney test shows that the diagnosis interferes with the Morisky test score ($U = 1,917.00$; $p < 0.05$) and on the BMQ - Regimen score ($U = 1,959.00$; $p < 0.05$). **Conclusion:** Patients with epilepsy demonstrated greater adherence to treatment the oldest, disease duration, education level and better control of symptoms.

Epilepsia

TL 1105122

LEVEL OF CONFIDENCE OF HEALTHCARE PROVIDERS IN DIAGNOSING AND TREATING FUNCTIONAL SEIZURES IN BRAZIL: A CROSS-SECTIONAL STUDY

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Background: Functional seizures (FS) are a common differential diagnosis of epilepsy. However, not all FS-involved healthcare providers (HCP) are confident in diagnosing and treating this disorder. **Objective:** To compare the level of confidence of HCP in diagnosing and treating FS in Brazil. **Methods:** Neurologists, psychiatrists, and psychologists who work in the health sector respond to an online questionnaire. Researchers publicized the link to the survey via email and social media. The level of confidence in diagnosing and treating PNEs were questioned separately and self-rated on a 5-point scale (1 = none; 2 = low; 3 = medium; 4 = high; 5 = very high). Kruskal-Wallis test was used to compare the confidence level distribution across categories of HCP ($p = .05$). Data were collected from February 22 to May 4, 2022. **Results:** A total of 515 HCP (mean age of 39.4 ± 11.4 yrs [range 22-75 yrs]; 68% female) respond to the survey: 190 neurologists (37%), 82 psychiatrists (16%), and 243 psychologists (47%). The respondents represented the public and private healthcare sectors from all geographical regions of Brazil. Fifty-four (28%) respondents were neurologists with expertise in epilepsy, 54% were psychologists with expertise in clinical psychology, and 26% were psychiatrists with expertise in psychotherapy. Almost all neurologists (94%) and psychiatrists (96%) have diagnosed or treated at least one adult patient with FS. In contrast, a minority (43%) of psychologists reported experience with these patients. Psychologists' level of training in FS was lower than psychiatrists and neurologists (Mdn 2 points vs. 3 pts vs. 4 pts, respectively) ($p < .01$). Also, psychologists' level of confidence in diagnosing FS in adults was lower than psychiatrists and neurologists (Mdn 2 pts vs. 3 pts vs. 3 pts, respectively) ($p < .01$). The level of confidence in treating FS in adults differed among psychologists, neurologists, and psychiatrists, who had the highest level of confidence (Mdn 2 pts vs. 3 pts vs. 3.5 pts, respectively) ($p < .01$). **Conclusion:** Psychologists' confidence in diagnosing and treating FS in adults is lower than neurologists and psychiatrists. Since psychological treatment is the treatment of choice for FS, patients will likely face difficulties in Brazil accessing psychologists with expertise.

Epilepsia

TL 1105124

INJURIES CAUSED BY FUNCTIONAL SEIZURES: A SURVEY OF PROVIDERS' PERCEPTION

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Background: Functional seizures (FS) commonly produce physical injuries but the stigma that injuries rarely occur during FS persists among providers. **Objective:** To describe providers' perception of the occurrence of injuries as a result of functional seizures. **Methods:** Professionals from the health sector involved in the care of patients with FS answered to an online questionnaire, publicized via email, social media and professional organizations. The closed-ended question "In your opinion, can an adult with FS be injured during an event?" was rated on a 5-point scale (1 = never; 2 = rarely; 3 = occasionally; 4 = often; 5 = always). Kruskal-Wallis H test was used to compare providers' perception at the alpha level of .05. Data were collected during the first semester of 2022. **Results:** A total of 515 healthcare providers (mean age of 39.4 ± 11.4 yrs [range 22-75 yrs]; 68% female) answered to the survey: 190 neurologists (37%), 82 psychiatrists (16%), and 243 psychologists (47%). The respondents represented the public and private healthcare sectors from all geographical regions of Brazil. The perception of most neurologists (53%), psychiatrists (59%), and psychologists (58%) was that an adult with FS can occasionally be injured when having a seizure. However, neurologists and psychiatrists tended to perceive the occurrence of injuries as less frequently than psychologists ($H(2) = 34.628$, $p = 0.001$, mean rank 215.31 vs 210.91 vs 281.67, respectively); none of psychologists respond "never" to the occurrence of injuries, in contrast to 1% of neurologists and 1% of psychiatrists. Thirty-three percent of neurologists and 30% of psychiatrists responded "rarely" for the occurrence of injuries, compared to 14% of psychologists. The option "often" was selected by 13% of neurologists, 10% of psychiatrists, and 24% psychologists. None of neurologists and psychiatrists respond "always", contrasting to 4% of psychologists. **Conclusion:** A significant percentage of providers underestimate the occurrence of injuries caused by functional seizures.

Epilepsia

TL 1105215

PROTEOMIC ANALYSIS OF SYNAPTOSOMES ISOLATED FROM BRAIN TISSUE OF PATIENTS WITH MESIAL TEMPORAL LOBE EPILEPSY

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Background: The synapses are electric, and chemical communications between the neurons and synaptosomes are membrane vesicles that contain the synaptic components. The study of synaptosomes can give clues about synaptic transmission and its abnormalities since they contain all the machinery involved in the release, reuptake, and storage of neurotransmitters. In addition, synaptic proteins are possible therapeutic targets in many neurological diseases. Mesial temporal lobe epilepsy (MTLE) is the most common type of focal epilepsy in adults. It presents a high proportion of patients who do not respond to antiseizure medication (ASM) treatment. However, patients with MTLE who are pharmacoresistant to treatment with ASM may benefit from epilepsy surgery. **Objective:** To characterize the protein content of synaptosomes isolated from tissue of patients with pharmacoresistant MTLE using mass spectrometry-based proteomics. **Methods:** We analyzed the synaptosomes isolated from brain tissue obtained from epilepsy surgery (hippocampus and temporal lobe) of patients with MTLE and compared it to normal tissue. We isolated the synaptosomes using the Syn-PER reagent, extracted and digested the proteins using the S-Trap columns, and labeled them with TMT11-plex. Data was acquired using a Thermo Scientific™ Orbitrap Eclipse™ Tribrid™ coupled to a Dionex UPLC system. We used the ProteomeDiscoverer and R software for bioinformatics analysis. **Results:** We identified 1,890 proteins, 7,521 peptides and 18,601 PSMs. Our foldchange (FC) analysis revealed some critical proteins that may be relevant in the epilepsy context. Some of them are Synapsin-3 (FC=104), Abl interactor 1 (FC=147), Sodium-driven chloride bicarbonate exchanger (FC=37.8), and Outer mitochondrial transmembrane helix translocase (FC=51). The expression of these proteins was higher in the tissue of patients when compared to controls. **Conclusion:** Our study explores for the first time the synaptosome proteome of patients with MTLE. We identified novel proteins that may have a role in disease mechanisms, or they may contribute to biological processes leading to pharmacoresistance in MTLE.

Epilepsia

TL 1105276

ROBUST CHANGES IN GENOMIC METHYLATION FOUND IN MESIAL TEMPORAL LOBE EPILEPSY

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Background: Mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE+HS) is the most frequent and severe epilepsy in adults. When patients with MTLE+HS do not respond to clinical treatment, one may consider epilepsy surgery. The tissue resected by surgery has been extensively studied, and we and others have demonstrated abnormal gene expression in MTLE+HS. Gene expression is complex and regulated by many molecular mechanisms, including epigenetic changes. Therefore, we aim to investigate whether epigenetic changes determined by DNA methylation are present in brain tissue resected from patients with MTLE+HS. **Methods:** We performed whole-genome bisulfite sequencing to analyze DNA from 11 brain samples obtained by epilepsy surgery from patients with medically refractory MTLE+HS. We compared these to data obtained from samples of four autopsy controls (hippocampus and dentate gyrus). Samples from patients were divided into two groups: i) patients with less than 20 years of disease duration (G1 – n = 5); and ii) patients with more than 20 years of disease duration (G2 – n = 6). Methylation was performed in an Illumina[®] platform, and differentially methylated regions (DMRs) were identified. We conducted two by two comparisons between data from patients and controls and between the two groups of patients. Furthermore, we catalog the putative genes located in the DMRs. **Results:** Overall hypermethylation was observed in tissue from patients. Also, DMRs had a heterogeneous distribution throughout the genome, and these were usually observed in more than one region, depending on their length, but introns and promoters had increased DMRs compared to other genomic regions. We identified many genes in the DMRs for the comparisons performed (6,371 in G1 x Controls; 5,548 in G2 x Control; and 4,088 for G1 x G2). **Conclusion:** Currently, DNA methylation can be analyzed using high throughput methods to determine its distribution in the entire human genome, the so-called methylome. This analysis gives a complete overview of gene regulation mediated by methylation. We found remarkable changes in the methylome of the brain tissue in patients with MTLE+HS, with thousands of DMRs in patients. These were related to many candidate genes potentially involved in the molecular mechanisms underlying MTLE+HS. Furthermore, our results indicate that the molecular abnormalities present in HS change over time.

Epilepsia

TL 1105361

COSTS AND CASE FATALITY RATES OF THE HOSPITAL TREATMENT OF EPILEPSY IN THE STATE OF BAHIA, BRAZIL

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Background: Epilepsy is one of the most common neurological disorders, characterized by a predisposition to the occurrence of unprovoked seizures. Worldwide, there are about 50 million people living with epilepsy and 125,000 deaths related to the disease per year, of which 80% occur in low- and middle-income countries. Due to its high prevalence and chronicity, epilepsy **Results:** in high costs for healthcare systems and patients. Thus, we aimed to evaluate the frequency of Hospitalizations, costs and deaths related to the public Hospital care of epilepsy in the state of Bahia, Brazil. **Methods:** Ecological study, carried out with data from SIH/SUS, extracted from the DATASUS platform. The period from January 2017 to December 2021 was considered. The ICD G40 was the selection parameter used. Data was collected on the number of Hospitalizations, total cost, mean cost per Hospitalization, mean length of Hospital stay (LOS), deaths, and case fatality rate (CFR) according to the variables gender, race and age group. **Results:** In the 5 year period, there were 16472 Hospitalizations for epilepsy in Bahia. The total sum spent was R\$ 12,833,908.38, and the mean cost per Hospitalization was R\$ 766.57. The mean LOS was 6.3 days and there were 650 deaths, with a CFR of 3.9%. Males were Hospitalized more frequently than females (59.5% vs. 40.5%), with similar costs (R\$ 802.75 vs. 713.40). Patients of mixed race were the majority (54.9%), followed by whites (3.5%) and blacks (3.4%). Compared to other race groups, the mean cost for black patients was the highest (R\$ 2392.33), as were the mean LOS (25.1 days) and the CFR (4.49%). Patients aged 1 to 4 years were the most frequently Hospitalized, corresponding to 19.9% of the total. The age group between 0 and 1 year had the highest mean cost (R\$ 1216.54). The highest CFRs were observed in patients aged 70 to 79 years (8.3%) and in patients that were 80 years old or more (13.4%). **Conclusion:** Black patients' Hospitalizations resulted in higher mean cost, longer LOS, and higher CFR, which may be related to inadequate access to primary health care to treat the underlying disease, but also to low income and deficient social support to maintain post-discharge care amongst this population. Infancy epileptic syndromes are more difficult to control, resulting in more frequent Hospitalizations and higher costs. Elderly patients have more comorbidities, as well as an inherent physiological fragility, which implies greater fatality.

Epilepsia

TL 1105407

WNT PATHWAY ALTERATION IN DISPLASIC CORTICAL TISSUE COMPARED WITH NO LESION ADJACENT TISSUE OF PACIENTS WITH FCD AND REFRACTORY EPILEPSY

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Background: Focal cortical dysplasia (FCD) is a malformation of the cortical development that cause medical refractory seizures and the only treatment may be surgical resection of the affected area of the brain. People affected by FCD may present seizures of variable severity since childhood. The physiopathology of the disease is not yet understood, however it is known that several genes alterations may play their role. The WNT/β-catenin pathway is associated with cell transformation and migration and for this reason may be crucial for understanding FCD's aetiology. The aim of this study was to explore genes related to the WNT/β-catenin pathway in patients with FCD type II. **Methods:** Dysplastic tissue and tissue adjacent to the primary dysplastic lesion of patients with FCD type II were obtained from two patients who underwent surgical treatment. The analysis of the relative expression of genes was performed by a qRT-PCR array containing 84 genes related to the WNT pathway. **Results:** In patient 1, the analysis showed a difference in the expression of seven genes, demonstrating an increase in AXIN2, FRAT2, FZD9, KREMEN1 and PP2R1A genes and a reduction in CSNK1G3 and PPP2CA genes in dysplastic tissue. In patient 2, the analysis showed increased expression of CSNK1A1, FZD4 and PPP2CA genes, as well as reduced of CTNNBIP1 gene in dysplastic tissue. **Conclusion:** Dysregulation in the expression of genes that control the receptors of the WNT pathway keeps it in an inactivated state. Therefore, a eventual manipulation of this pathway focusing on its activation may influence molecular manifestations underlying the epileptogenic status in injured brain tissue, which could act as a therapeutic alternative to FCD type II. The WNT/β-catenin signaling pathway is crucial for the control of embryonic development, which takes place through the regulation of cell differentiation, migration and proliferation, and apoptosis process.

Epilepsia

TL 1105610

EVALUATION OF ACCIDENTS OCCURRENCE IN PATIENTS WITH TEMPORAL LOBE EPILEPSY: COMPARISON BETWEEN SURGICAL AND CLINICAL TREATMENTS

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Background: Patients with Epilepsy have a greater risk of accidents, which associates with high morbidity. The causes of frequent accidents are multifactorial, including falls without previous warning related to seizures, incapacity of using primitive protective reflexes because of the unawareness during seizures, drugs' side effects (gait disturbances, disequilibrium and cognitive impairment). Mesial Temporal Lobe Epilepsy is recognized for being pharmacoresistant and, therefore, there will be a greater risk of accidents. Although the surgical treatment is recognized as effective in yielding seizure control, few studies have evaluated the impact of surgery on the frequency of accidents. **Objectives:** To compare the frequency of accidents in patients with Temporal Lobe Epilepsy undergoing clinical and surgical treatments. **Methods:** This is a retrospective, non-randomized study. Data were collected from medical records of individuals from outpatient Epilepsy Clinics of the University of Campinas (UNICAMP). Patients were divided into two groups: clinical and surgical. We searched for data related to falls, burnings, fractures, head trauma and car accidents. We performed chi-square and Fisher's exact tests to analyze categorical variables and survival analysis (Kaplan Meier) to evaluate the time between the beginning of follow-up and the event's occurrence. The comparison was made using Log Rank and Breslow tests. **Results:** The clinical group included 242 patients, while the surgical group had 197 patients. Accidents were more frequent in the clinical group (37 [15.3%]) when compared to the surgical one (9 [4.6%]) [$p < 0.001$]. During the follow up (median interval of 17 years [DP 6.78] in the clinical group; 8.6 years [DP 5.41] in the surgical group), 95.4% of the surgical patients were free of accidents, while in the clinical group, 84.7% [$p = 0.02$]. Falls were more frequent in the clinical group (90 [37.2%]) than in the surgical one (15 [7.6%]) [$p < 0.001$]. The survival analyses confirmed that over time, falls occurred in 36% of the clinical group compared to 8% in the surgical group [$p < 0.001$], which can act as a risk factor for major accidents. **Discussion:** Accidents can be frequent in people with epilepsy. Surgical treatment in pharmacoresistant Temporal Lobe Epilepsy may have an important role in accidents prevention. Therefore, surgery should be considered during clinical analyzes of these patients because of its significant impact on their quality of life.

Epilepsia

TL 1105871

ANALYSIS OF THE COST OF HOSPITALIZATIONS FOR EPILEPSY ACCORDING TO REGION AND AVERAGE LENGTH OF STAY

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Background: Epilepsy is defined as a reversible brain disorder characterized by the persistent predisposition of the brain to generate spontaneous epileptic seizures. **Objective:** Relate the cost of Hospitalization for epilepsy treatment with the type of epilepsy, geographic region and in-Hospital length of stay. **Method:** A quantitative, descriptive, ecological study was carried out, with data available on DATASUS, an online platform by the Ministry of Health. The data chosen between 2011 to 2021 were based on the ICD-10 with federation unit; total number of Hospitalizations; total cost; mean value of Hospitalizations; days of stay; and average length of stay. As public data and without identification of the individuals, there was no need for approval from an ethics committee. **Results:** In the evaluation of Hospitalizations in Brazil during 2011 to 2021, traumatic brain injury was the most prevalent neurological cause, followed by epilepsy. This one represents 561.953 Hospitalizations with an annual average of 51.086 and a 4,5% variation. Their mean value was R\$ 753,60, with an increase of 49% between the first and the last year of data, and the average length of stay was 5,8 days. The region with most Hospitalizations was the Southeast (43%). For the total and average costs per Hospitalization compared to the total number, the southeast continues to lead with 53%, such as about the average length of stay with 7,1 days, followed by the Northeast, North, Midwest and South. When comparing Hospitalizations for diseases of the nervous system, epilepsy only has higher mean value than migraine, multiple sclerosis and nerve disorders. **Conclusion:** With the analysis of the total number of Hospitalizations by region, a concentration of the number in Brazilian metropolises is seen, which can be related not only to the population density and number of stressors, but also to the major supply of Hospitals that assist these patients and avoid underdiagnosis. This fact shows the average cost and days of Hospitalization by region, except for the southern region, which has a higher cost of services per day of Hospitalization in the region. In addition, the low variation in Hospitalizations over the years is a response to their etiologies, since unlike having specific causes such as status epilepticus or first seizure, epilepsy not controlled by non-adherence to drug treatment is the main cause of Hospitalizations which tends to reverse with the improvement of outpatient care.

Epilepsia

TL 1106009

KNOWLEDGE ABOUT SUDEP: WHAT PEOPLE WITH EPILEPSY AND FAMILIES IN BRAZIL THINK ABOUT COUNSELING? IS IT DIFFERENT FROM OTHER COUNTRIES? WHEN, HOW, WHO AND WHAT MIGHT CHANGE AFTER INFORMATION

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Background: Sudden unexpected death in epilepsy (SUDEP) is the second leading cause of death in people with epilepsy (PWE), affecting 12% of those with drug resistant seizures. The concept that SUDEP counseling would cause fear, anxiety and finally not change outcome evolved due to recognition of effective prevention measures. Nevertheless, worldwide awareness is still insufficient, despite counseling is advised by international societies, such as American Academy of Neurology, American Epilepsy Society and National Institute for Health and Care Excellence – UK. **Objective:** Assess the knowledge, preferences and attitudes of PWE, family members and caregivers (FC) about SUDEP in Brazil. **Method:** An online questionnaire approved by ethics committee was available on social media of Brazilian Epilepsy Association. **Results:** 533 complete questionnaires (311 PWE; 222 by FC on behalf of patients), median age 33.5 (+9.64) and 18.8 (+15.03) years, respectively, most female (84,9% PWE; 57,2% FC). Mental/intellectual or physical disability was declared by 15% (PWE) and 32,4% (FC), active seizures by 61,1% and 71,6%, and polytherapy by 49,8% and 64,4%. Tonic clonic seizures were reported in the last 12 months by 56,9% and 81,7%. 48,6% PWE and 61,3% FC heard about SUDEP, rarely by clinicians (11,3% and 16,2%). No counseling by physicians were available in 89,6% and 76,4%, despite most wanted to know (76,5% and 77,9%), at the first consultation/diagnosis (34,1% and 41,2%) or during early follow-up (32% and 21,3%). Neurologists and epileptologists should be responsible for the information according to PWE (81,4%) and FC (82,9%). Most PWE and FC (96%) reported that they would change behaviors after being informed about SUDEP risk such as accepting devices to detect seizures, new treatments, lifestyle changes and better adherence to them. Fear and sadness were the most common reported feelings, but despite that, >80% responders believed that counselling should be done to all. **Conclusion:** PWE and FC in Brazil want to know about SUDEP, by neurologists, at the diagnosis or soon. They consider that treatment and behavior changes would potentially be used to prevent this issue. Our data suggest low adherence to international recommendations, and Brazilian neurologists should review their practices based on patient and family preferences, in addition to the possibility of preventing SUDEP.

Epilepsia

TL 1106091

CLINICAL AND NEUROPHYSIOLOGICAL EVALUATION OF EPILEPSY IN PATIENTS WITH CONGENITAL ZIKA SYNDROME

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Background: Congenital Zika virus syndrome (CZVS) is a pattern of congenital anomalies associated with ZIKV infection during pregnancy. The expanded spectrum of this anomaly includes neurological and non-neurological malformations, with epilepsy being of great prominence. There is wide variation in descriptions of epilepsy, between 9-88.3% of patients. The hypothesis of this investigation is that CZVS courses with epilepsy, at a higher frequency than previously documented, with alterations possibly documented through a 12-hour video electroencephalogram (VEEG) study. **Objectives:** The aim of this study is to describe the characteristics of epilepsy in SCZV, through clinical and electrographic evaluation. **Methods:** The evaluation was performed using a 12-hour video electroencephalogram, at the Hospital Universitário Walter Cantídio, and the characteristics of epilepsy were evaluated using a questionnaire. This is a non-randomized, cross-sectional study with consecutive inclusion, and the inclusion criteria were: confirmed SCZV and follow-up at the Hospital Infantil Albert Sabin. **Results:** Twelve patients with CZVS were evaluated, with a mean age of 4.94 years, eight males, all grade V (transported in a manual wheelchair) in the gross motor function classification (GMFCS) and eleven (91.6%), with microcephaly at birth. All patients in the sample had epilepsy, with a mean age of onset of epileptic seizures of 12.4 months. The most commonly observed seizures were: focal seizures (91.6%), followed by generalized tonic seizures, in 66.6% and gelastic seizures, in 25% of the sample. Three patients had, between two years and four years, reflex crises when positioned in the supine position. Polytherapy was used in 75% and monotherapy in 16.6%. As for the electroencephalographic characteristics, all patients had disorganized background activity. Interictal epileptiform paroxysms were present in 91.6%, of which 66.67% were activated by sleep. It was detected in 66.67% of electrographic seizures and in 41.67% of electrical sleep disorders. **Conclusion:** The frequency of epilepsy in the population of children with confirmed CZVS is very high (100% of patients), with varied and often refractory semiology, including subclinical and sleep-activated, being important longer and periodic electrographic evaluation.

Epilepsia

TL 1106186

ANALYSIS OF SEIZURE CONTROL AND RISK FACTORS FOR REFRACTORINESS IN PEOPLE WITH EPILEPSY IN AN ISOLATED INDIGENOUS POPULATION IN PARÁ, BRAZIL

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Background: The prevalence of epilepsy is variable, being higher in developing countries. Rates of 57/1000 were found within the Guaymí indigenous in Panama and 18,6/1000 within the Bakairi in Mato Grosso, central Brazil, but there was no study in isolated populations, which may have peculiar life habits and genetic features. **Objectives:** To evaluate risk factors, clinical and electroencephalographic characteristics and the evolution of people with epilepsy in a geographically isolated indigenous population. **Methods:** Retrospective, historical cohort study. It used the medical records of patients seen from 2003 to 2018 at the neurology outpatient clinic in the city of Oriximiná-PA, a reference for 2,503 Waiwai tribe's indigenous people. They are unmixed and inhabit a forest reserve in the Amazon. There were 25 patients with epilepsy among the 118 indigenous treated. Clinical aspects, electroencephalogram (EEG) and neuroimaging exams, family history, comorbidities, the treatment and the response were studied. A survival analysis was performed, whose outcome was seizure control. **Results:** The occurrence of epilepsy diagnosis in the Waiwai population was 10/1000, most cases started in childhood (68%), with no difference regarding gender. The number of seizures before the first consultation was high in 44%, family history was reported in 24%, perinatal hypoxia and febrile seizures in 20% each. There was intellectual disability in one-fifth of the patients. Neurological examination and neuropsychomotor development were abnormal in one third. EEG showed abnormalities in 68% of the patients, with a predominance of focal epileptiform discharges (44%). Focal epilepsies were 64% and probably genetic and unknown etiologies were prominent. Treatment controlled 72% of patients (monotherapy in 64%). The analysis of seizure control over time showed, through Kaplan-Meier estimation curves and Cox and Weibull regression models, that the factors most related to refractoriness with observation time of 24 months were abnormal neurological examination and the presence of a family history ($p=0.001$). Epilepsy classification and age had not statistical significance for the outcome (Log-rank=0.2 and 0.7, respectively). **Conclusion:** Unlike other indigenous peoples described in the literature, the Waiwai have a low incidence of epilepsy. Therapeutic results were similar to the general population.

Epilepsia

TL 1106210

COMORBIDITIES ASSOCIATED WITH EPILEPSY IN PEDIATRIC POPULATION

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Background: The presence of comorbidities epilepsy in the pediatric population is routinely evaluated in the follow-up, in such a way that the associated clinical condition can affect quality of life, response to treatment or even mortality. Proper diagnosis and treatment of comorbidities can influence the appropriate control and management of epilepsy. **Objectives:** To assess the prevalence and types of comorbidities (intellectual disability, migraine, behavioral changes and sleep Disorders); Analyze the association of comorbidities with types of seizures and use of anti-seizures medications. **Methods:** Retrospective study, carried out in the Child Neurology outpatient sector of a University Hospital from April 2020 to May 2022. Data were distributed in frequency, and the chi-square tests of independence were performed to verify the association between variables and Fisher's exact test, using IBM SPSS software, and adopted significance of 5%. **Results:** The sample consisted of 282 patients with epilepsy (50.7% male; 49.3% female). The most prevalent age group, when the disease started, was older than 11 years (31.2%), followed by those younger than 2 years (30.9%). Focal seizures in 77.7% of the sample, indeterminate seizures in 11.3% and generalized seizures in 11.3%. Some degree of intellectual disability represented 60% of the sample, and significant association with indeterminate epilepsy (53.1%; $p=0.007$). Temporal lobe seizures occurred in 21.6%, with seizures with behavioral arrest in 67.2%; the frontal crisis corresponded to 12%. In addition, the occurrence of temporal lobe crisis was associated with some degree of intellectual deficit (62.3%) with good correlation with the severity of the deficit ($p<0.001$). The presence of frontal seizures was more associated with the use of two or more medications (50%) compared to the group with other seizures (26.7%; $p=0.021$). Patients with migraine made up 34% of the sample, being more frequent in the non-frontal seizures group ($p=0.017$) or using more medications ($p=0.006$). Behavior disorders were observed in 21% of the sample, with a higher prevalence in the group that used 2 or more medications ($p=0.026$). **Conclusion:** The association of many common psychiatric and somatic conditions is increased in epilepsy. The use of 2 or more anti-seizures medications should reflect not only difficult in epilepsy control. These findings may have implications in diagnosis and management of epilepsy and coexisting conditions.

Epilepsia

TL 1106212

LOW BONE MINERAL DENSITY OCCURS IN MEN WITH EPILEPSY, REGARDLESS OF YOUNG AGE OR SURGICAL TREATMENT

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Background: Although some studies showed increased risk of developing metabolic bone disease in people with epilepsy, little is known about the multifactorial underpinning mechanisms, especially in men. Aspects of bone metabolism such as vitamin D, calcium homeostasis and bone turnover markers may be negatively impacted by the use of antiepileptic drugs (mainly the enzyme-inducers). **Objective:** To evaluate the bone mineral density (BMD) in men with epilepsy and correlate it with clinical data such as age, type of seizure and anti-seizure drugs (ASD). **Methods:** We evaluated 505 men previously diagnosed with epilepsy and exposed (minimum of five years of exposure) to previous or current antiepileptic drugs (phenobarbital, phenytoin, carbamazepine and valproic acid), followed at UNICAMP-Brazil in 2021. We identified 178 patients with BMD analysis (median age range, 50.5 (21-86 years). Individuals were split into two groups (young-group [21-49 years], 90 individuals; older group [50-86 years], 89 subjects). The BMD test evaluated t-score indexes from the femoral neck, whole femur and lumbar spine. Osteopenia was defined with a t-score of -1.0 to -2.4; osteoporosis, with T-scores lower than -2.5. Clinical data were extracted from medical records and analyzed with SPSS22. We performed chi-square tests for categorical variables. **Results:** BMD was reduced in 107/179 men (59.7%). High-levels of bone disease were identified in both young and older patients ($p=0.09$): young-group [37/90 normal (41.1%), 40/90 osteopenia (44.4%), 13/90 osteoporosis (14.4%); older-group [35/89 normal (39.3%), 30/89 osteopenia (33.7%), 24/89 osteoporosis (26.9%)]. As expected, the osteoporosis group (20.6% of patients) was older than the osteopenia group ($p=0.011$). About 90% of patients with abnormal BMD had focal epilepsy. Unfortunately, 33/53 (62%) operated patients presented abnormal BMD. **Conclusion:** We observed elevated levels of bone disease (regardless of the young age and surgical treatment) in men with epilepsy exposed to ASD. results Our results suggest that ASD exposure is associated with early BMD reduction, which evolves into osteopenia and osteoporosis. BMD evaluation in patients with epilepsy and appropriate treatment may be necessary to reduce the risk of fractures and related comorbidities. The selection of appropriate ASD is of extreme importance to avoid bone disease in people with epilepsy, who usually require long-term treatment.

Epilepsia

TL 1106345

BILATERAL, UNILATERAL AND ABSENT HIPPOCAMPAL ATROPHY EXHIBIT DIFFERENT GRAPH THEORY PATTERNS IN TEMPORAL LOBE EPILEPSY

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Background: Although temporal lobe epilepsy (TLE) with unilateral hippocampal atrophy (HA) has been extensively investigated, fewer studies evaluated the impact of bilateral HA and the absence of HA on the efficiency of brain connectivity and networks. **Objectives:** To investigate brain connectivity and network properties with graph theory **Methods:** in TLE patients according to the laterality of HA (RIGHT-HA, LEFT-HA, BILATERAL-HA, NEGATIVE-HA). **Methods:** We analyzed 216 3T MRI resting-state images of TLE patients followed at the University of Campinas (median age: 48 y.o.; 122 women): 92 with LEFT-HA, 70 with RIGHT-HA, 25 with BILATERAL-HA and 29 NEGATIVE-TLE. Groups were paired with healthy controls in terms of age ($p>0.05$) and sex ($p>0.05$). We used the CONN connectivity toolbox/MATLAB 2019 to preprocess and extract graph theory measures (global efficiency (G-eff), average path length (PATH), degree/cost (DEGREE), betweenness centrality (BC), local efficiency (L-eff) and clustering coefficient (CC), after a whole-brain analysis. We considered a cost of 0.15 (including only the 15% more statistically significant results) and reported p -values <0.05 . **Results:** The LEFT-HA and RIGHT-HA presented a trend to higher measures of global connectivity (G-Con), local connectivity (L-Con), and centrality in the brain (higher G-eff, DEGREE, BC, L-eff, CC and lower PATH) and a trend to lower G-Con, L-Con and centrality in the cerebellum. The LEFT-HA and RIGHT-HA presented higher DEGREE in the ipsilateral hippocampus and higher L-eff /CC in the contralateral hippocampus. The NEGATIVE-TLE group showed a trend to lower G-eff and higher average PATH, DEGREE and CC in the brain. The BILATERAL-HA presented a trend to higher L-eff /CC in the brain. **Conclusion:** All groups presented a trend of higher local connectivity to the detriment of the G-eff. While the unilateral groups showed higher connectivity levels in the brain (with higher ipsilateral hippocampal G-Con and probable compensatory higher contralateral hippocampal L-Con) and disconnection in the cerebellum, the NEGATIVE-TLE presented concordantly higher L-Con and lower G-Con both in the brain and cerebellum. BILATERAL-HA did not exhibit any other specific patterns. Overall, cerebral G-eff is reduced in all TLE groups, regardless of the presence of HA. The presence of unilateral HA adds specific dysfunction in bilateral hemispheric connectivity, which may yield poor performance of the global brain connectivity.

Epilepsia

TL 1106594

THE IMPACT OF PHARMACORESISTANT SEIZURES ON GRAY MATTER ATROPHY IN GENERALIZED GENETIC EPILEPSY

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Background: Genetic generalized epilepsies (GGEs) correspond to 20-25% of all epilepsies. They are genetically determined disorders with subtle encephalic alterations affecting the white and gray matter (GM), as demonstrated by modern quantitative neuroimaging techniques. Alterations involving the thalamo-cortical connections are well described; however, few studies have investigated the impact of pharmacoresistant seizures on gray matter atrophy (GMA) in patients with GGEs. **Objective:** To investigate gray matter atrophy in Generalized Genetic Epilepsy (GGE) patients according to seizure control. **Methods:** Fifty-six patients with GGE from UNICAMP's Epilepsy Clinic were recruited: 33 Seizure-Persistent ([SZ-Persistent] patients with current seizures during the previous year of MRI acquisition, 21 women, median of 30 years); 23 Seizure-Free ([SZ-Free] free of any type of seizures for at least one year before MRI, 17 women, median 34 years), compared to 63 healthy controls (43 women, median 34 years) matched for age ($p=0.51$) and sex ($p=0.83$). High-resolution 3T T1-weighted scans were firstly segmented on CAT12/SPM12/MATLAB 2019 software (<http://www.neuro.uni-jena.de/cat12-html/cat.html>). Then, voxel-based morphometry (VBM) and surface-based morphometry (SBM) were performed to search for areas of gray matter volume and cortical thickness reduction with an ANOVA-test (between the SZ-Persistent group and SZ-Free groups compared to controls) on SPM12. We used age, sex and intracranial volume as covariates for the VBM analysis and sex and age for the SBM analysis. Clinical data were analyzed with SPSS20. The results from MRI analyses were reported with $p<0.05$, corrected for multiple comparisons. **Results:** We identified cortical thickness atrophy only in the SZ-Persistent group, localized at the right precentral gyrus, while no alteration was observed in the SZ-Free. Regarding the analyses of cortical volume, we verified atrophy of the left thalamus in the SZ-Persistent group and the left precuneus region in the SZ-Free group. **Conclusion:** As previously demonstrated in the literature, our results indicate that thalamo-frontal networks are vulnerable to persistent seizures in GGE, with significant gray matter atrophy. However, we also find that different degrees and locations of GM abnormalities are related to seizure control, which may have implications for the understanding of the physiopathology of the GGE.

Epilepsia

TL 1106286

HOW FRANKLIN D. ROOSEVELT'S DIAGNOSIS INFLUENCED THE DEVELOPMENT OF POLIOMYELITIS VACCINE

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Background: Poliomyelitis is an acute infectious disease caused by an enterovirus that invades the central nervous system and damages the anterior horn cells of the spinal cord and brainstem. Progress to lower motor neuron cell death leads to disruption of motor units and subsequent muscle weakness or complete paralysis. Poliomyelitis already existed long time ago, but only when Franklin D. Roosevelt was diagnosed with the disease that the United States mobilized to try to eradicate it. He was an American lawyer and politician who served as the president of the United States from 1933 until his death in 1945. He was elected to four presidential terms, being the longest serving president. **Objective:** Describe Franklin D. Roosevelt's illness and the increase the research to develop a vaccine against poliomyelitis in the United States. **Method:** A comprehensive, critical and objective analysis of the current knowledge about poliomyelitis and Franklin D. Roosevelt disease by means of a narrative literature review. **Results:** In August 1921, Roosevelt was swimming with his son, and the next day he started to feel weakness in his left leg. He also reported fever and a severe pain in his legs and lower back. After 3 days, the politician had loss of sensitivity of the lower limbs. The professor Robert Lovett of Harvard confirmed the diagnosis of poliomyelitis. His diagnosis was essential to change the attention to public health. After the issue entered the national political agenda, the US tried to develop a vaccine against the disease. In 1933, Roosevelt created a commission to research infantile paralysis that in 1938 became the National Foundation for Infantile Paralysis (NFIP), whose awareness campaigns described polio as the greatest threat to public health. The NFIP has funded many studies that sought to develop a vaccine for polio. In 1952, Jonas Salk, funded by the NFIP, completed a study to create a vaccine and wanted to test it in children. 90% of respondents allowed their children to participate in the study, which involved 1,5 million people. In 1953, the University of Michigan announced that the vaccine was safe and effective against the disease. **Conclusion:** Roosevelt's diagnosis of poliomyelitis changed the American society's look to the disease by stimulating the improvement of research through the creation of specific institutions that increased resources to develop a safe and effective vaccine.

História da neurologia

TL 1106402

FRIEDRICH NIETZSCHE COLLAPSE: NEUROSYPHILIS OR FRONTOTEMPORAL DEMENTIA?

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Background: Friedrich Nietzsche was a German philosopher who influenced all Western philosophy during the 19th century. His death occurred in 1900, but it all started with a psychotic episode called "Turin Collapse" in 1899. In this episode, Nietzsche embraced the neck of a horse that was being whipped, unconsciously reproducing a scene from the book "Crime and Punishment" by Dostoevsky. After that, he was declining physically and mentally, dying a year after this episode. After several attempts at diagnosis, the main hypothesis is that he had neurosyphilis, a disease that can cause dementia in a more advanced stage of syphilis. However, new diagnoses have recently been raised such as Frontotemporal Dementia (FTD). **Objective:** To raise questions about the diagnosis of Nietzsche's death. **Methods:** Analysis of Nietzsche's death and the new diagnoses suggested, based on a narrative review of the literature. **Results:** In 1889, Nietzsche was admitted to a psychiatric Hospital in Basel, with loss of cognitive abilities, including language and episodes of aggression and disinhibition. Due to this, the philosopher was diagnosed with "General Paralysis of the Insane" (GPI), the parenchymal form of neurosyphilis. Symptomatic neurosyphilis can be divided into two main types: meningovascular and parenchymal, the latter being the most similar to Nietzsche's condition, starting with mood and personality disorders and worsening with the deterioration of higher mental functions. Although Nietzsche's condition was compatible with a diagnosis of GPI, many of the symptoms observed are not specific to GPI and are compatible with a variety of neurological conditions. Due to this and other symptoms such as migraine and stroke episodes, new diagnoses have been proposed, such as retrobulbar meningioma, CADASIL, MELAS and FTD. The FTD hypothesis was suggested due to the onset of the clinical condition of early decline in social interpersonal conduct, in personal hygiene, emotional blunting, hyperorality, behavior compulsions, perseverative and stereotyped behavior and mood with inappropriate jocularity, symptoms noted on his admissions. **Conclusion:** Even after more than 100 years of Nietzsche's death, there are still questions about what caused it. When analyzing its history, contradictions, and uncertainties about the diagnosis of neurosyphilis are perceived, contributing to the emergence of other diagnostic hypotheses, such as Frontotemporal Dementia.

História da neurologia

TL 1106475

PROFESSOR CHARCOT'S GENEALOGY

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Background: Professor Jean-Martin Charcot is widely recognized as one of the most influential physicians of the 19th century. Usually described as having an austere and reserved personality, his family not only influenced his personality but also helped tailor his career. **Objectives:** This work discusses Charcot's genealogy and the role played by this family in defining his personality and advancing his career. **Methods:** We reviewed historical evidence concerning Charcot's genealogy and his familial relationships. **Results:** Born on November 29, 1825, in Paris, France, Charcot was the eldest son of Simon-Pierre Charcot, a carriage builder, and Jeanne-Georgette Saussier. His mother, Jeanne, passed away when he was 14. Jean-Martin Charcot married in 1864, at the age of 38, to Madame Victoire-Augustine Laurent, who was 29 years old (born in 1834). She had one daughter from her first marriage, Marie Charlotte Thérèse Durvis, who was raised from the age of 10 by Jean-Martin Charcot and Victorie-Augustine. The couple had a couple of children: Jeanne Marie Amélie Claudine Charcot, and Jean-Baptiste Charcot. Jeanne married Sir Alfred Joseph Charles Pierre Edwards, but divorced soon after, and married in second nuptials with Sir Arthur Georg Hendry. Jean-Baptiste Charcot, a physician, trained in Neurology under the supervision of his father, abandoned his neurological medical career years after Professor Charcot's death, becoming a famous maritime explorer, known worldwide as Commander Charcot. Jean-Baptiste Charcot had a relationship with Miss Céline Ducassé and had a daughter named Marie-Louise (Marion Charcot). He married for the first time in the year 1896, to Miss Jeanne Léopoldine Hugo, daughter of the famous writer Victor Hugo, who had been married to Mr. Léon Daudet, son of the writers Alphonse Daudet and Julia Daudet, in the year 1891, and divorced in 1895. Jean-Baptiste Charcot and Jeanne Hugo Charcot, divorced in 1905, after Jean-Baptiste Charcot's trip to Antarctica, on the grounds of desertion. After that Jean-Baptiste Charcot, remarried in the year 1907, to Miss Marguerite Cléry. **Conclusion:** Professor Charcot's early life was marked by the premature loss of his mother and his brother Jean-Eugene. Although these events apparently did not take any toll on his career, they might have contributed to his reserved personality. His marriage to a widow from the aristocracy provided him with a quick social ascent, as he came from a modest family.

História da neurologia

TL 1106527

CHARCOT AND MIGRAINES: A PERSONAL RELATIONSHIP

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Background: Professor Jean-Martin Charcot is widely recognized as one of the most influential physicians of the 19th century. As head of the Neurology service of the Salpêtrière, Charcot developed a reputation as both a clinician and a professor. In his *Leçons du Mardi*, Charcot presented patient-guided discussions on neurological diseases, presenting the audience with his appraisal of the medical literature and his practical experiences on the subject. Although few of these discussions approached headache disorders, as a migraine sufferer Charcot was able to present his insights uniquely. **Objectives:** This work discusses Charcot's lecture on migraine and his personal experiences on the subject. **Methods:** We present historical evidence concerning Charcot's lecture on migraine, originally presented on November 22, 1887, including excerpts and original drawings representing his visual auras. **Results:** Charcot presented the case of a 37-year-old teacher who presented a 12-year-history of "seizures" accompanied by limb paresthesias and speech impairment. Although the patient reported that she never lost consciousness, she presented visual phenomenology that included scotoma and line of fortification, sometimes followed by severe headaches. Charcot exposed his own experiences as a migraine sufferer, describing the visual auras: "I know the shape of this picture for myself from having felt it, but the description comes from an astronomer [...] Sometimes the scotoma presents only the line of fortification of yellow tints, sometimes red and green tones, and inside the luminous zone, which remained dark, you can see a kind of smoke, vapor more or less thick, vibrating in a way reminiscent of what is seen in the atmosphere immediately above a working stove. The initial very narrow circle gradually widens and finally ends up disappearing, a new phenomenon succeeds it, hemiopia, which is to say that by looking someone in the face, we do not see distinctly half of his figure. From this moment, pain is felt in the affected eye and above. Gradually the ocular phenomena disappear but the pain persists, and vomiting may occur." **Conclusion:** As a professor and a migraine sufferer, Charcot offered a unique perspective on migraine phenomenology. Although none of these descriptions were new at the time, the richness of detail provided by Charcot certainly paints a picture of the experiences our migraine patients go through.

História da neurologia

TL 1106549

THE HISTORY OF NEUROLOGY IN PERNAMBUCO: A HISTORIOGRAPHICAL OVERVIEW IN PERNAMBUCO NEWSPAPERS

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Background: Neurology is a relatively recent specialty in Brazilian Medical History. Therefore, it is necessary to study the steps taken towards the status quo of this specialty. However, analyzing its entire history is a work that goes beyond its framework and for this study it was chosen to analyze the past and present of the Neurology in Pernambuco due to its relevance in the medical field. **Objective:** To make a general historical overview of the History of Neurology in Pernambuco. **Methods:** A bibliographic survey was carried out with the files of the Hemeroteca Digital Brasileira of available digitalized Brazilian newspapers in the year 2020. **Results:** Analyzing the historical process of Neurology in Brazil means to understand, above all, the links between it and Psychiatry, like Charcot who described the current Multiple Sclerosis at the same time that he was interested in hysteria. As evidence, the first Brazilian book that broke the paradigmatic model of "Psychiatry of the brain" was published in 1878 (*Licções sobre as moléstias do systema nervoso feitas na Faculdade de Medicina do Rio de Janeiro*). In a simple search in the records of newspapers in Pernambuco, the term "neurology" was first used on May 14, 1913, when a neurologist at the Hospital of Philadelphia showed Brazilian doctors a 6km extension of films about clinical cinematography, a practice that was not commonly applied in Brazil. The newspaper reports: "Certainly whoever sees them not only assists the phases of an etiology but also the development, the march of the morbid entity and can thus draw much more valuable conclusions [...]". Neurology in Pernambuco was initially linked to Psychiatry. It became an independent specialty due to the influence of the chair of neurology at the Faculdade de Medicina - UFPE, with the outstanding participation of Jarbas Pernambucano. Later, with Manoel Caetano de Barros, Jarbas Pernambucano's successor, we have the expansion of the field of neurosurgery. Subsequently, the area of ??Neuroradiology grew, with Glenystane Holanda. **Conclusion:** The history of Neurology in Pernambuco had a long trajectory to become what it is today. Understanding the sequence of steps that have been taken up to the present day makes the neurologist beyond a mere external observer of the phenomena to become an actor and participant in the story, as the links of a chain which contribute to its steadiness

História da neurologia

TL 1106641

MACBETH AND CREUTZFELDT-JAKOB DISEASE: THE FIRST RECORD OF HUMAN PRION DISEASE

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Background: William Shakespeare, born in 1564, was an English poet, playwright and actor, considered one of the greatest figures in world literature. After more than four centuries of his death, his works continue to be the subject of discussions, due to the more than 712 medical references described in the symptoms of his characters, when the writer did not attend university. These symptoms fulfill the diagnostic criteria of several pathologies, especially neurological disorders such as parkinsonism, dementia, epilepsy, sleep disorders, headache, paralysis and human prion disease. His references to neurological syndromes inspired Jean-Martin Charcot, in the 19th century, to use his literature in neurological teaching sessions. One of the outstanding works is the Shakespearean tragedy *Macbeth*, in which it is believed that there is a possible description of Creutzfeldt-Jakob disease, through the protagonist's symptoms in a soliloquy, this being the first record of the human variant of prion disease, three centuries earlier of the official record. **Objective:** To describe the symptoms portrayed in *Macbeth* about human prion disease and the importance of literature in the development of neurological clinical thinking. **Methods:** Clinical, critical and objective analysis of the work *Macbeth*, through a narrative literature review. **Results:** Human prion disease is a fatal spongiform encephalitis caused by a mutation that transforms the PrPC protein into PrPSc, which is deposited in tissues, causing dementia, neuromuscular symptoms such as ataxia and myoclonus, and psychiatric manifestations. In *Macbeth*, the protagonist presents a rapid cognitive deterioration, also presenting involuntary movements, such as myoclonus, visual and auditory hallucinations, and insomnia, symptoms of Creutzfeldt-Jakob disease. Another event that confirms the diagnostic hypothesis is the description of the ingestion of necromantic infusions that contained a variety of human and animal organs, such as neural tissues, which is a known form of transmission of infectious prions, which occurred prior to the onset of *Macbeth*'s symptoms. After this event, *Macbeth* claims to have a strange illness. **Conclusion:** Shakespearean literature reveals itself, in this way, not only as a literary object of entertainment, but as a vehicle of medical knowledge, which precedes official pathological records and which, in this case, contributes to the neurological teaching and improvement of the clinical view.

História da neurologia

TL 1105019

CHANGES IN CEREBROSPINAL FLUID FROM COVID-19

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Background: The new coronavirus (SARS-CoV-2) appeared in Wuhan, China, and quickly evolved into a pandemic. There have been reports of neurological manifestations associated with COVID-19. These manifestations include mild symptoms, such as headache, fatigue, hypogeusia and hyposmia; and severe manifestations, such as encephalitis, encephalopathy, Guillain-Barré syndrome (GBS) and stroke. Cerebrospinal fluid (CSF) analysis is essential in making diagnoses of infections of the central nervous system (CNS), once it can provide information about the inflammatory response and can identify the etiological agent. The aim of this study is to assess the profile of patients with suspected and confirmed COVID-19 for and verify the cytochemical changes in CSF. **Methods:** It is a cross-sectional, retrospective and multicenter study. Patients who were selected had suspected or confirmed COVID-19. Lumbar punctures were performed from May 2020 to April 2021 at the following locations: Hospital Português, Hospital Esperança Recife, Hospital da Unimed, Hospital Memorial São José, Hospital Santa Joana Recife and Hospital Esperança Olinda. All punctures were requested by the attending physician to assess neurological disease. The CSF samples were analyzed at the Fernando Travassos Laboratory. Data collection is being carried out in the medical records. The research project for the master's degree was accepted by the UFPE ethics committee. **Results:** The total number of patients was 116. Men represented 55.17% of the sample. The mean age was 49.874 years, which ranged from 10 days to 95 years. The main reasons for requesting the CSF analysis were: headache (32.4%), encephalitis (25.9%) and lowered level of consciousness (17.65%). During the procedure, 61.7% patients were alert and 32.2% were in a coma. Almost all collections were performed in the lumbar/lying position, except for 1 patient who was seated and another patient who sent the sample. The mean initial pressure was 19.9 cmH₂O. The average global white cell count was 34.38 mm³, in only 3 patients there was the presence of neutrophils. The mean total protein was 72.4 mg/dL, and in only 1 patient had glucose consumed. **Conclusion:** Analysis of cerebrospinal fluid is important to understand how SARS-CoV-2 acts on the nervous system.

Líquido cefalorraquiano

TL 1106295

KAPPA INDEX AND OLIGOCLONAL BANDS DETECTION IN CEREBROSPINAL FLUID SAMPLES OF PATIENTS WITH SUSPECTED MULTIPLE SCLEROSIS (MS)

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Background: The evaluation of the intrathecal production of IgG can be useful in the evaluation of MS suspected cases. The most important test to verify the existence of intrathecal IgG production is the detection of oligoclonal bands (OCBs) in cerebrospinal fluid (CSF) and serum. Recent studies have reported that measuring CSF IgG free light chains (FLC), especially the Kappa Index, is comparable with OCBs as markers of IgG synthesis. It is still unknown if diagnostic sensitivity be increased by performing these two methods in parallel. **Objective:** The aim of this study was to evaluate the results of the Kappa index and the oligoclonal bands (BOCs) in samples from patients with suspected multiple sclerosis (MS). **Method:** Patients with suspected MS underwent cerebrospinal fluid (CSF) analysis. Intrathecal synthesis of IgG was evaluated with BOCs, IgG index, Reiber nomogram, and kappa index. **Results:** One hundred and fifty-three serum and CSF samples were included, 70 of them had positive CSF BOC. Among the BOC+ cases, 44 (77.14%) were also positive with the Reiber nomogram, 48 (68.5%) were also positive with the IgG index, and 68 (97.1%) were also positive with the kappa index. Among the patients without BOCs, 5 (6.02%) cases had a positive Reiber, six had a positive IgG index, and seven (8.4%) had a positive kappa index. **Conclusion:** The Kappa index was the method with the highest association with BOCs, confirming that this method is at least as sensitive as the detection of BOCs. Positive kappa results found among cases without BOCs may be false positives but may also indicate false negative results with BOC. Therefore, it is possible that kappa index can help to diagnose cases of MS without detection of BOCs.

Líquido cefalorraquiano

TL 1106297

CEREBROSPINAL FLUID (CSF) CULTURE RESULTS IN 47,473 CSF SAMPLES WITH NORMAL CELL COUNT AND BIOCHEMICAL RESULTS

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Background: Some studies have shown there is a low probability of positive microbiological results in patients with suspected meningitis/encephalitis and with normal CSF cell count. However, there is not yet a clear recommendation of not performing CSF culture in such cases. **Objective:** To retrospectively analyze all the CSF culture results in samples obtained from patients with suspected meningitis and/or encephalitis sent to a specialized laboratory over a period of 3 years. **Method:** Culture results for bacteria, fungi, and mycobacteria in patients diagnosed or suspected of having meningitis and/or encephalitis were analyzed. CSF general analysis was considered altered when at least one of the following were identified: global count above 3 cells/mm³, protein above 45 mg/dL, glucose below 50 mg/dL, lactate above 19.6 mg/dL. **Results:** Five hundred and nine (509) culture samples (1.07%) were positive. CSF general analysis was altered in 495 (97.25%) and normal in 14 (2.75%) of them. In nine cases the identified bacteria were considered a sample contamination. In 5 there was a positive result for *Cryptococcus*. **Conclusion:** Our data suggest that except in cases of cryptococcal meningitis, in which the CSF culture may remain positive after CSF normalization and is a therapy parameter, to perform a microbiological evaluation in CSF samples with normal cell and biochemical parameters is not justified.

Líquido cefalorraquiano

TL 1106545

RARE CASE OF PANCREATIC CANCER WITH LEPTOMENINGEAL CARCINOMATOSIS

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Background: Leptomeningeal arcinomatosis (LC) occurs in 5-8% of cases with solid tumors such as lung, breast and melanoma. LC of pancreas adenocarcinoma is rare. We report a case of unresectable pancreatic cancer that progressed to LC, despite maintaining good oncological, clinical and radiological disease control, as well as a significant reduction in plasma tumor markers, with no evidence of local progression. **Case Presentation:** Male patient, 58 years old, followed up at the Oncology Department of IMIP (Instituto de Medicina Integral Professor Fernando Figueira), in Recife/PE, due to metastatic pancreatic adenocarcinoma to the liver, lymph nodes and osteoblastic lesions in several vertebral bodies, more evident in L1, without lesions in other organs, since February 2021. In May 2022, he presented with acute left hemiparesis and dysarthria. The cranial resonance showed 6 lesions suggestive of secondary neoplastic implants and the presence of meningeal enhancement. The result of the analysis of the cerebrospinal fluid was 27 cells/mm³ with 80% of lymphocytes and 20% monocytes, proteins: 560mg/dL, glucose: 10mg/dL. and there was the presence of neoplastic cells. Some of them were shaped like signet rings. **Conclusion:** Particularly, in patients with pancreatic cancer, the development of central nervous system metastases is rare (occurring in approximately 0.3% of cases). This case is remarkable in two ways. First, only 15 cases of LC in pancreatic cancer have been published so far. Second, neoplastic cells were seen in the first lumbar puncture. The diagnosis of LC is difficult in most patients because this requires the detection of malignant cells on CSF cytology. They are detected in the initial CSF sample in only 50% of patients with LC. Repeat CSF analysis has been found to improve the yield to 90%.

Líquido cefalorraquiano

TL 1104707

EVALUATION THROUGH THE STOP-BANG QUESTIONNAIRE FOR THE RISK OF OBSTRUCTIVE SLEEP APNEA AMONG MILITARY POLICE OF AMAZONAS

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Background: Obstructive sleep apnea (OSA) is a sleep disorder caused by total or partial cessation during sleep, associated with air hypoxia and recurrent awakenings; **Objectives:** To evaluate the risk of sleep apnea and sleep quality among the military police of Amazonas; **Methods:** We carried out a cross-sectional observational study from August 2019 to January 2020 with about ninety military police officers from the military police of the state of Amazonas (PMAM). The study was approved by CEP-UEA (CAAE: 13042019.0.0000.5016). Data were analyzed by the R software. The accepted level of significance was 5%. 47.8% (N=43) of military police officers are at high risk for OSA, 50% (N=45) have poor sleep quality according to the MSQ questionnaire and 41.1% (N=37) have excessive sleepiness. A significant association was found between the presence of snoring and HAS (p=0.005) and an association between poor sleep quality (MSQ) with anxiety and depression (p=0.001 and p=0.003). **Conclusion:** In our study, we observed that military police officers have poor sleep quality, excessive sleepiness, and snoring, in addition to being overweight, which may be associated with the presence of OSA. Public health policies aimed at this population are necessary.

Miscelânea

TL 1104957

TELECONSULTATIONS IN NEUROLOGY DEVELOPED AND IMPLEMENTED DURING THE COVID-19 PANDEMIC IN THE BRAZILIAN UNIVERSAL HEALTH SYSTEM

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Background: Teleconsultation gain strength in Brazil as an urgent response to the COVID-19 pandemic, aiming at providing alternative strategies of care in a context of overburdened health systems. Ever since Brazil's Unified Health System (Sistema Único de Saúde, SUS) conception, primary health care (PHC) was designed to be the main form of access to the health services, to facilitate a structured and coordinated care. Despite these efforts, there are still important gaps between primary and specialized care due to the fragmentation of healthcare networks and to the expressive demand for consultations in secondary and tertiary care, ultimately resulting in long waiting lists for many medical specialties, including neurology. In 2018, the Ministry of Health of Brazil started a project in partnership with private Hospitals aiming to improve efficiency in referring cases from PHC to specialized care, employing telehealth strategies. To minimize the impact of the pandemic on the access of patients to specialized care, the project expanded its activities by initiating the offer of teleconsultations. **Objective:** The aim of this study is to characterize the teleconsultations in neurology carried out by this project in Recife, capital of Pernambuco, a state in northeastern Brazil. **Methods:** Retrospective study evaluating teleconsultations in neurology that happened between May to September 2020. Teleconsultation was performed preferably in the video modality. Conditions eligible for teleconsultation were headache, epilepsy, and cerebrovascular disorders. **Results:** A total of 243 teleconsultations were analyzed, of which 76.95% was a first appointment. In 48.97% of cases, the teleconsultation represented the first opportunity for the patient to be consulted with the specialist. Among cases of first appointment, 20.16% were further referred to a face-to-face consultation and 21.81% could be redirected to primary health care. Headache disorders were the most predominant clinical conditions. **Conclusion:** The implementation and development of telemedicine in neurology during the Covid-19 pandemic represented an opportunity to assess the value of having teleconsultations added along the line of care from primary care to a medical specialty, promoting the coordination of care across different levels of complexity of care in the health system and improving access to specialized care.

Miscelânea

TL 1105240

DEVELOPMENTAL MILESTONES EVALUATIONS AND BRAIN MEASUREMENTS IN RATS EXPOSED TO THE LARVICIDE PIRIPROXYFEN DURING THE GESTATIONAL PERIOD

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Background: In 2015, there was an increase in the occurrence of microcephaly in Brazil, which was related to maternal contamination by Zika virus during pregnancy. However, there are inconsistencies between the Zika maternal infections rates in relation to microcephaly cases. Thus, other causes that may be related to the occurrence of this malformation have been studied, including the exposure of pregnant to pesticides. The pyriproxyfen is a pesticide used to control the *Aedes aegypti* mosquitoes, the vector of Zika virus. This pesticide is structurally similar to retinoic acid, a derived from vitamin A, involved in the control of neurogenesis during embryonic development. **OBJECTIVE:** This study evaluated if prenatal exposure to pyriproxyfen can affect the neonatal development and brain measurements in rats. **Methods:** Wistar rat pups were divided into negative control group (offspring of mothers that drink water during pregnancy, CT-); positive control group (offspring of mothers that drink water containing 2 g/L of vitamin A during pregnancy, CT+) and pyriproxyfen group (offspring of mothers that drink water containing 0.002 g/L of Sumilarv during pregnancy, a pesticide that uses pyriproxyfen as the active ingredient, PIR). From the 1st to the 21st postnatal day, the developmental milestones of pups were evaluated. At the 45th day of age, animals were euthanized and measurements of the brain volume, the brain and cerebral hemisphere length, the cerebellar vermis length, the maximum brain and cerebellar width were performed. **Results:** Neonatal developmental assessments showed that PIR group performed latter the forelimb grasping, but carried out early the negative geotaxis task compared to CT- and CT+ groups. CT+ group exhibited the audio startle response latter than the other groups. However, the eyelids opening and the hindlimb placing response were performed early by the CT+ group. A reduction in the maximum brain width was observed in PIR and CT+ groups when compared to CT-. **Conclusion:** The changes in neonatal developmental milestones observed in the PIR group were not the same as those seen in the CT+ group, showing that this pesticide does not appear to interfere with retinoic acid signaling during embryogenesis. However, pyriproxyfen affected the acquisition of part of the developmental milestones evaluated and reduced the maximum brain width of the offspring, suggesting that prenatal exposition to this pesticide can produce neurotoxic effects.

Miscelânea

TL 1105242

POSSIBLE RELATIONSHIP BETWEEN PRENATAL EXPOSURE TO THE LARVICIDE PYRIPROXYFEN AND DEFICITS IN MOTOR FUNCTION AND IN TIBIALIS ANTERIOR MUSCLE STRUCTURE IN RATS

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Background: Pyriproxyfen is a pesticide used in Brazil to control the *Aedes aegypti* mosquitoes. However, this pesticide has a similar molecular structure to the retinoic acid, a vitamin A derivative that regulates neurogenesis, skeletal striated muscle development and expression of myogenic specification genes. In this sense, it is postulated that exposure of pregnant women to this larvicide could interfere with retinoic acid signaling, impairing the motor function and muscle tissue structure. **Objective:** This study aimed to verify if the prenatal exposition to pyriproxyfen can produce motor deficits, as well as damage to the muscular tissue of the offspring. **Methods:** Male Wistar rat pups were divided in 3 groups: Negative control (CT-) - offspring of mothers who drunk water during pregnancy; Positive control (CT+) - offspring of mothers who drunk water containing 2 g/L of vitamin A during pregnancy and Pyriproxyfen (PIR) - offspring of mothers who drunk water containing 0.002 g/L of Sumilarv during pregnancy, a pesticide that uses pyriproxyfen as the active ingredient. The gait quality of the experimental animals was evaluated using Basso, Beattie and Bresnahan scale (1996). On the 40th postnatal day, animals were euthanized and the tibialis anterior muscle was dissected. Samples were stained with hematoxylin and eosin for analysis of the peripheral nucleus/muscle fiber ratio, capillary/muscle fiber ratio and cross-sectional area of muscle fibers. **Results:** In one animal from the PIR and one from the CT+ group it was observed external rotation of the hindlimbs. Deficits in motor coordination were also observed in one animal from the PIR and in four animals from the CT+ group. However, the BBB score was similar between CT-, CT+ and PIR groups. The peripheral nucleus/muscle fiber ratio and cross-sectional area of muscle fibers was also similar in the three experimental groups studied. On the other hand, capillary/muscle fiber ratio was lower in the CT+ and PIR compared to the CT-group. **Conclusion:** The motor and structure damage of the muscle observed in this study were subtle, which may be related to the administration of pyriproxyfen and retinoic acid having been performed on demand. This method of administration was used to mimic what occurs when pyriproxyfen is added to drinking water tanks to control the *Aedes Aegypti*. More studies are needed to confirm whether maternal exposure to pyriproxyfen produces more severe deleterious effects on the offspring.

Miscelânea

TL 1105329

FAT MASS CHARACTERIZATION IN POST-STROKE INDIVIDUALS

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Eighty percent of the risk factors for the development of stroke are modifiable, with obesity being among the top five – In the northern plateau of Santa Catarina, it one is among the three most prevalent. With this in mind, present study aimed to characterize and correlate in a pioneering way the body mass index (BMI) and percentage of fat mass in affected individuals and bedridden by stroke, during immediate Hospitalization. For that, data such as: sex; BMI; age; physical activity level; leg length (right, cm); and fat mass (%). For weight, self-reported values by the patient and/or caregiver were assumed, justified because 100% of the population was bedridden or unresponsive – for the same reason, height was predicted according to Chumlea et al. (1988), using knee height as an anthropometric measure. BMI was calculated and classified according to WHO recommended criteria. Fat mass was estimated using the tetrapolar electrical bioimpedance method (Maltrom, model BF906) – considering a desirable level for men when ?15%, borderline from 15.1-24.9% and elevated ?25%; and for women, desirable ?23%, borderline 23.1-31.9% and high ?32%. The sample refers to the three-month period of collection in the Unit-stroke in the northern plateau of SC; with a mean age of 70±11.1 years, and 57.1% of the individuals were female. The mean BMI found was 23.36±5.36 (normal) of the according to the considered criteria. However, the population presented an average of 35.18±7.65% of fat mass, which is considered high for both sexes. When the indices were statistically analyzed in relation to the distribution of means by sex, there were no significant differences between them or in relation to the mean, however, higher coefficients of variation were found for females. Interestingly, 100% of population self-reported being sedentary. These data demonstrate the discrepancy between the qualification that the BMI and bioimpedance measures generate, pointing out here that in immediate post-stroke population with a normal mean BMI, where obesity would not be characterized as a prevalent risk factor, there were quantitative findings of the percentage of fat mass beyond the limits considered high, regardless of sex. Therefore, it is proposed that other studies, with a larger sampled population, be carried out to highlight the importance of the anthropometric characterization of fat mass as a tool for analyzing a risk factor for stroke, as well as its reliable characterization and tracking.

Miscelânea

TL 1105474

INVESTIGATION BETWEEN A MOZART SONATA EFFECT ON CONTEXTUAL MEMORY EXTINCTION

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Background: Memory consists in the capacity of storing and recalling information acquired through experiences. After consolidated it can be kept, if repeatedly evoked, or extinguished. Memory extinction is as important as its formation, since it demonstrates the capacity of suppressing thoughts and/or behaviors no longer needed. This process does not imply experience elimination, but it's re-signification. Despite the known music beneficial effects on the brain, especially Mozart's K448 sonata, few is known about its potential on memory extinction. **Aims:** Investigate Mozart's K448 sonata effects on contextual memory extinction in mice. **Methods:** The project was accomplished with 41 C57BL/6J mice, divided into G1: Mozart (n=14), G2: Ambience (n=14) and G3: Control (n=13). Only the first group was exposed to music from 9:00 pm to 7:00 am throughout the project, since intrauterine life. Groups 2 and 3, however, were only exposed to ambient noise. From the 50th to the 54th day, the animals underwent Habituation, in order to control behavioral interferences in response to the testing ambient differences. On the 55th day the Aversive Training was performed with groups 1 and 2. On the 82nd day all groups began Extinction Test, which lasted five consecutive days. All groups then underwent Recall Test on the 107th day. The procedures were recorded for analysis. Repeated measures ANOVA Test was used to analyze Extinction Test and One-Way ANOVA Test to Recall Test. Values of $p < 0,05$ were considered significant. **Results:** The statistical analysis demonstrated that the fear extinction process varies amongst groups, despite the reduction of the freezing time for all of them during the tests. In general G1's freezing time was longer than G2's. On the first day of extinction G1's freezing time was longer than G2's e G3's, whilst on the second day it was only longer than G3's. At last, on the fifth day, G1's freezing time was shorter than G2's. Amongst the second and third day of extinction, G1's freezing time reduced more than G3's, whilst amongst the fourth and the fifth day, G1 upheld its freezing time, whilst G2's extended. On recall, G1's freezing time was longer than G3's. All analysis were statistically significant ($p < 0,05$). **Conclusion:** It was noted that mice exposed to Mozart's K448 sonata were more reactive to aversive stimulus. However, they exhibited faster contextual memory of fear extinction.

Miscelânea

TL 1106118

MEDICAL CANNABIS: SOCIAL HISTORY CONTEXT AND CURRENT APPLICATIONS IN NEUROLOGY – A REVIEW

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Background: Cannabis is a plant genus native to south-central Asia that includes three different species: Cannabis sativa, Cannabis indica and Cannabis ruderalis. More than a hundred compounds with physiological action have already been isolated from the plant, including cannabidiol and tetrahydrocannabinol. The discovery of these compounds allowed the subsequent discovery of a vast biological system called the endocannabinoid system (ECS), which promotes several interactions between different systems and organs. With this new knowledge, new drugs were created aiming at the treatment of all sorts of conditions, mainly neurological, psychiatric and gastric. Investigations of the safety and efficacy of these drugs are still at its beginning, even though its commercialization and use in different diseases are already a reality. **Objective:** To carry out a comprehensive literature review on the use of medical cannabis in various neurological diseases, discussing its social history context and elucidating the applicability of cannabinoids and their results in the light of current scientific evidence. **Methods:** A literature review was performed in 2021 using the MeSH terms "Neurology" and "medical cannabis" in PubMed. Clinical trials, systematic review and meta-analyses were included, without restrictions on date of publication and language. **Results:** Thirty-eight articles were found, from which twenty were analyzed for the writing of this work. Among the twenty articles selected, four were meta-analyses, nine were systematic reviews and seven were clinical trials. As for the diseases studied by each paper, three were about movement disorders, three about pain, three about multiple sclerosis, ten about epilepsy and one about various diseases. **Conclusion:** This work found little evidence in current medical literature that supports cannabis-based treatment for neurological conditions, except in specific epileptic syndromes and in spasticity in the context of multiple sclerosis, both which have been approved in some parts of the world and are already in use. However, the use for Parkinson's disease and chronic pain still needs further evidence. Therefore, the main critics and caveats of this study are that the irrational use of medical cannabis may promote short and long-term harm to patients and their families and that this drug may end in a new historical ostracism because of political and economic reasons.

Miscelânea

TL 1106130

OPSOCLONUS-MIOCLONUS-ATAXIA POST-INFECTIOUS COVID-19 SYNDROME: CASE PRESENTATION

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Case Presentation: A 65-year-old man came to the outpatient clinic reporting tremors for 30 days. The condition progressed with dizziness, feeling of malaise, vomiting, scant speech, opsoclonus and myoclonus, which were presented nine days after the mild coronavirus disease (COVID-19) was confirmed. He denies vertigo, convulsive crisis and history of cerebrovascular accident (CVA). Neurological examination showed bilateral dysmetria, dysidiadochokinesia, dysynergia, myoclonus, truncal ataxia, astasia and abasia. Brain Magnetic Resonance Imaging (MRI) revealed mild global atrophy and microangiopathy. An electroencephalogram (EEG) was requested, which showed normal baseline activity with the presence of multidirectional rapid eye movement artifacts, in addition to laboratory tests to screen for paraneoplasia and possible etiologies (CA 19.9, carcinoembryonic antigen, ESR, CRP, occult blood test, ANA, homocysteine, vitamin B12, vitamin E, folic acid, negative antiphospholipid antibodies, liver enzymes, hepatitis B virus, Hepatitis C virus, HIV Elisa, VDRL and FTA-ABS, chest and abdomen tomography). **Results:** The results did not show paraneoplasms or other infections, thus concluding the diagnosis of Kinsbourne Syndrome. Lumbar puncture was performed to assess cerebrospinal fluid (CSF), cultures for bacteria, fungi and tuberculosis were negative and pulse therapy with high-dose methylprednisolone. The patient showed complete improvement of the clinical picture. **Discussion:** Kinsbourne Syndrome or Opsoclonus Myoclonus Ataxia Syndrome is a rare neuroinflammatory disease with a variable combination of opsoclonus, myoclonus and ataxia¹. The etiology can be paraneoplastic, such as neoblastoma, parainfectious, after viral or bacterial infections, toxic and metabolic or idiopathic. The exact pathogenesis is not fully known, however, in recent years, the emergence of neurological disorders, such as Kinsbourne syndrome itself, after COVID-19 infection has been reported in several studies¹. It is believed that the syndrome may be an inflammatory phenomenon mediated by the autoimmune system, secondary to infection by the coronavirus². **Conclusion:** Despite being a rare disease, the diagnosis of Kinsbourne syndrome by the neurologist, in the present case, was verified from the neurological physical examination, mainly related to motricity and persistence of opsoclonus, as reported in the literature, in addition to a broad diagnosis differential.

Miscelânea

TL 1106152

PREVALENCE OF CERVICAL POSTURAL ALTERATIONS IN OCULOMOTOR DISTURBANCES

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Background: The human mechanism of adjustment the balance and the posture involves a reflex relation between the oculomotor system and the superior cervical, which **Results:** from the information coming from the vestibular and ocular systems. In the deficient posture, there is an abnormal relation between various body parts, resulting in excessive solicitation of the supporting elements and in the decrease of the perfect balance of the body over its supporting foundation. Among the cervical postural problems there is the torticollis, describes the condition of a cephalic deviation in relation to the torso. The oculomotor disturbance, which can cause torticollis, is known as squint. This pathology is an extraocular musculature dysfunction, congenital or acquired, which has high prevalence in the population, varying from 2% to 4%. **Objectives:** This research aims to evaluate the prevalence of cervical postural alterations in patients with oculomotor disturbance, through eye motricity tests and postural exams, showing the correlation between both pathologies. **Method:** The inclusive criteria to being a part of the research were: patients between the ages of four and fifty years old, carriers of oculomotor alteration and acquired cervical postural alterations. As the exclusive criteria, carrier patients of congenital postural disturbs which present profound amblyopia, monocular or subnormal vision. **Results:** The obtained results demonstrated that the oculomotor disturbers carrying individuals also presented postural alteration, the presence of several dysfunctions on segments responsible for posture in individuals with limitations, paresis and paralysis in the ocular muscles. This study demonstrated the it was reported a really frequent vicious head position condition is the eye adjustment for the purpose of compensating for a squint. Beside this, the literature is still scarce, in this perspective it is clear the need for further research and publications to clarify the association of the two pathologies. **Conclusion:** The present study conclude that patients who carry oculomotors disturbs, also present cervical postural alterations, which demonstrate a meaningful percentual results to alterations on the segments responsible to the maintenance of postural balance. Therefore, these patients should be referred to specialists for a postural assessment and, if possible, undergo postural reeducation treatment and treatment of the extrinsic eye muscles.

Miscelânea

TL 1106223

SAFETY AND EFFECTIVENESS OF COVID-19 VACCINATION IN MULTIPLE SCLEROSIS PATIENTS – INTERIM ANALYSIS OF A MULTI-CENTER STUDY

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Background: Multiple sclerosis (MS) is the most common immune-mediated demyelinating disease of the central nervous system. COVID-19 infected more than 535 million people worldwide. The most promising solution remains an effective vaccine. Given the autoimmune condition of MS, questions have arisen as to whether vaccination for COVID-19 confers effective immunity or whether it could represent a trigger for EM attacks. **Objective:** Report the clinical features and possible adverse effects of COVID-19 vaccination in patients with MS. Describe clinical events in patients diagnosed with MS that are subsequent to vaccination. Show possible changes in functional classification after vaccine exposure. **Method:** Cross-sectional clinical study. Applied questionnaire directed to patients diagnosed with MS of patients followed up at the Hospital Universitario Walter Cantídio and Hospital Geral de Fortaleza. Data collected between January and February 2022 includes age, gender, vaccine used, side effects, number of MS attacks, clinical form of MS, EDSS, COVID infection. Statistical analysis was descriptive. Continuous variables were described by their means and standard deviations (SD). **Results:** Total of 13 participants, 10 female and 3 male. Mean age of 43.7 years, SD of 16.9. Predominance of complete higher education (5 participants) and born in Fortaleza (6 participants). Only 1 patient had not been vaccinated with any dose and, of the 12 vaccinated, only 1 had not taken the booster dose. Vaccines applied in immunization: BNT162b1, ChAdOx1 and COVID-19 Adsorbed Vaccine (inactivated). 4 patients reported mild side-effects, all after ChAdOx1 vaccine. 2 vaccinated patients had COVID-19 after vaccination. No MS attack after any dose of vaccine. All of them use or have already used some disease-modifying drug, but 1 does not currently use it. 4 participants had attacks in the last 2 years (1 unvaccinated patient with 3 attacks; 1 patient with 3 attacks-only 1 attack after vaccination; and 2 patients with 1 attack before the beginning of the vaccination). The EDSS remained unchanged for all participants, except for one with a change in EDSS from 1 to 3 due its psychotic condition (psychosis with urinary incontinence). **Conclusion:** There was no triggering of new clinical MS attacks in patients vaccinated and no worsening of EDSS attributable to vaccination. Side effects to vaccination were present only after ChAdOx1 vaccination, with side effects already reported in the general population in phase III studies.

Miscelânea

TL 1106226

OPTICAL COMPRESSION CHANGES IN PATIENTS WITH SELAR INJURY: A TERTIARY HOSPITAL STUDY

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Background: Sellar tumors are classified according to size, presence of compressive symptoms and functionality. When the lesion is large and has a suprasellar extension, it can compress the optic chiasm and may evolve with ophthalmological changes. Optical coherence tomography (OCT) can determine the thickness of the nerve fiber layer (NFL) and its division by quadrants, being of significant relevance in chiasmic compressions due to its predilection for the nasal region of the optic nerve, and its correlation with data already classically obtained by campimetry. **Objective:** The present study aims to evaluate the changes in optical compression of patients with a sellar lesion, using visual computerized campimetry (VCC) and OCT. **Methods:** This is a longitudinal analytical observational study whose design is a retrospective cohort. By reviewing the attendance records, information was collected regarding demographic and clinical data, data from the magnetic nuclear resonance of the sellar region and the eye examination. The research was approved by the Research Ethics Committee of the Hospital, complying with ethical principles. **Results:** Seven patients were selected for the study, with ophthalmological assessment before and after treatment. Unilateral hemianopsia was the most common symptom, present in 57.1% of patients (n = 4), followed by bitemporal hemianopsia and headache. All cases (n = 7) had suprasellar extension and compression of the optic chiasm and only 14.3% (n = 1) of the cases had parasellar extension. In the analysis through VCC, the mean of Mean Deviation (MD) was -21.03 dB \pm 6.27 (-24.42 dB) in pre-treatment and 15.29 dB \pm 10.32 (-21, 51 dB) in post treatment. In optic nerve analysis through OCT, the mean NFL thickness was 73.86 μ m \pm 22.88 (7550 μ m) in the pre-treatment and 70.42 (69.50 μ m) in the post. Those who showed better thickness of the MD fiber layer in the pre-treatment, also showed better values in the recovery of campimetry and visual acuity. Patients with better NFL thickness on OCT also had the best results on MD in post treatment. **Conclusion:** These data reinforce the importance of a complete ophthalmologic evaluation as a means of determining the possibility of visual improvement in the treatment of pituitary tumors, in addition to demonstrating its effectiveness in decompressing the optic pathway, especially the chiasm. It also suggests that the OCT should be used as a tool to assess the patient's visual prognosis.

Miscelânea

TL 1106577

ROLE OF THE NERVOUS SYSTEM IN THE DEVELOPMENT OF LANGUAGE AND COMMUNICATION

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Background: Language is essential for communication between human beings, being responsible for the transmission of sounds, symbols, gestures and words. Several organs and systems work together in the development of speech, writing and body expression, such as the visual and auditory system, cerebral cortex. **Objective:** To understand the mechanism of action of the nervous system in language development. **Material and Methods:** This is a descriptive study, through a bibliographic review, carried out through books and scientific articles published between 2018 and 2022 available on the Scientific Electronic Library Online (SciELO), Latin American and Caribbean Literature in Science platforms. da Saúde (LILACS) available in full in Portuguese, English and Spanish versions. The keyword "neurophysiology" was used. 34 results were found, of which 5 works were selected to compose the bibliographic review. **Results:** Studies show that language and communication skills are the result of the combination of the performance of specific areas in the cerebral cortex: Broca's, Wernicke's, angular gyrus and part of the motor cortex. Broca's area, located close to the lateral sulcus, is responsible for the articulation and motor coordination of the pronunciation of words. Wernicke's area, located in the temporal lobe, provides understanding and meaning for the language used in communication, as does the area of the angular gyrus, which makes language intelligible through reading. In addition, there is also the participation of the motor cortex - which is divided into primary, pre-motor and supplementary areas, which enables speech vocalization and dominant body expression in non-verbal language. These areas work in complex neural networks together with sensory and memory information for the execution of the different types of language used in communication and send the information to the skeletal muscles, mainly through the corticospinal tract and the innervation from the brainstem. **Conclusion:** The neural circuitry involving the cerebral cortex and the conduction pathways make it possible to maintain the high complexity of the patterns of the skills in question, and the involvement of lesions in these places interferes with the quality of communication.

Miscelânea

TL 1104977

CLINICAL AND EPIDEMIOLOGICAL PROFILE OF PATIENTS DIAGNOSED WITH MULTIPLE SCLEROSIS IN CASCAVEL, PARANÁ, BRAZIL

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Background: Multiple sclerosis is a chronic autoimmune disease of the brain and spinal cord in which inflammation, demyelination and axonal loss occur from the early stages of the disease. The onset of multiple sclerosis has been typically observed in individuals aged from 20 to 40 years, with the female predominance. **Objectives:** The aim of the present study was to describe the clinical-epidemiological profile of patients with multiple sclerosis who were diagnosed and followed at a local neurology service. **Methods:** The model of this study was observational and retrospective descriptive, involving patients diagnosed with multiple sclerosis in the last two years and followed in a neurology service in the city of Cascavel, Paraná (PR). The epidemiological variables were gender, age, clinical presentation, patient age at diagnosis, patient age at onset, predominant symptoms, use of disease-modifying drugs and EDSS (expanded disability status scale). The clinical presentation was defined in the following categories: relapsing-remitting, secondary progressive and primary progressive. **Results:** The sample size presented a number (n) of 39 participants diagnosed with multiple sclerosis, with a predominance (79%, n = 31) of female patients (female: male ratio = 4: 1). The raw prevalence of multiple sclerosis was determined to be 11.6 per 100,000. Concerning the age, the average was 36 years and 69% (n = 27) were aged between 20 to 40 years. The relapsing-remitting clinical form was predominant (87%, n = 34), followed by primary progressive form (7%, n = 3) and only one case of secondary progressive form. The average of the EDSS score was 1. At the time of diagnosis they had a median age of 32 years and at the first onset they had a median age of 27 years. As initial signs and symptoms, 35% (n = 14) reported paresthesia, 28% (n = 11) reported hemiparesis, 23% (n = 9) reported dizziness, 20% (n = 8) reported diplopia or blurred vision, 7% (n = 3) had dismetria. Other symptoms include urinary urgency, holocranial headache and syncope. In the sample, 76% (n = 30) use some disease-modifying drug. Among these, 43% (n = 13) use dimethyl fumarate, 20% (n = 6) use glatiramer acetate, 16% (n = 5) use natalizumab. Other medications include ocrelizumab, interferon beta-1a, teriflunomide and fingolimod. **Conclusion:** The clinical-epidemiological profile and prevalence of multiple sclerosis in Cascavel (PR) is considered average in accordance with most Brazilian studies.

Neuroepidemiologia

TL 1105034

RELATIONSHIP BETWEEN PARKINSON'S DISEASE AND DEMENTIA IN A CITY IN THE INTERIOR OF MINAS GERAIS

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Background: Parkinson's Disease (PD) is the second most common neurodegenerative disease in the general population, after Alzheimer's Disease (AD), with Parkinson's Disease Dementia (PDD) being a well-established complication of PD. About 25 to 30% of patients with PD develop PDD, and the incidence of dementia in patients with PD is about six times higher when compared to the general population, which is cumulative with advancing age. **Objective:** To quantitatively correlate dementia as a complication of Parkinson's disease in a city in the countryside area of Minas Gerais, in addition to describing the clinical-epidemiological profile of these patients. **Method:** Descriptive, cross-sectional, documentary study, which used secondary data collected from 56 medical records of patients diagnosed with PD from two general neurology outpatient clinics in Alfenas and who underwent consultation during the years 2020 and 2021. **Results:** Of the 56 medical records reviewed, 31 had some degree of dementia on clinical examination, but 14 patients were excluded from the study because they did not meet the criteria for diagnosing PDD or had other clinical entities besides PD that could justify dementia. Therefore, of the 42 patients selected, 17 had dementia with probable cause of PD (40.48%). Furthermore, of these 42 patients, 25 were men and 17 women. Of the 17 patients with PDD, the mean age was 70.86 years, with a mean time of diagnosis of PD of 5.14 years, 9 were women and 8 men; of the 25 who did not have dementia, the mean age was 67.32 years, with a mean time of diagnosis of PD of 4.25 years, with 8 females and 17 males. Of the motor signs and symptoms, some degree of bradykinesia was observed in 42, plastic rigidity in 40, postural instability in 32, tremor at rest in 30, and difficulty in swallowing in 10 patients. Other frequent diagnoses in these patients were: arterial hypertension in 22, insomnia in 17, depressive disorder in 13, hypothyroidism and REM sleep behavior disorder in 12, anxiety disorder, hallucinations and excessive daytime sleepiness in 9, diabetes in 7, heart disease in 6 and arrhythmia in 4. **Conclusion:** The proportion of patients with PPD found in this study (40.48%) is in line with the global average. Resting tremor is the motor symptom that best responds to pharmacological therapy, and difficulty in swallowing is common in these patients.

Neuroepidemiologia

TL 1105054

EPIDEMIOLOGICAL PROFILE OF LEPROSY IN AN AMAZON HEALTH REGION

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Background: Leprosy is a chronic, granulomatous, infectious disease, caused by the bacterium *Mycobacterium leprae*, also called Hansen's bacillus, an alcohol-acid resistant bacillus, which has the characteristic of being obligatory intracellular, demonstrating a peculiarity of predilection for skin cells and cells. peripheral nerves, which settles in the infected person's body and can multiply. By 2005, leprosy elimination had been achieved in all countries except: Angola, Brazil, India, Madagascar, Mozambique, Nepal, Central African Republic, Democratic Republic of Congo and Tanzania. Brazil has an average detection rate of 12.2 cases/100,000 inhabitants, being classified as having a high burden for the disease and, in the world ranking, as the 2nd country with the highest number of new cases registered in the world, second only to India. **Objectives:** To trace the epidemiological profile of the leprosy in the 11th Health Region of Pará, from 2010 to 2020. **Method:** This is a cross-sectional, descriptive, retrospective epidemiological study with a quantitative approach, based on secondary data collections, reported in the System of Notifiable Diseases Information (SINAN), from 2010 to 2021. The data were organized in Excel spreadsheets and statistically analyzed in Bioestat 5.3. **Results:** During the period from 2010 to 2020, 9,863 cases of leprosy were reported in the 11th Health Region of Pará, with 7,966 new cases, with an average of 896 notifications/year. Despite the large decline in the annual detection rate of leprosy in the region, based on the first and last year of the survey (2010 - 2020), the region is still in the Very High Endemicity profile, with 29 cases per 100,000 inhabitants/year. The profile of leprosy cases in the region is predominantly male (62.4%), between 15 and 50 years old (60.6%), brown (67.8%) and with a low level of education (73.9%). Mostly Multibacillary (68%) and Dimorphic (57%), reflecting high percentages of disabilities (27%) and new cases in children under 15 years of age (12.6%). **Conclusion:** It is concluded that leprosy is a public health problem in the region, with high rates of disability, under 15 years-old and hidden cases, requiring interventions in various spectrums, such as active casework, better monitoring of the treatment, training of the professionals about the disease and its attributions, as well as health education for the population on the subject.

Neuroepidemiologia

TL 1105096

STROKE PROFILE AND CARE DURING THE COVID-19 PANDEMIC: THE JOINVILLE'S FIRST YEAR EXPERIENCE

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Background: The SARS-CoV-2 coronavirus pandemic (COVID-19) has become one of the hardest challenges nowadays. Public managers and health teams have concerned about social restrictions, lack of resources and a possible negative impact on incidence and quality of care for Stroke patients. **Objective:** We aim to investigate the impact of COVID-19 on incidence and management of Transient Ischemic Attack (TIA), Stroke and its subtypes in Joinville, Brazil. **Methods:** We did a prospective population-based stroke registry of all first-ever cases of Transient Ischemic Attack (TIA), Stroke and its subtypes admitted to health services in Joinville since the beginning of restrictions due to COVID-19 (defined as March, 2020) and the same period one year before. A comparative analysis was made between these two periods (12 months before versus 12 months after COVID-19 outbreak). The patients' profile, incidences of TIA, stroke and its subtypes, severity, reperfusion therapies, in-Hospital stay and workload and in-Hospital lethality in these periods were compared. **Results:** The analysis showed a significant incidence reduction of TIAs (32,8%; $p=0,002$). The rate of thrombolysis (IV) and trombectomy (MT) did not change. The intervals from admission to IV/MT were similar in both periods. Patients with ischemic stroke caused by atrial fibrillation had their in-Hospital stay abbreviated during the pandemic. Workout investigation was similar before and during the pandemic, with an increase in number of Cranial Tomography ($p=0,02$), transthoracic ecocardiogram ($p=0,001$), chest X-rays ($p<0,001$) and Transcranial Dopplers ($p<0,001$). During COVID-19 pandemics, in-Hospital lethality did not change. **Conclusion:** We concluded that COVID-19 Pandemic in Joinville was associated with lower TIA incidence, without negative influence over quality of care, workout investigation and in-Hospital management of stroke patients. Studies are needed to understand the real reasons for this epidemiological behavior. We believe that fast and strategic response to unusual pandemic challenges can explain good results e minor hazards in stroke prevention and caring.

Neuroepidemiologia

TL 1105125

THROMBOLYTIC THERAPY IN THE STROKE UNIT OF A HOSPITAL IN NORTHEASTERN BRAZIL – EXPERIENCE IN THE LAST 5 YEARS

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Background: In Brazil, the incidence of ischemic stroke ranges from 105 to 137 per 100,000 inhabitants (1). It is the second cause of death and the leading cause of permanent disability in the adult population. Thus, reperfusion treatment is a goal to be achieved in patients with acute ischemic stroke (2) and, stroke units have a fundamental role related to the organized and fast care of such individuals (3). However, the pandemic of the disease by coronavirus brought with it barriers for reperfusion therapies in acute stroke, such as the closing of several stroke units around the country (4). In this study we will present the profile of cerebral ischemic patients undergoing thrombolysis in the stroke unit of a large Hospital in the Northeast of Brazil in the last 5 years. **Objective:** To describe the evolution of thrombolytic therapy in the stroke unit of the Hospital da Restauração over the past 5 years. **Methods:** We conducted a cross-sectional, observational, retrospective study that included patients with ischemic stroke undergoing chemical thrombolysis in the period from June 2018 to May 2022 admitted to the neurological emergency of Hospital da Restauração, Recife, Pernambuco. **Results:** 269 patients underwent chemical thrombolysis in the period studied. There was a predominance of males (51.7%) and a mean age of 65 years. Our study, therefore, reaffirms the profile found in the literature (5). Despite the male predominance, in 2021 and in the first five months of 2022 there was a female predominance of 57.1% and 56.8%, respectively. Prior to the closure of the stroke unit due to the COVID-19 pandemic, from June/2018 to April/2020, 174 patients were thrombolized. In contrast, we noticed a sharp drop in the number of patients undergoing reperfusion therapy in the period when the unit was deactivated in May 2020 to the end of November 2021, totaling 30 patients. After reopening, we had a significant increase and by May 2022, 65 patients were thrombolized. **Conclusion:** Our study showed a significant increase in the number of patients undergoing thrombolysis over the years. The closure of the stroke unit resulted in a decline in the number of patients treated in the period of the COVID-19 pandemic. With this, it is shown the importance of the units for a better assistance to patients who need care in the acute phase of stroke.

Neuroepidemiologia

TL 1105159

IDIOPATHIC INTRACRANIAL HYPERTENSION MORTALITY IN BRAZIL: AN EPIDEMIOLOGICAL ANALYSIS

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Background: Idiopathic Intracranial Hypertension is an unexplained increase of intracranial pressure, which is defined by the relationship between the cranial components (brain, cerebrospinal fluid, and blood) and its volume, a constant. **Objective:** To compare, through a prevalence analysis, the mortality of Idiopathic Intracranial Hypertension among the Brazilian regions in the one decade (2011-2020). **Methods:** Epidemiological, retrospective, descriptive study, carried out with data obtained from the Mortality Information System (SIM/SUS) and data published by the Brazilian Institute of Geography and Statistics (IBGE). From these data, the number of deaths per million people of each Brazilian region per year of the period was calculated. For a more elaborate analysis, the variables age group of patients and gender were considered. **Results:** In 2011, the national average of deaths per million from Idiopathic Intracranial Hypertension in Brazil was 0,63. The Southern Region featured the lowest mortality rate, with 0,47 deaths per million, and the Northern Region featured the highest rate, with 0,68. Furthermore, males were proven more vulnerable than females, spanning 57% of total deaths that year. Regarding age group, in 2011, deaths were more prevalent in the groups ranging from 20-29 and 50-59 years old, while they were less prevalent in the group ranging from 10-14. In 2020, the national average of deaths per million was 0,40, representing a 37% drop when compared to the beginning, in which the Mid-Western Region had the biggest decrease (71%), and the Northeastern Region had the lowest decrease (17%). In that regard, the region with the biggest mortality rate is the Northeastern, with 0,58, while the lowest rate goes to the Mid-Western Region, with 0,18. In relation to gender, the male population had a reduction of 29% while the female had a reduction of 33%. Moreover, the age group of 20-29 years old had the biggest decrease in that period (76%), and the age group of 50-59 years old, besides having a reduction of 24%, kept the highest average, with 20,6 deaths. **Conclusion:** This analysis allowed a better understanding about the impact of Idiopathic Intracranial Hypertension in Brazil, since, despite an important decrease in mortality rate during the period of 2011-2020, keeps being a relevant cause of death among the population, mostly affecting the males, the age group between 50-59 years old and the country's Northeastern Region.

Neuroepidemiologia

TL 1105162

VIRAL MENINGITIS: AN EPIDEMIOLOGICAL ANALYSIS IN ONE DECADE IN BRAZIL WITH THE IMPACT OF THE COVID-19 PANDEMIC

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Background: Accordingly, to the Brazilian Ministry of Health between 2007 and 2020 more than 265 thousand cases of meningitis were confirmed. Viral meningitis was responsible for 45,9% of the confirmed cases. The social isolation of the COVID-19 could have affected this scenario. **Objective:** Analyze the latest epidemiological findings of viral meningitis among the Brazilian regions in the last decade (2012-2021) and the impact of the COVID-19 pandemic. **Methods:** Epidemiological, retrospective, descriptive study, carried out with data obtained from the Hospital Information System (SIH/SUS) and data published by the Brazilian Institute of Geography and Statistics (IBGE). From these data, the number of Hospitalizations per million people of each Brazilian region per year of the period was calculated. For a more elaborate analysis, the variables, age group of patients, gender, value of Hospital services, average length of stay and the mortality rate per year of care were considered. **Results:** In 2012, the Hospitalization rate for viral meningitis per million people was 20,70 in Brazil. The Southern region had the highest rate (24,92) and the Northern region had the lowest (9,44). The national cost per admission was R\$ 892,78, and the 1-4 years old age group was the most affected, representing 19% of the Hospitalizations. Males represented most admissions (58%), and the average duration of Hospital stay was 7,2 days with a mortality rate of 3,04. By the end of 2021, Hospitalization rate per million was 4,95, representing a 76% decrease. The Southern region continued with the highest rate, 8,26, while the Northeastern region had the lowest rate with 3,02. The average cost per admission in 2021 was R\$ 1341,33, 50% more expensive than in 2012. Moreover, the below 1 year old age group was the most affected, while the age group of 5-9 years old had the highest variation, with a decrease of 88%. The male sex remained the most affected, with 54%. Besides, the average duration of Hospital stay in 2021 was 8 days, and the national mortality rate was 5,88, 93% higher than in 2012. **Conclusion:** This study made it possible to identify the population most affected by viral meningitis in Brazil, and the changes sustained in the last decade. Moreover, the impact of the COVID-19 pandemic and its forced social isolation should be further analyzed since this study demonstrated a big decrease of the Hospitalization rate but an associated rise in mortality and length of stay.

Neuroepidemiologia

TL 1105316

EPIDEMIOLOGIC ASSESMENT OF HOSPITALIZATION RATES FOR MENINGOCOCCAL DISEASE IN BRAZIL

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Background: *Neisseria meningitidis* is an important cause of bacterial meningitis and sepsis worldwide, with twelve recognised serogroups that can lead to meningococcal infection. This disease represents a significant burden to society due to its high morbidity and mortality rates, thus it remains relevant to discuss its epidemiology. **Objective:** The aim of this study was to evaluate the annual variation of Hospitalization rates (HR) for meningococcal disease by age, in Brazil, from 2011 to 2020. **Methods:** Data were obtained from the Hospital Information System (SIH/SUS) and from the Brazilian Institute of Geography and Statistics (IBGE), both of which are governmental datasets fed with health reports and demographic information, respectively. Using the number of Hospitalizations and deaths due to meningococcal infection as well as population projections, it was possible to estimate the yearly Hospitalization rates for each age group. A temporal analysis was then performed using a joinpoint regression model, by means of which it was obtained the Annual Percent Changes (APC) with a 95% confidence interval (CI). **Results:** In the referred period, a total of 16,984 Hospitalizations for meningococcal disease were registered in the country. In 2011, Brazil presented a Hospitalization rate of 18,8 per one million inhabitants. Regarding the general population, the HR presented a decreasing trend the following years, with an APC of -16,8 (CI95%=-19,6;-13,8). Interestingly, if no inflection point is established in 2015, it is observed a more accentuated decline from 2011 to 2015 than from 2015 to 2020. Hospitalization rates have also been observed to follow a lowering tendency for each age group analysed. The greatest Annual Percent Change was found for the age range from 5 to 19 years old, with an APC of -21,29 (CI95%=-25,6;-16,5). In contrast, the age group of 60 or older displayed the smallest APC (APC=-9,63; CI95%=-12,2;-7,0). The age group from 0 to 4 years old held the highest Hospitalization rates throughout the period, with a HR of 12,86 per million at the end of the observed period. **Conclusion:** The results of this study showed that the Hospitalization rate for meningococcal disease has been decreasing steadily in the past decade. Nevertheless, this disease remains very relevant, particularly among younger age groups, pointing to the necessity of constant improvement in regards to its control and prevention.

Neuroepidemiologia

TL 1105405

MORTALITY FROM GUILLAIN-BARRÉ SYNDROME IN BRAZIL: A TEN-YEAR RETROSPECTIVE ANALYSIS

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Guillain-Barré Syndrome (GBS) is a polyradiculoneuropathy of autoimmune etiology, considered the leading cause of acute flaccid paralysis in the world. This syndrome is characterized by ascending motor and sensory symptoms, also presenting varied clinical forms in part of the patients, such as Miller Fisher syndrome and Bickerstaff encephalitis, whose presentations include cranial nerves involvement, altered coordination, and involvement of the level of consciousness. This study aims to evaluate the mortality from Guillain-Barré Syndrome during the last decade (2011-2020) in Brazil, comparing descriptive variables: region, age group and gender. The analysis was conducted through an epidemiological, retrospective, descriptive and cross-sectional study, carried out with information released by the Mortality Information System (SIM/DATASUS) and the Brazilian Institute of Geography and Statistics (IBGE). From these data, mortality per 1 million inhabitants of each region per year was calculated and adjusted for the descriptive variables. In the decade analyzed, 1875 deaths from GBS were recorded in Brazil, of which 47.4% occurred in the Southeast region. In 2011, the country had a mortality rate of 0.7 deaths per million inhabitants for this syndrome, with the Southeast region having the highest mortality rate (1), and the North region, the lowest (0.4). In 2020, an average of 1 death per million inhabitants was observed, showing an increase of approximately 40% compared to 2011. At the end of the decade, the Midwest region had the highest mortality rate per million inhabitants (1.6), representing an increase of 149% compared to 2011, while the Northeast region had the lowest rate (0.7), corresponding to an increase of 36% compared to 2011. Regarding age group, people aged over 50 years accounted for 40% of the deaths caused by GBS. Moreover, the average annual number of deaths per million inhabitants was 0.7 for women and 1.14 for men, an average annual difference of 62.8% between the two groups, there was also a difference of 56.5% between the number of deaths accumulated by each of the genders in this period. **Conclusion:** This study provided a better understanding about the mortality of the Guillain-Barré Syndrome in Brazil, showing particularities in the profile of patients and the heterogeneity of mortality rates among different regions. However, it was not possible to identify complete data about the incidence of GBS, limiting the scope of the study.

Neuroepidemiologia

TL 1105512

PROFILE OF PATIENTS ADMITTED TO A STROKE UNIT IN NORTHEASTERN BRAZIL AND ITS QUALITY INDICATORS

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Background: Stroke is one of the leading causes of mortality and morbidity worldwide (1). To improve this scenario, it is necessary to evaluate reliable data to improve the standards of Hospital care. Ordinance No. 665, published in 2012 by the Ministry of Health, provides for the implementation of stroke units, as well as the inclusion of quality indicators to monitor their management (2). **Objective:** To describe the characteristics of patients admitted to the stroke unit during the implementation of quality indicators in the service. **Method:** We conducted a cross-sectional, observational, retrospective study that included patients admitted to the stroke unit of the Hospital da Restauração, in Recife, Pernambuco, during the period from March 2022 to the beginning of May 2022. **Results:** The studied stroke unit has 12 beds, one of which is permanently available for venous thrombolysis. All beds are monitored, there is availability of daily physiotherapy and speech therapy assessment; nurse 24h/7 days and neurologist available for evolution and reassessments, as well as full time acute treatment. In the period studied, there were 123 Hospitalizations, of which 54.5% were for ischemic stroke (n=67) and 4.9% were TIA (n=6), and the rest were for HCVA. There was a predominance of females (52.8%) and a mean age of 68 years. The proportion of patients treated with reperfusion therapy was 14.6% (n=18). The mean score on the NIH stroke scale was 12.6 (SD: \pm 8.4). All patients were evaluated by physical and speech therapy. 40.7% of the patients admitted to the stroke unit had as a reason for discharge the transfer to other sectors of the Hospital, and only 1 patient died during Hospitalization. Many other data were collected, however, as it is a pilot project for the implementation of indicators, the collection was incomplete, mainly due to the doctors and nurses team not filling out the form. **Conclusion:** This study allowed corroborating the importance of the adequate filling of quality indicators in the stroke unit by the multiprofessional team, as a validated instrument to guide the care provided to patients with cerebral ischemia.

Neuroepidemiologia

TL 1105519

PREVALENCE AND CLINICAL ASPECTS ASSOCIATED WITH COGNITIVE DECLINE IN THE ELDERLY IN THE INTERIOR OF THE AMAZON

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Background: Dementia represents a worldwide public health problem, due to the extent and severity of its functional, psychological and social manifestations, both for the elderly and their families. Advanced age, low level of schooling and socioeconomic conditions have a great impact on the cognitive decline of the elderly. **Objective:** To analyze the prevalence and clinical aspects associated with cognitive decline in the elderly in the interior of Amazonas. **Method:** This is an observational, cross-sectional population-based study, approved by the Research Ethics Committee (CAAE nº 249.098/2013) and carried out in Coari/AM. After performing the sample calculation, cluster sampling was performed. Inclusion criteria were: 60 years or older and resident in the urban area of Coari. In the data collection, a socioeconomic, demographic, activities of daily living, cognitive status, depression, health status and independence measure were applied. Data were tabulated in electronic spreadsheets and analyzed using Jamovi software. **Results:** The study was carried out with 274 elderly people, 63.1% female, with a mean of 71.6 years (SD 8.03) and a mean of 3.01 years (SD: 4.03) of study. Most had a family income of 1 to 2 minimum wages (62.9%) and did not have caregivers (90.4%). Regarding the perception of health, 55.1% reported regular health and 24.8% good health. Among the reported comorbidities, Systemic Arterial Hypertension (48.2%), heart disease (17.2%), Diabetes Mellitus (16.1%), Stroke (12.8%), dementia (5.5%) prevailed.) and cancer (2.2%). About 31.2% had mild depression on the Geriatric Depression scale, 30.2% had mild cognitive loss on the Mini Mental State Examination (MMSE), and 28.1 had moderate cognitive loss. Both obtained, respectively, an average of 5.50 points (SD: 4.57) and 23.3 points (SD: 4.62). The MMSE showed a moderate negative correlation with knowing how to read and write (Pearson's r -0.501; p <0.001). Regarding the performance in Instrumental Activities of Daily Living according to the Lawton scale, 34% have mild dependence, with an average of 24.9 (SD: 2.84). **Conclusion:** It is observed in the study that the elderly presented moderate to mild cognitive loss, mild depression and mild dependence. These results: are similar to those found in other regions of the country.

Neuroepidemiologia

TL 1105563

PREVALENCE AND CLINICAL ASPECTS OF COGNITIVE DECLINE IN INSTITUTIONALIZED ELDERLY IN THE INTERIOR OF BAHIA

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Background: Aging, although inherent to life, is correlated to cognitive decline and establishment of comorbidities. This causes problems, such as forgetfulness; disturbances in attention, concentration and reasoning; physical dysfunctions; dependency; and institutionalization. **Objective:** To assess the prevalence of cognitive decline in institutionalized elderly people in a city in the interior of Bahia. **Method:** This is a cross-sectional, observational and descriptive study, approved by the CEP, through CAAE No. 92726218.0.0000.8467 and carried out in a long-term care institution for elderly people (LTCI) in Teixeira de Freitas/BA. It included elderly people of the age of 60 years old or older living in the LTCI for at least 6 months. Elderly people with severe mental disorders were excluded. Questionnaire about sociodemographic and health aspects was applied, and the Mini Mental State Examination (MMSE) for cognitive assessment. The sample is made up of 28 elderly people, which is higher than the estimated population for a confidence level of 95% and sampling error of 10%. Data were tabulated in spreadsheets and analyzed using the Jamovi software using the Spearman test, considering $p < 0.05$. **Results:** Most of the elderly were men (60.71%); browns (50%); coming from the interior of Bahia (100%); aged between 70-79 years (39.28%), ± 76.5 years (SD: 10.6); institutionalized for at least 3 years (39.28%); without a partner (89.28%); with no or only one child (60.71%) and no schooling (57.14%). Systemic Arterial Hypertension was the most prevalent pathology (60.71%), followed by Brain Stroke sequelae (32.14%) and low visual acuity (21.42%). It was found that 89.28% of the sample showed cognitive decline. The main correlation identified concerns age, being moderately negative (Spearman's $r = -0.424$ and $p = 0.027$). The most compromised domains, respectively, were 'registration' and 'attention and calculation', with r equivalent to 0.283 ($p = 0.034$) and 0.394 ($p = 0.042$), respectively, expressing moderate correlation with cognitive decline. On the other hand, the 'memories' and 'spatial orientation' domains showed less commitment, with r values equal to 0.615 ($p < 0.001$) and 0.510 ($p = 0.07$). **Conclusion:** It was verified a degree of cognitive impairment in a significant portion of institutionalized elderly, corroborating national studies on the subject. Investigations are necessary to develop assistance resources to stimulate the affected cognitive areas.

Neuroepidemiologia

TL 1105593

MULTIPLE SCLEROSIS MORTALITY IN BRAZIL: A RETROSPECTIVE ANALYSIS FROM 2016 TO 2020

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Background: Multiple Sclerosis (MS) is an inflammatory and chronic immune-mediated disease, being the most common primary demyelinating pathology of the central nervous system. It presents a high diversity of motor and sensory clinical manifestations, with a progressive degenerative evolution, and usually affects young and middle-aged adults. **Objective:** The aim of this study was to assess MS mortality rates in Brazil over the five years 2016-2020, with a comparative analysis of the descriptive variables sex, age, and region. **Methods:** The research was carried out through an epidemiological, retrospective, descriptive, and cross-sectional study of information available at SIM/DATASUS and IBGE. Subsequently, annual mortality per million inhabitants was calculated by region of the country, with subsequent adjustment of the descriptive variables sex and age. **Results:** There were 1518 deaths from MS during the 2016-2020 five-year period, among which 922 (60.74%) were female deaths, while 596 (39.26%) were male. In addition, there was a predominance of deaths in the Southeast region, with a total of 695 (45.78%). However, in the analysis of the number of deaths per million inhabitants, the South region, with an average of 2.13 deaths/million inhab., presented the highest rate over the five years of analysis. In sequence, in descending order, are the Southeast region (1.58 deaths/million inhab.), Midwest (1.55 deaths/million inhab.), Northeast (1.03 deaths/million inhab.) and, finally, in the North (0.95 deaths/million inhab.). It is worth noting that the mortality profile has changed little over the years of work analysis. Furthermore, in the analysis of mortality by age, the age range younger than 19 years old presented the lowest number of deaths in the period. From adulthood onwards, the constant growth in mortality was remarkable until reaching the peak, in the range of 50 to 59 years, with a total of 383 deaths from MS (25.23%), followed by the age cut from 60 to 69 years, with 342 deaths (22.52%), with a subsequent decline in rates with advancing age. **Conclusion:** Given the data analyzed, it was clear that the South region had the highest mortality per million inhabitants, followed by the Southeast region. In addition, mortality from MS in women was substantially higher than mortality in men. Finally, the age group that comprises the end of adulthood and the beginning of the elderly was the one with the highest number of deaths.

Neuroepidemiologia

TL 1105678

EPIDEMIOLOGICAL PROFILE OF CONGENITAL ZIKA VIRUS SYNDROME (SCZ) IN CEARÁ BETWEEN 2015 AND 2021: A CRITICAL VIEW ON THE NOTIFICATIONS

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Background: Zika Virus is transmitted by the *Aedes aegypti* mosquito and its infection has as its main symptoms myalgia, headache, and fever. During pregnancy, contamination by the Zika virus carries a high risk of developing a congenital malformation called microcephaly, characterized by Congenital Zika Virus Syndrome (SCZ), in which a newborn (NB) is born with a head circumference smaller than expected (32 cm). **Objective:** To delineate the epidemiological profile of SCZ in Ceará between the years 2015 and 2021, identifying the risk factors of this condition. **Methods:** This is a descriptive epidemiological study that used secondary data from Tabnet, being tabulated in Microsoft Excel, according to notifications of suspected cases of SCZ (DATASUS). **Results:** In the period analyzed, 924 cases of SCZ were notified. The most affected maternal ethnicities were brown (71%) and uninformed or ignored (23.38%). The average age group of the most affected mothers was 20 to 29 years (42.1%), with the two least affected being 45 to 49 (0.32%) and under 15 years (0.65%). The gestational age at which the most significant congenital abnormality was detected was >27 weeks (3rd trimester) (66.99%). Among the congenital alterations identified between 2015 and 2021, exclusive microcephaly was the most prevalent (63.43; 79.43%), followed by microcephaly with Central Nervous System (CNS) alterations (8.86; 11.09%). Almost all cases of SCZ had no detection of concomitant neurological impairment (910; 98.4%). In the year 2016 (449 cases), there was the most intense underreporting frequency by etiology, because there is an amount of unreported, ignored and/or unknown etiology in 372 cases (82.85%). From the studied period, 2018 to 2021, the underreporting of microcephaly cases became even more exorbitant, as 94.66% fell into the unknown etiology. The percentage difference between the genders of NB with microcephaly in the analyzed period is 15.91%, with males being the most affected (55.74%). However, in 4.44% of cases, there was no information, or the gender of the NB was ignored. **Conclusion:** It is not possible to delineate precisely the epidemiological profile of the SCZ given the intense underreporting, among them: etiology and ethnicity, being the most important in the diagnostic context. There was a relevant distinction between genders, which hypothesizes that males are more exposed to the condition.

Neuroepidemiologia

TL 1105801

EPIDEMIOLOGICAL PROFILE OF ACUTE FLACID PARALYSIS IN BRAZIL: 2015-2020

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Background: Guillain-Barré syndrome (GBS) is a acute inflammatory polyneuropathy, currently considered the most frequent cause of acute flaccid paralysis (AFP) in children after polio eradication in developed countries. It is characterized by acute paralysis with areflexia and liquor albuminocytological dissociation, being it a neurological emergency. The differential diagnosis of GBS offers a wide range of possibilities, among them stands out spinal canal compression, transverse myelitis, botulism and cerebellar ataxia¹. **Objective:** To characterize the epidemiological cases profile of notified acute flaccid paralysis between in Brazil 2015 and 2020. **Methods:** The data were extracted from Notifiable Diseases Information System (SINAN) and encompassed the period from 2015 to 2020. **Results:** Between 2015 and 2020, 2711 patients with AFP were notified in Brazil, and 2018 had the higher number of notifications (520/19.18%). The Northeast region showed the highest number of cases (1047/38.62%) and the Midwest was the region with the lowest cases of AFP (171/6.3%). As for age, 122 (4.5%) were under 1 year old, 966 (35.63%) were between 1 and 4 years, 805 were between 5 and 9 years and 818 were between 10 and 14 years. No patients over fifteen years was notified. In relation to gender, the male gender had the highest prevalence with 1545 cases (56.99%). The Brown color/race exceeded the other races. None cases were classified as "Confirmed Wild Poliovirus". **Conclusion:** It was found that AFP still occurs frequently in Brazil, especially in areas with lower socioeconomic status, in male children and in black race people. There was no poliovirus cases detected.

Neuroepidemiologia

TL 1105877

TEMPORAL TRENDS OF THE HOSPITALIZATION RATE OF MENINGOCOCCAL INFECTION IN BRAZIL BETWEEN 2011 AND 2021

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Background: Meningitis is one of the most feared issues in the medical community. The infection by the *Neisseria meningitidis* may be sudden and affect mostly young and children under 5 years old. It is endemic in Brazil, with an incidence of about 1.8 cases per 100,000 inhabitants, with lethality reaching 18-20% of the infected. The clinical features are alarming and the efficiency of protective measures may be monitored through the number of Hospitalizations in the national territory, which is usually, underreported. **Objectives:** analyze the data available on the DataSUS platform about the Hospitalizations by meningitis in Brazil and provide epidemiological information about this health issue. **Methods:** A retrospective, longitudinal, observational and analytic study. The number of meningitis Hospitalizations (CID 10-A36) were obtained from the Sistema de Informações sobre Mortalidade (SIM) and the population data from IBGE. That includes the number of Hospitalizations from 2011 to 2021, stratified by region. The Hospitalisation rate (HR) was calculated per 100,000 inhabitants. The HR trends of each region during the study time were determined by segmented linear regression (joinpoint regression), with the dependent variable the natural logarithmic transformation of the HR and the year for the regression variable. Thus, the Annual Percent Changes (APCs) were obtained with a confidence interval of 95% (CI95%). **Results:** Hence, HR by meningitis in Brazil from 2011 to 2021 was reduced nationwide and in all regions, evaluated individually through data stratification. In Brazil the results of simple linear regression point to an annual reduction of 17% (APC = -17.0), with a p-value <0.1, with a confidence interval (IC) = -19.7 to -14.3, which shows the assessment was statistically significant. Concerning the rates by region, the North had APC = -6.4 until 2019, in which rates began to reduce by approximately 43% per year (APC = -43) until 2021. The Northeast had APC = -14.8. In the Central-West region, there was a decrease from 2011 to 2021, with an average APC = -14.5 in the HR. The analysis of the Southeast shows an APC = -20.8 for the same period, the highest of all Brazilian regions. Finally, in the South, there was a continuous reduction from 2011 to 2021 APC = -13.9. **Conclusion:** In summary, the Hospitalizations due to meningitis nationwide had a significant reduction. The Southeast region had the highest HR reduction with a decrease of 20.8% per year.

Neuroepidemiologia

TL 1105889

TRENDS OF THE MORTALITY RATE OF TRAUMATIC BRAIN INJURY IN BRAZIL BETWEEN 2011-2021

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Background: Traumatic Brain Injury (TBI) is a disorder caused by a sudden and external assault responsible for either anatomical lesions or functional impairment of the brain. TBI is a common cause of disability and death, with a significant impact on Brazil's public health system and its policies. **Objective:** To analyze the mortality rate due to TBI among the Brazilian population, from 2011 to 2021, through data collected from the Informatics Department of the Unified Health System (DataSUS). **Methods:** A retrospective, longitudinal, observational and analytic study. Intracranial trauma deaths (ICD-10 S06) data were obtained from the Mortality Information System (SIM) and the population data from the Brazilian Institute of Geography and Statistics (IBGE). The number of deaths from 2011 to 2021 was stratified by age group, and the mortality rate (MR) was calculated per 100,000 inhabitants. Age-specific MR trends were determined by joinpoint regression. The dependent variable was the natural logarithmic transformation of the MR, and the year of analysis was the regression variable. The Average Annual Percent Changes (AAPC) were obtained with a confidence interval of 95%. **Results:** The AAPC in mortality rate due to TBI registered in Brazil was -0.7% (95%CI: -1.2, -0.1; P<0.1), indicating a statistically significant reduction in the number of deaths due to intracranial trauma from 2011 to 2021. Analysis of age-specific trends in MR explains the apparent controversial results observed in Brazil. From 2014 to 2021, the age group 15-19 years revealed a statistically significant decline in MR, with an AAPC of -8.9% (95%CI: -11.3, -6.4; P<0.1). A different trend was observed in the age group 70-79 years, in which a statistically insignificant AAPC was recorded in both periods of analysis: an increase of 1.6% (95%CI: -1.0, 4.3; P<0.1) from 2011 to 2016, and a reduction of -2.1% (95%CI: -4.4, 0.2; P<0.1) from 2016 onwards. The age group 70-79 years showed stable rates of intracranial trauma mortality over the ten years analyzed. The age group of individuals over 80 years old indicated a statistically significant increase in the MR due to TBI only from 2011 to 2016, with an AAPC of 4.2% (95%CI: 1.0, 7.4; P<0.1). **Conclusion:** Analysis of data systems such as DataSUS is relevant in establishing policies to prevent TBI in age groups with higher incidence. Identifying its symptoms, as well as correlating its signs with imaging exams, is essential for adequate treatment decisions.

Neuroepidemiologia

TL 1105898

EPIDEMIOLOGICAL PROFILE OF STROKE IN CHILDREN AND ADOLESCENTS IN BRAZIL BETWEEN 2012 AND 2021

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Background: Stroke among pediatric patients is a rare event, but when it occurs it is severe and has the potential to leave sequelae that compromise neuropsychomotor development, leading to low schooling, reduced life expectancy, incapacity to work and burdening the health and social security systems. Still, the rarity of this event can make it difficult to suspect of, delaying diagnosis and treatment. **Objective:** To describe the epidemiological profile of stroke in children aged 0 to 19 years in Brazil. **Methods:** Observational, retrospective, cross-sectional and descriptive study carried out with data from the Hospital Morbidity System (SIH/SUS - DATASUS) from the years 2012 to 2021 considering the number of Hospitalizations due to stroke in Brazil according to place of residence. The variables analyzed were sex, race, mortality rate and year of Hospitalization. Data analysis was performed using the software Microsoft Office Excel 2016. **Results:** Based on the data collected, there was a total of 7031 Hospitalizations due to childhood and adolescent stroke in Brazil between 2012 and 2021, with São Paulo being the state with the most Hospitalizations (19.69%), followed by Minas Gerais (11%), Bahia (8.6%) and Pernambuco (8.5%). Among the states with fewer Hospitalizations are Sergipe (0.73%) and Ceará (3.88%). The epidemiological profile of Hospitalizations nationally was composed, above all, by females (50.23%), brown people (36.90%), without identification (31.67%) and white people (25.44%). In the temporal analysis of the last decade, the year with the most Hospitalizations was 2014 with 731 Hospitalizations and the year with the fewest Hospitalizations was 2020 with 637 Hospitalizations. The national mortality rate was 8.03%, with the states with the highest mortality rate being Sergipe (21.15%), Ceará (17.39%) and Tocantins (15.38%), while in São Paulo the mortality rate was 4.59%; 7.06% in Minas Gerais; 10.42% in Bahia and 8.51% in Pernambuco. **Discussion and Conclusion:** The mortality rate was higher in states with few Hospitalizations, as in the case of Ceará and Sergipe, suggesting disparities in access to health services, especially the high-complexity services responsible for handling these cases, which when associated with the limitations of therapeutic approaches for stroke within this age group points to the need for further studies to expand management of stroke in children and adolescents.

Neuroepidemiologia

TL 1105902

TEMPORAL TRENDS ANALYSIS OF THE MORTALITY RATE DUE TO CEREBRAL INFARCTION IN BRAZIL BETWEEN 2011-2021

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Background: Cerebral infarct results from ischemic necrosis caused by an interruption or reduction of the blood supply of the damaged area. According to the World Health Organization, 6 million people die annually from cerebrovascular diseases, corresponding to 11% of all deaths, becoming the second major cause of annual mortality. **Objective:** To analyse the MR due to Cerebral infarction among the Brazilian population. **Methods:** A retrospective, longitudinal, observational and analytic study. The number of brain infarction deaths (CID-10 I63) were obtained from the Sistema de Informações sobre Mortalidade (SIM) and the populational data from IBGE. That includes the number of deaths between 2011 and 2021, stratified by region. The mortality rate (MR) was calculated per 100.000 inhabitants. The MR trends of each region during the study time were determined by segmented linear regression (joinpoint regression), with the dependent variable the natural logarithmic transformation of the MR and the year for regression variable. The Annual Percent Changes (APCs) were obtained with a confidence interval of 95% (CI95%). **Results:** 31857 cases of stroke were notified in Brazil between 2011-2021, with 2019 and 2021 being the years with the highest incidence. The average annual mortality presents 1,4 events for every 100000 people. The progression of MR in the Brazilian population revealed a rising tendency: APC=4,36%/year (CI 95%= 3,4-5,3 P<0,05). The Southeast was the only one that presented a rising tendency during the whole period (APC 7,6%/year, CI 95% 5,2-10,1 p<0,05), although the progression of the deaths could only be identified between 2014 and 2021 (APC = 9,76%/year, CI 95% 7,8-11,8 p<0,001). The South region presented a significant progression until 2017 (APC 5,4% CI 95% 1,8-9,1 p=0,01), while analysing the whole period, the absolute evolution of deaths doesn't show a significant difference (p>0,05). Similar to what happened to the North region, with an increasing tendency only during 2011 and 2019 (APC 7,5%/year CI 95% 1,8-13,5 p=0,017). The trends were stationary for Center-West and Northeast during the whole period without significant difference. **Conclusion:** Two types of epidemiologic patterns in the MR for cerebral infarcts in Brazil are observed, a stationary and progressive evolution. In Center-West and Northeast regions follow the stationary pattern, while the South, Southeast and North regions follow the progressive and significant profiles.

Neuroepidemiologia

TL 1106117

DECREASING ON WAITING TIME TO START TREATMENT FOR MALIGNANT BRAIN NEOPLASM DURING THE PANDEMIC IN BRAZIL

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Background: The priority of care given to SARS-COV-2 patients during the COVID-19 pandemic has configured itself as a challenge to the care provided in oncologic treatments. Thus, studies highlight the change in the surgical burden as well as in the time pattern of brain tumor treatments. Thus, it is necessary to understand the impact of the pandemic on the waiting time for the treatment of this disease. **Objective:** Analyze whether there was a change in the waiting time for brain tumor treatment during the pandemic of COVID-19. **Method:** This is an ecological computational program-based time-series study. The Health Information System (TABNET) database was used, relying on health workers to input data accurately, thus having the corresponding limitations arising from it. Two spreadsheets were generated to look for diagnosis and the treatment start date for malignant brain neoplasm in different ranges. The first is from January 2018 to December 2019, being the pre-COVID period. However, the TABNET system generated it until February of 2020 due to some treatments only being started in this period. The second spreadsheet was generated with data from January 2020 to March 2022 (during COVID). After that, a script written in Python was built to calculate the time from diagnosis to the start of the treatment, providing weighted average and standard deviation of this time for each spreadsheet. **Results:** In the pre-COVID period, on average, there was a 2.56 months interval from the diagnosis to the start of the treatment with a 4.61-month standard deviation (SD) and 7140 patients. During the pandemic, the month interval from the diagnosis to the start of the treatment was 1.58 with a SD of 2.43 months and a total of 6696 patients. This result of 1 month less wait during pandemics is truly surprising due to the tendency of postponing surgical treatments, necessary in most cases of brain neoplasm. Although our study is not capable of point the cause of this reduction we elected some possible reasons: lower demand in the health system due to fear of covid, lower number of elective surgeries providing beds for the not-elective ones, and efforts to remove immunosuppressed patients, like oncologic ones, as quickly as possible from Hospital settings. **Conclusion:** It's unclear if the reduction of the time from diagnosis to treatment has improved survival or life quality of patients. Therefore, further studies are needed to understand this complex situation of neuro-oncology during pandemics.

Neuroepidemiologia

TL 1106143

EPIDEMIOLOGICAL PROFILE OF ACUTE FLACCID PARALYSIS IN BRAZIL: 2015-2020

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Background: Guillain-Barré syndrome (GBS) is an acute inflammatory polyneuropathy, currently considered the most frequent cause of acute flaccid paralysis (AFP) in children, after the eradication of poliomyelitis in developed countries. It is characterized by acute paralysis with areflexia and albuminocytological dissociation in the CSF, being a neurological emergency. The differential diagnosis of GBS offers a wide range of possibilities, among them spinal canal compression, transverse myelitis, botulism and cerebellar ataxia. **Objective:** To characterize the epidemiological profile of cases notified in Brazil of acute flaccid paralysis. **Methods:** The data were extracted from the Notifiable Diseases Information System (SINAN) and encompassed the period from 2015 to 2020. **Results:** Between 2015 and 2020, 2711 patients with AFP were notified in Brazil, and 2018 had the higher number of notifications (520/ 19.18%). The Northeast region showed the highest number of cases (1047/38.62%) and the Midwest was the region with the lowest cases of AFP (171/6.3%). As for age, 122 (4.5%) were under 1 year old, 966 (35.63%) were between 1 and 4 years, 805 were between 5 and 9 years and 818 were between 10 and 14 years. None patient over fifteen years was notified. In relation to gender, the male gender had the highest prevalence with 1545 cases (56.99%). The Brown color/race exceeded the other races. None cases were classified as "Confirmed Wild Poliovirus". **Conclusion:** It was found that AFP still occurs frequently in Brazil, especially in regions with lower socioeconomic status, in male children and in brown people. There were no reported cases of poliovirus.

Neuroepidemiologia

TL 1106193

GENERAL NEUROLOGY OUTPATIENT CLINIC: EPIDEMIOLOGICAL PROFILE

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Background: The present study is intended to identify the profile of neurology events in a specific period of time in a general neurology outpatient clinic of the Brazilian public health system. In fact, it is a frequent question during the training of the neurologist about the prevalence of the main diseases in the consultations in the neurological area. Thus, we seek to trace this reality by surveying the various diagnoses in neurology in outpatient care, elucidating questions about the field of work itself. **Objective:** To identify the profile of care in general neurological clinics in a given period of time, elucidating the most and/or less commonly identified diseases and, consequently, the profile of work in the neurological field in the outpatient environment of the unified health system. **Method:** The neurological diagnoses were collected in a neurology outpatient clinic of the public health system in a Municipal Hospital of the health network of the city of Poços de Caldas-MG, covering the period from April 2016 to April 2017. In this way, it was possible to trace the profile of disease groups in general neurology care in the public health system. **Results:** A total of 1236 persons were evaluated from April 2016 to April 2017, 1043 male patients, corresponding to 84.38% of the patients in this outpatient clinic in this period. Such care includes: headache: 215 (17.39%), epilepsies: 180 (14.56%), dementia syndromes: 171 (13.83%), cervical-dorsal-lumbar discopathies: 140 (11.32%), neurovascular diseases: 120 (9.70%), movement disorders: 102 (8.25%), peripheral nerve diseases: 64 (5.17%), vestibular disorders: 48 (3.88%), spinal trauma spinal cord and cranioencephalic: 41 (3.31%), mood disorders: 35 (2.83%), central nervous system neoplasms: 25 (2.02%), CNS demyelinating diseases: 23 (1.86%), neuromuscular diseases: 15 (1.21%), sleep disorders: 13 (1.05%), neuro-infectious diseases: 12 (0.97%), other diagnoses: 32 (2.58%). **Conclusion:** An interesting epidemiological data can be seen with the majority of male patients in this study. It is possible to link this fact to diseases related to the occupational aspect (cervico-dorso-lumbar discopathies, spinal cord and cranioencephalic trauma). On the other hand, aspects already known in the literature are confirmed, such as the high prevalence of headaches and epilepsies, now objectively explained.

Neuroepidemiologia

TL 1106196

EPIDEMIOLOGICAL PROFILE OF NEONATES BORN WITH CONGENITAL HYDROCEPHALUS IN BRAZIL: ANALYSIS OF A DECADE

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Congenital hydrocephalus (CH) is the condition in which there's an abnormal build-up of cerebrospinal fluid in the cerebral ventricles or the subarachnoid space, resulting in possible impairment of both physical and mental development of children born with it. This study aims to describe the epidemiological profile of newborns with congenital hydrocephalus (ICD-10 Q03) in Brazil from 2011 to 2020. Data from the Brazil Live Birth Information System (Sistema de Informações sobre Nascidos Vivos/SINASC) were used. This study descriptively evidenced the prevalence of congenital hydrocephalus, gender distribution of cases, type of delivery, weight, and asphyxia scores in the first and fifth minutes of life in the period from 2011 to 2020. Prevalence analysis also included the assessment of annual percent change in the period. In parallel, independent T-tests were used to compare the group of neonates with CH and those born alive without the condition. In the period from 2011 to 2020, a prevalence of 26,52 cases per 100.000 births (7699 births with CH) was observed, with its peak incidence occurring in the year 2011 with 32,2 per 100.000 births. The Average Annual Percent Change was -3,6%, with a decrease of 18% in prevalence from 2011 to 2020. Approximately 54% of the newborns with congenital hydrocephalus were male and 88,5% were born from cesarean delivery. 74,4% of the neonates with CH presented more than 2500g at birth, while 6,7% of them presented less than 1500g. Regarding asphyxia scores, only 29% of newborns with CH presented moderate to severe asphyxia (APGAR score 0-4) at 1 minute, and 11% at five minutes from birth. In the comparison between the CH group and the neonates without the condition, the first ones presented with higher rates of moderate to severe asphyxia at both 1 minute ($p < 0,001$; 95% CI 25,2-28,2%) and 5 minutes ($p < 0,001$; 95% CI 9,5-11,9%), of cesarean delivery ($p < 0,001$; 95% CI 32,7-35,1%) and of very low weight at birth ($p < 0,001$; 4,5-6%). It was also observed that the group of newborns with CH had a higher male-to-female ratio compared to the general population ($p < 0,001$; 95% CI 7,4-17,6%). **Conclusion:** This study presented the profile of congenital hydrocephalus in Brazil over the last ten years, showing a reduction in the prevalence of the disease, along with the characteristics of the newborns affected. The comparative analysis also showed that CH neonates are more likely to present with indicators of poor prognosis.

Neuroepidemiologia

TL 1106209

INCIDENCE OF MIGRAINE AND CEPHALIC PAIN SYNDROMES IN PEDIATRIC PATIENTS IN BRAZIL

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Migraine is classified as a type of recurrent primary headache, characterized by recurrent attacks of pulsating headache, preceded by photophobia, phonophobia, nausea and/or vomiting. It is the most common acute headache in children. Although its etiology is still uncertain, the family history is positive in most patients. The aim of this study was to analyze the incidence of migraine and other pain syndromes in pediatric patients, around Brazilian regions, between January 2017 and February 2022. Quantitative data was extracted from the Ministry of Health's database (DATASUS) through the SUS Hospital Information System (SIH/SUS). It was decided to include patients aged from 10 to 14 years, in all Brazilian federations and both genders without distinction. The search was made by selecting the options provided in the system: 'Migraine and other cephalic pain syndromes', from January 2017 to February 2022. Between January 2017 and February 2022, there were 2,609 Hospitalized children. In descending order of incidence, there was the Southeast region (35.87%), Northeast (28.74%), South (22.23%), Central-West (6.89%) and North (6.24%). Among the years analyzed, 2019 stood out as the year with the highest number of cases and 2020 the year with the lowest number of cases. It was found that the Southeast region has the highest incidence of migraines and cephalic pain syndromes in childhood. Therefore, there is a clear need for preventive activities in these age groups and better preparation of health professionals with regard to a fast management in emergencies.

Neuroepidemiologia

TL 1106214

INCIDENCE OF TRANSIENT ISCHEMIC ATTACK BEFORE AND AFTER THE PANDEMIC

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COVID-19 is an infectious disease caused by SARS-CoV-2. The most important clinical manifestation is the respiratory one, although, an increase in neurological features and cerebrovascular disorders have been reported as increased risk of stroke and transient ischemic attacks (TIA). This study aims to evaluate the change in the incidence of TIA regarding to the coronavirus pandemic context experienced in Brazil. This is a cross-sectional, quantitative epidemiological study, with analysis of data collected by the SUS Department of Informatics (DATASUS). In 2020 and 2021, there was a significant reduction in cases when compared to 2018 and 2019, with an average reduction of 15%. The average number of cases in the last two years analyzed was close to 18,261 cases. The region with the highest incidence of cases over the years analyzed was the Southeast region, representing 30,829 cases, followed by the South region with a total of 20,308 cases. North and Midwest were the least incident regions, representing 6,392 and 4,492 cases, respectively. Although there is an association between COVID-19 and an increase in the risk of cerebrovascular events, such as TIA, the number of cases did not increase in the years 2020 and 2021, which may reflect both underreporting and overload of the health care system during the pandemic.

Neuroepidemiologia

TL 1106335

MORTALITY BY CEREBRAL CRYPTOCOCCOSIS IN BRAZIL: A FIVE-YEAR RETROSPECTIVE ANALYSIS

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Cryptococcosis is a fungal disease that manifests as a chronic meningoencephalitis in its neurological form, having unspecific symptoms, such as headache, fever, cranial nerves paralysis, sensory, motor and memory deficits. This disease usually occurs in immunosuppressed patients, where *Cryptococcus neoformans* is the main agent. This research aims to trace the epidemiological profile of mortality by cerebral cryptococcosis in Brazil, comparing the data from the 5 national regions from 2016 to 2020. A cross-sectional, descriptive, and retrospective study was conducted using data from the Mortality Information System (SIM/Datasus) and from the Brazilian Institute of Geography and Statistics (IBGE). We calculated the mortality rate per 10 million inhabitants in Brazil and in the five regions individually for each year analyzed. Furthermore, the profile of mortality by age and gender was also accessed. 419 deaths were recorded. The national average mortality rate per 10 million inhabitants per year was 4.01, with the North Region presenting the highest average (8.03) and the Northeast Region the lowest (2.96). In 2016, Brazil presented 3.59 deaths per 10 million inhabitants. The North Region had presented the highest average with 7.34, and the Northeast Region the lowest with 1.93. By the end of 2020, Brazil showed a mortality rate of 3.87, with a 7.8% growth compared to the beginning of the period. The Northeast region remained with the lowest mortality (2.09), with an increase of 8.22%, and the Southeast region showed the highest mortality (4.83), with an increase of 26.42%, which was the second-highest increase recorded. The most expressive increase was 40.84%, which occurred in the South Region. There was a higher number of deaths caused by cerebral cryptococcosis among men (69.45%) than women. Moreover, the 30 to 59 age group was the most affected, concentrating the majority (58.47%) of deaths. **Conclusion:** This study allowed a better understanding of the mortality by cerebral cryptococcosis in Brazil, which showed a growth trend in the country. A significant interregional discrepancy was observed, with the South and Southeast regions showing the highest growth rates. It is questioned that this disparity may be associated with several factors, such as underreporting and endemicity of the pathology, but further studies are needed to elucidate the relevance of these factors. Moreover, it was possible to analyze the age range and the most affected gender.

Neuroepidemiologia

TL 1106339

A PANORAMA OF NEUROLOGICAL EMERGENCIES DURING COVID-19 PANDEMIC IN BRAZIL

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Background: Population aging changed the demographic profile of the Brazilian population, generating an epidemiological transition. Hence, there was an increase in the incidence of neurological emergencies such as stroke, meningitis, epilepsy, and headache. COVID-19 changed those dynamics, making it necessary to understand the new panorama of neurological emergencies. **Objective:** Analyze how the panorama of neurological emergencies has changed with COVID-19, in respect to Hospitalizations, deaths, and mortality rates. **Method:** This is an ecological time-series study, which used the Health Information System database (TABNET), made available by the Department of Informatics of the Unified Health System (DATASUS). In the SUS Hospital Information System (SIH/SUS), Hospitalizations, deaths, and mortality rates of the major neurological emergencies in the country (stroke, meningitis, epilepsy, and headaches) were collected, using secondary data from the public domain, dispensing ethical appreciation. The range was from 02/2020 (beginning of COVID-19 in Brazil) to 04/2022. **Results:** For strokes, Hospitalizations peaked 10/2021, with 16951 cases and an average (avg) of 14272.85, and a linear trendline shows a slightly decreasing trend (SDT); for deaths, a peak of 2759 in 07/2021 with an avg of 2157.3, SDT; mortality rates peaked with 17.33 in 03/2021, avg of 15.06, and SDT. For meningitis, Hospitalizations peaked 03/2020, with 556 cases and an avg of 369.67, SDT; for deaths, a peak of 62 on 01/2022 with an avg of 32.74, SDT; mortality rates peaked at 12.55 on 01/2022, avg of 8.83, and slightly increasing trend (SIT). For epilepsy, Hospitalizations peaked 12/2021, with 5189 cases and an avg of 4199.70, SIT; for deaths, a peak of 153 on 06/2021 with an avg of 110.63, SIT; mortality rates peaked at 3.28 on 01/2022, avg of 2.63, SIT. For headaches, Hospitalizations peaked 02/2020, with 1000 cases and an avg of 683.40, SDT. For headaches, only Hospitalization data was available. **Conclusion:** Strokes had a SDT in all analyzed factors. For meningitis, mortality rates increased. Epilepsy increased in all factors that were analyzed. The rate of Hospitalizations due to headaches slightly decreased. These results show that meningitis was deadlier and that epilepsy cases may require more attention. A hypothesis would be that patients chose to wait longer to receive proper care due to the pandemic. More studies are necessary to better understand these dynamics.

Neuroepidemiologia

TL 1106446

EPIDEMIOLOGICAL PROFILE OF MENINGOENCEPHALIC TUBERCULOSIS IN BRAZIL: 2011 – 2021

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Background: According to the World Health Organization (WHO), approximately 10 million cases of tuberculosis (TB) are recorded worldwide every year, being it the infectious disease that kills the most young people and adults, surpassing HIV/AIDS, which is one of the main risk factors for developing extrapulmonary TB forms. In Brazil, there were approximately 69,000 new cases and 4,500 deaths of the disease in 2020¹. Tuberculous meningitis or meningoencephalic tuberculosis (MT) is a type of extrapulmonary tuberculosis that develops in the nervous system due to the dissemination of *M. tuberculosis* in the subarachnoid space, being the worst form of the disease. **Objective:** To characterize the epidemiological profile of Meningoencephalic Tuberculosis cases notified in Brazil from 2011 to 2021. **Methods:** This is a quantitative observational study of the analytical cross-sectional and retrospective type developed from information obtained from the records of the Notifiable Diseases Information System (SINAN). **Results:** Between 2011 and In 2021, there were 9,221 confirmed cases of MT in Brazil, the peak being in 2018. More cases were recorded in the state of São Paulo with 2,544 notifications (27.56%), followed by Rio Grande do Sul (12.5%) and Pernambuco (7.27%). Roraima was the only state that did not register any cases in the last 11 years. The most affected age group was between 20 and 39 years old (4,023/43.62%), under five years old kids presented an incidence of only 1.91% with 177 registered cases. About genders, there was a higher frequency in males. 248 expressed other extrapulmonary forms of TB, mainly being the pleural (64/25.8%) and miliary forms (50/2.16%). 4,735 (51.35%) of the reported patients had HIV/AIDS co-infection at the time of diagnosis. 36.13% evolved with cure, 9.45% left the treatment and 9.22% died. **Conclusion:** Finally, it was observed that the Meningoencephalic tuberculosis is more common in males, among adults and young people, and that the incidence in children under 5 years old has decreased, probably in response to good coverage of BCG vaccination at birth. There was a significant reduction in the number of deaths in relation to the previous century, but it was not possible to find data in SINAN of the frequency of sequelae, which limits the analysis of the clinical outcome of the cases.

Neuroepidemiologia

TL 1106544

CHANGES IN STROKE HOSPITALIZATION EPIDEMIOLOGY DURING PANDEMICS IN BRAZIL

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Background: According to the literature it is possible to elucate 3 main alterations caused by pandemic on stroke epidemiology: possibility of stroke as a complication of COVID-19, decreased number of admission of stroke patients and difficulties to maintain full range of stroke treatment services in periods of worsening in the pandemic scenario. These alterations were seen in most Brazilian services, but there are some particularities of the stroke epidemiology on pandemics in the country that remain in need of investigation. **Objective:** This study aims to understand epidemiology of stroke Hospitalizations in Brazil before and during pandemics and compare it with the global findings. **Methods:** This is an ecological time series study, which used the Health Information System database (TABNET), made available by the Department of Informatics of the Unified Health System (DATASUS). In the SUS Hospital Information System (SIH/SUS), information about Hospitalizations due to stroke were collected in two periods of time, the first (pre-pandemics) ranging from november 2017 to january 2020 and the second (during pandemics), from february 2020 to april 2022. The data were divided by Brazilian region and transferred for the software "Microsoft Excel" to be quantitatively analyzed. **Results:** A reduction of 9463 (3,4%) strokes was noticed when comparing the 2 years and 2 months pre-covid period with the pandemic period, confirming the global tendency of less internations. Another finding was that the average Hospitalization time decreased from 7,13 days, with a standard deviation (SD) of 0,51, in pre-covid to 6,83 days, with a SD of 0,43, currently. Before pandemics the linear trendline already shows a slightly decreasing tendency ($R^2=0,27$) that became even lower ($R^2=0,12$), but it is still decreasing in pandemics. The lowest Hospitalizations time averages (6,2 and 6,7 days) happened simultaneously to the peaks of Hospitalizations due to COVID-19 in march and april of 2020 and 2021. **Conclusion:** COVID-19 pandemics altered the epidemiology of Hospitalizations due to stroke in Brazil. The ideal post-stroke Hospitalization time is still unclear, but the reasons for this tendency of decreasing this time in pandemics can be seen as an effort to remove the patient's from Hospital as quickly as possible due to the risk of contracting COVID-19. Although, it is not clear if this tendency, benefits patients, therefore more studies are needed to clarify this point.

Neuroepidemiologia

TL 1106571

LOWER CRANIAL NERVE INVOLVEMENT IN PATIENTS WITH BASILAR INVAGINATION: A SERIES OF 65 CASES

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Background: Basilar invagination (BI) corresponds to a bony malformation that flattens the skull base over the cervical spine, displacing the odontoid process through the foramen magnum and compressing brainstem, cerebellum, and cervical spinal cord structures. Lesions in this region can imply alterations of the last cranial nerves. **Objective:** Describe the cranial nerve changes in people who have basilar invagination. **Methods:** This is a descriptive, retrospective and quantitative study. The sample was composed of 65 medical records with diagnosis of IB and information regarding the time of pre-surgical evaluation seen between 1996 to 2020. Data were collected with presurgical changes of the last 6 cranial nerves. **Results:** In 65 medical records, 35 % of cases with reduced sensitivity of the nauseous reflex, 35 % with rhinolalia, 34 % with decreased soft palate reflex, 19 % with soft palate paralysis, 15 % with nasal reflux, 07 % with prognathism and 03 % with retrognathism were evidenced. Dysphagia was also evidenced in 32 % of the cases and dysarthria in 10 % of them. As for other anatomical changes in the craniofacial region, 7 % had hypoglossal nerve damage. **Discussion:** It was found that 100% of the population studied presented some type of Orofacial and Cervical Myofunctional Disorder (OOFM), showing lesions of the VIII to the XII cranial nerves. The decrease in nausea and soft palate reflexes occurred in a significant number of cases, and in the literature it is one of the most present symptoms both before and after surgery. Nasal reflux impairment, soft palate paralysis, hypotonia, deficit in sensitivity and mobility of the orofacial muscles are also commonly described. Rhinolalia and dysarthria present, are not evidenced in the literature. There was a higher number of disorders related to phonoarticulatory organs (OFA's), and a prevalence of alterations related to dysphagia. **Conclusion:** The results suggest a relationship between IB and the lower cranial nerves involvement, being the disorders related to the phonoarticulatory organs prevalent. Thus, preoperative evaluation by a speech therapy professional and evaluation of the permanence or not of these alterations in the postoperative period is indicated.

Neuroepidemiologia

TL 1106581

CLINICAL-EPIDEMIOLOGICAL PROFILE OF PATIENTS WITH SPONTANEOUS CEREBROSPINAL FLUID FISTULA ASSOCIATED WITH IDIOPATHIC INTRACRANIAL HYPERTENSION IN A UNIVERSITY HOSPITAL

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Background: This descriptive and retrospective study aims to describe the analysis of medical records of patients enrolled in a university Hospital diagnosed with cerebrospinal fluid (CSF) fistula resulting from intracranial hypertension (ICH). **Methods:** Data from patients treated at Hospital Universitário Walter Cantídio at Ceará state, Brazil were collected. This study descriptively evidenced the clinical and epidemiological data of individuals with the studied condition. The inclusion criteria used were adult patients with a diagnosis of spontaneous nasal CSF fistula. A positive history of skull surgery or head trauma was used as an exclusion criterion to define the spontaneous fistula. The present study represents the partial results of the major research, as all the patients with the disease are yet to be accessed. **Results:** All these patients were diagnosed with the support of both magnetic resonance imaging (MRI) and computerized tomography and 2 of them were submitted to lumbar puncture to evaluate the presence of high opening pressure. Only one patient received a proper diagnosis of idiopathic intracranial hypertension from the attending physician. The mean age at diagnosis was 36,5 (ages of 26, 28, 57 and 35 at the first presentation) and all the patients were female. The main clinical manifestations of the disease were nasal CSF leak and headache, present in all four patients. Other signs and symptoms observed were paresthesia (1 patient), ear pain (1 patient), dizziness (1 patient), tinnitus (1 patient), and epistaxis (1 patient). Half of the patients had recurrent meningitis at some point in their disease, while 1 of them progressed with seizures. The four patients were submitted to surgical procedures, but only one of them evolved without relapse. The absence of symptoms was achieved only with the use of acetazolamide in the remaining three patients, with one of them requiring the setup of a ventriculoperitoneal shunt. The mean time to relapse after surgical procedure was 2 months. **Conclusion:** This study presented a partial profile of patients with spontaneous nasal CSF fistula in a Brazilian teaching Hospital, showing the demography of individuals with the disease, as well as the difficulty in treating and recognizing this condition, and a high relapse rate. The apparent success of the use of pharmacological agents in the treatment opens new possibilities for use and further research.

Neuroepidemiologia

TL 1106608

NEUROLOGICAL CAUSES OF CHILD MORTALITY IN 10 YEARS, HAS SOMETHING CHANGED?

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Background: Child mortality (CM) is a relevant indicator that analyzes the living conditions of a country because its high values are alarming, and the national panorama is precarious, both from a social and economic point of view. In Brazil, defects in the nervous system comprised 3,5% of infant deaths in the last ten years. Encephalitis and meningitis are the dominant viral infections related to the central nervous system that affect children and that, when aggravated, currently confer a high risk of mortality. **Objective:** The project aims to analyze the situation of CM resulting from neurological causes in the last 10 years, observing aspects such as the avoidability of deaths and using the TABNET tabulator for this analysis, which generates information linked to the Unified Health System (SUS). **Methods:** This is an ecological time-series study, which used the Health Information System database (TABNET), made available by the Department of Informatics of the Unified Health System (DATASUS). In the SUS Hospital Information System (SIH/SUS), information about infant (children with less than 1 year) deaths due to nervous system diseases (chapter VI of ICD-10) was collected. The period considered ranges from January/2010 to December/2020 and the data were divided by Brazilian region. All the information was transferred for the software "Microsoft Excel" to be quantitatively analyzed. This work used secondary data from the public domain, therefore dispensing ethical appreciation. **Results:** In the southeast region of Brazil, a significantly higher decrease in the number of CM related to neurological causes was observed ($R^2=0,79$) in comparison to other regions of the country, although all of them had fewer reported deaths at the end of the timeframe in comparison to the beginning (average of 35,8 less deaths). 34% of death causes were considered evitable, with 3% being reducible by immunization actions and 97% by adequate treatment and diagnosis, among these only epilepsy had an increase (50%) in the number of fatalities. **Conclusion:** This study provides insights regarding neurological causes of child mortality as well as their epidemiological rates. Although the total deaths had decreased in the chosen timeframe, the panorama appears to have changed in the later years with decreasing importance of meningitis and special attention needing to be given to epilepsy.

Neuroepidemiologia

TL 1106623

MENINGITIS PANORAMA IN 10 YEARS

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Background: Meningitis consists of an inflammation of the meninges, which are the lining and protective layers of the human brain. This condition can cause sudden high fever, severe headache, emesis and stiffness in the back of the neck. This disease can be caused by viruses, bacteria or fungi. Currently, it is possible to be immunized against it. In Brazil, meningitis is a notifiable disease, which allows the accompaniment of this pathology over the years and how it affects the country's population. **Objective:** The project aims to analyze the situation regarding deaths related to meningitis in the last 10 years, by observing aspects such as possibility of avoiding death and utilizing, for this analysis, the TABNET tabulator, responsible for generating information vinculated to SUS. **Methods:** This is an ecological time series study, which used the Health Information System database (TABNET), made available by the Department of Informatics of the Unified Health System (DATASUS). In the SUS Hospital Information System (SIH/SUS), information about meningitis was collected. Were considered the variables "year of the first symptom", deaths and evolution. The period of time ranges from January/2012 to January/2022 and the data were divided by Brazilian region, state and city. All the information was transferred for the software "Microsoft Excel" to be quantitatively analyzed. This work used secondary data of public domain, therefore dispensing ethical appreciation. **Results:** Between the years 2010 and 2022, there were 196741 confirmed and reported cases of meningitis in Brazil, with an emphasis on the southeast region, with 106063 of those. There have been fluctuations in the regional curves, reaching a peak of 21807 cases in 2012. Since then, the curve has declined ($R^2=0,71$). Considering only complete years, 2021 had the lowest number of confirmed and reported cases, which was 6223. As for the Federative Units, from 2010 to 2022, São Paulo was the state that had the highest number of confirmed and reported cases of meningitis, totaling 79747, representing 40,53% of the total in Brazil in the same period. **Conclusion:** This study provides data that reveal the decline in confirmed and reported cases of meningitis in Brazil. The highest percentage in the analyzed period was in the Southeast region (53.91%), mainly in São Paulo (40.53%). Future information must be gathered and analyzed to accompany the development of this scenario.

Neuroepidemiologia

TL 1106624

EPIDEMIOLOGICAL PROFILE OF MENINGOENCEPHALIC TUBERCULOSIS IN BRAZIL: 2011-2021

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Background: According to the World Health Organization (WHO), approximately 10 million cases of tuberculosis (TB) are registered every year in the world. Main risk factors for the development of extrapulmonary forms. In Brazil, in 2020, approximately 69,000 new cases and 4,500 deaths from the disease were reported. ¹ Tuberculous meningitis or meningoencephalic tuberculosis (MT) is a type of extrapulmonary tuberculosis that develops in the nervous system, due to the dissemination of M. tuberculosis in the subarachnoid space, being the most severe form of the disease. **Objective:** characterizing the epidemiological profile of cases of Meningocephalic Tuberculosis reported in Brazil from 2011 to 2021. **Methods:** this is an analytical, cross-sectional, retrospective, quantitative observational study developed from information obtained from the records of the Notifiable Diseases Information System (SINAN). **Results:** Between 2011 and 2021, there were 9,221 confirmed cases of MD in Brazil, the peak being in 2018. More cases were recorded in the state of São Paulo with 2,544 notifications (27.56%), followed by Rio Grande do Sul (12.5%) and Pernambuco (7.27%). Roraima was the only state to register no cases in the last 11 years. The most affected age group was from 20 to 39 years old (4,023/43.62%), children under five had an incidence of only 1.91%, with 177 registered cases. Regarding gender, there was a higher frequency in males. 248 had other forms of extrapulmonary TB, mainly the pleural (64/25.8%) and miliary (50/2.16%) forms. 4,735 (51.35%) of the reported patients had HIV/AIDS co-infection at the time of diagnosis. 36.13% evolved with cure, 9.45% with abandonment and 9.22% with death. **Conclusion:** It was observed that approximately half of the cases are concentrated in the states of São Paulo, Rio Grande do Norte and Pernambuco; Meningocephalic Tuberculosis is more common in males, among young adults and that the incidence in children under 5 years of age has decreased, possibly in response to good coverage of BCG vaccination at birth. There was a very significant reduction in deaths compared to the previous century.

Neuroepidemiologia

TL 1104831

RESEARCH OF THE RELATIONSHIP BETWEEN MOZART'S SONATA K. 448 AND THE EXTINCTION OF THE MEMORY OF FEAR TO SOUND

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Background: Memory is the process of storing and evoking information learned from previous experiences. The memory process is divided into phases. Extinction is the final phase, in which previously established information is suppressed, starting with a new learning process. Music, (especially Mozart's Sonata K. 448) has several effects on the brain, due to its neuronal plasticity. Although there are studies showing positive effects of Mozart's sonatas on short-term memory, few investigate its effect on memory extinction. **Objective:** To analyze the exposure to Mozart's Sonata K. 448 in the extinction of fear memory to sound in mice. **Methods:** Twenty C57BL/6J mice were used, divided into: G1: Mozart (n=5), G2: Ambience (n=8) and G3: Control (n=7). The G1 group was the only one exposed to music from the intrauterine life onwards, from 9:00 pm to 7:00 am, throughout the project. The other groups remained exposed only to ambient sound until the end of the experiment. On the 50th to 54th days, the animals were submitted to the Habituation of the Arena, which aims to control the interference of behaviors related to the novelty of the experimental environment during training. On the 55th day, the Aversive Training was performed only with the G1 and G2 groups. On the 56th day, the Extinction Test started for 13 consecutive days with the three groups. On the 81st day, all groups were submitted to the Recall Test. The tests were videotaped for analysis. The Extinction Test and the Recording Test were analyzed by the repeated measures and one-way ANOVA tests, respectively. Values of $p \leq 0.05$ were considered significant. **Results:** From the statistical analysis, it was shown that the fear extinction process was different between the groups over time. Regarding the Extinction Test, the Mozart group showed a significantly higher percentage of freezing than the others, especially on the first day ($p \leq 0.05$). In addition, freezing reduction was faster for this same group. Finally, in the Recall Test, the freezing behavior in the Mozart group was significantly higher than in the other groups ($p \leq 0.05$). **Conclusion:** Our results suggest that mice exposed to music - Mozart's Sonata K. 448 - had a positive effect on fear memory, since music accelerated the extinction of fear memory to sound.

Neurofisiologia Clínica

TL 1104979

DYSAUTONOMIA IN RFC1-RELATED DISORDER: CLINICAL AND NEUROPHYSIOLOGICAL EVALUATION

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Objectives: to determine the frequency and pattern of autonomic complaints in patients with RFC1-related disorder. An additional goal was to characterize and quantify the involvement of cardiac and sudomotor branches of the autonomic nervous system. **Methods:** We enrolled 16 subjects with biallelic RFC1 (AAGGG)_n expansions and 16 age and sex-matched healthy controls that underwent comprehensive clinical and neurophysiological evaluation. SCOPA-AUT score was used to assess autonomic symptoms. Electrophysiological testing included assessment of heart rate variability and quantitative sudomotor axon reflex test (QSART). Between-group comparisons were assessed using non-parametric tests. **Results:** In the patient group, there were 9 men/7 women and the median age was 60.5 years. SCOPA-AUT scores were significantly higher in the RFC1 group compared to controls. Major differences were noticed for gastrointestinal, urinary and cardiovascular subscores. In neurophysiology, there was resting tachycardia combined with abnormal responses during Valsalva maneuver and deep breathing among patients. QSART responses were also significantly reduced in the RFC1 group, especially in the lower limbs. **Conclusion:** Autonomic dysfunction is frequent, clinically relevant and involves multiple domains in RFC1-related disorder. Patients have both sympathetic and parasympathetic involvement. From a topographical perspective, this condition is characterized by a small fiber (possibly length-dependent) autonomic axonopathy.

Neurofisiologia Clínica

TL 1105335

ELECTRODIAGNOSTIC CHARACTERISTICS OF SENSORY GANGLIONOPATHIES

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Background: the term sensory ganglionopathy refers to the pathological process in which there is involvement of the sensory component, both central and peripheral. In most cases patients will have preserved strength despite sensory involvement. To recognize this pathology, associated with malignancies and other potentially treatable diseases, allows the patient to undergo specific treatments. The differentiation from polyneuropathies is important and nerve conduction studies are particularly useful, since the distribution of ganglionopathy is not length-dependent and often asymmetrical. Goals: describe the electrodiagnostic (EDX) findings on a series of patients cases with ganglionopathy, obtained in the NCS/EMG laboratory. **Methods:** retrospective study based on EDX exams from the Hospital das Clínicas of the University of São Paulo in the last 8 years. Electrophysiological criteria similar to those used by Camdessanche were applied. Sensory changes were considered length-dependent if the sural/radial ratio was below 20% bilaterally. Sensory asymmetry was considered when we observed a side-by-side difference above 50% of the sensory potential amplitude. The following data were evaluated: sex, age, etiology, presence of ataxia and characteristics obtained from EDX: sensory and motor conduction amplitudes and velocities, late responses and assessment of the blink reflex. **Results:** 27 patients were identified between November 2014 and April 2022. There was a predominance of females 77% (20). The age of the patients ranged from 11 to 86 years (mean: 53.5). About 88% of the cases (26) met Camdessanche criteria for ganglionopathy. We observed asymmetry in the sensory action potentials in 53% (14) of the cases. Six cases (23%) had a length dependent distribution according to sural/radial ratio. There was a predominance of Sjögren's syndrome in 34% (9) and undetermined etiology in 34% (9), neoplasms in 27% (7), celiac disease in 7% (2), in addition to isolated cases of other etiologies. The H reflex was not obtained in 88% (23) of the cases. The blink reflex was altered in 75% of the cases, but the test was done in only 8 cases. **Conclusion:** Our cases were compatible with Camdessanche criteria. It was also possible to observe non-length dependence, predominance of this pathology in females and motor involvement in paraneoplastic etiologies. An important finding is that sural/radial ratio may not differentiate sensory ganglionopathy from polyneuropathy in isolation.

Neurofisiologia Clínica

TL 1106600

MOTOR UNIT NUMBER ESTIMATION IN PATIENTS WITH SPINAL MUSCULAR ATROPHY USING THE CMAP SCAN TECHNIQUE

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Background: 5q-Spinal Muscular Atrophy (SMA) is one of the most prevalent neuromuscular diseases in our country, and still an important cause of lethality, due to genetic disease, in its most severe forms. From a genetic point of view, it is already known that the number of copies of the SMN2 gene drastically influences the phenotype in an inverse relationship with the severity of the disease. Several studies show the reduction of motor unit counts by different techniques. A new Motor Unit Number Estimation (MUNE) technique described in 2016 by Bostock, MScanfit (CMAP Scan MUNE), uses a mathematical model that considers the stimulus-response curve of the compound muscle action potential (CMAP) to estimate the number of motor units. Studies have shown that this technique has greater sensitivity than other conventional techniques (MUNIX and MPS), in addition to not requiring voluntary activation, facilitating its use in children or patients with marked weakness. **Objectives:** The present study aims to evaluate the usefulness of Mscanfit in patients with SMA at the Hospital das Clínicas-FMUSP neuromuscular diseases outpatient clinic compared to other techniques (MUNIX and CMAP amplitude). **Methods:** Forty-seven patients with SMA were evaluated, CMAP scan values were obtained with surface electrodes on the abductor pollicis brevis (APB) and abductor digiti minimi (ADM) muscles. MUNIX values were obtained in the same muscles, for comparison, of 40 collaborative patients for the technique. In 8 patients, the same exams were performed with one year of follow-up. **Results:** Seven patients were SMA type 1, 25 patients were SMA type 2 and 15 were SMA type 3. Mean CMAP scan MUNE values correlated inversely with disease severity, with patients with SMA type 1 having lower values while those with patients with type 3 SMA have higher MUNE values. Among the patients who were controlled at 1 year of follow-up, only 1 patient was SMA type 1, being treated with nusinersen, and the MUNE values obtained by the CMAP scan were the same after 1 year of follow-up. The remaining seven patients who underwent the technique were SMA type 2 or 3 and the values did not differ significantly between the two exams in relation to the use or not of disease-modifying therapy. **Conclusion:** CMAP Scan can be used to count motor units in patients with spinal muscular atrophy. More ongoing studies should assess its usefulness as a biomarker of disease progression and treatment response parameter.

Neurofisiologia Clínica

TL 1105167

NON 5Q SMA: A BRAZILIAN COHORT STUDY

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Spinal muscular atrophy (SMA) describes a group of hereditary motor neuron disorders, characterized by progressive muscle weakness due to the degeneration of the anterior horn cells. This process is a consequence of structural alterations of the lower motor neurons, 95% of the time caused by loss-of-function SMN1 mutations on chromosome 5. Recently, with the advance of next generation sequencing, several potentially causative genes outside the chromosome 5 have been recognized. This subgroup of wide phenotypic variability and inheritance pattern, in which proximal muscle weakness predominates, is known as non-5q spinal muscular atrophy. We sought to describe the clinical and neurogenetic spectrum of non 5q proximal SMA patients in a Brazilian cohort of patients. Cases from a neuromuscular clinic were reviewed. Patients with proximal neurogenic weakness (SMN1 testing negative) or those with clinical evaluation suggestive of motor neuronopathy were included. So far 95 patients from 48 families were identified, 45 (48%) of them, male. Eleven of the patients (12%) started symptoms at birth, 50 patients (53%) had from 1 to 30 years and 30 (32%) had more than 30 years when symptoms started. The inheritance pattern was autosomal dominant in 51 (53%), recessive in 9 (10%), X-linked in one (1%), mitochondrial in 2 (2%) and sporadic in 32 (33%). Molecular analysis is ongoing and, so far, 52 (55%) have a definitive diagnosis. The phenotype is complex in 71 patients (75%) and many associated features have been described: disautonomia, pyramidalism, ophthalmoplegia, scoliosis, artrogriposis, cognitive impairment, sensory neuropathy, ataxia, among others. Non-5q SMA is genetically diverse, and many genes will be novel. Our cohort will be fully genetically investigated using next-generation sequencing techniques with targeted panels and/or whole exome sequencing. So far, the results highlights this heterogeneity of genes and associated features.

Neurogenética

TL 1105186

CLINICAL VARIABILITY IN A LARGE GERSTMANN-STRAUSSLER-SCHINKER (P.P102L) KINDRED FROM BRAZILIAN NORTHEAST

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Gerstmann-Sträussler-Scheinker syndrome (GSS) is a rare autosomal dominant hereditary prion disease caused by a mutation at codon 102 at PRNP protein gene. The disease has high penetrance, and symptoms may vary among affected individuals. The most common presentations are ataxia, dementia, spasticity, nystagmus, and behavioral abnormalities. This study describes the phenotypic variability within kindred from Bahia state (Brazilian northeast). We used neurologic examination to map clinical symptoms and Addenbrooke's Cognitive Examination-Revised to assess cognition. We used clinical history and genetic panels to map 26 affected individuals, 7 of whom are still alive. In our cohort, we have individuals in different stages of the disease, ranging from pre-symptomatic to bedridden. We also identified 28 individuals at risk of having the mutation who declined genetic testing or could not be tested yet. The age of onset was between the 3rd and 5th decade of life. Some individuals might have started even earlier if we considered their history of neuropsychiatric symptoms. Gait abnormalities, broad base, and unsteadiness were the first signal observed in all the patients evaluated. During disease progression, ataxia and spasticity become prominent. The Babinski sign is present very early in those patients. On the other hand, cognitive symptoms occurred much later in disease evolution. All the deceased individuals presented ataxia at some moment in their disease progression, along with spasticity and loss of ambulation. Neurosensory deafness was observed in 2 out of 7 alive individuals and reported in five other deceased individuals, contrasting with other cohorts in which deafness was reported in less than 2% of individuals. MRI was normal in most patients; some presented mild cerebellar atrophy later in the disease. The causes of death were infection, complications related to neurodegeneration and immobility, and suicide. Since its first description in 1992, much has been studied in prion protein and its related diseases. Studies of natural history are important to a better understanding of disease progression. They may play an important role in knowing other neurodegenerative disorders with similar progression patterns and pathophysiological substrates. Therefore, there is still much to be investigated in this field. Genetic counseling is very important to reduce the number of affected individuals in the family.

Neurogenética

TL 1105288

LONG-TERM CARDIAC MAGNETIC RESONANCE IMAGING STUDY IN FRIEDREICH'S ATAXIA

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Background: Friedreich's ataxia (FRDA) is an autosomal recessively inherited disorder related to abnormally expanded GAA repeat in intron 1 of FXN gene (9q21.11), and characterized by cerebellar ataxia, spasticity and areflexia of lower limbs and loss of position and vibration sense. The disease is also related to cardiomyopathy in about two thirds of individuals, and heart failure is the main cause of death in these patients. There is a lack of biomarkers for clinical care and assessment in clinical trials, and previous data might suggest that cardiac magnetic resonance imaging (cMRI) might be sensitive for early damage and clinical follow-up. **Objective:** The present study aimed to assess the usefulness of cMRI as a clinical marker in FRDA. **Methods:** Eighteen healthy controls and 42 FRDA patients were included. cMRI was performed in a 3T Phillips scanner. Among patients, 17 also underwent a 5 year-interval repeat cMRI. The following imaging data were assessed: left ventricle ejection fraction (LVEF), mass and volumes, late gadolinium enhancement (LGE), extracellular volume fraction (ECV) and intracellular water-lifetime (T₂*). The FRDA rating scale (FARS) was determined to quantify neurological decline. An ANCOVA test, using age, sex and follow-up time as covariates, was performed between groups, and also a Pearson correlation coefficient to determine relationship between clinical and imaging data. **Results:** The FRDA group had increased LV volumes and thickness, ECV and T₂* with preserved LVEF, when compared to controls in cross-sectional analyses. Nine patients (21,9%) had epicardial LGE. FARS scores did correlate LV volumes. Follow-up analyses evinced an increase of the LV diastolic volume and decrease of the LV volume/mass ratio, but no significant LV decline. Also, 5 additional patients presented epicardial LGE. **Conclusion:** FRDA patients have an expansion of the myocardial interstitium and an increase in cardiomyocyte size, with increased LV volumes and thickness, and a preserved LVEF. The cMRI might be useful not only for cardiac assessment but for general clinical evaluation in FRDA since LV volumes correlate with ataxia severity. The follow-up analyses indicate a switch from a hypertrophic cardiomyopathy phenotype towards a dilated cardiomyopathy phenotype, suggesting progressive heart damage. Thus, our data suggest that cMRI might be a useful biomarker in FRDA.

Neurogenética

TL 1105371

GASTROINTESTINAL TRACT MANIFESTATIONS IN PATIENTS WITH A DIAGNOSIS OF FABRY'S DISEASE IN FOLLOW-UP CARE BY A NEUROLOGY TEAM IN CUIABÁ

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Background: Fabry disease (FD) is an inborn metabolic error linked to the X-chromosome, affecting the GLA gene, characterized by a lysosomal storage disorder of the globotriaosylceramide (Gb3). Its pathophysiology is based on the absence or dysfunction of alpha-galactosidase, with protein accumulation in a variety of cells in the body, with cytotoxic, pro-inflammatory and pro-fibrotic effects. Likewise, the gastrointestinal tract (GIT) is affected, with deposit primarily in the autonomic intestinal ganglia, generating intestinal dysmotility, and associated manifestations. It is considered a rare disease, predominantly diagnosed in men, affecting one in every 117,000 live births. **Objective:** The aim is to describe specific manifestations of the GIT, such as abdominal pain, nausea, and constipation, in patients with confirmed FD followed up at a neurology outpatient clinic. **Methods:** A systematic literature review was conducted based on the PUBMED and UPTODATE databases using keywords as "Fabry disease", "Gastrointestinal manifestations" and "Neurological Disorders" to compare data acquired from standardized GIT symptoms questionnaires that were regularly applied. **Results:** Six patients diagnosed with FD were interviewed of whom 2 were men and 4 women, aged between 37 and 66 with a mean age of 49 years, presenting a mean time of diagnosis of 7.6 years; of these 5 (74 %) were on enzyme replacement therapy (ERT) with alpha-galactosidase. It was observed that 100% of the patients had some gastrointestinal manifestation in their lives, dyspepsia being the main symptom, which remains a lifelong symptom in 34% of them with a frequency that varies from weekly to daily, appearing after main meals. In addition, 16% of the interviewed who had not yet started ERT reported nausea and vomiting, as well as abdominal cramps since childhood, lasting approximately a week, with a recurrence of every two months. Along the years, this slowly evolved to daily cramps associated with constipation at the age of 39, with a 4-day stool interval. **Conclusion:** In sum, it has been observed that the use of ERT is important to prevent progression of various manifestations, in particular GIT disorders, caused by FD that threaten patients' lives and well-being. In addition, only patients who had not been on ERT progressed to more severe symptoms compared to those that were on it.

Neurogenética

TL 1105545

RFC1-RELATED DISORDER: IN VIVO EVALUATION OF SPINAL CORD DAMAGE

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Background: RFC1-related disorder is a recently described neurodegenerative condition related to biallelic intronic expansions in RFC1, leading to CANVAS syndrome. This condition is typically characterized by a clinical triad of cerebellar ataxia, sensory neuropathy and vestibular areflexia, but also with a broad phenotypic spectrum, including dopa-responsive parkinsonism, spasmodic cough and dysautonomia. Cerebellar and encephalic MRI changes have been described in this disease, but little is known regarding spinal cord involvement. **Objectives:** The present study aims to determine spinal cord alterations in RFC1-related disorder and its clinical correlates. **Methods:** Seventeen subjects with biallelic RFC1 (AAGGG) n expansions and 11 age and sex-matched healthy controls were enrolled between 2019 and 2021. All individuals underwent multimodal MRI spinal cord acquisitions in a 3T Philips Achieva scanner and patients were evaluated including SARA scores and time from ataxia onset (disease duration) to quantify disease severity. The Spinal Cord Toolbox (SCT) was used to assess spinal cord cross-sectional area (CSA), eccentricity (ECC), spinal cord gray matter area and diffusivity parameters of fasciculus cuneatus, fasciculus gracilis and lateral corticospinal tract across C2-C5 levels. Between-group comparisons were assessed using Mann-Whitney U test, employing age and gender as covariate. A Bonferroni test correction was used to control for multiple comparisons. Correlations between MRI findings and clinical data were assessed through Spearman correlation test. **Results:** RFC1 group had significant spinal cord CSA reduction along all cervical levels, increased ECC in lower cervical levels and gray matter atrophy when compared to controls. Regarding white matter alterations, diffusivity abnormalities were found only in the dorsal columns. There was no significant correlation between clinical data and imaging parameters. **Conclusion:** RFC1-related disorder patients have prominent gray matter atrophy and white matter degeneration in cervical spine, mostly on dorsal columns, showing that spinal cord damage is a hallmark of this disease. More studies are needed to assess the usefulness of these findings as clinical biomarkers.

Neurogenética

TL 1105588

MUTATIONAL SCREENING IN BRAZILIAN PATIENTS WITH ACUTE INTERMITTENT PORPHYRIA DISEASE

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Background: Acute Intermittent Porphyria (AIP) is a rare, genetically determined neurometabolic disorder with an autosomal dominant inheritance pattern and reduced penetrance. Pathogenic variants in the HMBS gene lead to a partial deficiency of the enzyme hydroxymethylbilane synthase, also called porphobilinogen deaminase (PBGD), the third enzyme in the heme group biosynthetic pathway. Deficiency in HMBS production leads to the accumulation of heme precursors, which are toxic to cells. During puberty, the disease manifests itself and is characterized by intermittent neurovisceral attacks, including severe abdominal pain, peripheral neuropathy, and psychiatric manifestations. The combined prevalence of acute porphyrias is 5 cases per 100,000. The Brazilian epidemiology is unknown. In a study carried on 2002 it was found seven mutations and in a 2015 study it was found nine variants. **Objectives:** To evaluate the prevalence of genetic variants in the HMBS gene in a Brazilian cohort of patients admitted to a tertiary Hospital presenting with an acute neuropathy associated with abdominal pain, psychiatric manifestations, or laboratory abnormalities suggestive of AIP. **Methods:** Forty-nine consecutive probands were tested, of which thirty-four were women. Sanger sequencing from all HMBS exons was performed. Variants detected were classified according to ACMG criteria. **Results:** A pathogenic/likely pathogenic variant was found in twelve probands. Segregation analysis revealed additional seven carriers, of which three had clinical manifestation. Eight variants found according to (NM_000190.4/ENST00000652429.1) were: three missense alterations (c.346C>T probably pathogenic; c.517C>T pathogenic; c.77G>A probably pathogenic), two nonsense alterations (c.973C>T pathogenic; c.912_913insT pathogenic); two splice site alterations (c.652-1G>A pathogenic; 499-1G>A pathogenic) and one frameshift variant (c.854_855insC pathogenic). The remaining thirty-seven probands had no variants, the remaining for genes are being tested for this cohort. **Conclusion:** In this study, the molecular diagnosis was possible in 24.5% of probands tested with a clinical diagnosis of AIP. We found the most frequent abnormality (c.973C>T), a nonsense pathogenic variant observed in five unrelated individuals. We are now using a next-generation sequencing methodology to search for the remaining genes associated with acute porphyria

Neurogenética

TL 1105595

CARCINOGENIC EVALUATION OF LISDEXAMFETAMINE DIMESYLATE THROUGH BIOASSAY WITH DROSOPHILA MELANOGASTER

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Lisdexamfetamine Dimesylate is a psychotropic drug used as first-line therapy for Attention Deficit Hyperactivity Disorder. The lack of bio-safety studies on this compound demonstrates the need to verify long-term adverse effects. This study aimed to evaluate the carcinogenic effect of this amphetamine through the Epithelial Tumor Test (ETT) in *Drosophila melanogaster*. This test uses the interpretation of malignant phenotypes in the body segments of *Drosophila*, being used to study the mechanisms involved in tumor formation and the assessment of drug biosafety. ETT is used to assess the carcinogenic and anticarcinogenic effects of various chemical compounds since about 70% of cancer-causing genes have homologs in *Drosophila*, including the *wt*s gene homologous to human *LATS1*. In the present study, *D. melanogaster* larvae from the crossing of *wt*s/TM3 and *mwh*/mwh mutant strains were treated with different concentrations of Lisdexamfetamine Dimesylate (0.125; 0.25; 0.5; 1.0 and 2.0 mM). Doxorubicin was used as a positive control, and for the negative control, ultrapure water was used. The results show a significant increase ($p < 0.05$) in tumor frequencies in individuals treated with concentrations of 0.5; 1.0 and 2.0 mM of Lisdexamfetamine Dimesylate when compared to the negative control suggesting a dose-dependent carcinogenic effect in *D. melanogaster*. However, this assay contrasts with some previous studies for carcinogenic evaluation of the active ingredient d-amphetamine. Carcinogenicity studies indicated negative results upon continuous administration of this compound. Nonetheless, the authors cited various capacities of the active principle in indirectly inducing the formation of neoplasms. This is due to the substance's adverse effects, such as increased body temperature and anorexia, causing loss of adipose tissue, increasing the incidence of adenomas in the testicles, and myeloproliferative diseases. Therefore, it can be concluded that Lisdexamfetamine Dimesylate had a carcinogenic effect in vivo analysis model with *D. melanogaster*, but further investigations involving other biological models in vivo and in vitro are needed to verify drug safety in humans.

Neurogenética

TL 1105695

DISTAL HEREDITARY MOTOR NEUROPATHY: CLINICAL AND MOLECULAR CHARACTERIZATION OF A BRAZILIAN POPULATION

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Background: The distal hereditary motor neuropathy (dHMN) is a rare form of hereditary neuropathy presenting with a length-dependent motor / predominant motor neuropathy. Additional features as upper motor neuron signs (UMN), respiratory distress, vocal cord involvement may be seen, and together with the inheritance pattern are routinely used for clinical classification. **Objectives:** To describe the clinical and neurogenetic spectrum of 127 dHMN proband / familie's diagnosed in reference centre for neuromuscular disorders. **Methods:** 148 patients from 127 families were included (85 males; mean age 33.7y, range 2-78y). Cases were genetically investigated using next-generation sequencing technics – targeted panel and / or WES. WES was performed in 25 probands. Target panel with 27 genes (AAAS, AARS, BICD2, BSLC2, DCTN1, DYNC1H1, DYNC1H1, FBLN5, FBXO38, GARS, GBE1, HINT1, HSPB1, HSPB3, HSPB8, IGHMBP2, KIF1A, LAS1L, MYH14, PLEKHG5, REEP1, SETX, SLC52A1, SLC52A2, SLC52A3, TRPV4, VRK1) Sanger sequencing was performed for segregation analysis. **Results:** The inheritance pattern observed was autosomal recessive in 20, dominant in 18 and sporadic in the remaining cases. Ten cases had also UMN signs, 19 had some proximal involvement, 6 had upper limb predominance and 1 had vocal cord involvement. A class 4 or 5 variant was identified in 26% and a VUS in 20% of proband / families. BSLC2 and DNAJB2 were the most frequent cause dHMN in our cohort, another 13 genes have one or two proband. **Conclusion:** Distal hereditary motor neuropathies encompass a rare neuropathy group. Several causative genes have been identified in the Brazilian cohort, but only a few were found in more than one proband/family. The spectrum of dHMN genes differs considerably between populations. We did not found any predominant causative gene. Additional studies are needed to better determine the genetic profile of our population. It is expected that additional tests for the cases underwent the panel analysis may increase our positivity rate and may give a more accurate genetic picture from our cases.

Neurogenética

TL 1105714

BIALLELIC PATHOGENIC VARIANTS IN VACCINIA RELATED KINASE-1 GENE IS A CAUSE OF MOTOR NEURONOPATHY WITH UPPER MOTOR NEURON SIGNS

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Background: Recessive mutations in VRK1 (human vaccinia-related kinase 1) gene have been reported in a small number of patients with spinal muscular atrophy with pontocerebellar hypoplasia (SMA-PCH). More recently, a pure axonal neuropathy has been reported in association with VRK1 mutations. **Objective:** To describe clinical, neurophysiological and image findings in cohort of patients with VRK1-related disorder. **Methods:** Clinical, MRI and electrophysiological data were collected in pre-defined preform from patients with with genetic diagnosis. **Results:** Six affected patients from 5 different families were included. Four were female, and there was history of consanguinity in just one family. All cases presented in the first decade of live with difficulty walking or frequent falls, and no patient had sensory abnormalities. One patient presented with fasciculation, and all patients presented with brisk reflexes. Brain and spinal MRI was normal in all cases. Involvement of the posterior compartment of the leg was greater than the anterior compartment in four cases detected by clinical examination and confirmed by lower limb MRI. One case has a homozygous missense variant and other cases were compound heterozygous carrying one missense variant in association with a loss of function mutation. When available, analysis of parental DNA showed the mutations were on separate alleles. **Discussion:** This study expands the clinical spectrum of VRK1-related disorders, and suggests that mutations in VRK1 should be considered in patients with motor neuropathy. We propose that missense mutations in VRK1 manifest a milder phenotype compared to nonsense mutation characterized by a combination of upper and lower motor neuron signs mostly confined to the lower limbs, with normal brain development.

Neurogenética

TL 1105797

HYPOTHALAMIC STRUCTURAL ALTERATIONS IN HUNTINGTON'S DISEASE PATIENTS

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Background: Huntington's Disease (HD) is a genetically determined neurodegenerative disorder, with dominant autosomic transmission, provoked by unstable trinucleotide repeat expansion in the huntingtine gene. In addition to the classic features of involuntary movements, cognitive decline and neuropsychiatric disorders, non motor symptoms are frequently reported. Hypothalamic involvement might explain these findings. **Objective:** Assess strutral hypothalamic involvement in Huntignton's Disease patients. **Methods:** Magnetic ressonance with volumetric acquisition was performed in 35 HD patients and 35 healthy controls paired by age and sex. The hypothalamus was segmented utilizing a Deep Learning based network. Volume and texture was compared between the two groups. **Results:** There was no significant difference in hypothalamic volume between HD patients and healthy controls. However, there was significant difference between texture parameters between the two groups ($p=0,048$). **Conclusion:** We didn't detected hypothalamic atrophy in HD patients, however, there is indirect evidence of microstructural alterations that might justify the non motor features observed in Huntington's Disease patients.

Neurogenética

TL 1105837

CLINICAL AND ELECTROPHYSIOLOGICAL FEATURES OF A BRAZILIAN COHORT OF CHARCOT-MARIE-TOOTH TYPE 4C (CMT4C)

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Background: Charcot-Marie-Tooth (CMT) is a clinical and genetically heterogeneous group of inherited neuropathies with an estimated prevalence of 1 in 2500 individuals. CMT4 is a group of demyelinating neuropathy with autosomal recessive inheritance. To date, there are 11 causative genes. CMT4C is caused by pathogenic variants in SH3TC2. It usually presents in the first decade of life with distal muscle weakness, pes cavus, difficulty walking and slow nerve conduction velocities, additional features include scoliosis. **Objective:** We aim to describe the clinical and electrophysiological features of a case series of CMT4C from a tertiary Hospital. **Methods:** Clinical and electrophysiological data were retrospectively collected from clinical records. Seven patients from five different families with molecular diagnosis were included. **Results:** Four out seven were female. The most frequently presenting symptoms were frequent falls and imbalance (42,8%). In five patients symptoms started in the first decade. Other symptoms were described, as feet deformities, distal weakness, distal paresthesia, scoliosis, difficulty walking and difficulty to run. Deep tendon reflexes were absent or reduced throughout, and vibratory sensation was abnormal on the lower limb in all patients. Nerve conduction studies were characterized by patchy sensory and motor reduction of the conduction velocity with at least one nerve presenting with temporal dispersion (TD). **Conclusion:** Biallelic pathogenic variants in the gene SH3TC2 are a known cause of demyelinating neuropathy, being the most common subtype of CMT4 in several populations. TD indicates desynchronization of components CMAP due to different rates of conduction. Recently, nerve pathology from SH3TC2-knockout animal models revealed loss of internodal architecture that could ultimately cause TD. Suggests an important role of this gene in peripheral nerve maintenance, which may be a frequent finding in patients with CMT4C that may differentiate it from other CMT subtypes.

Neurogenética

TL 1106372

GENETIC COUNSELING AND EMPOWERMENT IN WOMEN WITH PATHOGENIC VARIANTS ASSOCIATED WITH DUCHENNE AND BECKER MUSCULAR DYSTROPHY

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Background: In recent years, with the emergence of next-generation sequencing and advanced therapies for Duchenne muscular dystrophy (DMD), access to genetic testing for diagnostic confirmation and detection of mutations amenable to specific therapies has been expanded in Brazil. Such programs favor genetic testing of boys with DMD and Becker muscular dystrophy (BMD) suspicion, with little attention being given to women testing. **Objectives:** to evaluate the process of genetic counseling and empowerment of genetic information for women at risk of carrying variants related to DMD/BMD in Brazil. **Methods:** a cross-sectional study was carried-out, in which an online questionnaire was applied through Google Forms with the application of the Genomic Outcome Scale (GOS) that evaluates empowerment of genetic information. The main inclusion criteria were being female and being at risk of carrying or being known to carry variants that cause DMD/BMD. **Results:** Ninety-five participants were included, with a mean age of 41.6 years. In 72.6% of the cases, genetic testing was offered, with 60% of them reporting having performed the test and of those who did not, 87.3% mentioned they would have liked to perform it. 63.2% said they received genetic counseling and 68% decided not to have children after being diagnosed as a carrier. The mean GOS score was 20.03 (4.1) points. There was a difference in the GOS regarding schooling ($p=0.021$), with greater empowerment of women with complete higher education compared to those with complete elementary school or lower education ($p=0.017$). Per capita family income correlated with GOS ($R=0.338$, $p=0.008$), with higher income being associated with greater empowerment, as well as women who participated in patient associations had higher GOS scores (2.72 points, 95% CI -0.90 to 4.55, $p=0.004$). **Conclusion:** Our research found that most of the women interviewed have genetic empowerment to deal with the diagnosis of their children; however, they do not have it to deal with the prevention and care of dystrophies in themselves. It is necessary to instruct and refer these women about the manifestation of DMDs in their bodies, in order to prevent complications, and so that they can obtain a better quality of life.

Neurogenética

TL 1106575

IMPACT OF RELIGIOSITY ON THE HEALTH OF CHILDREN WITH DIFFERENT LEVELS OF ASD IN AN ASSOCIATION FROM PIAUÍ

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Background: Spirituality and religiosity interfere with the health-disease process and patient longevity. In general, the impact is positive and associated with protection against diseases and better quality of life (MISHRA, 2017). Spirituality, which may or may not involve religious practices and groups, shows great potential for coping with stressful experiences (MARTÍNEZ, 2014). As an example, spirituality can be used as an ally in the traditional treatment of epilepsy and other neurological disorders, such as autistic spectrum disorder (ASD) (VANCINI, 2016). Thus, it is essential to establish the impact of spirituality and religiosity, also, on the quality of life and development of patients with ASD. **Objectives:** to correlate the impact that religiosity causes on the health of children of different levels with ASD. **Methods:** Cross-sectional, analytical study with a qualitative approach. The sample is composed of 42 parents whose children have been diagnosed with some degree of ASD and who attend the Associação de Pais e Amigos dos Excepcionais (APAE) in Teresina. The research instruments chosen were the Duke Religiosity Scale (DUREL) and the Childhood Autism Rating Scale (CARS). DUREL (HGK), contains 5 questions that capture 3 of the dimensions of religiosity that are most closely related to health outcomes: organizational, non-organizational, and intrinsic religiosity, in which the first two are related to indicators of physical and mental health and social support. The CARS is done through 15 questions related to different aspects of the child's life, so that it is possible to diagnose and define the degree of his or her ASD. After obtaining the data from the questionnaire, the data were filled in a Microsoft Excel spreadsheet and later analyzed using Spearman software. **Results:** The grouped data were analyzed using Spearman's correlation tests. The correlation coefficients of the CARS with the DUREL scale were: RO and RNO (-0.043; $p = 0.788$) and RI (0.169; $p = 0.314$). Therefore, there was no significant association between the autism scale score and intrinsic religiosity nor with organizational and non-organizational religiosity. **Conclusion:** The study showed no association between the two variables analyzed, evidencing the complexity of the factors linked to the psychosocial and familial course of Autism.

Neurogenética

TL 1104939

PROFESSIONAL TRAINING PROGRAM FOR PEOPLE WITH MULTIPLE SCLEROSIS

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Background: Multiple Sclerosis (MS) is an autoimmune disease that affects the nervous system of people of working age, with an impact on employability. **Objectives:** To present a professional skills development program for people with MS. **Methods:** The free program was developed through a partnership between two Brazilian entities, one focused on the rehabilitation of people with MS and other focused on training and promoting professional development. Seven remote meetings were organized and publicized throughout the national territory, which addressed the topics of emotional intelligence, marketing, entrepreneurship, finance, business idea and company formalization. All those with MS, as well as their relatives and caregivers, who expressed interest, were included. **Results:** 755 registrations were obtained from people with MS, residing in 22 Brazilian states. Of these, 418 (55.36%) regularly participated in the meetings, completed the program and received a Certificate of Participation. All received didactic material and had a technical team for the application of classes, lives and calls. The Training Program promoted entrepreneurship, increased self-knowledge, enabled a greater network of contacts, awakened competitiveness in business and guided financial credit. **Conclusion:** The professional skills development program for people with MS is a pioneer in Brazil and was successfully implemented as a result of the partnership. Participants acquired knowledge both to optimize their preserved skills and to develop new ones for the professional market.

Neuroimunologia

TL 1105001

ANTI-LGI1 ENCEPHALITIS: A CASE SERIES FROM BRAZIL

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Background: Anti-LGI1 antibody causes an autoimmune encephalitis characterized by seizures of variable types, cognitive impairment, limbic encephalitis, and rarely Morvan syndrome. Faciobrachial dystonic seizures (FBDS) are the hallmark of the disease, occurring in 47% of cases. Anti-LGI1 encephalitis is more common in men, is rarely paraneoplastic, and responds well to steroids. Data from developing countries are lacking. **Objective:** To describe epidemiologic and clinical features of a case series of anti-LGI1 encephalitis. **Methods:** We reviewed charts of patients with anti-LGI1 antibodies from BRAIN (Brazilian Autoimmune Encephalitis Network) database from 2017 to 2022. Information on clinical, epidemiological, and treatment features was compiled. **Results:** Of the 130 patients with AIE, we found 13 (10%) patients with anti-LGI1 antibodies. Most were female ($n=9$, 65%), with a mean age of 61.9 ± 13.2 years. Two patients had myasthenia gravis (MG), thymoma, and developed Morvan syndrome with anti-CASPR2 antibodies; one had breast cancer. All the patients met the criteria for possible autoimmune encephalitis, with 2 patients with criteria for limbic encephalitis (15%). The most frequent symptoms were behavioural changes ($n=13$, 100%), epilepsy (76%, $n=10$), memory disturbances (69%, $n=9$), impaired level of consciousness (38%, $n=5$), language disturbances (23%, $n=3$) and autonomic stability (15%, $n=2$). Reported abnormal facial movements were: FBDS ($n=3$, 23%), and other types in 4 (30%) including tremor, myokymia, and dyskinesia. We observed variable taxonomy in describing Morvan Syndrome and abnormal facial movements. Pleocytosis was found in 3 (23%) patients and abnormal EEG findings in 12 (92%). Approximately 92% had abnormal MRI, and of those 53% ($n=7$) with mesial temporal T2/FLAIR hyperintensities – 4 of them with this finding happened bilaterally. **Conclusion:** We report variable findings amongst Brazilian patients with anti-LGI1 encephalitis: they were more commonly female, older, and with a higher frequency of abnormal CSF when compared to prior series. We found limbic encephalitis phenotype in only 15%, and prior reports indicate 90%. Patients with Morvan syndrome and thymoma may harbour anti-LGI1 antibodies. Although our data need to be confirmed in larger Brazilian cohorts, our findings may indicate the occurrence of genetic variability in the disease. Nevertheless, our results indicate the need for neurology training in recognizing anti-LGI1 encephalitis.

Neuroimunologia

TL 1105040

SAFETY AND EFFICACY OF INEBILIZUMAB IN NMOSD OVER A MEAN TREATMENT DURATION OF 3.2 YEARS: END OF STUDY DATA FROM THE N-MOMENTUM TRIAL

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Background: Inebilizumab is approved in the USA and Japan for aquaporin 4 immunoglobulin (Ig)G seropositive neuromyelitis optica spectrum disorder (NMOSD). **Objective:** To report final safety and efficacy data from the N-MOMentum trial of inebilizumab in NMOSD. **Methods:** Participants with NMOSD (aged 18+, EDSS score of ≥ 8 , recent history of attacks) were randomized 3:1 to inebilizumab or placebo monotherapy for 28 weeks or up to attack occurrence; the randomized controlled period (RCP). Primary outcome was time to adjudicated attack. Participants could then enter the inebilizumab open label period (OLP). Final study data are presented, including attack risk and safety outcomes. **Results:** Of the 230 participants randomized and dosed, 216 (93.9%) entered and 174 (80.6%) completed the OLP. In the RCP, 87.0% were attack free with inebilizumab and 59.9% with placebo (72.8% risk reduction, $p < 0.001$). In the OLP, 87.7% were attack-free in those continuing inebilizumab and 83.4% in those switched from placebo. Regardless of randomization, 225 participants received inebilizumab. Mean (SD) treatment duration was 3.2 (1.4) years; 36.8% were treated for > 4 years (maximum of 5.5 years). Total exposure was 730.36 person-years (py) with an annualized attack rate of 0.092; 40/63 (63.5%) attacks occurred in the first year. Treatment-emergent adverse events (AE) were reported by 89 (39.6%) participants, most frequently urinary tract infection (26.2%), nasopharyngitis (20.9%) and arthralgia (17.3%). Infusion-related reactions with inebilizumab occurred in 28 (12.9%) participants (rate per 100-py: whole study, 11.1; RCP, 37.6). The rate (95% confidence interval) of infections per 100-py did not increase with continued treatment: year 1, 116.3 (102.4-131.6); year 2, 68.1 (57.2-80.6); year 3, 61.9 (50.3-75.5); year 4, 55.1 (41.7-71.4). 105 participants had transient low IgG (< 700 mg/dL) during treatment, but no correlations were found between the worst IgG, IgM or IgA levels recorded and the occurrence of any infection or an infection grade 3 (Fisher exact test, all $p > 0.05$). Three trial participants died: one from complications of NMOSD attack, one from a CNS event of unclear etiology and one due to COVID-19, after 9, 224 and 1225 days of inebilizumab treatment, respectively. **Conclusion:** During the 5.5 years of N-MOMentum, the risk of attack in participants receiving inebilizumab remained low with no evidence of unexpected serious adverse events, including serious infection.

Neuroimunologia

TL 1105112

LOWER GLOBUS PALLIDUS VOLUME IN MULTIPLE SCLEROSIS PATIENTS ASSOCIATED WITH DISEASE DURATION: A POSSIBLE DISTINCT NEURODEGENERATIVE PROCESS

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Background: Multiple Sclerosis (MS) is a chronic inflammatory disease of the central nervous system, characterized essentially by white matter lesions, or plaques, and acute relapses with focal neurological deficits. However, a chronic disease course with neurological and radiological deterioration has been described with limited biomarkers and predictors. **Objective:** The present study aimed to evaluate volumetric analyses of magnetic resonance imaging in multiple sclerosis (MS) patients with the purpose to establish associations or predictors of poor outcome/severity of this disease in a reference center in Pará state. **Methods:** a descriptive and transversal study with patients' records evaluation and usage of imaging of magnetic resonance to generate volumetric data in VolBrain platform. Thirteen patients with MS were evaluated and fifteen imaging of healthy control subjects paired by age were obtained from online databank. **Results:** the brain volume and other diverse structures, in cm³, showed significant reduced values in MS patients compared to healthy controls, with relative preservation of cortical frontal structures and normal appearing white matter. None volumetric variable correlated with age, age at diagnosis and disability (EDSS). The Globus Pallidus volume was the only structure that showed strong correlation and significant association in regression analyses with disease duration, showing the dependency between these two variables. **Conclusion:** this study concludes that, just as reported in scientific literature, MS patients showed diminished volume in diverse brain structures, suggesting a neurodegenerative process with relative sparing of frontal and normal appearing white matter that occurs independently of disability. Yet, the Globus Pallidus volume, in isolation, what diverges from scientific literature, could be a potential biomarker of disease progression, though, more studies are necessary to establish better relationship between these variables.

Neuroimunologia

TL 1105269

NEUROFILAMENT LIGHT CHAIN LEVELS DURING RELAPSING PHASES OF MULTIPLE SCLEROSIS AND NEUROMYELITIS OPTICA SPECTRUM DISORDER

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Background: Multiple Sclerosis (MS) and Neuromyelitis Optica Spectrum Disorders (NMOSD) are autoimmune diseases that present different morbidity. NMOSD patients reach higher levels of EDSS and lethality rate in a shorter interval of time when comparing with MS, due to more aggressive relapsing. Brazilian population is characterized by high miscegenation and ethnicity is also associated to disease severity. Serum Neurofilament light chain (NfL) level is a noninvasive tool to measure neurodegeneration. **Objective:** Measuring Serum Neurofilament Light Chain level during relapsing phases of MS and NMOSD patients. **Methods:** We include five of 60 NMOSD patients, nine of 250 MS patients during relapsing phase and 24 healthy control matched by age. Plasma samples were underwent by single molecule array (SIMOA) to investigate NfL level according to the manufacturer's instructions, with limit of detection 0.05 pg/mL. **Results:** Among NMOSD patients, all were female with mean age of 44.4 years, and among MS patients six were female with mean age of 42.8 years. Healthy control mean age was 40.5 years. NfL level MS patients was 26.99 pg/mL, NfL level in NMOSD patients was 244.64 pg/mL and in healthy control was 6.77 pg/mL. **Conclusion:** Significant difference between MS and NMOSD NfL levels ($p < 0.01$) seem to be in according to more severity pathophysiology of NMOSD and reinforce the necessity of aggressive treatment to prevent and treat autoimmune diseases associated to neurodegenerative process. Yet, our NfL results in Brazilian NMOSD patients can possibly be associated to the population ethnicity and need to be repeated in a larger number of patients and population background.

Neuroimunologia

TL 1105297

BRAZILIAN AUTOIMMUNE ENCEPHALITIS NETWORK (BRAIN) STUDY: ANTIBODY PROFILE, CLINICAL CHARACTERISTICS FROM A NATIONAL WIDE NETWORK

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Background: Autoimmune encephalitis (AIE) is a group of inflammatory diseases characterized by prominent neuropsychiatric symptoms associated with neuroglial. Most common AIE is anti-NMDAR encephalitis, followed by anti-IgI1 and anti-CASPR2. Data on clinical features from developing countries are lacking. **Objectives:** To describe the clinical and epidemiological profile of AIE in the Brazilian population and to verify the sensibility of the criteria methods Brazilian Autoimmune Encephalitis Network (BrAIN) is a prospective study on AIE involving 17 centers from all Brazilian regions, in partnership with the Medical University of Vienna. Inclusion criteria are possible AIE according to Graus and agreement with informed consent. Demographic data, clinical presentation, presence of neoplasia, and treatment were compiled by the attending physician using REDCap. CSF analysis, EEG and brain MRI were also recorded. CSF and serum samples were tested using TBA for intraneuronal and surface antigens, followed by CBA or immunoblot. **Results:** We included 547 patients and tested 512 for ANeA, which 402 (78.5%) fulfilled the probable AIE criteria and 103 (25.6%) were confirmed as seropositive AIE. The following ANeA were found anti-NMDAR (n=53, 51%), anti-MOG (n=12, 11.5%), anti-GAD (n=10, 9.6%), anti-LGI1 (n=7, n=6.7%), anti-Caspr2 (n=4, 3.8%), anti-AMPA (n=4, 3.8%), anti-GABA-BR (n=3, 2.9%), anti-glyR (n=3, 2.9%). In pediatric population, it was detected only two antibodies: 21 anti-NMDAR (75%) and 7 anti-MOG (25%). Sensitivity of clinical criteria for possible AIE was 94%, and Specificity was 26%; Negative predictive value was 95%. Predictable variables of AIE in pediatric population were decreased level of consciousness (p=0.042), chorea (p=0.008), and dystonia (p=0.018) and in adults, movement (p=0.016) and memory impairment (p=0.042). **Conclusion:** Frequency of antibodies in AIE vary among populations, and we found unexpected high rates of anti-glycine and anti-MOG. In our population sensitivity for AIE clinical criteria is low (26%) with high negative predictive value. Clinical variables that could help better predict AIE in children were decreased level of consciousness, chorea and dystonia. Among adults, movement and memory. Future scores for better AIE prediction should consider specific clinical presentation.

Neuroimunologia

TL 1105412

SERUM PRESENCE OF ANTINUCLEAR ANTIBODIES IN NMOSD PATIENTS RELATES TO AQP4-IGG REACTIVITY

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Background: Neuromyelitis optica spectrum disorder (NMOSD) is an autoimmune and demyelinating disease of the central nervous system (CNS), whose pathophysiology is related to the anti-aquaporin 4 autoantibody (AQP4-IgG). Patients with NMOSD tend to present with concurrent autoimmune diseases, such as systemic lupus erythematosus (SLE), Sjögren's syndrome (SS) and Hashimoto's thyroiditis (HT). These patients may also express autoimmunity biomarkers, such as antinuclear antibodies (ANA), even without a given diagnosis of other autoimmune diseases. Therefore, we aimed to evaluate the influence of the serum presence of ANA on clinical and prognostic aspects of NMOSD patients. **Methods:** Cross-sectional study carried out in a public outpatient clinic (Ambulatório Magalhães Neto, HUPES-UFBA) in Salvador, Brazil. Clinical and demographic data were collected using a standardized questionnaire. Laboratory and MRI results were obtained through a retrospective search of medical records. EDSS, annualized relapse rate (ARR) and progression index (EDSS/disease time) were used as prognostic parameters. **Results:** 106 patients were enrolled, of which 85 (80.2%) were female and 84 (79.2%) were afrodescendants. The mean age at disease onset was 36.5 (±13.6) years. Sixteen (15.6%) patients were diagnosed with a concomitant autoimmune disease, the most frequent being HT (6.8%) and SLE (3.8%). 41 patients had documented ANA results ANA was positive in 23 (52.3%) patients. Serum presence of ANA was related to reactivity for AQP4-Ig with a trend towards statistical significance (85.7% vs 57.1%; p=0.089). ANA positive patients had a higher frequency of concomitant autoimmune diseases, although without statistical significance (28,6 vs 14,3; p=0.406). There were no significant differences between the groups regarding age at onset (39,4 ± 8,4 vs 38,5 ± 17,0; p=0.871), sex (85,7% vs 82,1%; p=1.000), initial clinical core syndrome (p=1.000 for transverse myelitis; p=1.000 for optic neuritis), EDSS (4,5±2,4 vs 5,2±2,8; p=0.424), ARR (1,6±1,6 vs 2,0±1,8; p=0.749) or progression index (4,9±5,6 vs 3,7±5,5; p=0.938). **Conclusion:** The serum presence of ANA was not related to specific clinical syndromes at disease onset nor to a worse prognosis in patients with NMOSD. An interrelationship between ANA and AQP4-IgG was observed, ratifying the humoral autoimmunity predisposition in NMOSD patients. HT and SLE were the most prevalent diseases concurrent with NMOSD in our sample.

Neuroimunologia

TL 1105479

RADIOMICS IN THE STUDY OF SPINAL CORD INJURIES CAUSED BY MULTIPLE SCLEROSIS, MNOSD AND HTLV-I VIRUS INFECTION

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Background: Radiomics is an emerging field that uses Artificial Intelligence for medical research. Radiological imaging exams can be statistically analyzed in a way that allows extracting multiple graphical elements not visible in routine assessments and transforming them into numerical data. It is a method that extracts a large number of features from medical images using data-characterization algorithms. The main steps of radiomics analysis include image collection as well as reconstruction, segmentation of the region of interest (ROI), feature extraction as well as quantification, and establishment of the predictive as well as prognostic models. Through this technology, it is possible to analyze the heterogeneity of tissues and lesions at the cellular level, helping to distinguish pathophysiological processes in a non-invasive way. Compared to biopsy studies, in radiomics, the whole tissue or the whole lesion is available for analysis, and not just a short sample. In this way, radiomics plays an important role in daily practice, once it can distinguish diseases that eventually look alike radiologically, but hold completely different pathophysiology and treatments. In our case, the diseases under study are MS, NMOSD, and HTLV-I virus, a common infectious disease in our country. **Objectives:** Our objective is to use the technology of radiomics to analyze, in a non-invasive way, spinal cord injuries caused by Multiple Sclerosis, MNOSD and HTLV-I virus infection. Thus, it is possible to identify the structure and components of the lesions, helping to distinguish the different medical conditions and guiding to adequate treatment, as well as accompanying the structural differences during treatment or surveillance. The need for a consistent number of patients for the study brings to us a necessary inclusion of patients from different Centers in Brazil, thus comprising a multicenter study. **Methods:** This is an ongoing study in which patients enrolled are those in whom there is a spinal lesion with common characteristics for MS, NMOSD, or HTLV-1 disease and who will be submitted under radiological investigation with the aid of radiomics technology to differentiate in a non-invasive and unequivocal way these three conditions. **Conclusion:** This study will allow us to demonstrate the great potential of radiomics in the diagnosis and monitoring of neurological diseases. We invite neurologists from other centers to take place in this project.

Neuroimunologia

TL 1105494

HETEROGENEOUS PHENOTYPES OF INFLAMMATORY DEMYELINATING DISEASES AND TGF- β 1 AS POTENTIAL BIOMARKER OF DISEASE PROGRESSION

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Idiopathic inflammatory demyelinating diseases (IIDDs) are a group of Central Nervous System disorders which have white matter lesions as common feature. Neuroimaging and molecular assays led to the establishment of more discernible diagnosis in this field, such as multiple sclerosis (MS), neuromyelitis optica spectrum disorders (NMOSD) and MOG associated diseases (MOGAD), each with their own clinical course and therapeutic interventions. In this work, we aim to determine the clinical profile and disease course in a population of patients from an IIDDs reference center. To explore new tools for course evaluation, we used in vitro assays to propose potential markers. In preliminary analysis, we had 60 patients enrolled. From those, 46 patients were diagnosed with MS, being 43 with relapse-remitting (RRMS), 2 with secondary progressive disease (SPMS) and 1 with primary progressive (PPMS). 8 presented with NMOSD, 1 with optic neuritis, 1 with acute disseminated encephalomyelitis and 3 with a clinically isolated syndrome. Another patient was still with diagnosis pending. We determined 59,7% of patients presented with a multifocal disease as early as clinical onset, with sensitive (44%), pyramidal (30,4%) and brainstem (23,2%) being the most committed functional systems. And although 60% of patients got an EDSS equal or higher than 3,0 during onset, 77,8% got total remission, even if 35,2% didn't get any medical care. At time of current assessment, though, 25% percent already got to that level. From these, 6 (10% from total) were equal or higher than 6,0. Investigating which CNS molecules could be used for disease evaluation on those patients, we performed in vitro assays: astrocytes from healthy mice, cells linked to inflammation and synaptic regulation, were cultured and treated with myelin proteins. While MOG exerted little response on those cells, 24 hour treatment of cultures with Nogo-A led to defective expression of pro-synaptogenic factors. Among them, we found a nearly 30% reduction in TGF- β 1, a cytokine that in CNS exerts a neuroprotector effect, being currently considered both as therapeutic target and biomarker for an array of neurodegenerative diseases. Thus, here we identified TGF- β 1 as a player in demyelination synaptopathy, with potential for investigation as a biomarker in IIDDs. New assays are still needed to functionally relate that molecule to different stages of disease progression, helping to better define its clinical employability.

Neuroimunologia

TL 1105566

THE VARIABILITY OF SUBACUTE ONSET HTLV-1 MYELOPATHY

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HTLV-1 infects around 10-20 million people worldwide and despite its prevalence only a small fraction of the infected individuals ever manifest HTLV-1 related pathology. The most common neurological manifestation of HTLV-1 is the tropical spastic paraparesis, a chronic spastic paraparesis of insidious onset. The clinical manifestations include spasticity and/or hyperreflexia of lower extremities, urinary disturbances and lower extremity weakness. Radiologically, brain white matter lesions and mild atrophy of the spinal cord can be observed. Symptomatic treatment remains the mainstay of therapy. To report 3 cases of subacute onset myelopathy due to HTLV-1 infection. A retrospective study was performed on 3 patients with subacute onset myelopathy, positive serology for HTLV-1 and absence of other causes of acute myelopathy. The clinical history, physical examination, CSF, MRI, treatment and disclosure of the cases were all reviewed. All 3 patients were male, the age ranged between 29-41 years old. They were admitted to our Hospital with a history of paraparesis, difficulty in walking and urinary disturbances and/or constipation over the last 2-5 months. The spinal MRI showed no significant alteration in one case, one presented as longitudinally extensive myelitis with swollen and high T2 signal in the cervical and thoracic spinal cord and the other with discrete degenerative alterations in cervical and lumbar spine, in addition to small foci of signal alterations in the high thoracic region. CSF examination revealed pleocytosis in all cases range between 15-28 leukocytes, with a lymphocytic-mononuclear pattern. All three patients had a positive serology for HTLV-1, and one was positive since childhood but had no symptoms until 2 months before admission. The two patients with abnormal MRI had negative AQP-4 IgG and one negative anti-MOG. Two patients received cycles of pulse therapy with IV mPSL with only insignificant or no improvement at all. One of these patients was still submitted to 5 plasmapheresis without any improvement. These two patients maintained urinary incontinence, erectile dysfunction and constipation, and, from these, one became totally plegic in inferior limbs. The patient which did not receive the mPSL treatment, had mild improvement of the symptoms. Despite an uncommon presentation, HTLV-1 can cause subacute paraparesis and thus, represents a differential diagnosis for neuromyelitis optica and other inflammatory myelitis.

Neuroimunologia

TL 1105607

NEUROFILAMENT LIGHT CHAIN LEVELS AS BIOMARKER OF SEVERITY ACCORDING TO THE PRESENCE OF ANTI-AQP-4 IN NEUROMYELITIS SPECTRUM DISORDER PATIENTS

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Neuromyelitis Optic Spectrum Disorder (NMOSD) is a severe Central Nervous System (CNS) inflammatory and autoimmune disease, preferentially affecting the spinal cord and optic nerves. Advances in diagnostic criteria have allowed greater diagnostic sensitivity and specificity and the inclusion of patients without anti-aquaporin-4 antibody (AQP-4 ab). Neurofilament light chain (NfL) is an important biomarker of neurological damage in a wide spectrum of neurological diseases. NfL levels may contribute to evaluate disease severity. Here, we analysed samples from a cohort of 59 follow-up NMOSD patients in treatment at HUCFF Demyelinating Diseases outpatient Clinic. All these patients match the 2015 diagnostic criteria for NMOSD, and they underwent anamnesis, laboratory tests and neuroimaging. They were divided into 2 groups: anti-AQP4 positive and anti-AQP4 negative. This anti-aquaporin-4 status was achieved by CBA (Cell-Based Assay), prior to the beginning of NfL tests. To evaluate the neuroinflammation targeting NfL as biomarker, we collected blood samples by venipuncture in EDTA tubes and plasma was isolated. Furthermore, we used the NfL kit for SIMOA (Single Molecule Array), an ultrasensitive immunoassay, for detecting and quantifying this neurological biomarker in plasma. We observed that the proportion of patients with positive anti-AQP4+ (65.7%) was higher than negative ones (34,21%). Also, during relapse NMOSD, they presented an increase in NfL levels when compared to non-relapse patients and healthy controls ($p < 0.001$). Independently of relapse moment, AQP-4 ab (+) patients present significant higher level of NfL compared with AQP-4 ab (-) ($p < 0,05$). NfL high level may represent not only a biomarker of active disease, but also an important biomarker for a more severe and neurodegenerative course in AQP-4 ab (+) patients.

Neuroimunologia

TL 1105651

FACTORS AGGRAVATING QUALITY OF LIFE IN AUTOIMMUNE DISEASES OF THE CENTRAL NERVOUS SYSTEM: A COMPARATIVE STUDY BETWEEN NEUROMYELITIS OPTIC SPECTRUM DISEASES (NMOSD) AND MULTIPLE SCLEROSIS (MS)

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Background: Neuromyelitis optical spectrum diseases (NMOSD) and multiple sclerosis (MS) are autoimmune diseases of the central nervous system (CNS) that establish a chronic inflammatory and demyelinating process. The limitations caused by the active disease, or the sequelae, can negatively impact the quality of life, in addition to causing disorders such as depression and anxiety. Comparing the quality of life between the diseases is a point to be explored, as the comparative studies carried out differ both in terms of the presence of differences between groups of diseases and the severity of the factors. **Objective:** To compare the quality of life in patients with NMOSD or MS, as well as to determine aggravating factors. **Methods:** Cross-sectional, single-center study conducted with patients who comply with the 2015 International Consensus criteria for the diagnosis of NMOSD or the 2017 McDonald criteria for the diagnosis of MS. The quality of life was quantified by the EQ-5D Quality of Life Questionnaire. Depression and anxiety were measured by the Hospital Anxiety and Depression Scale (HAD). Fatigue was measured by the Modified Fatigue Impact Scale (MFIS). The degree of disability was expressed by the Expanded Disability Status Scale (EDSS). **Results:** 229 patients were enrolled, of which 69 (30.1%) were diagnosed with NMOSD and 160 (69.9%) with MS. The mean quality of life by EQ-5D was 0.59 (SD \pm 0.23). Forty-three patients (18.8%) were classified as having probable symptoms for anxiety and thirty-four (14.8%) as having probable symptoms for depression. One hundred forty-four patients (62.9%) reported symptoms compatible with fatigue on the MFIS scale. Patients with NMOSD had worse quality of life [0.51 (\pm 0.23) vs 0.62 (\pm 0.22); p = 0.001]. We did not detect any difference between anxiety symptoms [7.60 (\pm 4.60) vs 7.60 (\pm 4.60); p = 0.978], depressive symptoms [6.7 (\pm 4.90) vs 6.1 (\pm 4.10); p = 0.353] and fatigue [43.8 (\pm 17.3) vs 43.5 (\pm 19.9); p = 0.916]. In multivariate analysis, higher scores on the EDSS, MFIS, older age, and employment status were independently associated with worse quality of life. There was no association between diagnosis of NMOSD and quality of life. **Conclusion:** NMOSD and MS have similar impacts in quality of life after adjusting for disease severity. However, disease severity is clearly worse in NMOSD when compared MS, secondarily impacting quality of life.

Neuroimunologia

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FACTORS RELATED TO JC VIRUS SEROPREVALENCE AND SEROCONVERSION IN PATIENTS TREATED WITH NATALIZUMAB

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Background: Natalizumab (NTZ) is a monoclonal antibody effective in treatment of relapsing-remitting multiple sclerosis (RRMS), however high in title the antibody against the John Cunningham virus (JCV), the antibody can often limit the drug use due to the risk of developing progressive multifocal leukoencephalopathy (PML). **Objective:** This retrospective study has the objective of evaluating the relationship between sero-prevalence and epidemiological factors, previous therapy used and treatment time in the MS population studied. **Methods:** Anti-JCV levels were evaluated in 30 patients diagnosed with multiple sclerosis and followed up at a treatment center in the city of Recife, and with known serological status at the initiation of treatment, and who kept follow-up in that service. All patients studied had no symptoms suggestive of PML and were on Natalizumab monotherapy. Serum tests were performed using the enzyme-linked immunosorbent method (ELISA) at the initiation of treatment, followed by annual control tests. **Results:** The majority (73.4%), were female. The highest percentage of patients (36.6%) were over 50 years old at the time of the survey. Most patients (46%) were dark-skinned. Baseline positivity was 16.6%, and overall seroconversion was 23%. Of the baseline positive patients, 100% had been using immunosuppressants for a period greater than 2 years. In the group of patients who seroconverted during treatment, 76% had also used immunosuppressants previously. The most used immunosuppressant was interferon beta 1a (INF- β 1a) 22 or 44 μ g subcutaneously three times a week. It not possible to have a relationship between seroprevalence and increasing age, as also no relationship was identified between the time of exposure to medication and seropositivity. **Conclusion:** There was variation in seroconversion rates in the different cohorts studied, but the rate found in this study is like those found in European and American countries. There is a strong correlation between seroprevalence and previous use of immunosuppressants. None of the patients in the group developed PML. For patients with anti JC titers above 1.5 associated with infusion time longer than two years, a modification of the interval between infusions was defined.

Neuroimunologia

TL 1105792

ANTI-NMDA AUTOIMMUNE ENCEPHALITIS: DESCRIPTIVE ANALYSIS OF 75 PATIENTS FROM BRAIN DATABASE

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Background: Anti-NMDAR encephalitis is the most common type of autoimmune encephalitis (AIE), and clinical picture involves prodromic symptoms followed by a neuropsychiatric syndrome. Patients present acute behavioral changes, epilepsy, memory impairment, reduced level of consciousness movement disorders, language disturbances and autonomic instability. Pediatric population (<13 years old) usually present seizures and movement disorders, while adult patients tend to have behavioral changes. Data from the Brazilian population are lacking. **Objective:** To describe clinical and epidemiological profile from 75 anti-NMDAR encephalitis patients and to compare with prior reports. **Method:** Chart of patients with anti-NMDAR antibodies from BRAIN (Brazilian autoimmune encephalitis network) database from 2017 to 2022 were reviewed. Information on clinical, epidemiological and treatment were compiled. Patients were divided in two age groups: pediatric (younger than 13) and adult (13 or older). **Results:** Of the 130 patients with AIE, we found 75 (58%) patients with anti-NMDAR antibodies. Approximately 69% were female, mean age was 13.8 \pm 10.1 years. There were 28 (37%) pediatric patients. Thirty-five (47%) patients presented prodromes, most commonly headache (n =17, 23%), fever (n =17, 23%) and nausea/vomiting (n =10, 13%). The most reported symptoms were behavioral, psychiatric, and cognitive changes (99%; n =74), epilepsy (85%; n =64) and movement disorders (81%; n =61). Most common symptoms in adult and pediatric population were movement disorders. Pleocytosis was reported in 56%. Abnormal EEG was present in 87% (n =65) and 18% (n =12) had extreme delta brush. Thirty-two patients (43%) had abnormal brain MRI, ten of them were children. Two patients reported neoplasia, one ovarian teratoma and the other a pulmonary mass under investigation. Methylprednisolone was the most common first line treatment (n =54, 52%) followed by intravenous immunoglobulin (n =35, 47%). **Conclusion:** In Brazil, anti-NMDAR encephalitis follows the typical clinical picture, being more frequent among young females. We found a lower association with neoplasm in our population when compared to other series.

Neuroimunologia

TL 1105806

CLINICAL DIFFERENCES AMONG CHILDREN WITH SERONEGATIVE NORSE AND ANTI-NMDA ENCEPHALITIS

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Background: NORSE patients often fulfill the criteria for possible AIE, nonetheless nearly half of the cases are cryptogenic. NORSE secondary to anti-NMDAR encephalitis have different treatment options and response rates, and data comparing both diseases are scarce. **Objective:** To compare clinical and paraclinical characteristics of pediatric patients with NORSE and anti-NMDAR. **Methods:** Chart of patients > 13 years with NORSE that fulfill the AIE criteria included in BrAIN (Brazilian autoimmune encephalitis network) database from 2017 to 2022 were reviewed. Information on clinical, epidemiological and treatment data were compiled. NORSE was defined as new-onset refractory status epilepticus in patients without active epilepsy or other preexisting relevant neurological disorder, according to 2018 consensus. Seronegative NORSE were compared to a cohort of pediatric anti-NMDAR, using T-test and Chi square tests. Statistical significance was set at $p < 0.05$. **Results:** We found 19 patients with NORSE, and 5 of them that had AIE antibodies identified were excluded. The remaining 14 patients constituted the pediatric NORSE cohort. There were no differences between groups regarding female gender (NORSE: 36% x NMDA: 56%, $p = 0.23$) or age (NORSE: 6.64 ± 2.87 x NMDA: 5.62 ± 3.27 , $p = 0.3$). Prodromal symptoms were more frequent in NORSE (NORSE: 93% x NMDA: 52%, $p = 0.009$), of which the most common was fever (NORSE: 71% x NMDA: 37%, $p = 0.04$). NORSE patients presented less frequently with behavioral, psychiatric, and cognitive changes (NORSE: 50% x NMDA: 96%, $p = 0.0005$) and movement disorders (NORSE: 64% x NMDA: 93%, $p = 0.02$), like orofacial dyskinesia (NORSE: 29% x NMDA: 44%, $p = 0.035$), dystonia (NORSE: 0% x NMDA: 56%, $p = 0.0005$) and chorea (NORSE: 0% x NMDA: 37%, $p = 0.0009$). NORSE patients also showed less frequent CSF pleocytosis, defined as $> 5 \text{ cells/mm}^3$ (NORSE: 21% x NMDA: 52%, $p = 0.059$), and normal brain MRI (NORSE: 21% x NMDA: 64%, $p = 0.009$). **Conclusion:** We found that NORSE patients have more frequently prodromal symptoms. It's possible that the high frequency of orofacial dyskinesia in NORSE patients might be result of misinterpretation of focal seizures. Moreover, movement disorders, behavioral and cognitive changes were more common in anti-NMDAR, indicating that cognitive symptoms may be a clue in differentiating anti-NMDAR from seronegative NORSE.

Neuroimunologia

TL 1105817

COMPARISON OF VISUAL ACUITY RECOVERY IN 3-, 5-, AND 7-DAY SCHEDULES OF INTRAVENOUS METHYLPREDNISOLONE IN PATIENTS WITH OPTIC NEURITIS: A CASE-CONTROL STUDY

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Background: Intravenous methylprednisolone (IVMP) is the main therapy for optic neuritis to accelerate and improve final visual outcomes. However, systematic reviews demonstrated studies used different schedules of IVMP 1 g/day and the optimal duration of IVMP is unknown. **Objective:** We aimed to investigate if 3-day, 5-day, and 7-day schedules of IVMP could lead to different recovery of visual outcomes. **Methods:** We performed a case-control study in a tertiary center in São Paulo, Brazil. We selected from a registry of consecutive patients with optic neuritis, patients that received 7-day and 3-day schedules of IVMP 1 g/day. We chose controls with 5-day schedules of IVMP, matching the groups with a propensity score for significant prognostic variables in optic neuritis – age, baseline severity of disease, the presence of anti-AQP4 antibody, the time from symptom onset to IVMP, and the co-intervention with plasma exchange. Our primary outcome was the percentage of patients with incomplete recovery of the visual acuity at 6-12 months, using the Rosebaum Chart. As sensitivity analyses, we tested if the results were consistent using an estimated LogMar scale and evaluating two different timepoints: the discharge and one month after symptom onset. **Results:** We screened 107 patients for eligibility. We found 24 patients that received 7-day schedule of IVMP and 11 patients that received 3-day schedule of IVMP. After exclusion of patients due to missing of significant data in confounding variables, we matched 22 patients with 7-day with 22 patients with 5-day IVMP and 11 patients with 3-day with 11 patients with 5-day IVMP schedules. We found that the percentage of patients with incomplete visual acuity recovery, adjusted by prognostic variables, was similar at 6-12 months when we compared 3-day versus 5-day and 7-day versus 5-day schedules. Our results remained in different scale (LogMAR) and timepoints (at discharge and one month after symptom onset). **Conclusion:** Shorter IVMP 1 g/day schedules appears to be as effective as longer schedules in the recovery of visual acuity in patients with optic neuritis. We should consider 3-day instead of 5-day schedules for mild cases and 5-day instead of 7-day schedules for severe cases.

Neuroimunologia

TL 1105819

FATIGUE IN MULTIPLE SCLEROSIS PATIENTS

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Background: Multiple Sclerosis (MS) is an inflammatory demyelinating disease that affects about 2.3 million people worldwide. Fatigue is characterized by lack of physical and mental energy, and can be of primary cause as a result of the underlying demyelinating disease or of secondary cause to comorbid conditions. Among the various impairments to the quality of life of patients with MS, this is one of the most frequent and debilitating conditions. **Objective:** This study aims to evaluate the impact of fatigue on patients monitored in a reference outpatient clinic for Demyelinating Diseases. **Methods:** Cross-sectional, single-center study conducted with patients meeting the 2017 McDonald criteria for the diagnosis of MS. Fatigue was measured by the Modified Fatigue Impact Scale (MFIS), with the patient being classified with fatigue at scores > 38 and high-impact fatigue > 59. The degree of disability was expressed by the Expanded Disability Status Scale (EDSS). Quality of life was quantified by the EQ-5D Quality of Life Questionnaire. Depression and anxiety were measured by the Hospital Anxiety and Depression Scale (HAD). **Results:** 195 patients were included with a mean age of $39.9 (\pm 11.7)$, most female (71.8%). The clinical forms in the sample were relapsing-remitting (RRMS) and primarily progressive (PPMS), with the majority being RRMS (90.1%). The average MFIS in these patients was $44.8 (\pm 21.1)$. Most patients (65.1%) achieved scores for presence of fatigue, of which 43.3% had high-impact fatigue. Patients with fatigue had worse quality of life [$0.49 (\pm 0.20)$ vs $0.80 (\pm 0.17)$; $p < 0.001$], major functional disability [$3.4 (\pm 2.4)$ vs $1.4 (\pm 1.9)$; $p < 0.001$], higher number of bouts [$3.7 (\pm 4.0)$ vs $2.4 (\pm 1.8)$; $p = 0.014$], higher HAD scale scores for anxious symptoms [$9.3 (\pm 4.6)$ vs $5.4 (\pm 3.7)$; $p < 0.001$] and depressive symptoms [$7.5 (\pm 4.0)$ vs $4.0 (\pm 3.1)$; $p < 0.001$]. We detected no difference related to length of illness [$10.2 (\pm 7.5)$ vs $8.8 (\pm 6.3)$; $p = 0.201$], and age [$40.6 (\pm 11.2)$ vs $38.6 (\pm 12.4)$; $p = 0.263$]. **Conclusion:** Fatigue is a common symptom in MS patients and manifests itself mainly in those with major disease severity and more psychiatric symptoms. It is still a poorly measured condition in clinical practice, delaying access to available treatments. For being a cross-sectional study, it cannot state the primary or secondary origin of fatigue. Therefore, prospective studies are needed to clarify how fatigue and comorbid conditions are truly related in these patients.

Neuroimunologia

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COMPARATIVE ANALYSIS OF THE PROFILE OF HOSPITALIZATIONS AND DEATHS FROM MULTIPLE SCLEROSIS IN THE PRE AND INTRA PANDEMIC PERIOD IN BRAZIL

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Background: Multiple Sclerosis (MS) is the most prevalent chronic inflammatory disease of the CNS, and those with the disease require periodic specialized follow-up. The COVID-19 pandemic has had an impact on this population on access to health care and the outcome of the disease. **Objective:** Comparatively describe the profile of Hospitalizations and deaths related to MS in Brazil in the pre (Apr/2019-Jan/2020) and intra (Feb/2020-Nov/2020) pandemic period by COVID-19. **Methods:** Descriptive and retrospective study carried out from DataSUS between Apr/19 and Nov/20. The variables analyzed were number of Hospitalizations, age range, character of care, gender, race/color, and deaths. **Results:** In pre-pandemic, with a total of 3,864 Hospitalizations, there was a peak in Jan/20 (518) and the lowest value in Dec/19 (208). Of 1551 in capital cities, 1074 occurred in São Paulo. 30-39 years old led with 1180 cases and 5-9 years was the lowest (4). There were 1872 urgent and 1992 elective admissions. There were 1286 male and 2578 female Hospitalizations (2.004: 1). By color/race, there were 2374 white and 895 brown people. There were 41 deaths in the country, 7 in capitals. The 50-59 age group led (13 deaths). 25 deaths occurred in men and 16 in women (1.56: 1). By color/race, 22 deaths were in whites and 12 in browns. In intra-pandemic, with a total of 3105 Hospitalizations (19.64% less), there was a peak in Apr/20 (498). Of 1272 in capitals, 815 occurred in São Paulo. 30-39 years old led with 947 cases and 5-9 years was the lowest (2). There were 1640 urgent admissions (12.39% less) and 1465 electives (26.45% less). In Apr/20 (498 Hospitalizations), 350 were elective (70.29 %). On Sep/20, of the 190 Hospitalizations, 170 were urgent (89.47%). There were 1003 men and 2102 women notified (2.09: 1). By color/race, there were 1933 white and 724 brown people. There were 21 deaths in the country (48.78% less), 5 in capital cities. 40-49 and 50-59 years old presented 5 deaths each. 8 deaths occurred in men and 13 in women (1.625: 1). By color/race, there were 11 deaths in whites and 7 in browns. **Conclusion:** White women aged 30-39 form the population most affected by MS in Brazil in the pre-pandemic period, while in deaths, white men aged 50-59. In the intra-pandemic period, there was a reduction in Hospitalizations, but the epidemiologic profile regarding sex, race, and age remained the same. In deaths, however, white women aged 40-59 were the most affected.

Neuroimunologia

TL 1105990

CLINICAL AND EPIDEMIOLOGICAL PROFILE OF PATIENTS DIAGNOSED WITH MULTIPLE SCLEROSIS IN A SINGLE CENTER

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Background: Multiple Sclerosis (MS) is a Central Nervous System demyelinating disease with a varying clinical presentation. Some studies have statistically validated an epidemiological profile regarding prevalence between genders, age of onset, initial symptoms and clinical classification. These studies, however, applied older criteria for classification and diagnostic. There is also a common clinical perception that MS is more frequent among white people. In this context, we investigated the epidemiological profile of MS patients in a single reference center clinic of demyelinating diseases. **Objectives:** Comparatively evaluate the clinical and epidemiological profile of MS patients in a reference center clinic of demyelinating diseases. **Methods:** Three articles published on Brain were chosen for comparison. We reviewed the electronic records of patients seen or prescribed in this clinic at least once within the last year (May 17th, 2021 – May 17th, 2022), assessing the following parameters: gender – male or female; age of onset – years; skin color – white, black, brown, yellow or indigenous; initial symptoms – isolated optic neuritis (ION), isolated brainstem/cerebellum dysfunction (IBCD), isolated long tracts dysfunction (ILTD) or combined dysfunction (CD); clinical classification (Lublin's 2013 classification) – Clinically Isolated Syndrome (CIS), Relapsing-Remitting Multiple Sclerosis (RRMS), Secondary Progressive Multiple Sclerosis (SPMS) or Primary Progressive Multiple Sclerosis (PPMS). We excluded from the study cases which did not meet the McDonald's 2017 diagnostic criteria for MS, and those without defined initial symptoms. Cases were organized accordingly into Relapsing Onset (RRMS and SPMS) or Progressive Onset (PPMS), as they were in the reference studies. All data was organized and individually compared between the three articles before mentioned. **Results:** In the RRMS subgroup (46 of 54 cases) we found a 2.83: 1 F/M Ratio; a 30.24 years mean age (11.85 SD; 1,748 SE); a 26 years median age, and a comparable proportion of initial symptoms – 17.79% ION, 30.43% IBCD, 41.30% ILTD, 10.87% CD. The comparison of other subgroups was hindered by a lack of cases. The skin color of all cases pooled was: 31 brown (57,40%); 18 white (33,33%); 5 black (9,26%). **Conclusion:** Despite the small case pool, our subpopulation matches the large cohorts' epidemiological profile, except for the skin color, which could be explained due to Brazilian miscegenation.

Neuroimunologia

TL 1106158

EPIDEMIOLOGICAL, CLINICAL AND THERAPEUTIC PROFILE OF PROGRESSIVE FORMS OF MULTIPLE SCLEROSIS FOLLOWED UP AT A BASED REFERENCE CENTER IN THE STATE OF CEARÁ

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Background: Multiple Sclerosis (MS) is an immune-mediated, chronic inflammatory disease of the Central Nervous System (CNS), characterized by multifocal areas of demyelination in the brain and spinal cord, with infiltration of inflammatory cells, reactive gliosis and axonal degeneration. There is a knowledge gap in understanding the disease, particularly the progressive forms in different regions of the country, such as the Northeast. **Objective:** To report the profile of progressive MS at a reference center in Ceará. **Methods:** This observational and cross-sectional study was conducted through a review of medical records and interviews at a Reference Center for Demyelinating Diseases in the city of Fortaleza, Ceará. The population was composed of patients with the progressive forms of Multiple Sclerosis, registered in that center, whose diagnoses meet the criteria of McDonald 2017. **Results:** From 283 patients regularly followed with MS diagnosis according to the 2017 McDonald criteria, 36 (12.7%) had progressive phenotype, 20 (56%) secondary progressive and 16 (44%) primary progressive. From those 36 patients currently followed up, 25 (69,4%) were women, 19 (52,8%) were Caucasian, 18 (50%) were high educated and the majority lived in Fortaleza 25 (69,4%). The peak incidence was in the third decade of life and the peak prevalence was between the 4th and 5th decades. The median length of illness is 193 months. It is important noting that patients are many with an undiagnosed disease for years. In turn, the follow-up time in a tertiary service, when most of them had a closed diagnosis, is 61 months. Regarding disability, 27,8% already used support in the first assessment (EDSS 6.0), and in the last assessment, considering the last EDSS recorded, the percentage of patients in need of support almost doubled: 52,8%. Regarding the first disease-modifying drugs (DMD), there was a prevalence of interferons, considering that it was the most used class in the 1990s. Considering the current DMD, there is a prevalence of monoclonal antibodies (47%), with Natalizumab (25%) and Ocrelizumab (22%). In a comparative analysis, there is a drop in the use of interferons (8%). **Conclusion:** Understanding the epidemiological, clinical and therapeutic profile of MS patients with a progressive phenotype is essential to guide the propaedeutic reasoning, define conduct and restructure the dynamics of health services.

Neuroimunologia

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AUTOIMMUNE ENCEPHALITIS: HOW TO RECOGNIZE?

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Autoimmune encephalitis (AS) comprises a group of immune-mediated inflammatory diseases of the brain parenchyma, usually lasting less than 3 months. A monophasic course is common in AS by surface antibodies; a progressive course can occur in paraneoplastic encephalitis – such as paraneoplastic cerebellar degeneration (anti-Yo) –, hence the importance of screening for neoplasms. The objective is to elucidate diagnostic forms and the importance of a detailed history and physical examination in the management of these patients. After clinical suspicion, investigation with magnetic resonance imaging (MRI) of the brain and analysis of cerebrospinal fluid (CSF) is usually carried out. It is known that the presence of bilateral limbic encephalitis is the only isolated MRI finding that is sufficient for a definitive diagnosis, within the correct clinical context, even in the absence of measured antibodies. All other suggestive MRI patterns (cortical/subcortical, striatal, diencephalic, brainstem, encephalomyelitis, and meningoencephalitis) require antibody positivity. Furthermore, the electroencephalogram (EEG) provides evidence of focal or multifocal brain abnormality when MRI is negative. AS is the leading cause of refractory de novo status epilepticus (NORSE) and suggestive findings include focal deceleration/seizures and periodic lateralized discharges (Delta Brush). In addition, all patients with suspected AE require lumbar puncture (LP), unless contraindicated. In the analysis of the CSF, usually performed after MRI, the presentation of an inflammatory character may be the only abnormality found, serving as an indication for empirical immunotherapy, after ruling out infection. Common findings include: mild-to-moderate lymphocytic pleocytosis (commonly 20-200 cells, can be as high as 900 cells), hyperproteinorrea, and sometimes an elevated IgG index. These findings, in the setting of negative infectious and oncologic cytology studies, support an immune-mediated etiology, but do not differentiate AS from other immune-mediated conditions, hence the need for clinical correlation. In addition, CSF and serum antinuclear antibody (NAA) testing and monitoring of the sodium level (showing hyponatremia in certain subtypes, such as LGI-1 antibody AS) may increase sensitivity. Thus, it is observed that the wide diagnostic approach and early identification are bases for diligent treatment, maximizing the chances of full recovery of the patient.

Neuroimunologia

TL 1106421

THE ASSOCIATION OF DIFFERENT ACUTE MANIFESTATIONS OF MS ON QUALITY OF LIFE AND FUNCTIONAL OUTCOME

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Background: Multiple sclerosis (MS) is an inflammatory demyelinating disease that affects about 2.3 million people worldwide, with a direct impact on quality of life, causing an important degree of disability. The first manifestations of the disease can be subdivided into optic neuritis, with visual blurring or blindness; myelitis, with paresis, plegia, hypoesthesia, and sphincter incontinence; trunk syndrome, with ataxia and cranial nerve syndrome; and mixed syndrome, when symptoms from more than one group are present. **Objective:** The present study aims to analyze the relationship of the first clinical manifestation on the quality of life and functional outcome of MS patients. **Methods:** Cross-sectional study, conducted with patients diagnosed with MS according to McDonald 2017 criteria and followed in a neuroimmunology referral service in Bahia from July 2021 to May 2022. Quality of life was quantified based on the EQ-5D Quality of Life Questionnaire and functional outcome was rated by the Extended Disability Status Scale (EDSS). Individuals were classified into 4 groups according to the first manifestation of MS: optic neuritis, myelitis, trunk syndrome, and mixed syndrome. **Results:** 212 patients participated in this study, of which 59 (27.8%) were male and 153 (72.2%) were female. The mean EDSS was 2.62(±2.4) and the mean EQ-5D 0.6 (±0.24). Regarding the first clinical manifestation, we obtained: nine in optic neuritis (4.2%); thirty in myelitis (14.2%); thirteen in trunk syndrome (6.1%) and one hundred and sixty in mixed syndrome (75.5%). The Kruskal-wallis test showed that there is an effect of the different syndromic manifestations on the EDSS at admission [$\chi^2(3) = 12.8$; $p < 0.01$] but not on the EQ5D score [$\chi^2(3) = 7.7$; $p = 0.052$]. Pairwise comparison showed differences between the median EDSS scores of the mixed syndrome and trunk syndrome groups [2(IQR 1-4) vs 0(IQR 0-1); $p = 0.03$], but not between the other groups. **Conclusion:** The different acute MS conditions have similar impact on the quality of life of patients, but interfere differently in the functional capacity of these individuals. This important divergence between the groups with trunk syndrome and mixed syndrome can be interpreted by the fact that the EDSS is a scale that refers to the functional capacity, especially regarding mobility, therefore, the most affected are those with mixed syndrome and the least affected are those with symptoms related to the trunk syndrome.

Neuroimunologia

TL 1106428

CLINICAL SIGNIFICANCE OF THE CEREBRAL LESION'S SIDE IN ISCHEMIC STROKE

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Background: Stroke is a cerebrovascular disease that can affect both hemispheres and cause functional dependence. The structural and functional symmetry of the right and left sides of the brain is not absolute. Some differences in the two hemispheres have already been demonstrated. The left side of the brain is more associated with the language, logic and problem solving, while the right side is associated with emotions, empathy and visuo-spatial tasks. Furthermore, about 90% of individuals have the right hand as dominant, which is controlled by the contralateral motor cortex. Structural and physiological changes are involved in this differentiation, for example, there is an increase in the length of the central sulcus in the left hemisphere for right-handers and in the right for left-handers demonstrated in some studies. **Objective:** Evaluate possible changes in the functional outcome of right-handed patients who had an ischemic stroke in the right hemisphere compared to the left. **Methods:** Prospective cohort with patients from the Stroke Intensive Care Unit of Hospital Geral Roberto Santos. Patients were analyzed in relation to their handedness, a question asked directly to the patient or family members. Right-handed patients were kept for analysis, while left-handed patients were excluded due to their reduced number. The affected hemisphere was collected from medical records. For the outcome, functional dependence (mRS>2) was evaluated using the modified Rankin scale performed 90 days post-ictus. Patients with previous stroke and who died were excluded from the analysis. **Result:** Of the 132 patients analyzed, 69 (52.3%) were male with an average age of 58.1 (±40.5), those with stroke in the right hemisphere had a higher risk of functional dependence (42.3% vs 27.8%, RR=1.52). With the chi-square test, there was no statistical significance in the association of the affected hemisphere and the functional outcome ($p = 0.088$). **Conclusion:** The percentage of patients with functional dependence was higher in patients with stroke in the right hemisphere, but the p-value didn't show statistical significance. Patients with ischemic stroke in the dominant hemisphere may be more likely to develop functional difficulties. The dominant hand is used for activities that require precision, such as writing. Mild functional independence can lead to disabling deficits in patients post-stroke.

Neuroimunologia

TL 1106432

PREDICTION MODEL FOR THE DIFFERENTIAL DIAGNOSIS OF MOG-IGG ASSOCIATED DISEASE AND MULTIPLE SCLEROSIS AT FIRST CENTRAL NERVOUS SYSTEM DEMYELINATING EPISODE IN PAEDIATRIC PATIENTS

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Background: Anti-myelin oligodendrocyte associated disease (MOGAD) is more frequent in pediatric patients than in adults. In international cohorts about 30% of pediatric patients with central nervous system inflammatory diseases have MOGAD. Since MOG-IgG testing is not yet widely available worldwide, the identification of patients that might benefit from testing at first presentation is recommended specially in low-income countries. Also, the correct differential diagnosis of MOGAD and multiple sclerosis (MS) prevents unnecessary and potentially harmful treatments. **Objective:** Our aim is to propose a predictive score based on the clinical characteristics at first clinical attack for the differential diagnosis between MOGAD and MS in pediatric patients. **Methods:** This is a nested case-control study of patients ≥ 18 years of a Brazilian pediatric cohort of 6 reference centres in neuroimmunology in 5 states. We selected patients that tested positive for MOG-IgG at the incidental inflammatory attack and those diagnosed with MS at the first year of follow-up in the study. The clinical characteristics at first presentation were evaluated identifying those more strongly associated with MOGAD. Maximum likelihood or exact logistic regressions were used to investigate associations of clinical characteristics and the diagnosis of MOGAD or MS. The cut-off for the predictive score was estimated based on the receiver operating characteristic (ROC) analysis. **Results:** We found that younger age at presentation (1 point), male sex (1 point), bilateral optic neuritis (1 point) and multifocal presentation with encephalopathy or isolated optic neuritis (1 point) were associated with MOGAD and contributed for the composite score. Having ≥ 2 points in our proposed clinical composite score has 80% sensitivity (95% CI, 0.56-0.93) and 66% specificity (95% CI, 0.35-0.89) for the diagnosis of MOGAD. **Conclusion:** Combined clinical and demographic characteristics at first attack might be used to guide serologic testing for MOG-IgG and help to differentiate pediatric MS from MOGAD. The score might support treatment decisions and optimize the use of health resources.

Neuroimunologia

TL 1106524

REPORT OF MULTIPLE SCLEROSIS HOSPITALIZATIONS IN BRAZIL BETWEEN JANUARY 2010 AND DECEMBER 2020

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Background: Multiple Sclerosis (MS) is the leading autoimmune, inflammatory, demyelinating, chronic disease of the central nervous system. MS affects about 2.3 million people worldwide and is the most common cause of nontraumatic disability in young adults. It can cause a progressive neurological dysfunction in this population, leading to significant functional and economic damage. The disease presents itself through outbreaks, which lead to frequent hospitalizations and consequent important socioeconomic implications. **Objective:** Describe the epidemiological profile of Hospitalized patients for MS. **Methods:** A descriptive epidemiologic study was carried out with data collected from the Departamento de Informática do Sistema Único de Saúde (DATASUS) system, in which the report of Hospital admissions for MS between January 2010 and December 2020 was evaluated. The Hospitalizations were analyzed by age group, ethnicity, region, care scheme, gender, total value, and number of days of stay. Patient mortality rate, deaths and the predominant age group were also described. The average was calculated, along with the standard deviation (SD) to describe the continuous variables and the relative frequency for the categorical variables. Since this is a public domain database, submission to the Research Ethics Committee was not necessary. **Results:** There were 30,768 Hospitalizations. Most were white (58.26%), women (69.7%), between 30 and 39y (29.6%) and in the southeast region of the country (64.98%). About the care regimen, 70.3% were ignored at Hospital admission; however, there was no significant difference between public and private. The total value of Hospitalizations in these 10 years was R\$1,757,360.311 (SD: R\$714,199.89), with the mean price per Hospitalization being R\$686.19 (SD: R\$129.44). With the total number of deaths in this time frame being 328 individuals, the year with the highest incidence was 2019 (N: 51), with an average of 27.5 patient deaths/year. The average mortality rate is 1.14% with a total of 0.92% over 10 years. The predominant age group in the deaths was 50-59 years (19.51%). **Conclusion:** The data of patients Hospitalized for MS match with the epidemiology for the disease itself. Over the years, the number of Hospitalizations remained high, making MS a high-cost disease, both because it affects an active population and because of the total costs of Hospitalizations due to its progressive characteristics and frequent outbreaks.

Neuroimunologia

TL 1106531

STUDY OF MULTIPLE SCLEROSIS BY FOURIER-TRANSFORM INFRARED VIBRATIONAL SPECTROSCOPY TECHNIQUE

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Background: Multiple Sclerosis (MS) is a chronic, immune-mediated, inflammatory and degenerative disease characterized by demyelinating lesions of the central nervous system. The development of new **Methods:** that can extract biochemical information from biological fluids and promote an effective and rapid diagnosis is of great importance to improve patient management and treatment. The vibrational infrared spectroscopy technique with Fourier transform (ATR-FTIR) has been approached as an alternative for the identification of biomarkers and diagnosis in several areas. The present study aims to distinguish individuals with MS from the control group by ATR-FTIR spectra through pattern recognition (PR). The sample consisted of serum obtained from fifteen control subjects (mean age 35.0 ± 13.7 ; 11 females) and fifteen subjects with relapsing-remitting MS (mean age 32.0 ± 11.7 ; 10 females), from the Neurology outpatient clinic of the Cassiano Antonio Moraes University Hospital, following the 2017 McDonald's criteria. The EDSS score in subjects with MS ranged from 1 to 7 (mean 1.8 ± 1.5) and the number of outbreaks from 1 – 8 (mean 3 ± 1.5). FTIR spectra were obtained from 10 μ L pipetted onto an aluminum plate and dried for at least 2 hours in triplicate. The spectra were preprocessed and normalized to selected regions. For PR, Principal Component Analysis (PCA) and Unsupervised Random Forest (URF) **Methods:** were used. Using the PCA method, it was possible to observe a tendency to distinguish between the groups, but with a greater number of combinations up to the main component 4 (total spectrum 89.22%; high number of waves 98.68%; fingerprint 86.46%). Then, the URF method was performed, where it was possible to distinguish samples from the EM and control groups both in the regions of high number of waves (48.35%), fingerprint (47.84%) and total spectral (62.86%) with 3 main components. Combinations of regions (high number of waves and fingerprint) by the URF method were performed, with a distinction between the samples (50.42%). **Conclusion:** The results showed a distinction between the two groups, which suggests the presence of structures in the data capable of biologically distinguish between MS and the control group. This indicates a high potential of the ATR-FTIR technique associated with machine learning methods for the study and diagnosis of MS. Ethical approval: (CAAE n.44387521.8.0000.5071).

Neuroimunologia

TL 1106543

CORRELATION BETWEEN THE DEGREE OF DISABILITY AND DEPRESSIVE AND ANXIOUS SYMPTOMS IN MULTIPLE SCLEROSIS

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Background: Multiple Sclerosis (MS) is a chronic multifactorial disease characterized by inflammation, demyelination and gliosis mainly in the white matter of the Central Nervous System (CNS). Among these individuals, depression and anxiety are quite common disorders, the former condition affecting up to 54% of patients, and can be caused and aggravated by several factors. **Objective:** To evaluate the association between the degree of disability and depressive and anxious symptoms in patients with MS. **Method:** Cross-sectional, single-center study conducted with patients diagnosed with multiple sclerosis according to the 2017 McDonald criteria. The degree of disability was assessed using the Expanded Kurtzke Disability Status Scale (EDSS). Depressive and anxious symptoms, on the other hand, were assessed using the Hospital Anxiety and Depression Scale (HAD), which assesses the preexistence of anxiety and depression separately through 14 questions. The score for each modality ranges from 0-21, where: 0-7 unlikely, 8-11 possible, 12-21 likely. **Results:** We evaluated 196 patients, of which 70.5% were diagnosed as unlikely, 16.7% as possible and 12.9% as probable for depression. And for anxiety 50.5% had unlikely diagnosis, 31.5% possible diagnosis and 18.6% probable diagnosis. The mean EDSS among the participants was 2.60 (+/- 2.39). Kruskal-wallis test was performed between the groups categorized by HAD scale previously mentioned and there was difference of median EDSS between depression groups [$\chi^2(3) = 9.58$; $p < 0.01$] and Pairwise comparison showed the difference was found between Improbable-Probable groups [p adjusted < 0.05]. Whereas among anxious symptoms, no difference in degree of disability was found between the groups [$\chi^2(3) = 1.23$; $p > 0.05$]. **Conclusion:** Through the results of the study, it is possible to conclude that there is a difference in the degree of disability between patients with a probable diagnosis of depression and patients with an unlikely diagnosis. However, it is not possible to state that there is any difference in the degree of disability regarding the anxious symptoms of patients with multiple sclerosis.

Neuroimunologia

TL 1106559

NEUROLOGICAL COMPLICATIONS POST-VACCINATION AGAINST SARS-COV-2: A CASE SERIES FROM NORTHEASTERN BRAZIL

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Background: The SARS-CoV-2 virus caused the coronavirus disease pandemic in mid-2020 with millions of deaths, which led to an urgency in the development of vaccines against the virus. Neurological complications resulting from vaccination against SARS-CoV-2 are increasingly evident worldwide and require further clinical investigation. Vaccination-related neurological manifestations are rare but already reported for other vaccines, such as Guillain-Barré syndrome after influenza vaccination. **Objective:** To describe the clinical presentation of sixteen cases of patients exhibiting neurological manifestation after SARS-CoV-2 vaccination. **Methods:** This is a case series report nested within the Brazilian multicenter case-control NeuroCOVID study (Fiocruz-PE). Sixteen adult patients with neurological manifestations referred to Hospital da Restauração (Recife, Pernambuco, Brazil) in 2021 were studied. Physical examination, imaging test (when necessary), and laboratory tests were performed. All the patients (or legal representatives) signed the study consent form. **Results:** Most of the patients were of the female gender (10/16) and the median age was 63.5 years (mean \pm SD = 58.25 \pm 21.03 years). Of the sixteen patients, eleven patients were vaccinated with an adenoviral vector-based vaccine encoding the spike glycoprotein of SARS CoV-2 (ChAdOx1 nCov-19, Oxford/AstraZeneca/Fiocruz), and six individuals received the inactivated virus-based vaccine (Sinovac/Coronavac/Butantan). The delirium-associated encephalopathy was the major neurological manifestation among the patients (4/16), followed by optic neuritis (3/16), and headache/migraine (2/16). Other neurologic complications were benign paroxysmal vertigo, ischemic stroke, encephalitis, Guillain-Barré syndrome, myasthenia gravis, sixth nerve neuritis, and transverse myelitis. The interval between the vaccine dose and the onset of the symptoms varied from zero to ninety-five days. These cases had no other cause associated with the neurological diagnosis and were reported to the government surveillance system. Moreover, our study team is part of the Expert Committee on Adverse Events of the COVID-19 vaccine of the Pernambuco State Health Department. The notification and evaluation of these cases are of great importance for a better understanding of the clinical picture and management of the patients. Financial Support: Fiocruz-PE.

Neuroimunologia

TL 1106579

THERAPEUTIC INERTIA AND FUNCTIONAL DISABILITY IN MULTIPLE SCLEROSIS

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Background: Multiple sclerosis (MS) is an autoimmune demyelinating neurological disease that develops a chronic degenerative and inflammatory condition. After the onset of symptoms, it can take time before diagnosis and initiation of treatment. This therapeutic inertia can have deleterious impacts on the individual's functional capacity. **Objective:** To analyze the association between therapeutic inertia time and functional disability in MS. **Methods:** Cross-sectional, single-center study conducted with patients diagnosed with multiple sclerosis according to the 2017 McDonald criteria. The time of therapeutic inertia was calculated based on the dates of symptom onset and start of pharmacological treatment for MS. The degree of disability was expressed according to the EDSS. The sequelae were grouped into syndromes, namely optic neuritis syndrome, transverse myelitis syndrome, brainstem syndrome, mixed syndrome, and other symptoms. **Results:** 187 patients were admitted, of whom 20 (10.7%) were diagnosed with primarily progressive multiple sclerosis (PPMS) and 154 (82.4%) with relapsing remitting multiple sclerosis (RRMS). The mean time to therapeutic inertia was 31.96 (SD \pm 45.56) months, with 110 (58.8%) patients with inertia less than 32 months and 56 (29.9%) patients with inertia greater than or equal to 32 months. The median EDSS score was 2 (IQR 1-4). Regarding the presence of sequelae: 1 (0.5%) was classified with optic neuritis syndrome, 10 (5.3%) with transverse myelitis syndrome, 2 (1.1%) with brainstem syndrome, 154 (82.4%) with mixed syndrome and 13 (7%) with other symptoms. 7 (3.7%) patients had no sequelae. Time of therapeutic inertia greater than or equal to 32 months was associated with higher EDSS scores [median 4 (IQR 1-6) vs 1.75 (IQR 0-3.5); $p = 0.001$] and these more often had EMPP ($\chi^2 = 11.402$, 70.6% vs 29.5%, $p = 0.001$). **Conclusion:** Greater therapeutic inertia was associated with more impaired functional capacity and a clinical form with worse disease progression. Thus, possible factors that delay the initiation of pharmacological therapy should be investigated in clinical practice, considering their impacts on the natural history of the disease. Prospective studies can better explore this relationship and the possibilities of intervention on therapeutic inertia in MS patients.

Neuroimunologia

TL 1106588

ADVERSE REACTIONS TO COVID-19 VACCINES IN PATIENTS WITH DEMYELINATING DISEASES

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Background: Vaccine efficacy is heterogeneous and can be modified by multiple external factors; it is noteworthy that, although vaccines have demonstrated high efficacy or effectiveness in preventing Hospitalizations and deaths, all of them have some adverse event. **Objectives:** To analyze the occurrence of adverse reactions to COVID-19 vaccines in patients with demyelinating diseases. **Methods:** A retrospective, cross-sectional study carried out by reviewing the medical records of 115 patients, selected in a non-probabilistic manner and for convenience, with demyelinating diseases at the Multiple Sclerosis Reference Center of the State of Paraíba, using the variables sex, age, diagnosis, COVID-19 vaccination, and adverse reactions after COVID-19 vaccination. **Results:** Among the 115 medical records analyzed, there was a predominance of females (70.43%), in addition, a distribution of 101 (87.82%) patients with Multiple Sclerosis (MS) and 14 (12.12%) with Neuromyelitis Optica (NMO). In categorization by age group, 3% belonged to the age group under 16 years old, 9% from 16 to 25 years old, 34% from 26 to 35 years old, 31% from 36 to 45 years old; 10% from 46 to 55 years old; 10% from 56 to 65 years old and 3% from those over 65 years old. Regarding the vaccination of COVID-19, 100% performed the first dose, 99% the second dose and 74% the third dose. Regarding vaccine reactions, reactions were reported in 49% of patients, with 36% reporting body pain, 30% pain at the injection site, 16% fever, 16% headache, 11% chills, 9% worsening of MS symptoms, 5% fatigue, 4% nausea, 2% vomiting and diarrhea. Among the patients who had vaccine reactions, 11% had NMO and 89% had MS. **Conclusion:** The effectiveness of vaccination varies according to the vaccine applied, and may present vaccine reactions, despite being extremely effective in preventing deaths resulting from infection by SARS-CoV-2. We can observe that no patient had serious adverse reactions. Thus, vaccination against Sars-COV 2 is extremely important, since the benefits will be greater than the harm, as observed in the study.

Neuroimunologia

TL 1106591

THE PRESENCE OF DEPRESSIVE AND/OR ANXIOUS SYMPTOMS IS CORRELATED WITH FATIGUE AND ITS INTENSITY IN PATIENTS WITH MULTIPLE SCLEROSIS

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Background: Multiple sclerosis (MS) is an inflammatory disease that affects the central nervous system and is the leading cause of disability in young people in developed countries. MS significantly affects the quality of life of patients and has great importance for public health. Further, the pathophysiology is complex and the clinical presentation is heterogeneous, where fatigue is the most prevalent symptom among patients with the disease, being significantly experienced in up to 92% of sick patients. Worldwide researchers have already tried to investigate the impact of fatigue on the quality of life of those patients, however, especially in Brazil, there's still a gap. **Objective:** To evaluate impact of fatigue in depressive and anxiety symptoms in MS patients. **Methods:** A prospective study was conducted with consecutive patients who met McDonalds diagnostic criteria. Clinical and epidemiological characteristics were described. Fatigue was measured with Modified Fatigue Impact Scale (MFIS) and depressive and anxiety symptoms with the Hospital Anxiety and Depression Scale (HADS). Patients were separated in six categories: absence of fatigue (MFIS score 0-38) and fatigue clinically significant (MFIS score 39-84), probable depression (HAD-depression 0-12), improbable depression (HAD-depression 13-21), probable anxiety (HAD-anxiety 0-12) and improbable anxiety (HAD-anxiety 13-21). The descriptive analysis was performed through clinical information, their frequencies, means and standard deviations. The inferential analysis was performed using Pearson's chi-square test and its respective odds ratio. **Results:** 204 patients were included in study. The frequency among men was 58 (28.4%) and women was 145 (71.1%). The mean of age was $39,78 \pm 11,89$ years. The Pearson Chi-square test demonstrated that there is an association between the presence of depressive symptoms among patients with clinically significant fatigue [$\chi^2(1) 13,494$; $p < 0,001$] with 10,010 of odds ratio (CI 95%: 2,3-43,47) and also anxiety symptoms [$\chi^2(1) 11,803$; $p < 0,001$] with 4,546 of odds ratio (CI 95%: 1,81-11,41). **Conclusion:** the study concludes that there is a correlation between the presence of clinically significant fatigue and the presence of important depressive and anxious symptoms in patients with MS. In addition, it was noticed that patients with significant fatigue are 10,01 times more likely to have major depressive symptoms and 4,54 times more likely to have major anxiety symptoms.

Neuroimunologia

TL 1106611

PREVALENCE OF DEPRESSION IN PATIENTS WITH RELAPSING REMITTING OR PRIMARY PROGRESSIVE MULTIPLE SCLEROSIS

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Background: Multiple sclerosis is a demyelinating autoimmune disease of the central nervous system (CNS). May cause visual, motor, cognitive and dysautonomic symptoms. The main clinical presentations are relapsing-remitting (RRMS) or primary-progressive (PPMS). Due to the debilitating symptoms and the unpredictability of new outbreaks, in addition to the neurological dysfunctions associated with the condition, MS patients are susceptible to depression, which can compromise the individual's quality of life and prognosis. **Objective:** To compare the prevalence of depression in patients with RRMS or PPMS. **Methods:** Cross-sectional study, single-center, conducted with multiple sclerosis patients at the Magalhães Neto ambulatory in Salvador from august 2021 to may 2022. Patients were asked about their clinical history, whether they had a diagnosis of depression were also measured by the Hospital Anxiety and Depression Scale (HADS). **Results:** Of the 178 patients that were enrolled, 89.3% (n = 159) had RRMS, 10.6% (n = 19) had PPMS. Of the 159 RRMS patients, 22.6% (n = 36) had depression, 76.7% (n = 122) did not. Of the 19 PPMS patients, 47.3% (n = 9) had depression, 52.6% (n = 10) did not. The HADS was applied in 105 RRMS patients. 74.2% (n = 78) had a normal value of 0-7, 8.6% (n = 9) had a borderline value of 8-10 and 17.14% (n = 18) had an abnormal value of 11-16. The HADS was applied in 15 PPMS patients. 60% (n = 9) had a normal value, 20% (n = 3) had a borderline value and 20% (n = 3) had an abnormal value. **Conclusion:** Patients with PPMS had a higher prevalence of depression and abnormal values on the HADS compared to patients with RRMS. The patient with PPMS presents progressive worsening, with the accumulation of neurological deficits over time, in addition, there are few medications available for these patients, leading to a higher occurrence of depression in this group.

Neuroimunologia

TL 1106659

ACUTE TRANSVERSE MYELITIS ASSOCIATED WITH VACCINATION AGAINST COVID-19: A SYSTEMATIC REVIEW OF THE LITERATURE

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Background: Longitudinally extensive transverse myelitis (LETM) is a rare complication of different vaccines. Due to the pandemic, several COVID-19 vaccines have been used worldwide. Among the reported side effects, LETM deserves to be highlighted, being a condition with high morbidity. **Objective:** To conduct a systematic review of the literature on transverse myelitis associated with vaccination against Covid-19. **Method:** A systematic review of the literature was carried out, that included articles published between March 2021 and January 2022. Initially, 86 articles were found. After applying the inclusion and exclusion criteria, we encountered 21 publications. **Results:** There were a total of 23 cases, with the highest number in Germany, followed by Japan. The remaining cases were reported in Italy, United States, China, United Kingdom, Malaysia, Iran, Brazil, Portugal, Turkey, India. The vaccines involved were: AZD1222 AstraZeneca, ModernRNA-1273, the BNT162b2 vaccine, BBIBP-CorV and Ad 26.COV2.S, Johnson and Johnson. 34.7% of the cases occurred in female patients and 47.8% in male. Hypertension and diabetes were the most commonly present. The interval between vaccination and the LETM ranged from 24 hours to three weeks. The most common radiological findings on MRI were: extensive involvement of the cervical cord up to the thoracic spine, involvement of the cervical and thoracic cord separately. Among the alterations found in the CSF, mild hyperproteinorrea up to above 500mg/dL with unaltered cellularity were reported. Among the clinical manifestations, tetraparesis, paraparesis or paraplegia were reported, with or without paresthesia, sensory level and urinary sphincter dysfunction. The majority of patients received pulse therapy with methylprednisolone 1g/day, from three to five consecutive days. In 26% of cases, there was additional therapy with plasmapheresis. In one case, the patient spontaneously improved. In five cases reported there was complete remission of symptoms, in three cases the clinical outcome after treatment was not reported. And in the remaining cases there was partial improvement. In only one case, the patient died. **Conclusion:** Vaccines against COVID-19 have a significant impact on morbidity and mortality, being indispensable in the fight against the pandemic. Post-vaccination LETM is a rare event, but early recognition is important as treatment can reduce long-term disability.

Neuroimunologia

TL 1104800

REGION IN THE AMAZON HAS HIGH RATES OF HIDDEN PREVALENCE OF LEPROSY AND HIGH CASES WITH DISABILITIES

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Background: The method to estimate the hidden prevalence of leprosy, was proposed by Gil Suárez and Lombardi, in 1997, and is used and suggested by the Pan American Health Organization (PAHO) and used by the WHO (2016), it is based on the percentage of cases with some incapacity in the diagnosis (in relation to those evaluated), and applied to the total of new cases in the last 05 years of the research. At the end, the hidden prevalence is added to the new cases and the estimated real prevalence is obtained. **Objectives:** Evaluate the hidden prevalence of leprosy in the 11th Health Region of Pará, Brazil, between 2010 and 2020. **Method:** This is a cross-sectional and retrospective study, with data from the information system of notification disputes in Brazil. The data were organized in worksheet and analyzed according to the methods proposed by Gil Suárez and Lombardi (1997), and recommended by the World Health Organization and the Pan-American Health Organization. **Results:** The 11th Health Region of Pará was classified in 2010 as a hyperendemic area, with a detection rate of 90/100.000 inhabitants. Over the years, this rate has decreased, and in 2020 it was 29/100.000 inhabitants, where much of this decrease may be related to the Covid-19 pandemic. However, even with a significant decrease in cases ($p < 0.05$), the region is classified in the parameter of very high endemicity, with high rates of cases in children under 15 years old (11.3%) and some degree of disability (27%). Regarding occult leprosy, 792 cases were not diagnosed in the study region, thus suggesting an occult prevalence of 8.19/10,000 inhabitants. In other words, it can be said that 23% of the cases were not diagnosed and, consequently, went untreated, maintaining the active cycle of leprosy in the region. **Conclusion:** It is concluded that the 11th Health Region of Pará has a high rate of leprosy in children under 15 years of age, and in this region it is a very disabling disease with many hidden cases, keeping the transmission cycle active, resulting in a region with difficult to control leprosy.

Neuroinfecção

TL 1105138

NEGLECTED TROPICAL DISEASES AND THEIR NEUROLOGICAL MANIFESTATIONS: IMPORTANT DIFFERENTIAL DIAGNOSES FOR NEUROLOGY

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Background: Neglected tropical diseases (NTDs) consist of a group of 20 infectious diseases caused by bacteria, helminths, protozoa, and viruses, affecting more than one billion people worldwide, mostly in the context of social disparities. Many of these pathologies cause systemic repercussions, including a wide variety of neurological manifestations. **Objective:** To investigate the frequency and types of neurological involvement among NTDs in Brazil from January 2012 to December 2021 according to government databases. **Methods:** This is an epidemiological, descriptive, and retrospective study. Data were extracted from the SUS Hospital Information System (SIH/SUS). In addition, a review of the literature on the neurological manifestations of NTDs was performed. **Results:** The study revealed 507,469 Hospitalizations by NTDs in Brazil between 2012 and 2021, with the Northeast and Southeast regions being the most affected. The number of deaths in this period was 5,232, with higher burden among adults and those living in the Southeast region. The most relevant NTDs in the Brazilian context were Dengue, Leprosy, Visceral Leishmaniasis, Helminthiasis, and Trypanosomiasis. Neurological manifestations of varying degrees of severity and pattern of involvement have been described, including damage to the peripheral nervous system (plexopathy, Guillain-Barré syndrome, polyneuropathies, etc.) and central nervous system (encephalopathy, meningoencephalitis, stroke, epilepsy, etc.) The frequencies of the manifestations were not described in the SIH/SUS database. These findings are in accordance with our review of the literature, which emphasizes Hanseniasis as an important cause of peripheral neuropathy in low and middle income countries, as well as neurocysticercosis as a significant cause of epilepsy in south-eastern countries like Brazil. **Conclusion:** The neurological forms of NTDs contribute substantially to their morbidity and mortality, with heterogeneous manifestations being reported in the present survey. The frequencies of the findings found were not described, which points to the need for more research in the area and for improvements in the NTDs notification systems.

Neuroinfecção

TL 1105449

HOSPITALIZATION FOR MENINGITIS IN THE NORTHEAST REGION OF BRAZIL IN THE PRE-PANDEMIC AND PANDEMIC PERIOD (2018-2021): AN ECOLOGICAL TIME SERIES STUDY

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Background: The meningitis refers to an inflammatory process in the meninges. It can be caused by an infectious etiology, whose pathological agents are bacteria, viruses and fungi; as well as it can be a non-infectious etiology, being triggered by trauma, drugs and other mechanisms. The meningitis has the highest morbidity and mortality rates among central nervous system infections, and is therefore considered a severe public health problem. **Objective:** To analyze the variation in the number of Hospitalization for meningitis in the Northeast of Brazil during the pre-pandemic (2018-2019) and pandemic period (2020-2021). **Methods:** This is an ecological time-series study that estimated the number of Hospitalizations and Hospital detection rate of meningitis using data from the SUS's Hospital Information System (SIA/SUS). The data were obtained from the Tabnet Platform (DATASUS), being analyzed according to the variables age, sex, color/race, comparing those findings in the pre-pandemic and pandemic period. **Results:** 2,756 Hospitalizations were registered in this period, with a decrease of 0.2 in incidence per 10,000 inhabitants, between 2020 and 2021. Hospitalizations for meningitis due to parasitic infections represented 0.6% of all causes, showing a 21% drop in the number of Hospitalizations corresponding to this etiology in the years 2020 to 2021. Other causes of meningitis were responsible for 114 Hospitalizations, which 46% occurred in the pandemic period. Men, people under 70 years of age and people of mixed race represented the largest number of Hospitalizations, corresponding to 56.09% (1,546), 96.84% (2,669) and 58.27% (1,606), respectively. **Conclusion:** The lower number of Hospitalizations for meningitis, may have been a reflection of the underreporting that occurred during the years 2020 and 2021 as a result of the attention paid to cases of Covid-19. Besides, the viral form of meningitis is the most prevalent in Brazil. Therefore, infections by SARS-Cov-2 could justify the large number of Hospitalizations in the pandemic period. The lower rates of meningitis due to parasitic infection may have been a consequence of the unrestricted use of antiparasitic drugs for prophylactic purposes of Covid-19 during the last two years analyzed. Considering the importance of meningitis, the results obtained in the present analysis denote the need for more research to investigate and deepen the associations between the different variables of the condition.

Neuroinfecção

TL 1105510

DETAILED CLINICAL EVALUATION IN ASYMPTOMATIC SEROPOSITIVE HOUSEHOLD CONTACTS OF PATIENTS DIAGNOSED WITH LEPROSY – THE IMPORTANCE OF THE NEUROLOGIST IN EARLY DIAGNOSIS

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Background: The long incubation period of leprosy, its insidious signs and symptoms produce difficulties in its diagnosis and correct clinical classification. The early recognition of neural involvement in leprosy, especially in household contacts with subclinical infection, in which the classic clinical and laboratory findings of the disease are, by definition, absent, represents a major challenge in clinical practice. **Objective:** To characterize the clinical, molecular, serological and neurophysiological aspects in the early diagnosis of leprosy neuropathy in household contacts with subclinical infection (positive anti-PGL1 ELISA serology). **Method:** 361 asymptomatic seropositive household contacts (SPHC), defined as subclinical infection, followed up in a national reference center for leprosy in Brazil, from 2016 to 2020, were recruited. All individuals underwent a clinical, laboratory and neurophysiological evaluation. **Results:** 361 SPHC were evaluated, with a mean age of 35.7 years and female predominance (66.2%; 239/361). The qPCR analysis was positive in 35.5% (128/361) in dermal scrapings and in 25.8% (85/361) in skin biopsy of SPHC. In electroneuromyographic evaluation, 23.5% (93/361) of SPHC showed signs of neural involvement, with an average of 2.1 compromised nerves per SPHC. 62.3% (53/93) showed a mononeuropathy pattern on electroneuromyography (ENMG). The group of SPHC with altered ENMG had a higher qPCR positivity of dermal scrapings (47.0%; 40/85) when compared to the group with normal ENMG (31.9%; 88/276) ($p=0.01$), conferring a greater chance of neural involvement (OR, 1.8; $p=0.02$). **Conclusion:** The annual monitoring of SPHC showed a considerable prevalence of peripheral nerve impairment assessed by ENMG, favoring the early initiation of treatment. It is essential the role of neurologists in the control protocols of this endemic, favoring the break of the transmission chain of the disease.

Neuroinfecção

TL 1105530

SERUM LEVELS OF NEUROFILAMENT LIGHT CHAIN, GLIAL FIBRILLARY ACIDIC PROTEIN, UBIQUITIN CARBOXY-TERMINAL HYDROLASE L1 AND TAU PROTEIN IN COVID-19 INFECTED PATIENTS DURING THE PANDEMIC FIRST WAVE: EARLY NERVOUS SYSTEM INVOLVEMENT AND SEVERE OUTCOME

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SARS-CoV-2 infection can lead to highly heterogeneous clinical outcomes, ranging from no or mild symptoms to severe cases with need for life support in intensive care units (ICU) and death risk. The search for predictors able to forecast disease severity at patient admission to direct clinical choices would be beneficial to set priorities and optimize interventions. Central nervous system biomarkers might reflect early multi-organ involvement of Coronavirus Disease 19 (COVID-19), typical of patients with more severe disease evolution. Neurofilament light chain (NFL) is a protein of central and peripheral neurons validated as a nervous system damage biomarker in a variety of neurological diseases. Glial fibrillary acidic protein (GFAP) is highly expressed in astrocytes and increasingly used as a serum biomarker of astrocytic activation/injury. Ubiquitin carboxy-terminal hydrolase L1 (UCH-L1) is a neuronal specific protein frequently used for indicating brain damage induced by viral infections. Tau protein is mostly expressed by neurons, implicated in the pathogenesis of brain damage, especially in neurodegenerative diseases. This study aims to investigate whether NFL, GFAP, UCH-L1 and Tau protein provide non-redundant prognostic value to COVID-19 severity. So, the objective of this study is to investigate these biomarkers (NFL, GFAP, UCH-L1 and Tau) in the plasma of patients with severe and mild COVID-19. An observational study with 106 participants, comprising 73 patients with COVID-19 admitted to intensive care unit (ICU), 9 COVID-19 patients with mild symptoms and 24 healthy controls. Blood samples were taken from all participants and Single molecule array (SIMOA) was used to analyze NFL, GFAP, UCH-L1, and total Tau. The ICU participants were collected at the moment of the admission in the ICU and the COVID-19 outpatients at the beginning of symptoms. Graph prism was used to compare results of the ICU group, outpatients and healthy controls, in each protein. In our findings, NFL, GFAP, UCH-L1, and total tau levels were significantly increased in patients with severe or critical COVID-19 compared to outpatients ($p<0.0001$) and controls ($p<0.0001$), independently of nervous system involvement. No relevant difference was observed between outpatients and control groups. These results indicate the early involvement of the nervous system measured by serum neurodegeneration biomarkers as an important risk factor in prognosis of COVID-19 outcome.

Neuroinfecção

TL 1105532

FREQUÊNCIA DE NEUROCISTICERCOSE EM DOENTES ADMITIDOS NO HOSPITAL AMÉRICO BOAVIDA DE 2012-2014

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Background: Neurocysticercosis (NCC) is a worldwide public health problem, being the most common parasitic infection of the Central Nervous System (CNS), caused by the larval form of *Taenia solium*, the cysticercus cellulosae. It is the most frequent cause of epilepsy worldwide and has a lethality rate ranging from 16.4% to 25.9% in regions considered endemic, which include countries in Southern Africa, Asia, South and Central America, mainly affecting rural communities in developing countries. **Objective:** To describe the Frequency of Neurocysticercosis in patients admitted to the Neurology Service of Hospital Américo Boavida from 2012 to 2014. **Methods:** Retrospective descriptive study, the sample selection was by the non-probabilistic method for convenience, where all the Clinical Processes of patients admitted to the Neurology service of Hospital Américo Boavida in the period from 2012 to 2014, of both sexes, with aged over 18 years who were accompanied by computed tomography of the skull. **Results:** 923 clinical files were analyzed, of which 31 (3.3%) presented calcifications suggestive of NCC. Of these (51.6%) were female. The age of the patients ranged from 25 to 85 years, with a predominance of the age group from 66 to 75 years (25.8%); (74.2%) of the patients were single, and most of them lived in the municipality of Rangel (25.8%), followed by the municipality of Cacauco (22.6%). Regarding the number of calcifications in the tomographic findings, most had 1-2 calcifications (46.7%). Headache (32.3%) and epileptic seizures (25.8%) were the most frequent symptoms. As for the form and stage of the cysts, (96.8%) of the patients had the cysts in the calcification stage and only one (3.2%) in the active stage. **Conclusion:** In this study, the frequency of Neurocysticercosis is 3.3%, which we can consider high. The female gender was more representative; The Municipalities of Rangel and Cacauco had higher numbers of cases of NCC; Headaches and seizures were the most frequent symptoms.

Neuroinfecção

TL 1105583

IMPACT OF COVID-19 ON VIRAL ENCEPHALITIS CASES IN BRAZIL

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Background: At the imminence of the COVID-19 pandemic, it was found that the pathogen has an infectious potential of neuroinvasion and neurotropism, resulting in high rates of morbidity and mortality. In this context, its association with neurological conditions emerges, such as viral encephalitis, whose manifestation is mediated by a diffuse inflammatory process and affects mainly the extremes of age and immunosuppressed patients. In addition, the etiology also covers enteroviruses, arboviruses, herpesviruses and adenovirus. **Objective:** To verify if there was an increase in the diagnosis of viral encephalitis during the pandemic. **Method:** This is an analytical observational ecological study based on data on viral encephalitis obtained from the Hospital Morbidity System (SIH/SUS) of DATASUS. The period analyzed was from March 2018 to February 2022, using the following variables: Year/month of processing, age group 2 and Hospitalizations. Microsoft Excel version 2204 was also used to analyze the results, using graphs. **Results:** During the study period, 7754 cases of Hospitalization were recorded, of which 4578 occurred in the period before the pandemic and 3176 during the pandemic, showing a reduction of 18%. In this context, there was a prevalence in the age group of 0-9 years, with 1581 and 1124 cases in the pre- and post-pandemic period, respectively. Thus, it is inferred that the observed results contradict what was expected, as they portray a reduction in cases. Such a reduction may be related to the adoption of biosecurity measures to mitigate the contagion and prevention of COVID-19, also acting to reduce the spread of other viruses that can cause the pathology. In addition, broad-spectrum vaccination for SARS-CoV-2, which began in January 2021 in Brazil, and other immunization campaigns have a direct impact on reducing viral encephalitis. **Conclusion:** It was found that there was a reduction in the number of cases of viral encephalitis during the COVID-19 pandemic in Brazil. This was possibly due to biosecurity measures and widespread vaccination. In addition, age is an important determinant in the diagnostic profile of viral encephalitis in the country. However, the lack of data on Coronavirus viral encephalitis and the possibility of underdiagnosis and underreporting were limiting. In this regard, there is a lack of studies at the national level that better characterize the relationship of the virus as a potential agent of the pathology in question.

Neuroinfecção

TL 1105631

NEUROLOGIC MANIFESTATIONS IN PATIENTS WITH ACUTE CHAGAS DISEASE TREATED WITH BENZNIDAZOLE

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Background: Chagas disease (CD) is a serious public health problem and more than 70% of transmission in Brazil occurs by consumption of foods contaminated. Neurologic manifestations in the acute phase are rare, with a predominance of headache. Common side effects of Benznidazole include headache and peripheral neuropathy. Given the infrequency of neurologic findings associated with acute CD, it is important to differentiate between manifestations associated with the disease itself or the imidazole treatment. **Objective:** Describe the neurologic manifestations in patients with acute CD treated with Benznidazole. **Method:** Cross-sectional observational study with 32 patients treated at a referral Hospital for infectious and parasitic diseases in the city of Recife-PE between May and July 2019 during an outbreak in the backlands of the state. Previous, post-infection and post-use antiparasitic neurologic symptoms were evaluated, such as headache, tremor, drowsiness, dyskinesia, syncope, or pre-syncope and sensory complaints. The project was approved by the local Ethics Committee under No. 2,107,217. **Results:** Mean age was 30.7 years (range: 9-56 years), 50% female. From a personal history, 15 patients (46.8%) had a headache, predominantly migraine (60%), 6 (18.7%) drowsiness, 3 (3.3%) postural tremor in upper or lower limbs and 2 (6.2%) paresthesia. After infection, 26 (81.2%) had a new headache, mostly with a migraine pattern (76.9%). With the introduction of the antiparasitic drug at a dose of 200-300 mg/day, 7 patients (21.8%) had a new headache with a migraine pattern in 5 cases (71.4%) and 17 (53.1%) had tremor in their majority of postural character (82.3%) and may affect upper and lower limbs and head. Different sensory complaints were also reported: 7 cases of hypoesthesia (21.8%), 3 of pain (9.3%) and 7 of paresthesia (21.8%), all symptoms predominantly in the upper limbs. A 10-year-old child, after 300 mg/day of Benznidazole, developed dyskinesia on the seventh day of treatment. Dose reduction to 150 mg/day led to full recovery. **Conclusion:** Acute CD is infrequent and neurologic manifestations may be a consequence of the infection itself or treatment. Headache and tremor were the predominant findings, the latter being associated with Benznidazole use.

Neuroinfecção

TL 1105697

ENCEPHALITIS ASSOCIATED WITH PARVOVIRUS B19 INFECTION: A CASE PRESENTATION

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Case Presentation: A 62-year-old female patient was admitted to the emergency department with epigastric pain, hypoactivity, fever, chills, nausea, arthralgia and decreased consciousness for 2 days. Two days later, she evolved with headache, vomiting, decreased level of consciousness and tonic-clonic seizure. Petechial rash, papules, livedo reticularis, hyperemia and jaundice were also present. Head MRI and EEG were normal. Cerebrospinal fluid (CSF) analysis revealed xanthochromia, hyperproteinorrachia (98 mg/dL), normal glucose (66 mg/dL), 21 cells/m³ (69% neutrophils; 10% macrophages). Diagnosis of meningoencephalitis was made. Empirical treatment with ceftriaxone, ampicillin and acyclovir was initiated. Serologic tests for Zika, Dengue, CHIKV, HIV, VDRL and hepatitis were negative. Acute renal failure, septic shock, leukocytosis (27.350), thrombocytopenia developed rapidly, and the patient was transferred to the ICU. She woke up 12 days later, with delirium, diffuse weakness and mild right VI and VII cranial nerve palsy. New CSF was collected which remained xanthochromic and showed hyperproteinorrachia (138 mg/dL), glucose of 83 mg/dL and 23 cells/m³ (77% lymphocytes; 18% macrophages). XGEN MULTI N9 PCR was performed on CSF for Varicella zoster, HSV 1, 2, 6 and 7, CMV, EBV, enterovirus, adenovirus, parechovirus and parvovirus (PVB19) and only PVB19 was positive. EBV was detectable (296 IU/mL) by PCR in serum. Six days later, the CSF showed reduced cellularity (4 cells/m³) and proteinorrachia (92 mg/dL). There was improvement on neurological examination with normal cognition and mild generalized weakness. She was then referred for physical rehabilitation. After 2 months she returned to clinical office with normal neurological examination. **Conclusion:** The most frequent clinical presentation of PVB19 infection is erythema infectiosum, a self-limited disease which is very common in children. Encephalitis due to PVB19 infection is uncommon and should be considered in cases of encephalitis whose main etiologies have been ruled out. When available, multiplex-PCR should be performed, as it allows for a faster and wider etiologic diagnosis.

Neuroinfecção

TL 1105698

NEUROLOGICAL MANIFESTATIONS OF LONG-TERM COVID-19 IN PATIENTS WITH SARS-COV-2 INFECTION: DESCRIPTIVE STUDY

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Background: The clinical scenery of COVID-19 was initially described as a respiratory infection with possible severe and systemic manifestations. Studies point to chronic complications that affect neurological system of patients infected with the virus. Complaints may appear after the period of acute infection, with persistent and prolonged neurological manifestations being perceived. **Objectives:** To investigate, in a population sample from the State of RJ, the frequency of different types of neurological manifestations after COVID-19 infection and their medium and long-term outcomes. **Methods:** Cross-sectional descriptive study carried out in an outpatient clinic for post-COVID-19 neurological manifestations of an university Hospital in the city of Rio de Janeiro. A total of 233 patients were analyzed from September 2020 to April 2022 using socio demographic questionnaires to collect clinical-epidemiological data, including gender, age, comorbidities, time of onset of symptoms, most prevalent complaints in the acute phase, Hospitalization, complications and possible neurological manifestations of long-term COVID-19 in patients with SARS COV-2 infection. **Results:** Among the 233 patients evaluated, 182 were women and 51 were men. In addition, the mean age was 33.97 years (RI: 18 to 72) and 45.92% of them had comorbidities, the most prevalent being obesity (20.06%), followed by systemic arterial hypertension (18.45%) and diabetes mellitus (10.42%). The mean time between symptom onset and assessment was 7.45 months (RI: 1 to 25). The main symptoms in the acute phase of the disease were, respectively, fatigue (79.39%), anosmia (68.66%), headache (67.38%) and ageusia (64.37%). In addition, 93.13% did not need Hospitalization. It is noteworthy that 78.54% of the sample remained with some symptoms in the chronic phase of the disease, namely chronic fatigue (43.20%), cognitive deficit (42.06%), anosmia/hyposmia (38.19%), ageusia/hypogeusia (33.04%) the most relevant. **Conclusion:** The prevalence of the neurological condition in the acute phase of the disease was remarkable in the results presented. It is also worth mentioning that the main symptoms throughout the period after infection by the virus were predominantly neurological, with the most frequent being fatigue, anosmia, ageusia and cognitive deficit. Thus, it is evident the need for more studies that can improve the biopsychosocial impact that these late manifestations can have on the patient's life.

Neuroinfecção

TL 1105719

EFFICACY OF THE PHOTOBIO-MODULATION THERAPY ON RECOVERY OF OLFACTORY AND GUSTATORY DYSFUNCTION IN PATIENTS WITH LONG-COVID-19. A SERIE OF CASES

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Background: Smell and taste disorders are frequent symptoms during acute COVID-19 and may persist long after the resolution of the initial phase. Many studies have reported persistence of these symptoms more than 6 months after the onset, with female patients more likely to report persistent symptoms. Photobiomodulation Therapy (PBMT) represents a new and emerging therapeutic modality for the treatment of olfactory impairment after COVID-19, as it is able to modulate inflammatory processes and improve tissue healing in general. **Objectives:** Report a series of cases using PBMT for the management of COVID-19-related olfactory and taste impairment. **Methods:** Six females (average: 46.6 years \pm 13.7) and one male patient were seen at the post-COVID-19 Outpatient Clinic of a University Hospital in Rio de Janeiro, Brazil, from march to December 2021. COVID-19 infection was confirmed by biomolecular assay (RT-PCR) and all patients had anosmia and ageusia since infection. Data about the exact initiation and duration of all signs and symptoms related to COVID-19 was collected. Olfactory and gustatory function was evaluated separately using visual analog scales (VAS) ranging from 0 (normal smell/taste) to 10 (complete anosmia/ ageusia). Scale was applied at the first clinical appointment (baseline) and after the end of the treatment. Two questions related to quality of life were applied before and after treatment. An intranasal PBMT protocol was applied using Laser DUO* (MM Optics Ltda São Carlos, SP, Brazil) at 660 nm, on contact mode, with 100 mW of power, and 8J of energy in each nasal mucosa and 6J in ten points of the tongue. A total of sixteen laser sessions, twice a week and with a 48-hour interval were done. The paired t test was used to compare the pre- and post-treatment VAS means. **Results:** All patients completed the protocol (16 sessions). On average, the olfactory and gustatory VAS scale scores after treatment (media = 3,66, standard error = 0,55; media = 8,09, stand error = 0,85 respectively) was lower than the pre-treatment score (media = 8,00, stand error = 0,85; media = 3,50, stand error = 0,42 respectively), t (5) = 6,50, p < 0,01; t (5) = 5,89, p < 0,01. Patients also reported an improvement in quality of life after treatment. **Conclusion:** Given the lack of information about the available treatments for olfactory and gustatory dysfunctions in patients with long-term covid, PMB appears to be a promising therapeutic modality.

Neuroinfecção

TL 1105736

NEUROPSYCHIATRIC DISORDERS OF COVID-19 AFTER MILD SARS-COV-2 INFECTION

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Background: COVID-19, a disease caused by the SARS-CoV-2 virus, started in December 2019 in Wuhan, China. It is a disease that affects multiple systems, including neuropsychiatric manifestations such as anxiety, depression, fatigue and sleep disorders. These disorders may persist after the acute phase, leading to impairment of cognition and life quality. **Objectives:** To analyze the prevalence of persistent neuropsychiatric manifestations (fatigue, depression and anxiety) after primary COVID-19 infection. **Methods:** This is a descriptive cross-sectional study carried out in a cohort of 146 patients who had mild COVID-19 infection. Patients were seen at the Post-COVID-19 Outpatient Clinic of a university Hospital in Rio de Janeiro, Brazil, from September 2020 to May 2022. Tests were performed to screen for neuropsychiatric symptoms, including the fatigue severity scale (FSS), Hospital Anxiety and Depression Scale (HADS), in addition to surveys to collect socio demographic data. The HADS classifies patients into unlikely, possible and probable diagnoses of depression and/or anxiety. **Results:** Tests performed an average of 6.95 (RI: 1 – 24) months after COVID-19 infection. The patients average age was 22 (RI: 19 – 71). 75.34% of patients were female. 47.26% of patients have previous comorbidities. 54.79% of patients reported presence of subjective cognitive deficit after COVID-19 infection. Per the evaluation of the FSS, 71.91% of the patients presented fatigue after infection by COVID-19. Among the evaluated patients, 19.86% had probable depression, with the same value for possible depression. 30.13% of the patients had probable anxiety, while 20.54% had possible anxiety. **Conclusion:** The scales used showed that patients with mild COVID-19 infection had frequent complaints of chronic fatigue, anxiety and depression. In addition, more than half of the patients reported some component of subjective cognitive deficit, which may be related to the referred alterations. Further investigations and evaluations are needed regarding the origin of the reported complaints, since the period of confinement and social isolation can contribute to the emergence of neuropsychiatric complaints evidenced in the study.

Neuroinfecção

TL 1105745

PREVALENCE OF SPEECH-LANGUAGE SYMPTOMS RESULTING FROM COVID-19 OVER TIME IN A 2-YEAR CROSS-SECTIONAL STUDY IN RIO DE JANEIRO

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Background: The emergence of the disease known as COVID-19, caused by the virus SARS-CoV-2, was reported in China in 2019. It spreads rapidly and devastatingly around the world, infecting people in mild, moderate and severe ways. It is known that the virus uses the receptors of the enzyme angiotensin 2 (ACE2), which is present in various organs of the human body (lungs, brain, kidneys, heart), as an entry into cells, allows us to understand the diversity of symptoms of this infection. Although the main symptoms of this respiratory syndrome are cough, fever, dyspnea, which are marked factors for the severity of the condition, other speech and language symptoms can occur, such as: dysphonia, dysphagia, hypoacusis and chemosensory changes over time, as markers of symptoms of COVID-19. **Objectives:** Identify the prevalence of speech-language symptoms resulting from COVID-19 infection over time. **Methods:** This is a 2-year-long cross-sectional, quantitative and descriptive study. Patients with COVID-19 infection by PCR, rapid test or serology, of both genders, living in the State of Rio de Janeiro, selected by convenience, participated in the study. To determine the prevalence of speech-language symptoms, the questionnaire was designed by the author and applied electronically through Google Forms. It is composed of 10 open and closed questions, classified according to the presence of the symptom and its duration, focusing on the evaluation of the symptomatology report. Statistical analysis was performed using SPSS 22.0 version. **Results:** A total of 403 patients were included in the study. Median age was 37 (interquartile range (IQR) 19-77) years; 328 patients (81.4%) were female. Olfactory and Gustatory Dysfunction were the most frequently reported, followed by vocal and cognition impairment (80.9%, 73.7%, 28.0% and 23.8% respectively). The median duration of ageusia and anosmia were 58 (IQR 1-695) and 76 (IQR 1-695) days, respectively. Median duration of cognitive impairment was 173 (IQR 5-820) days. Standing out 18.4, 14.1% and 11.7% of the sample that kept Gustatory, Olfactory and Cognitive Dysfunction for more than 3 months. **Conclusion:** The prevalence of speech-language symptoms resulting from COVID-19 infection is high, highlighting the changes in smell and taste with varied durability. More studies are needed, especially regarding the impact of these changes on the quality of life of patients.

Neuroinfecção

TL 1105753

CAUSE AND EFFECT RELATIONSHIP BETWEEN PARKINSON'S DISEASE AND COVID-19

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Background: Parkinson's disease (PD) is one of the most common neurodegenerative disorders, affecting dopaminergic neurons in the substantia nigra. Patients can present motor and neuropsychiatric complications, besides sleep disorders and sensory abnormalities. The Braak hypothesis suggest that PD may originate from an external pathogen, whom would enter the body through the nasal cavity or the vagal nerve at the gut. This would help link the neurological complications that followed COVID-19, mainly the worsening of symptoms of PD and your appearance in patients who never had it before. **Objective:** This work includes evidence of the relationship between apparition of PD and the worsening of its symptoms after COVID-19. **Methods:** This study consists of a literature review of articles in English and Portuguese in PUBMED and GOOGLE SCHOLAR databases, published between 2020 and 2022 with the following descriptors: Parkinson's disease, COVID-19. **Results:** The studies analysed showed that close to 34% of COVID-19 survivors received a neurological or psychiatric diagnosis in the six months that followed their infection. This occurs because COVID-19 may have a neurodegenerative function, due to cytokine storm, triggering an autoimmune reaction, brain damage and making it more priming to other neurotoxic processes. This also happened in the Spanish-flu pandemic, in which there was a wave of new cases of encephalitis lethargica, while in the COVID-19 pandemic, non-motor symptoms in PD patients were exacerbated or make them appear as an early state of PD in other patients. The two main attack zones of COVID-19 are the nasal cavity and the gut. In the first one, the virus induces neuroinflammation of the olfactory bulb and reduces neural stem cells, causing anosmia. Meanwhile in the second, the patient may present alteration in the microbiota, causing dysbiosis, dysphasia and constipation. This, as said in the Braak hypothesis, is similar to the clinical presentation of PD, which can explain the link between this two, seeing the COVID-19 as a trigger to future complications and it's not being explored in enough studies to make it relevant. **Conclusion:** It is of utmost importance to bring this group to the fore, producing more evidence of the correlation between PD and COVID-19 by monitoring patients with neurological diseases after. This will allow the medical community to prepare itself for the upcoming of new cases creating a proper conduct and treatment method.

Neuroinfecção

TL 1105780

COVID-19 NEUROLOGICAL MANIFESTATIONS IN PATIENTS WITH INFLAMMATORY BOWEL DISEASE TREATED IN A TERTIARY HOSPITAL IN CEARÁ

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Background: COVID-19 has been frequently associated with a wide range of neurological disorders. However, it is unknown whether COVID-19 exacerbates neurological symptoms in patients with Inflammatory Bowel Disease (IBD). **Objective:** To identify possible worsening of gastrointestinal and neurological manifestations in patients with IBD due to COVID-19 infection. **Methods:** Research was carried out in 146 patients from the gastroenterology outpatient clinic, from June 2020 to June 2022, in a tertiary Hospital in Ceará. Evaluation was conducted in person or by phone and addressed, among other aspects, whether the patient was infected by COVID-19, if there was worsening of IBD symptoms and if there was neurological manifestation. **Results:** Among 146 patients interviewed during a 2 year period (28 in 2020; 36 in 2021; 82 in 2022), 78 had Crohn's disease and 68 had ulcerative colitis. Of this total, 36 patients reported COVID-19 infection with positive test confirmation, comprising about 24% of the sample, with 16.6% of the patients responding that there was a worsening of IBD symptoms during COVID. Regarding neurological symptoms, 72.2% of patients reported headache during COVID infection. In comparison, among patients who didn't have COVID with positive test confirmation, about 57% of patients reported headache. Other neurological symptoms reported by patients affected by COVID were anosmia (47.2%), ageusia (47.2%), tingling (22.2%), numbness (19.4%) and disorientation (5.5%). Surprisingly, no patient reported syncopal spell during COVID-19 infection. **Conclusion:** The results indicate that 83.3% of patients who claim to have COVID had at least one of the aforementioned neurological symptoms. Furthermore, the prevalence of neurological manifestations of these patients with IBD during SARS-CoV-2 infection is in line with the literature. In this sense, it is relevant to point out that the percentage of patients with headache was higher in the group of patients with COVID than in the uninfected group. Finally, the patients with GI worsening also had neurological exacerbation.

Neuroinfecção

TL 1105782

MOLECULAR MIMICRY AS ONE OF THE POSSIBLE MECHANISMS ASSOCIATED WITH PHENOTYPES OF DEMYELINIZING EVENTS IN PATIENTS INFECTED WITH ZIKA VIRUS

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Evidences observed during the Zika Virus (ZikV) epidemic in Brazil indicate a strong link between the Zika infection and neurological complications. Acute myelitis, optic neuritis, polyneuropathy and encephalomyelitis that mimic inflammatory idiopathic demyelination disorders (IIID) of the Central Nervous System (CNS) after ZikV infection have been reported. The present study aims to investigate the possible occurrence of molecular mimicry between ZikV antigens and Multiple Sclerosis (MS) autoantigens, the most frequent IIID of the CNS. A retrospective cohort study with 305 patients admitted due to suspected arbovirus infection in 3 university Hospitals in Rio de Janeiro was performed. All patients were submitted to neurological examination and biological sample was collected for serologic and molecular diagnostic by a multidisciplinary team. Bioinformatics tools were used to analyze the peptides shared between ZikV antigens and MS autoantigens and their conformation. Of 305 patients, twenty-six were positive for ZikV and 4 presented IDD pattern found in MS cases. Sequence homology comparisons by bioinformatics approach between NS5 ZikV and PLP MS protein revealed a homology of 5/6 consecutive amino acids (CSSVPV/CSAVPV) with 83% identity. Analysis of the 3D structures revealed a similar conformation with alpha helix presentation in a perfect align, which can deduce a molecular mimicry. Molecular mimicry between NS5 Zika virus antigen and PLP MS autoantigens emerge as a possible mechanism for IDD spectrum in genetically susceptible individuals.

Neuroinfecção

TL 1105812

NEUROLOGICAL AFFECTS IN THE PEDIATRIC POPULATION INFECTED BY SARS-COV-2 IN MARANHÃO

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Background: In 2019, the SARS-CoV-2 coronavirus was identified. It is important to conduct studies reporting the behavior of this virus in Maranhão in the pediatric population since the epidemiological profile may be different from the rest of the world. **Objective:** To delineate the clinical epidemiological profile of neonates, children and adolescents infected with SARS-CoV-2 in the state of Maranhão. **Methods:** This research was approved with CEP 4.315.245. It constitutes a cross-sectional observational study from March 2020 to August 2021. Secondary data from neonates, children and adolescents affected by SARS-CoV-2 in Maranhão were used. The analysis was performed using descriptive statistics. 260 data were collected, 99 with incomplete information were excluded and 161 were selected for analysis. **Results:** Regarding neurological involvement, 82.61% (n=133) did not present and 17.39% (n=28) expressed some of the analyzed types. Neurological involvement was considered as the presence of at least one: sign and symptom, neuropathy, neuroimaging, treatment, neurological coinfection. Regarding neurological signs and symptoms, 87.58% (n=141) did not and 12.42% (n=20) did. Among the signs and symptoms, 4.35% (n=7) each presented with seizure and decreased level of consciousness. In addition, 3.11% (n=5) presented with headache, 1.86% (n=3) with encephalitis and motor loss, respectively. 1.24% (n=2) presented anosmia, hypotonia and stiffening of the limbs, each. Also, 0.62% (n=1) presented hyponosmia, decreased reflexes, dysgeusia, and areflexia, respectively. No patient presented ageusia, hypogeusia and neuralgia. Regarding neuropathy (comorbidity), 93.79% (n=151) did not and 6.21% (n=10) did. Regarding the type of neuropathy, 2.48% (n=4) had encephalopathy and epilepsy, respectively, 1.86% (n=3) cerebral palsy, 1.24% (n=2) hydrocephalus, and 0.62% (n=1) myelomeningocele. As for the type of neurological treatment, 93.79% (n=151) did not perform and 6.21% (n=10) did. In addition, 98.14% (n=158) did not have neurological coinfection and 1.86% (n=3) did. Regarding the type of neurological coinfection, 1.24% (n=2) had PVD infection and 0.62% (n=1) had meningitis and brain abscess, respectively. Regarding the use of neuroimaging, 99.38% (n=160) did not perform it, with only 0.62% (n=1) performing it. **Conclusion:** Most patients had no neurological involvement, the most frequent being the presence of signs and symptoms.

Neuroinfecção

TL 1105849

CEREBELLAR SCHISTOSOMIASIS: AN UNUSUAL PRESENTATION

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Case Presentation: A previously healthy 23 year-old man from a known endemic area for schistosomiasis was admitted at the Neurology Emergency Department. Two months before admission, he developed severe daily headache, which was refractory to common analgesics. Moreover, he complained about weight loss. One month later, he presented with a newly onset cerebellar ataxia, more pronounced on the right side. Initial Magnetic resonance imaging (MRI) revealed high T2 signal areas with a slight expansive effect in the right aspect of the medulla; right cerebellar hemisphere and pons, and meningeal enhancement. Cerebrospinal fluid (CSF) analysis showed normal opening pressure, slight pleocytosis (16 cells) 75% lymphocytes, protein 35mg/dL. CSF Serologies for tuberculosis, cryptococcosis, Syphilis, HIV and cultures were negative. Computed tomography of the chest and abdomen showed only nonspecific alterations. MRI Spectroscopy suggested a pattern compatible with inflammatory disease in detriment of a tumor lesion. An inflammatory disease hypothesis was made and the patient was discharged with oral corticosteroids. However, some weeks later, he developed a right peripheral facial palsy and he was readmitted to the Hospital. A new MRI showed progression; Enhancement of right VII and VIII cranial nerves and a recent ischemic event on the left lateral aspect of the pons. Finally, a cerebellar biopsy was performed and revealed a Schistosomal granulomatous cerebellitis. The patient was treated with Praziquantel and high dose glucocorticoids with partial improvement. **Discussion:** Schistosoma mansoni is endemic to many Brazilian states. Its eggs may be found throughout the Central nervous system (CNS). They have been reported into the brain, cerebellum, leptomeninges and choroid plexus. However, the most common presentation is the schistosomal myeloradiculopathy. CNS lesions may cause cerebral small vessels endarteritis with fibrinoid necrosis, or immunological reactions secondary to toxin release from the granuloma, leading to astrocytic proliferation, neuronal necrosis and demyelination. **Conclusion:** This case shows the challenge of an atypical presentation of neuroschistosomiasis, which could have been easily misdiagnosed as a noninfectious inflammatory or neoplastic disease. A high index of suspicion is necessary, mainly in those patients coming from endemic areas, and histological diagnosis remains crucial.

Neuroinfecção

TL 1106010

HOSPITALIZATIONS FOR VIRAL ENCEPHALITIS: AN ANALYSIS OF THE EPIDEMIOLOGICAL PROFILE IN BRAZIL IN THE YEARS 2019 TO 2021

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Background: Infectious encephalitis is defined as a group of diseases characterized by an inflammatory process of the brain caused by microorganisms, especially viruses, with evidence of neurological dysfunction. The clinical presentation may involve acute fever, headache, altered consciousness and focal neurological deficits, with high morbidity and mortality. **Objective:** To analyze the epidemiology of Hospitalizations for viral encephalitis in the SUS in Brazil between the years 2019 and 2021. **Methods:** This research is a descriptive, observational, cross-sectional study of Hospitalizations for viral encephalitis in SUS between the years 2019 and 2021. Data were collected in the DATASUS database, with investigation of the variables: region, age group, gender, number of Hospitalizations, total value, mortality rate and days of stay. **Results:** There were 5741 Hospitalizations in the period, with 54% men and 46% women, with predominance in the Northeast (41.9%). As for age, the cases occurred more in the range between 1 and 4 years (16.8%) and less in the range of 80 years or more (2.3%). The total number of Hospitalizations in the first year was 2436, being higher than the following years, which had 1689 and 1616 cases. The total costs were R\$11.7M, with higher spending in the Northeast region (40.9%). The average total amounts per Hospitalization were R\$1913, R\$2103, R\$2153, respectively, in the years 2019, 2020, and 2021. The average mortality rate was 5.4%, with higher rates in those aged 60 to 69 years (10.5%), 70 to 79 years (10.3%), and 80 years or older (18.4%). The number of days of stay was highest in the 1- to 4-year age group, with 9121 total days, and lowest in the 80 years and older populations, with 1470 days. **Conclusion:** Based on the data, we noted a significant difference in the total number of Hospitalizations throughout the years of the period, which makes it possible to suggest interference from the pandemic, requiring further studies on the subject from now on. It is also observed that, despite the lower number of Hospitalizations among the elderly, this group had the highest mortality rates in the period, so as to suspect the importance of age during Hospitalization and early diagnosis. Finally, it is worth mentioning the high financial investments evaluated, raising the importance of more extensive research capable of assessing both direct and indirect costs resulting from severe cases of viral encephalitis in Brazil.

Neuroinfecção

TL 1106074

REDUCED WORKABILITY ASSOCIATED WITH INCREASED LEVELS OF FATIGUE AND ANXIETY IN PATIENTS WITH POST-COVID SYNDROME

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Background: Although the neuropsychiatric symptoms of fatigue, depression, anxiety and sleepiness are part of the post-covid syndrome, little is known about their negative impact on work capacity. Here we quantified these symptoms with validated questionnaires and analyzed the relationship with reduced work capacity. **Objective:** To investigate the impact of persistent neuropsychiatric symptoms (and cognitive dysfunctions) on the occupational vulnerability of survivors in a homogeneous group of bank workers. **Materials and Methods:** We analyzed 626 individual bank workers diagnosed with confirmed COVID19 (Hospitalized (17%) and non-Hospitalized (83%)). The information was extracted from the database of a follow-up study. Patients answered the "Chalder Fatigue Scale (CFQ)", "Epworth Sleepiness Scale (ESS)", "Hospital Anxiety and Depression scale (HADS)" and "Work Ability Index (WAI)". We used SPSS22 for statistical analysis with Chi-Square tests for analyses of proportions. **Results:** Our patients presented average age 42 years (22 to 66 years old), sex (53.5% female), education (48.9% complete undergraduate, 35.6% graduate and 8.1% complete high school), vaccination (83.4%) and function at work (74.6% contact with the public and 25% administrative work). With an average interval of 200 days between diagnosis and online interview, subjects SELF-reported several symptoms, including related fatigue (37%), anxiety (36%), depression (17%) and cognitive dysfunction (29%). Approximately 20% reported no post-COVID symptoms at the interview. The quantification of symptoms with questionnaires revealed excessive somnolence (ESS) (45.1%), fatigue (CFQ) (79.2%), symptoms of anxiety (HADS) (60.6%) and symptoms of depression (HADS) (65.9%). In addition, the comparison of symptoms between the group with normal WAI (37.5%) and reduced WAI (62.5% of participants) revealed high levels of excessive sleepiness (25% without somnolence versus 56%; $p < 0.05$), fatigue (54% no fatigue versus 93.2% with fatigue; $p < 0.05$), symptoms of depression (18% versus 68.2%; $p < 0.05$) and anxiety (30% versus 76.8%, $p < 0.05$). **Discussion and Conclusion:** Our results reveal a negative impact of neuropsychiatric symptoms on reduced work ability months after the acute infection. These findings point to the urgent need to provide specific treatment for patients to minimize the individual burden and the economic loss.

Neuroinfecção

TL 1106103

ATYPICAL NEUROLOGICAL MANIFESTATIONS POST-COVID-19: A CASE SERIES

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Background: SARS-COV-2 infection has affected millions of individuals to date. This disease has variable manifestations from an asymptomatic condition up to a rapidly evolving severe. Moreover, this infection may lead to multisystemic involvement, including the central and peripheral nervous systems. We report below three cases of atypical neurological manifestations post- COVID-19 and their clinical follow-up. **Case Presentation:** 27-years-old male patient, previously healthy, who had symptoms of COVID-19 confirmed by oropharyngeal and nasopharyngeal swab tests. On the tenth day of symptoms, the patient started to experience acute paraparesis, urinary retention, constipation, and hypoesthesia, up to T4 level. The second patient is a 50-years-old male, previously healthy, who had symptoms of the flu-like syndrome. The diagnosis of COVID-19 infection was confirmed by oropharyngeal and nasopharyngeal swab test. On the tenth day of symptoms, the patient started to experience paraparesis, urinary incontinence, and hypoesthesia up to T6 level. The neuroimaging and cerebrospinal fluid (CSF) analysis of both patients confirmed acute transverse myelitis, after COVID-19 infection. High-dose corticosteroid therapy was started, and both patients showed rapid recovery from their deficits. The third patient is a 45-year-old male patient who presented an 8-month history of progressive gait disorder and cognitive impairment after being Hospitalized for SARS-CoV-2 infection. Magnetic resonance imaging (MRI) was compatible with NPH. A spinal tap test was positive and there was a progressive improvement after shunting, with complete resolution of symptoms after 30 days of follow-up. **Conclusion:** We reported two cases of patients who developed acute transverse myelitis after COVID-19 infection that had a good and rapid response to high doses of methylprednisolone, unlike the previous reports. The differences in the doses and the treatment time may explain this. The third patient is a previously healthy patient who developed NPH two months after the COVID19 infection. Given the significance of this condition as a cause of reversible dementia, it is important to consider the possibility of a causal association with COVID19.

Neuroinfecção

TL 1106168

FREQUENCY OF EPSTEIN-BARR VIRUS (EBV) AND HUMAN HERPESVIRUS 6 (HHV-6) INFECTION IN PATIENTS WITH RELAPSING-REMITTING MULTIPLE SCLEROSIS

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Multiple sclerosis (MS) is a chronic neuroinflammatory and neurodegenerative disease of the central nervous system (CNS). The etiology of MS is not well understood, but it's likely one of the genetic and environmental factors. Approximately 85% of patients have relapsing-remitting MS (RRMS), while 10-15% have primary progressive MS (PPMS). Epstein-Barr virus (EBV) and Human herpesvirus 6 (HHV-6), members of the human Herpesviridae family, are strong candidates for representing the macroenvironmental factors associated with MS pathogenesis. Antigenic mimicry of EBV involving B-cells has been implicate in MS risk factors and concomitance of EBV and HHV-6 latent infection has been associated to inflammatory MS cascade. To verify the possible role of EBV and HHV-6 as triggering or aggravating factors in RRMS and PPMS, we compare their frequency in blood samples collected from 166 MS patients. The presence of herpes DNA was searched by real-time PCR (qPCR). The frequency of EBV and HHV-6 in MS patients were 1.8% (3/166) and 8.9% (14/166), respectively. Among the positive patients, 100% (3/3) EBV and 85.8% (12/14) HHV-6 are RRMS and 14.4% (2/14) HHV-6 are PPMS. About clinical phenotype of these patients, incomplete multifocal myelitis, and optic neuritis were the main CNS manifestations. These are the first data about concomitant infection of these viruses in MS patients from Brazil. Up to date, our findings confirm a higher prevalence in female with MS and a high frequency of EBV and HHV-6 in RRMS patients.

Neuroinfecção

TL 1106306

EVALUATIVE STUDY OF THE MOST PREVALENT INFECTIOUS AGENTS ASSOCIATED WITH MENINGITIS/ ENCEPHALITIS BEFORE AND DURING THE COVID-19 PANDEMIC. EXPERIENCE IN THE CITY OF RIO DE JANEIRO

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Infectious meningitis/encephalitis (EM) are pathological conditions associated with high rates of morbidity/mortality, where the examination of the cerebrospinal fluid (CSF) is essential in the diagnosis. In 2020 and 2021, the world was faced with a pandemic caused by the SARS-CoV2 virus. We selected CSF samples in 2 different periods: Before the pandemic and during the pandemic. CSF samples collected were sent to the Neurolife Laboratory immediately after lumbar puncture for diagnostic investigation. CSF was analyzed by conventional and molecular tests. The screening of the panels was performed according to the clinical indication, epidemiology and morbid history of the patients. In 2019, 737 PCR tests/month were performed for the main pathogens related to meningitis/encephalitis. In 2020, this monthly average was lower (8%) due to the reflection of the pandemic. It is worth mentioning that from April onwards the increase in PCR requests for SARS-CoV-2 in CSF was also perceptible, including becoming among the top 5 of the PCRs most requested by the medical community. Although the number of PCR tests requests has returned to the equivalent in the period before the pandemic, the lockdown schemes and restrictions due to the pandemic showed a drop in the numbers of infectious agents reported in cases of meningitis/encephalitis. A reduction from 354 positive cases (in 2019) to 124 (in 2020) and 141 (in 2021) can be observed. This is evident when looking at positive Enterovirus cases in 2019 compared to the pandemic years. The numbers have dropped by almost 80%. In 2019, there were 151 patients, while in 2020 there were 19 and in 2021 there were 24 patients. The top 5 pathogens found in 2019 were Enterovirus (151), Herpes simplex 2 (34), Streptococcus Pneumoniae (34), Varicella Zoster (23) and Neisseriae Meningitidis (17). In 2020 and 2021, the top 5 were, respectively, Enterovirus (19) and (24), Varicella Zoster (18) and (26), Streptococcus Pneumoniae (16) and (6), Herpes simplex 2 (8) and (12) and Neisseriae Meningitidis (2) and (5). **Conclusion:** It is evident that the decrease in the movement of people was reflected in the drop in cases of meningitis/encephalitis throughout the pandemic.

Neuroinfecção

TL 1106353

GREY MATTER ATROPHY ACCORDING TO FOUR COVID-19 STRAINS: FRONTAL LOBE AND LIMBIC ATROPHY ARE COMMON TO DIFFERENT STRAINS

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Background: The presence of neuropsychiatric symptoms in the post-COVID syndrome seems frequent and debilitating and represents a major public health concern. Unfortunately, neither the physiopathology nor the underlying cerebral alterations are understood. **Objectives:** To investigate structural brain alterations (gray matter atrophy) associated with post-COVID syndrome in patients with different strains of SARS-CoV-2. **Methods:** We analyzed 3T T1 MRI images of 300 post-COVID patients followed at the University of Campinas (median age: 40 y.o.; 209 women): 188 infected with the Alpha strain, 29 infected with the P.2-Gamma strain, 61 infected with the P.1+P.1.*-Gamma strain, 22 infected with the Delta strain and 90 healthy controls scanned during the pandemic. The determination of strain was according to the Fiocruz database (<http://www.genomahcov.fiocruz.br/dashboard/>), which exhibits the Brazilian SARS-CoV-2 predominant strains by intervals. We used the CAT12 toolbox/SPM12/MATLAB 2019 to perform Voxel-Based Morphometry (VBM) analysis (to detect grey matter atrophy (GMA)) and statistical analyses. We reported p-values < 0.001, we used sex, age and total intracranial volume as covariates and corrected the results for multiple comparisons. **Results:** Overall, all four strains presented GMA atrophy of bilateral anterior cingula and ventromedial frontal lobes. In addition, Strain P.2-Gamma showed GMA of the right cuneus and left superior occipital gyrus. The P.1+P.1.*-Gamma Strain presented more widespread atrophy, mainly in the right angular gyrus, fusiform gyrus, frontal operculum and precuneus and left frontal operculum and precuneus. Interestingly, the Alpha and Delta Strains revealed exclusive frontal lobe GMA. **Conclusion:** Despite the vaccination and some differences, in acute presentation, all four groups presented GMA of the anterior cingula and ventromedial frontal area. Although different strains may show individual variations, post-COVID syndrome seems to exhibit some commonalities of GMA. These findings suggest similarities in terms of cerebral injury and physiopathology across different SARS-CoV-2 strains. Given the location of GMA, these areas may be somehow associated with the highly prevalent neuropsychiatric symptoms (dysexecutive syndrome, depression and anxiety) in post-COVID syndrome. Further research in this area is necessary to correlate with neuropsychiatric dysfunction and to detect therapeutic targets.

Neuroinfecção

TL 1106491

HYPEROXIA BY SHORT-TERM PROMOTES OXIDATIVE DAMAGE AND MITOCHONDRIAL DYSFUNCTION IN RAT BRAIN

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Background: Oxygen (O₂) is an essential molecule for aerobic life, being the basis of several metabolic reactions. Oxygen therapy is used as a therapeutic protocol to prevent or treat hypoxia. However, a high inspired fraction of O₂ (FIO₂) promotes hyperoxia, a harmful condition for the central nervous system (CNS). **Objectives:** The present study evaluated parameters of oxidative stress and mitochondrial dysfunction in the brain of rats exposed to different FIO₂ for a short period. **Method:** Male Wistar rats were exposed to hyperoxia (FIO₂ 40% and 60%) compared to the control group (FIO₂ 21%) during 2 hours. Oxidative damage in lipids and proteins, the activity of the antioxidant enzyme catalase (CAT), nitrite/nitrate (N/N) concentration, neutrophilic infiltration, and mitochondrial respiratory chain enzymes were determined in the hippocampus, striatum, cerebellum, cortex, and prefrontal cortex after 120 minutes of exposure. **Results:** The animals exposed to hyperoxia showed increased lipid peroxidation, formation of carbonyl proteins, N/N concentration, and neutrophilic infiltration in some brain regions, like hippocampus, striatum, and cerebellum being the most affected. Furthermore, CAT activity and activity of mitochondrial enzyme complexes were also altered after exposure to hyperoxia. **Conclusion:** rats exposed to FIO₂ > 21% by 2 hours showed increase in oxidative stress parameters and mitochondrial dysfunction in brain structures.

Neuroinfecção

TL 1106540

BRAIN MAGNETIC RESONANCE IMAGING (MRI) FINDINGS IN PATIENTS WITH INFECTIOUS CEREBROSPINAL FLUID (CSF): A SINGLE-CENTER EXPERIENCE FROM A TERTIARY CARE HOSPITAL IN NITERÓI, BRAZIL

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Central nervous system (CNS) infections are a significant cause of mortality and morbidity world-wide. Infectious diseases of the CNS include a wide spectrum of infections caused by various pathogens affecting one or more of these components. Despite advances in medicine, the accurate diagnosis of a specific disease of CNS infections has proved to be very challenging, as its clinical variable severity and radiological presentations are very general and nonspecific. In most cases, the etiological diagnosis is only confirmed with the investigation of the pathogen in the cerebrospinal fluid. This study aimed to know and describe, through the cases, the MRI findings in patients diagnosed with infectious diseases through the investigation of the pathogen in the cerebrospinal fluid and followed up in a tertiary Hospital in Niterói-Brazil, from December 2019 to March 2022. Among the 17 patients studied, the most common etiologic agent was varicella zoster virus (5 cases). Other etiological agents found in the study were Epstein Barr virus, herpes virus type 1, herpes virus type 2, Candida spp, Pseudomonas aeruginosa, Klebsiella pneumoniae and Enterovirus. what was most observed were images suggestive of diffusion restriction with an infarct pattern, some being a confounding factor, due to the fact that the CNS infection was still unknown at the time of the first MRI. We can then conclude that in critically ill patients, with suspected CNS infection, the imaging appearance may appear as recent infarctions.

Neuroinfecção

TL 1106595

CHARACTERIZATION OF INDIVIDUALS AFFECTED BY POST-SURGICAL INFECTION IN THE NERVOUS SYSTEM IN A REFERRAL HOSPITAL

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Background: Surgical procedures in the nervous system have a number of possible complications. In this context, the development of a surgical site infection (SSI) is capable of causing high morbidity and mortality, psychosocial impacts on the life of the infirm and their families, in addition to increasing treatment costs for the health institution. The literature related to SSI in neurosurgery is scarce. **Objective:** To detect the incidence of SSI, predisposing factors and, finally, to evaluate the forms of prevention. **Method:** This is a descriptive, cross-sectional and retrospective study to assess the incidence of SSI in neurosurgery at a referral Hospital in the interior of the state of São Paulo. Secondary data contained in the medical records of patients undergoing neurosurgical interventions at Hospital Santa Casa de Araraquara-SP, a tertiary referral Hospital for elective and emergency procedures, from 2017 to 2020, were used. **RESULT:** We obtained an incidence of SSI in neurosurgeries of 6.5%, in 651 surgeries, mainly wall infection (35.9%) and ventriculitis/meningitis (35.9%). The mean age of affected individuals was 53.8 years and 79.5% of patients who developed infection had comorbidities. Elective surgeries and clean procedures had a higher frequency of infection than non-elective and potentially contaminated procedures (53.8%). Trauma and hemorrhage interventions showed a greater presence of SSI compared to other types of surgeries; 53.8% of SSIs were diagnosed during the outpatient follow-up of the patient; 66.7% of the acts used some type of drain or graft device. *S. aureus* was the most frequently isolated pathogen. The study showed an incidence of infection similar to other national and developing country studies, but with a higher incidence in elective surgeries, which may be related to the lack of institutional protocols in the postoperative period and neurosurgical dressings. **Conclusion:** The investigation demonstrated the need to develop institutional protocols that indicate the adoption of pre, intra and post-surgical measures, such as: use of intranasal mupirocin, meticulous aseptic technique and exchange of gloves before the placement of prostheses, as well as the maintenance of prolonged outpatient follow-up and patient guidance on surgical wound care.

Neuroinfecção

TL 1106604

PREVALENCE OF ANOSMIA IN LONG-COVID PATIENTS: A COHORT OF 221 PATIENTS

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Background: Olfactory dysfunction (OD), including anosmia and hyposmia is a frequent symptom among upper respiratory symptoms in COVID-19 patients. Long-COVID refers to the persistence of symptoms beyond the initial phase of infection. Of note OD is a frequent symptom in Long-Covid. The suggested pathophysiology of hyposmia and anosmia in Covid-19 involves the injury caused by SARS-CoV-2 to nasal mucosa and olfactory bulb, considering its high expression of angiotensin converting enzyme 2 (ACE2), as the virus has a high neurotropism for it. **Objective:** To analyze quantitatively the epidemiology of OD as a long-term effect of coronavirus infection in a cohort of patients. **Methods:** We performed a prospective cohort study with patients followed in a post-COVID neurological outpatient clinic. Patients were recruited based on positive COVID-19 PCR and post-COVID neurological symptoms. Patients were evaluated by neurological examination, Addenbrooke Cognitive Scale (ACE-R) and laboratory tests for APOE genotyping. OD was evaluated by clinical complaint and simple olfactory examination. The duration of the OD was estimated in basis of clinical history and day of evaluation. **Results:** The final sample consisted of 221 patients. Nearly half of patients (N=101; 45.7%) had OD during COVID-19 infection. More than a half of these patients (N=65; 64.34%) persisted with OD for an average of 6.3±3.8 months. Subjects with persistent OD had 40.6±13.3 years and those without persistent OD had 48.8±14.2 years (p<0.01). Gender (p=0.54), ACER score (p=0.26), allele frequency in Apolipoprotein epsilon genotype test (p=0.30) showed no significant difference between groups. **Conclusion:** Persistent of OD is a common clinical manifestation in patients with COVID-19. Our study identified that younger age is a risk factor for persistent olfactory dysfunction. It should be noted that patients with dementia and elderly patients may not have brought this complaint to the consultation, raising the age of this group of participants.

Neuroinfecção

TL 1106607

ABSENCE OF STRAIN-SPECIFIC EFFECTS ON COGNITIVE DYSFUNCTION AFTER COVID-19: ANALYSES OF 454 SUBJECTS

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Background: Long-term effects on cognitive function are present even after mild COVID-19 infection, leading the WHO to coin the term "long COVID". Recent reports suggest that the viral strain plays a role in determining long-COVID symptoms. **Objective:** This study aims to evaluate the effect of the presumed viral strain on different cognitive domains. **Methods:** Data from UNICAMP's NeuroCOVID cohort was used in the study. The date of the positive test was used as a proxy to separate subjects in different presumed strain groups according to the prevalent strain Fiocruz database (<http://www.genomah-cov.fiocruz.br>) Subjects were evaluated with a neuropsychological battery consisting of the following tests: phonetic fluency (FAS), semantic fluency (SF: animals), logic memory (immediate and late recall), Rey-Osterrieth complex figure test (copy and recall), colored trails test A and B, 9-hole peg test (dominant and nondominant hand) and the five digits test (reading, counting, choice, alternation, inhibition and flexibility), comprising 16 cognitive measures. We converted Test scores to Z-scores using normative tables for sex, age and education. The MANOVA test was initially used to test for strain effects on any cognitive domain, followed by one-way ANOVA tests for each domain. **Results:** 454 subjects (15 years of education; 88 days after diagnosis) were divided into 5 presumed strain groups: original strain (247), original+P2 (36), P2 (86), gamma+P2 (50) and delta (6). Data for all tests were available for 141 subjects. One-way MANOVA in this subset showed significant strain effects (p=0.046). Nonetheless, post-hoc tests showed no strain effects on any of the cognitive tests. SF showed a marginally significant effect (n=392, p=0.056) due to better performances in the original+P2 and P2 strains. FDT reading subtest was marginally significant (n=323, p=0.073) due to better test performances in the P2 and delta presumed strain groups. **Discussion:** although a global test effect was observed, individual test data could not show significant effects of the presumed strain. Marginal significance in semantic fluency and FDT reading subtest mainly accounted by presumed P2 strain subjects' better performances, suggesting P2-related post-COVID syndrome may relatively spare processing speed. Regardless of the absence of strain-specific effects, post-COVID subjects in the cohort showed significant cognitive deficits, which may impair workability and quality of life.

Neuroinfecção

TL 1106114

GLASGOW SCALE: WHAT IS IT AND HOW TO USE IT?

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The Glasgow Coma Scale (GCS) is widely used to assess and calculate the patient's level of consciousness, due to the simplicity of the scoring system and rapid application at the bedside. The objective is to outline a strategy that combines the main indicators of severity in traumatic brain injury (TBI) in a simple scale. There are three criteria: eye opening (up to 4 points), verbal response (up to 5 points) and motor response (up to 6 points). Added together, they give a score between 3 and 15. Eye opening, when spontaneous, takes 4 points. If not, commands are requested, such as calling the patient by name, setting the score to 3. Without response, a painful stimulus is applied (supra orbital notch pressure or trapezius pinching), receiving 2 points. A minimum score of 1 indicates that there are no openings. To assess the verbal response, one asks, for example: "what month is it in?", "what is your name?". Responding, the maximum score is given. Otherwise, the extent of their verbal skills is assessed. Sentences can be coherent and complete, but with confusion about time, place or person (4 points). Inappropriate use or disorganized words refers to audible, random and out of context words (3 points). The patient may make noises or moans, without forming any words (2 points). If there is no audible response, the score is minimal. For the motor response, those who fit the maximum score obey commands such as: "close your hands", "show your tongue". If negative, the pain response is evaluated with the aforementioned stimuli. In this case, the patient can locate it correctly (5 points); flexing or withdrawing a limb away from the pain stimulus (4 points); perform adduction and internal rotation of the upper limbs (3 points) - "decortication posture" (suggesting damage above the level of the red nucleus) - or even, extend upper and lower limbs (2 points) - "decerebration posture". With no response, the patient receives 1 point. The GCS is just one part of the neurological system exam. It is also important to assess pupillary reactivity, brainstem reflexes, tendon reflexes, signs of meningismus, and plantar skin response. Together, they can help localize the neurological lesion. Even in the regular observations of an inpatient, attention is paid to the trend of GCS scores, since an abrupt decrease may represent neurological deterioration, requiring urgent additional evaluation.

Neurointensivismo

TL 1106569

THE COGNITIVE IMPACT OF POST-INTENSIVE CARE SYNDROME: A REVIEW OF RECENT PERSPECTIVES

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Background: Mortality from critical illness has decreased in recent decades. Intensive care unit survivors experience cognitive impairments and mental health problems, referred to as part of post-intensive care syndrome (PICS). **Objective:** In this review, we summarize recent data on PICS cognitive impairment and its new challenges. **Methods:** This is a literature review article that studied patients in the Intensive Care Unit. The data search used LILACS, SciELO and Cochrane databases. **Results:** In current studies, questionnaires such as the Short-Memory Questionnaire (SMQ) were used. Cognitive impairment occurs in approximately 40% of ICU survivors within 3-6 months of critical illness and persists for at least 1 year in some patients². Areas commonly affected are attention, concentration, memory, mental processing speed, and executive functions. Among them, the most commonly affected domains are memory and executive functions. Risk factors for cognitive impairment after admission to the Intensive Care Unit are preexisting cognitive dysfunction, delirium, sepsis, hypoxia, and invasive mechanical ventilation. It has been seen that patients face the difficulty of returning to their original lives due to disabilities that persist for up to years after discharge from the ICU.¹ Also, it was seen that bundles like ABCDEF can help to prevent PICS. **Conclusion:** Survivors of critical illness are at high risk of developing post-intensive care syndrome. These findings highlight the importance of adequate post-ICU care planning to diagnose and treat this population.

Neurointensivismo

TL 1104780

IMPACT OF THE SARS-COV-2 PANDEMIC ON ALZHEIMER'S DISEASE PATIENTS: A RETROSPECTIVE AND EVOLUTIONARY ANALYSIS OF COGNITIVE ASPECTS

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Alzheimer's Disease (AD) is a neurodegenerative disorder that manifests as deterioration of multiple cognitive domains. These patients are at greater risk of Covid-19 and suffering with the pandemic, which can lead to accelerated functional and cognitive impairment. Thus, it is necessary to understand the clinical and social impact of the SARS-Cov-2 pandemic in this group. The aim of the study was to describe the evolution of cognitive and functional performance of AD patients from 2018-22. A retrospective cohort of 77 AD patients treated at a tertiary Hospital in Fortaleza was carried out from 2018 to 2022. Cognitive (cross-sectional and retrospective) assessment was performed with the Mini Mental State Examination (MMSE), Semantic Verbal Fluency (SVF) and Clock Drawing Test (CDT) and functional assessment with the Clinical Dementia Rating Scale (CDR) and Functional Staging Scale (FAST), in addition to clinical aspects, caregiver stress, satisfaction with telemedicine and social isolation. We used MMSE and FAST 2 and 3 [(1-17) and (1-7), respectively, discrete numerical variables] to define significant impairment. The evolution of the data in relation to the beginning of the pandemic was recorded as: before (2018-19), during (2020-21/22), and in the transition (2019-20). The three annual evolutions were then compared to each other. The results showed that in four years of outpatient follow-up, there was a functional deterioration, assessed by an increase in CDR [1(0.5-2) to 3(2-3)], FAST 2 [4(3-5) to 8(5-11)] and FAST 3 [4(3-5) to 6(5-7)] ($p < 0.05$). The last two configured significant functional impairment. There was an increase in the prevalence of anticholinesterase, anti-NMDA and antipsychotic use (36.5% to 74.3%; 20.3% to 45.9%; 18.9% to 44.6%, respectively) and its side effects. We found an increase in the prevalence of urge incontinence (1.4% to 37.8%) and of neuropsychiatric symptoms such as anxiety (35.1 to 58.1%) and insomnia (17.6 to 40.5%). Cognitive worsening was evidenced through the MMSE, SVF and CDT scores (15 to 11; 6 to 4.5; and 2 to 0, respectively; $p < 0.05$), the first of which configured significant cognitive impairment. There was an increase in caregiver stress (10.8 to 74.3%). It was possible to conclude that AD patients showed significant cognitive and functional impairment after the beginning of the pandemic. The follow-up of these patients is necessary in order to better describe the long-term extent of the effects of the pandemic in this group.

Neurologia Cognitiva e do Envelhecimento

TL 1105029

EFFECTS OF THE MOZART'S SONATA K448 ON THE EXTINCTION OF AVERSIVE MEMORY

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Background: Memory is the ability to acquire, to store and to recall information. Memory can be classified as short-term and long-term memory. It is known that music has an effect on almost the entire brain and its neural subsystems. In addition, it seems to play a crucial role in the treatment of phobias and post-traumatic stress disorder. There is evidence of positive effects of Mozart's Sonata K448 on short-term memory. However, little is known about the effect of memory extinction to the environment. **Objective:** To investigate the influence of Mozart's Sonata K448 in the extinction of the aversive memory. **Methods:** Twenty mice were used, divided into three groups: G1: Mozart (n=5), G2: Ambience (n=8) and G3: Control (n=7). Only the G1 group was exposed to music from intrauterine life onwards, from 9:00 pm to 7:00 am. From days 50 to 54, the animals were submitted to Habituation. On the 55th day, the Aversive Training was performed only with G1 and G2 groups. On the 56th day, the Extinction Test started and was repeated for 12 consecutive days for the three groups. On the 81st day, all groups were submitted to the Recall Test. The tests were recorded for analysis. The Extinction Test and the Recall Test were analyzed by the repeated measures ANOVA and one-way ANOVA tests respectively, with values of $p < 0.05$ being considered significant. **Results:** The Mozart group had higher freezing time in the Extinction Test when compared to the other groups ($p < 0.05$), especially on the first 3 days. In addition, the decrease in freezing time was greater for the Mozart group in relation to the other groups ($p < 0.05$). Finally, the Mozart group showed a significant increase in freezing time compared to the other two groups in the Recall Test. **Conclusion:** Mice submitted to Mozart's Sonata K448 during pregnancy had a longer freezing time at the beginning of the Extinction tests, with a decrease in this freezing time over the days. Therefore, music seems to play a positive effect on short-term memory as well as on the extinction of aversive memory, since it causes this memory to be more rapidly extinguished.

Neurologia Cognitiva e do Envelhecimento

TL 1105056

CHRONIC STRESS MEDIATORS IN PATIENTS WITH MILD COGNITIVE IMPAIRMENT: A TRANSVERSAL ANALYSIS OF THE BRAZILIAN MEMORY AND AGING STUDY

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Background: Several studies suggest that chronic stress and persistently high levels of cortisol are related to cognitive dysfunction and neurodegenerative processes. The Brazilian Memory and Aging Study (BRAMS) is a research initiative for the longitudinal follow-up of individuals in a tertiary center specialized memory outpatient clinic. **Objectives:** to investigate the relationship between markers of chronic stress and cognitive status in BRAMS, making comparisons between the control, subjective cognitive decline (SCD) and mild cognitive impairment (MCI) groups (amnestic, non-amnestic, single domain or multiple domains). Secondly to correlate the markers with the amyloid status of the participants. **Methods:** observational study of the cross-sectional type with a subgroup analysis of the BRAMS cohort, in which individuals aged > 60 years who had access to medical, neuropsychological and amyloid PET assessment. The present analysis seeks to measure the allostatic load index (ALI) through anthropometric, endocrinological, cardiovascular, metabolic and inflammatory markers of chronic stress, correlating the variables with cognitive status and amyloid status. **Results:** Between the years 2018 and 2021, 77 participants were evaluated in the present protocol, of which 65 met the inclusion criteria. The mean age was 70.2 (± 6.01) years, with 72% being female with 11 years of schooling (interquartile range 7 – 15). There were 42 subjects classified as MCI, 15 as SCD and 8 as controls. There was a significant difference between HDL measurements (55.6 in the MCI group vs. 66 in the SCD group vs. 69 in the control group, $p = 0.04$), waist/hip ratio (0.94 in the MCI group vs. 0.88 in the SCD group, $p = 0.03$) and also for the measurement of the ALI in percentage value considering the extremes of cortisol (ALI % cortisol $p < 12.5$ or $p > 87.5$) (36.9% in the MCI group vs. 27.2% in the SCD group, $p = 0.04$). In the multivariate analysis, age, education and economic class played a moderating role. **Conclusion:** The present work probably represents one of the first research initiatives to address measures of chronic stress beyond cortisol in elderly participants diagnosed with SCD and MCI, with increased markers in the MCI group. Due to the cross-sectional nature of the study, the associations found do not allow inferring causality between the measures, but they can be better explored in the longitudinal follow-up of the participants.

Neurologia Cognitiva e do Envelhecimento

TL 1105062

CAUSES OF RAPIDLY PROGRESSIVE DEMENTIA: A MULTICENTRIC STUDY IN TERTIARY MEDICAL CENTERS OF SANTA CATARINA

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Background: rapidly progressive dementia (RPD) is characterized by accelerated cognitive decline leading to functional impairment. There is no clear accepted definition of RPD, with most studies using a time frame of 1 to 2 years from symptom onset to dementia. There are many causes of RPD, but few studies have examined the epidemiology of different RPD etiologies. **Objectives:** this study aimed to identify the relative frequency of different causes of RPD among inpatients from four tertiary medical centers in Florianópolis, Santa Catarina, Southern Brazil. This is an ongoing study and we report its preliminary results. **Methods:** this is a cross-sectional retrospective study. Medical records of patients admitted from 2001 to 2020 and registered under ICD-10 codes potentially linked to RPD were identified and searched for RPD cases. RPD was defined as any disorder fulfilling the National Institute on Aging – Alzheimer's Association criteria for dementia whose course from symptom onset to dementia was less than or equal to 2 years. Patients diagnosed with a primary psychiatric disorder or delirium were excluded. The etiological RPD diagnoses registered at Hospitalization were reported. Each medical record was reviewed and the authors reclassified the RPD etiology according to established diagnostic criteria. **Results:** 4714 medical records were identified. As for May 2022, 2502 medical records have been analyzed, and 68 RPD patients were found, 46 (67.6%) of which were male. Mean age of onset was 65.5 years ($SD = 15.3$), and mean time from onset to functional impairment was 222.3 days ($SD = 210.1$). At Hospitalization, the RPD diagnoses were: mixed dementia (MD; $n = 16$; 23.5%), vascular dementia (VD; $n = 15$; 22.1%), nonprion neurodegenerative disorders (NPND; $n = 9$; 13.2%), autoimmune encephalopathies (AIE; $n = 5$; 7.4%), neoplasms (NEO; $n = 4$; 5.9%), prion diseases (PRD; $n = 3$; 4.4%), carence diseases (CD; $n = 3$; 4.4%), infectious diseases (ID; $n = 3$; 4.4%), toxic-metabolic dementia (TMD; $n = 1$; 1.5%) and genetic diseases (GD; $n = 1$; 1.5%). No cause was registered in 8 cases (11.8%). The revised diagnoses were: NPND ($n = 17$; 25.0%), VD ($n = 17$; 25.0%), MD ($n = 8$; 11.8%), AIE ($n = 6$; 8.8%), TMD ($n = 4$; 5.9%), NEO ($n = 4$; 5.9%), ID ($n = 4$; 5.9%), PRD ($n = 2$; 2.9%), post-traumatic dementia ($n = 2$; 2.9%), CD ($n = 2$; 2.9%), GD ($n = 1$; 1.5%) and normal pressure hydrocephalus ($n = 1$; 1.5%). **Conclusion:** the etiologies of RPD are many and tertiary center studies may help clarify their epidemiology and clinical profile.

Neurologia Cognitiva e do Envelhecimento

TL 1105227

EVALUATION OF MEEM AND CDR IN ELDERLY WITH AND WITHOUT DEMENTIA FOLLOWED AT A HOMEOPATHY SERVICE IN RIO BRANCO, ACRE

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Background: Dementia is a clinical condition in which there is a significant cognitive decline in relation to a previous level, causing difficulties in occupational, social and domestic life. The Mini Mental State Examination (MMSE) and the Clinical Dementia Rating (CDR) are useful tools that can be applied to patients with dementia for screening and staging purposes, respectively. **Objective:** Evaluation and comparison of the results of the MMSE and CDR in elderly patients with and without dementia, undergoing complementary homeopathic treatment. **Methods:** Study participants were 43 elderly under follow-up for cognitive or motor complaints, with and without dementia, at Hospital do Idoso – Fundação Hospitalar do Acre. Dementia diagnoses were made according to well-defined etiological criteria. Participants were interviewed with individual and socioeconomic questionnaires, cognitive assessment by the MMSE and assessment of dementia severity using the CDR. Data were entered and analyzed using the SPSS 20 program. **Results:** The most frequent diagnoses of dementia were frontotemporal dementia, Alzheimer's dementia and vascular dementia. The results of the CDR were in agreement with those of the MMSE in 24 patients with dementia and 3 patients without dementia. Of 16 patients classified as "questionable dementia" by the CDR, 10 were classified as "dementia" by the MMSE and 6 were classified as "no dementia" by the MMSE. If we consider as having dementia confirmed by the CDR only scores from 1 to 3, the agreement rate between the CDR and the MMSE is 76.74%. **Discussion:** The MMSE result worsens as the CDR staging levels increase, which is in agreement with other studies. The data also point to a strong influence of the level of education on the MMSE, an association that is already well described in the literature. In addition, in the direct comparison of the MMSE with the degrees of the CDR, there is interference, even in patients without dementia, in attention, calculation and visuospatial ability, suggesting that these domains can be affected early even in patients with complaints that are limited to deficits in memory. **Conclusion:** The results of the present study suggest that the mean MMSE scores follow a pattern of agreement in relation to the staging degrees recommended by the CDR and tend to remain regular in returns, with some exceptions that can be explained by the progression of the disease, by the effect of medications and by external factors.

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TL 1105384

AVAILABILITY AND ACCESS OF PALLIATIVE CARE IN ELDERLY WITH ALZHEIMER'S DISEASE IN THE PUBLIC HEALTH SYSTEM: A REVIEW OF THE LITERATURE

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Background: Dementia syndromes are among the leading causes of death and have similar symptom control challenges as cancer patients. However, dementias tend to receive less attention in palliative care planning. According to the Brazilian Alzheimer's Association (ABRAZ, 2010), there are approximately 1.2 million elderly people with Alzheimer's Disease (AD). AD is characterized by a prolonged course of progressive disabilities that bring a significant responsibility to the public health system and families. **Aims:** Describe the availability and access of palliative care for elderly Alzheimer's patients using the SUS (Brazilian Unic health system). **Methods:** A literature review was developed in three steps: Development of the research question, search for scientific articles in the Pubmed database, and critical analysis of included articles. The search was conducted in March 2022, and articles between 2012 and 2022 were selected, for a total of 30 articles, of which 8 were used. **Results:** AD is one of the types of dementia that received the worst quality of end-of-life medical care. It is noted that it is not established as a routine of care in this group of patients even though the recognition of terminality is proposed as a fundamental prerequisite for better end-of-life care in demented patients. Effective care management for demented patients can be provided from specialized centers to complement usual care and mitigate the growing social and economic burden of dementia. Although, it is not recognized as a protocol within the healthcare system that provides outpatient care as well as, there is no unit and facility to which patients and their families can be referred for support and better care planning. **Conclusion:** Neuropalliative care should be available on the public health system (SUS) and offered to all patients who need it, regardless of diagnosis, in order to screen for psychosocial factors that worsen the disease in addition to promote better symptom control and recognize caregiver exhaustion.

Neurologia Cognitiva e do Envelhecimento

TL 1105413

AEROBIC EXERCISE AND IMPROVEMENT OF QUALITY OF LIFE IN PEOPLE WITH ALZHEIMER'S DEMENTIA: SYSTEMATIC REVIEW

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Background: Alzheimer's dementia (AD) corresponds to 90% of dementia cases in the elderly, and is expressed by functional, cognitive and behavioral impairment and consequent reduction in life quality. Because it is associated, among other things, with low levels of physical exercise, they can lead to a positive effect on the many losses related to the disease. **Objective:** To evaluate evidence of aerobic exercise efficacy in improving life quality of people with AD. **Method:** A systematic review was performed, following the PRISMA check-list, in the Scielo, PubMed, VHL, Medline, Cinahl, PEDro and Cochrane databases, using descriptors. Randomized clinical trials with the variables AD, aerobic physical exercises and life quality were included. The methodological quality was evaluated with the JADAD scale and the data were analyzed with descriptive statistics. **Results:** Six articles were evaluated, published between 2009 and 2020. The sample consisted of 314 elderly people of both sexes, between 65-80 years. The studies obtained an average of 4.5 points on the JADAD quality scale. The diagnostic criteria were established by the DSM IV and the National Institute of Neurologic and Communicative Disorders. About 66.6% of the investigations reported mild dementia, 33.3% moderate and 33.3% did not report the degree. There are reports of Chronic Noncommunicable Diseases in 33.3% of the investigations. The presence/severity of AD was analyzed by the Mini Mental State Examination, and in relation to life quality, the Quality of Life-AD was used, both used in 66.6% of the studies in the pre-test and post-test. Regarding the intervention, 50% of the studies analyzed walking and 50% aerobic exercises of strength, endurance, balance and flexibility, 83.4% with mild to moderate intensity and 16.6% with moderate to high. On average, the programs lasted from 8-12 weeks, ranging from 2-5 weekly sessions, the most common being 3 days/week (50%), and 41.3 minutes/session (± 11.17). It was found that 83.3% of the interventions were able to positively modulate the dependent variable Quality of life, evidencing medium-term observation of the effects of aerobic exercises on the improvement of cognitive and functional function. **Conclusion:** In elderly with AD, aerobic physical exercises led to changes in quality of life and improvement in physical and cognitive performance. However, further investigations are needed comparing interventions of high relevance in a larger sample size.

Neurologia Cognitiva e do Envelhecimento

TL 1105501

ADUCANUMAB AS A NEW THERAPEUTIC POSSIBILITY FOR ALZHEIMER'S DISEASE

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Background: Alzheimer's disease (AD) corresponds to a cognitive and functional decline associated with age, specific neuropathology and in its earliest stages it can be characterized by a deficit in the ability to encode and store memories. It is characterized by deposition of amyloid- β (A β) plaques and neurofibrillary tangles in the brain, accompanied by neurodegeneration. Regarding therapeutic options, there is still a relevant search for improvements in the treatment of this disease, and for that, the path that leads to the use of anti-amyloid agents showed significant efficacy in clinical trials. Agents with potential for short-term approval meet these criteria: the injectable antibodies, aducanumab, gantenerumab, and BAN 2401, and an oral small-molecule agent, ALZ-801. **Objective:** To analyze the new therapeutic possibilities in the treatment of Alzheimer's Disease and to understand the relevance of this new therapy for patients affected by this disease. **Methods:** This is a systematic literature review based on data extraction in indexes such as PUBMED and MEDLINE. Publications from 2018 to 2022 were considered, in English and with the following descriptors: Aducanumab, Alzheimer's, Neurology. **Results:** Alzheimer's disease currently ranks as the most common form of neurodegenerative disease, and its symptomatic treatment is able to offer only a modest and measurable effect on cognition. Regarding the aforementioned anti-amyloid agents, Aducanumab showed significant efficacy in clinical and biomarker results: o aducanumab atua ligando-se e reduzindo o A β solúvel e insolúvel de maneira dependente da dose e com cerca de um ano de infusões intravenosas mensais de aducanumab foi capaz de reduzir o A β cerebral, o que resulta em uma desaceleração do declínio clínico medido. **Conclusion:** In this way, it is noticed that new possibilities for the treatment of AD appear on the horizon and create new hopes to guarantee a better quality of life and who knows even evolve to a clinical decline of this pathology. Evidently, there is still a long way to go, mainly due to the complexity of the pathophysiological factors of this comorbidity, but the possibility of an effective treatment opens up new hopes.

Neurologia Cognitiva e do Envelhecimento

TL 1105536

MOLECULAR MECHANISMS IN NEURODEGENERATION AND PROTEIN ACCUMULATION: A REVIEW

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Background: The abnormal accumulation of proteins is a common feature of neurodegenerative diseases. Novel information regarding the mechanism of aggregation include dysfunction of proteins involved in membraneless organelles formation, which have a chemical structure that is prone to aggregation. Once assembled, these aggregates spread in the central nervous system through a prion-like mechanism that involves the exportation of seeding nuclei to nearby neurons. **Objective:** This work aims to review the main protein misfolding mechanisms involved in neurodegeneration, as well as to explain the recent prion-like spreading of such misfolding patterns throughout the central nervous system. Moreover, this review dives into chemical structure particularities of some common proteins identified in neurodegenerative diseases. **Methods:** Articles produced between 2010 and 2022 were searched using keywords related to the field of study. The selection of included studies was based on whether the papers' theme was neurodegeneration, protein aggregation and/or prion-like mechanism of spreading. **Results:** The reviewed studies showed that the mechanisms of protein aggregation seem to be closely related to membraneless organelles dysfunction. Such organelles have important physiological functions. However, under certain conditions, they might form irreversible solid aggregates, whose toxicity is related to neuronal dysfunction and death. The studied proteins have low complexity domains that predispose them to misfold, with a particular chemical structure identified. Moreover, we discussed the prion-like mechanism theory for neurodegeneration spreading, allowing novel approaches to this group of diseases. This theory states that transference of prionic nuclei may occur among neural cells, which would induce protein misfolding and aggregation in naive cells. **Conclusion:** Abnormal accumulation of protein aggregates in neurodegenerative disorders has been extensively studied in the last years. Although intrinsically disordered regions have been evolutionarily selected for the assembly membraneless organelles, they are related to toxic protein aggregation within neurons. Furthermore, the prion-like spreading mechanism of such aggregates seems to play a role in the dissemination of neurodegeneration. Identifying the mechanisms and structures involved in this process might contribute to the development of new therapeutic approaches against such disorders.

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TL 1105562

DIAGNOSTIC DISCLOSURE OF ALZHEIMER'S DISEASE IN BRAZIL: CURRENT PRACTICE OF PHYSICIANS WHO DIAGNOSE AND TREAT DEMENTIA

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Background: Dementia is a syndrome characterized by cognitive and functional decline. In Brazil, the estimated number of individuals with dementia in 2019 was 1.7 million, with an estimate that it will reach 3.7 million in 2050. Legal, ethical and moral issues permeate diagnostic disclosure of Alzheimer's disease (AD), the main cause of dementia, to patients and families. The right to know the diagnosis, the autonomy of those involved and the principles of non-maleficence and beneficence are mixed with values and attitudes that often do not correlate with bio-ethical and legal ones. **Objective:** To investigate the current practice of physicians who diagnose and treat dementia in Brazil. **Methods:** Observational cross-sectional study. Electronic survey was sent to specialists who are members of the national medical societies of Geriatrics, Neurology and Psychiatry (psychogeriatricians). **Results:** During 41 days (5/9/2022-6/18/2022), 218 responses were obtained. Of the total, 52.8% are men, whose main medical specialties are Neurology (58.9%), Geriatrics (30.7%), Psychiatry (6.9%), Internal Medicine (1.4%), others (2.1%). These professionals are aged between 26 and 84 years (mean = 46.2 years). The majority (58.7%) of the participants have been following patients with AD for more than 10 years. Concerning the disclosure of the diagnosis of AD, only 16.5% have the habit of always revealing it, 50.9% affirm that they usually do it, 20.6% sometimes, and 11.9% rarely. Despite this, 73.4% of physicians believe that patients, in general, want to know their diagnosis. The reasons why professionals decide to reveal or not the diagnosis of AD to their patients are related to the patient's desire to know it, the opinions of the patient's family about the disclosure, and the severity of dementia, which were indicated by 73.9%, 62.4% and 56.4% of the participants, respectively. On the other hand, 96.3% of physicians would like to know their diagnosis, if they were diagnosed with AD. Of these, 50.9% would like to be better prepared for the future. **Conclusion:** The diagnostic disclosure of AD by specialized physicians is still not very common in Brazil, which is related to several reasons. More studies and initiatives are necessary to discuss the benefits and harms that diagnostic disclosure can bring and to better understand the demands of patients and families.

Neurologia Cognitiva e do Envelhecimento

TL 1105623

COGNITIVE COMPLAINTS ASSESSMENT AND NEUROPSYCHIATRIC DISORDERS AFTER MILD COVID-19 INFECTION

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Background: There are an increasing number of reports of persistent and prolonged effects after the acute phase of COVID-19. Damage to the CNS can have a long-term negative impact on cognitive function, daily functioning, and quality of life, even several months after recovery from COVID-19. Reports of acute cognitive complications, such as attention and dysexecutive symptoms, are also emerging. **Objectives:** To analyze cognitive impairment related to long term COVID-19 and its correlation with anxiety, depression, and fatigue in patients with mild COVID-19. **Methods:** This is a cross-sectional study performed in a cohort of 127 patients with COVID-19. These patients were regularly followed up from September 2020 to September 2021. Tests to screen for neuropsychiatric symptoms included Mini-Mental State Exam 2 (MMSE-2), Symbol Digit Modalities Test (SDMT), and Hospital Anxiety and Depression Scale (HADS). **Results:** Tests were performed a median of seven months after COVID-19 infection, with the test application time varying between one and 18 months. SDMT was abnormal in 22%, and the BV version of MMSE-2 was abnormal in 16.5%, while the EV and SV versions were normal. There were significant positive correlations between the versions of the MMSE-2 and SDMT ($p < 0.001$ for all tests). In contrast, there were significant negative correlations between the different versions of the MMSE-2 and SDMT with depression ($r = -2.88$, $p < 0.001$ and $r = -0.397$, $p < 0.001$, respectively) and anxiety ($r = -0.175$, $p = 0.049$ and $r = -0.198$, $p = 0.026$, respectively). There was a significant negative correlation with SDMT ($r = -0.212$, $p = .017$), but not with MMSE-2 ($r = -0.149$, $p = 0.094$). However, the presence of a greater degree of these neuropsychiatric conditions correlated little with worse performance on the test. **Conclusion:** Even in those with a mild presentation, cognitive impairment complaints were frequent in patients with COVID-19. Screening tests, such as SDMT, helped confirm changes in the attentional domain and processing speed, especially in patients with cognitive impairment complaints after infection. Although manifestations such as fatigue, depression, and anxiety were frequent in the post COVID-19 phase, especially in those with complaints of cognitive deficits, these three conditions that are known to contribute to cognitive impairment showed little correlation with worse performance on rapid screening tests.

Neurologia Cognitiva e do Envelhecimento

TL 1105636

ASSESSMENT OF QUALITY OF LIFE AFTER MILD COVID-19 INFECTION

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Background: The incidence of post-COVID-19 sequelae is estimated between 10 and 35%. The COVID-19 infection affects people of all ages, generates a disease burden that can reduce work skills, cause social, psychological and neuropsychiatric challenges. These long-term symptoms could cause a significant effect on the Post-COVID-19 quality of life. **Objectives:** Evaluate the quality of life of patients after being affected by COVID-19, which domains are most prevalent in Short Form-36 (SF36) Scale and the presence of anxiety, depression, fatigue, chronic headache, chronic myalgia, ageusia and olfactory disorders after infection. **Methods:** This is a cross-sectional study performed in a cohort of 143 patients with COVID-19. These patients were regularly followed up from September 2020 to September 2021. Tests to screen for symptoms included the SF36 which range from 0 to 100, fatigue severity scale (FSS), Hospital Anxiety and Depression Scale (HADS) and yes and no questionnaires. **Results:** Median age was 41 (interquartile range (IQR 29-50) years; 108 patients (75.5%) were female. In the assessment of quality of life by the SF-36, the domain most affected was the physical aspects, with an average of 41. Then, the domain's vitality and emotional aspects showed low performance, with an average of 45 each one. The best scored domain was the functional capacity, with an average of 67 points. More than a half of the sample reported cognitive impairment (55.9%) and a minority reported chronic headache (23.1%) and myalgia (11.2%). Just 35.7% complained of chronic fatigue, but 71.3% scored in the FSS with the presence of fatigue. Regarding screening for depression and anxiety, 40.6% and 52.5% scored positively, respectively. **Conclusion:** The prevalence of fatigue, anxiety and reported cognitive impairment resulting from COVID-19 infection is high. The aspects of quality of life deserve attention which are observed in low scores in SF-36. More studies are needed, especially regarding the causes of impact quality of life and possible causes correlated.

Neurologia Cognitiva e do Envelhecimento

TL 1105660

USING AN EXTERNAL CONTROL TO CONTEXTUALIZE EFFICACY DATA FROM PATIENTS WITH PRODROMAL AND MILD ALZHEIMER'S DISEASE TREATED WITH GANTENERUMAB IN SCARLET ROAD AND MARGUERITE ROAD OPEN-LABEL EXTENSION STUDIES

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Gantenerumab, a human monoclonal antibody targeting aggregated beta-amyloid (Aβeta), is a potential disease-modifying treatment for early (prodromal-to-mild) Alzheimer's disease (AD). SCarlet RoAD (SR) (NCT01224106) and Marguerite RoAD (MR) (NCT02051608) were two Phase III trials with open-label extensions (OLEs), during which participants were treated with up to 1,200 mg/month of subcutaneous gantenerumab for up to 5 years. This work aims to evaluate the gantenerumab treatment effect during the SR and MR uncontrolled OLE studies by using an external control group based on the Alzheimer's Disease Neuroimaging Initiative (ADNI) cohort. Participants from the ADNI studies were weighted using the inverse probability of treatment weighting method to make their demographics and baseline characteristics more comparable to the patients in the OLE stages of the pooled SR and MR studies. In the weighted sample, the treatment effects of gantenerumab, per change from the OLE Baseline in CDR-SB, ADAS-Cog13, MMSE at Weeks 104 and 156 were investigated by a mixed-effect model of repeated measure. Participants (N = 164) from the SR and MR OLE studies and 1,218 participants from ADNI were selected. After weighting, the demographics and baseline characteristics were comparable between the two populations. Relative reductions between the pooled participants in SR and MR OLE studies and weighted ADNI participants were found for CDR-SB and ADAS-Cog13 at weeks 104 and 156, with greater reductions in SR and MR participants. **Conclusion:** When comparing participants in the SR and MR OLE studies to a matched ADNI control group, a slower progression was observed with gantenerumab treatment.

Neurologia Cognitiva e do Envelhecimento

TL 1105665

NEUROFILAMENT LIGHT CHAIN, ?-AMYLOID 40, ?-AMYLOID 42 AND TAU LEVELS IN PARKINSON'S AND CHAGAS DISEASE

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Background: The use of serum biomarkers as Neurofilament Light Chain[NfL], ?-amyloid40[?A40], ?-amyloid42[?A42] and Tau protein have been addressed for diagnosis risk and prognostic assessment of neurologic disorders, as marker of neurodegeneration. It is known that Chagas Disease (CD) is associated to stroke and cognitive impairment independently of cardiac disease severity. In this context, some of serum biomarkers may be altered in CD. **Objectives:** To determine if serum NfL, ?A40, ?A42 and Tau levels are altered in patients with CD when compared to controls and Parkinson's Disease (PD). **Methods:** Cross-sectional study of adult (? 18 years) patients from a heart failure (HF) and movement disorders outpatient clinics. CD and control patients fulfilled Framingham criteria for HF; and had no history of stroke, recent thrombotic event or neurodegenerative diseases. CD was defined by a positive ELISA serologic test. The PD group fulfilled UK Bank criteria for PD; and had no dementia or parkinsonism of any other etiologies. NfL, ?A40, ?A42 and Tau were analyzed by single molecule array (SIMOA) technique. Multivariable analysis was performed using linear regression adjusted by age and sex. **Results:** NfL was obtained from 22 patients with CD, 51 with PD and 24 controls. The median NfL was 7.73pg/ml in CD, 18.84 in PD and 8.81 in controls. NfL concentration adjusted for age/sex was 13.54pg/ml higher in PD compared to controls ($p=0.006$), but not different in CD group comparing to controls ($p=0.24$). ?A40 was analyzed in 11 patients with CD, 50 with PD and 6 controls. Median ?A40 was 0.73pg/ml in CD, 209.8 in PD and 3.01 in controls. Adjusted ?A40 concentration was 207.07pg/ml higher in PD compared to controls ($p<0.001$) and there was no difference of ?A40 levels in CD patients ($p=0.77$). Regarding ?A42, were 14 samples of CD, 50 of PD and 5 of controls. The median was 0.41pg/ml in CD, 10.29 in PD and 0.75 in controls. Adjusted ?A42 was 9.54pg/ml higher in PD comparing to control ($p<0.001$). CD had no effect on ?A42 values ($p=0.70$). Finally, Tau presented median of 0.72pg/ml in CD, 2.17 in PD and 0.48 in controls, with 20 samples in CD, 50 in PD and 15 in control. In the adjusted analysis, CD and PD had no effect on protein Tau levels comparing to controls. **Conclusion:** NfL, ?A40 and ?A42 were increased in PD when compared to controls. None of the serum biomarkers were different in CD compared to controls, suggesting that CD may affect cognition through different pathways.

Neurologia Cognitiva e do Envelhecimento

TL 1105723

ASSESSMENT OF MESIAL TEMPORAL LOBE ATROPHY USING THE MEDIAL TEMPORAL LOBE ATROPHY SCORE AND HIPPOCAMPAL VOLUMETRY IN THE BRAZILIAN POPULATION

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Background: The reference studies to determine cut-off parameter for biomarkers of neurodegeneration, such as the medial temporal lobe atrophy (MTA) score and hippocampal volumetry, in Alzheimer's disease (AD), were performed in high-income countries. There are few studies in low-income countries, and it was not found previous national research to establish the cut-off parameter for the Brazilian population. **Objective:** To establish diagnostic criteria for the MTA score and hippocampal volumetry based on the Brazilian population and to determine if there is a correlation between these two techniques for measuring the hippocampus. **Method:** The study is composed of 149 participants (73 controls, 44 with mild cognitive impairment (MCI) and 32 with probable early-stage AD from NAPNA and Foss co-hort). The automatic measurement of the hippocampal volume was performed (FreeSurfer software version 6.0) and two independent observers assigned the MTA score. **Results:** Based on the AUC-ROC, it was proposed as the cut-off value for the individuals aged < 75 years: MTAm_{mean} ?2 (sensitivity (Sens): 61% and specificity (Spec): 85%) and MTAw_{mean} ?3 (Sens: 47% and Spec: 82%) whilst for those ? 75 years: MTAm_{mean} ?3 (Sens: 64% and Spec: 91%) and the MTAw_{mean} was not statistically significant. The cut-off by the AUC-ROC of volumetry showed statistical significance only in the mean volume of those aged < 75 years being 3430mm³ (Sens: 56% and Spec: 88%). Considering the 95th percentile of controls, the proposed normality value suggested for individuals < 75 years: a score of 0 and 1 of the MTAw_{mean} and a mean volume of the hippocampal > 3013mm³ and for those ? 75 years: a score of 0 to 2 of the MTA worst and a mean volume of the hippocampal > 3115mm³. The premorbid volume of the left hippocampus is lower than the right (VL = 3893mm³ vs. VR = 3729mm³, D de Co-hen's D = 0.73, $p<0.001$). There is moderate correlation (RS = -0.496, $p<0.001$) between mean hippocampal volumetry and MTA Scale score. Moderate inter-observer agreement (Kappa (k) = 0.578) and substantial, nearly perfect intra-observer agreement were obtained from the radiologist (k = 0.674) and neurologist (k = 0.871) respectively. **Conclusion:** Considering the diagnostic performance of the Scheltens Scale, we suggested to use a MTAm_{mean} ?2 (Sens: 61% and Spec: 85%) in those aged < 75 years and a MTAm_{mean} ?3 (Sens: 64% and Spec: 91%) in those aged ? 75 years. Moreover, the MTA cut-off score of the Brazilian population was slightly higher than the one described on the literature and new studies with larger samples are needed to confirm these findings. The normality values based on the healthy population studied were similar to those described in the literature.

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TL 1105747

POST-STROKE COGNITIVE ASSESSMENT IN PATIENTS FROM A SPECIALIZED OUTPATIENT CLINIC

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Background: The occurrence of cognitive impairment after stroke is already studied in the literature. 50% of stroke survivors will have some degree of cognitive impairment, of these approximately 66.7% will have mild cognitive impairment (MCI). Some risk factors, such as the NIHSS, are associated with greater impairment. **Objective:** To assess the prevalence and predictors of cognitive dysfunction in a stroke outpatient clinic in a reference Hospital in Salvador - Bahia. **Method:** This is a cross-sectional, descriptive and analytical study carried out through data collection via medical records at the stroke outpatient clinic located at the Professor Edgard Santos University Hospital Complex of the Federal University of Bahia - Professor Francisco Magalhães Neto Outpatient Clinic, of patients treated during the period of January 2021 to October 2021. Among the variables analyzed are: Presence of systemic arterial hypertension, Diabetes Mellitus, acute coronary disease, current smoking, type of stroke if ischemic or hemorrhagic, use of antihypertensive drugs and which class, Pre-stroke rankin, NIHSS Stroke Scale, Anxiety and Depression Scale (HAD) and Mini Mental State Examination (MMSE) scores and **Results:** of complementary exams which can be cranial tomography or cranial resonance through which the location of the stroke will be identified. **Result:** There was a 71.4% prevalence of post-stroke cognitive impairment and the most important predictor was low education and prior cognition. **Conclusion:** There is a high prevalence of post-stroke cognitive impairment. Thus, it is important to act on early diagnosis and rehabilitation.

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TL 1105772

EPIDEMIOLOGICAL PROFILE OF PATIENTS HOSPITALIZED FOR ALZHEIMER'S DISEASE IN BRAZIL OVER THE LAST 10 YEARS

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Background: Alzheimer's disease (AD) is a progressive neurodegenerative disorder, which is manifested by cognitive degradation. From a neuropathological point of view, the brain of individuals presents diffuse cortical atrophy, neuronal loss, an excessive number of senile plaques, and neurofibrillary tangles. Knowledge of the epidemiological panorama of patients Hospitalized for this neurological pathology is of fundamental importance since it is the main and most common representative of the class of dementia, as well as being essential for the implementation of public policies and more effective social measures in the population-target. Since it is still a recent post-COVID period, we have some greater care to be taken regarding this population, whereas this period has further worsened their vulnerability and mortality. In addition to cognitive changes, motor abnormalities may also be observed in AD patients. Gait disturbances, decreased limb strength, and changes in postural control may be present in the initial phases of dementia or even in the pre-clinical stage of AD, thus referring to a greater need for support, especially from March 2020, due to the social distancing produced, and thus more unreported or notified cases already in urgency. **Objective:** Analysis of the epidemiological profile of patients who required Hospitalization for Alzheimer's disease, between 2012 and 2021, in Brazil. **Methods:** Quantitative and retrospective study, performed using secondary data from DATASUS on the monthly/annual frequency of admissions related to AD from July/2012 to June/2021. Sex, race/color, age range, the character of care, mean Hospitalization rate, and deaths were analyzed. **Results:** 12246 admissions were reported, with 2382 belonging to capitals. Of the total, we have 7969 female and 4277 male Hospitalizations, the majority being white (5294 cases). The most affected age group is 80 years and over (7073), with a peak in 2019 (932). Out of these, 9957 were urgent and 2289 elective cases. The average spending was R\$1832.23, with men generating higher expenses (R\$1921.28 versus R\$1784.43), as well as elective care (R\$5157.06 versus R\$1067.89). The total number of deaths was 551, led by São Paulo (185). **Conclusion:** It is concluded that white women, 80 years old and over, living in the country and under emergency care are the group with the greatest vulnerability to this disease.

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TL 1105776

CLINICAL AND EPIDEMIOLOGICAL PROFILE OF PATIENTS DIAGNOSED WITH TRANSIENT GLOBAL AMNESIA IN CASCAVEL, PARANÁ, BRAZIL

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Background: Transient global amnesia (TGA) is a neurological syndrome defined by sudden and severe alteration of recent memory, with resolution within hours. During the event, the patient has impaired memory of recent events, as well as becoming repetitive. However, despite the temporo-spatial disorientation, the other cognitive abilities remain preserved. Deficits tend to resolve within 24 hours and the prognosis is favorable in most cases. Although there is no specific treatment, it is necessary to treat any cardiovascular risk factors that may be present. MRI can reveal a focus of hyperintensity in the hippocampus through diffusion-weighted sequence (DWI). **Objective:** The objective of this study is to identify the epidemiological profile of patients who had TGA and were treated at a private neurology clinic in the city of Cascavel (PR), from 2014 to 2022. **Methods:** This is an observational, retrospective and descriptive study involving patients who presented transient global amnesia during the period of 2014 to 2022 and were treated at a private neurology clinic in Cascavel, Paraná. The variables analyzed were sex, age, risk factors, initial symptom, approximate duration of the symptom and changes in magnetic resonance imaging. **Results:** 19 (n=19) patients diagnosed with transient global amnesia were identified, with a predominance of females (78,94%, n=15). The age had a median of 64 years. The main symptom mentioned was "forgetfulness", followed by "becoming repetitive" and "confusion". The duration of the symptoms had a median of approximately 10 hours. The most lasting symptom had 24 hours and the most fast had three hours. MR, in most cases, did not show any alteration. In a few cases (n=4), microangiopathy was evidenced. The comorbidities related to the patients were hypertension (n=9), Dyslipidemia (n=3), and less often, hypothyroidism, hyperthyroidism, migraine, diabetes melitus, Anxiety, rheumatoid arthritis and arrhythmia. Only two patients had a repeat episode of amnesia. The neurological exam was normal in almost all patients, except by one, who had tremor at rest. **Conclusion:** Transient global amnesia is a syndrome whose pathophysiology is not fully understood. Its evolution tends to be benign, and there are usually no recurrences. It is highly related to cardiovascular risk factors, especially hypertension. More studies are needed to understand its development.

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TL 1105851

ALCOHOL AS A RISK FACTOR FOR DEMENTIA SYNDROMES

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Background: Alcoholism is a chronic disorder characterized by long periods of abstinence associated with periods of alcohol consumption relapses. It is already known that exposure to excess alcoholic beverages for a long period will lead to brain damage, such as reduced concentration, recent memory and can lead to pathologies such as dementia. Dementia is a persistent cognitive syndrome, with decreased functionality and changes in at least two cognitive domains. Under this question, many studies have done analyzes on the effects of chronic alcohol use and the brain damage that may be related. Because of these studies, several pathways for this damage have been suggested, including the neurotoxic effects of thiamine, ethanol, and acetaldehyde deficiency. **Objetivo:** Avaliar a relação entre o consumo inadequado de álcool e sua relação com a progressão das Síndromes Demências, investigando a quantidade e os limites entre o benefício e o dano no consumo de álcool. **Methods:** This is a systematic literature review based on data extraction in indexes such as PUBMED and MEDLINE. Publications between 2018 and 2022 were considered, in the English language and with the following descriptors: Alcohol, Dementia, Neurology. **Results:** The beginning of the discussion about the positive or negative effects of alcohol is based on the lack of basic guidelines on the amount and frequency of alcohol intake that would be adequate to guarantee health benefits or harm. To fill this gap, the research cites important points about the levels of alcohol consumption, and indicates that light to moderate consumption of alcohol does not imply a high risk for causes of dementia, however when this intake goes to a high level, the risks of dementia have already been shown to be significantly higher. 8 drinks per week for women and 15 drinks per week for men is found to be significantly associated with a consistently high trajectory of cognitive function and a lower rate of cognitive decline. **Conclusion:** Notably, the relationship between alcohol and progression of dementia syndromes is related to the level of ethanol intake, in which low to moderate intakes present a lower risk for progression of brain damage than people with high alcohol intakes.

Neurologia Cognitiva e do Envelhecimento

TL 1105856

PARKINSON'S DISEASE AND PSYCHOSIS: A SYSTEMATIC REVIEW

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Background: Parkinson's disease (PD) is a loss of dopaminergic neurons in the substantia nigra present in the midbrain and in the development of Lewy corpuscles. The pathophysiology of PD may be related to environmental factors (aging, family factors) and genetics. Diagnosis is based primarily on clinical findings and can be characterized by motor and non-motor symptoms, classically resting tremor, rigidity, bradykinesia, and hunched posture. When related to non-motor factors, PD can be associated with depression, dementia, autonomic dysfunction and psychosis. Parkinson's disease psychosis (PDP) encompasses minor phenomena such as illusions, hallucinations, and non-visual delusions and can commonly be associated with poorer quality of life. **Objective:** To analyze the presence of Psychosis in patients with Parkinson's Disease, to point out ways to improve these symptoms and consequently guarantee a better quality of life for both the patient. **Methods:** This is a systematic literature review based on data extraction in indexes such as PUBMED and MEDLINE. Publications between 2018 and 2022 were considered, in languages and with the following descriptors: Parkinson disease, neurology, psychosis. **Results:** The presence of symptoms of psychosis in patients with PD are indicative of a worsening in quality of life, because this symptom is related to sleep disorders, cognitive impairment, depression and, consequently, with the advancement of the disease's pathology. Because of this multifactorial relationship, the treatment of PDP requires investigation of the triggering symptoms, reducing or discontinuing measurements that can induce psychosis, and starting with non-pharmacological treatments. As for pharmacological medications that can prevent this condition, clozapine remains the most indicated drug for the treatment of PDP. As for pharmacological medications that can prevent this condition, clozapine remains the most indicated drug for the treatment of PDP. **Conclusion:** It is concluded that the way to treat psychosis in patients with PD involves the need to ensure quality of life for patients. As there is no complete cure for the condition, what becomes important in these conditions is to delay the evolution of the pathogenesis so that the patient has a longer and better survival.

Neurologia Cognitiva e do Envelhecimento

TL 1105890

A REVIEW OF THE LITERATURE ABOUT DEMENTIA IN AGED PATIENTS POST-COVID 19 INFECTION

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Background: With the pandemic, frail elderly people, with various comorbidities and more susceptible to having complications related to infection, were targeted with demanding restrictions and with delays in socializing. Therefore, there is a hypothesis of cognitive worsening in the elderly with dementia, as well as increased manifestation of behavioral and psychiatric symptoms. **Aims:** Analyze the impacts related to cognition in the elderly in the Covid-19 pandemic. **Methods:** A literature review was developed in three steps: Development of the research question, search for scientific articles in the Pubmed database, and critical analysis of included articles. The search was conducted in March 2022, and articles between 2019 and 2022 were selected, for a total of 34 articles were obtained of which 9 were used. Of these, 7 were cohort studies and 2 cross-sectional studies. **Results:** The effect on the cognition of the elderly in isolation is evident, due to reduced contact with family and society, and restricted access to health services during the pandemic, making it difficult to monitor cognitive disorders. Studies on the elderly have shown cognitive worsening and increased behavioral changes such as depression, apathy, anxiety, irritability, sleep disturbance, and agitation. **Conclusion:** More studies are needed to better understand the worsening of dementia post-covid. Furthermore, care alternatives for monitoring the elderly with dementia, such as telemedicine, should be improved.

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TL 1105914

IMPACT OF THE SARS-COV-2 PANDEMIC ON ALZHEIMER'S DISEASE PATIENTS: A RETROSPECTIVE AND EVOLUTIONARY ANALYSIS OF COGNITIVE AND FUNCTIONAL ASPECTS

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Background: Patients with Alzheimer's Disease (AD) are at greater risk not only of contracting COVID-19, but of increased physical and psychological suffering from the pandemic and greater vulnerability to accelerated functional and cognitive decline. Thus, it is necessary to understand the clinical and social impact of the SARS-Cov-2 pandemic in this group. **Objective:** The objective was to evaluate an evolution of the cognitive and functional performance of AD patients from 2018-22. **Methods:** A retrospective cohort of 74 AD patients treated at a tertiary Hospital in Fortaleza was carried out, from 2018-22. Cognitive assessments were carried out, longitudinally and retrospectively, with the Mini-Mental State Examination (MMSE), Semantic Verbal Fluency (SVF) and Clock Design Test (CDT); with the Clinical Dementia Rating Scale (CDR) and Functional Assessment Staging (FAST), in addition to assessments that sought to characterize neuropsychiatric symptoms, medication use, social isolation, caregiver stress and satisfaction with telemedicine care. We used MMSE, FAST 2(1-17) and 3(1-7) to define significant decline; the last two being, respectively, numerical encodings of the FAST categorical variables. The evolution of the data in relation to the beginning of the pandemic was recorded as: before (2018-19), after (2020-22) and during the transition to the beginning of the pandemic (2019-20). The annual evolution was then compared: 2018-19x2019-20x2020-22. **Results:** We evidenced that in four years of follow-up there was a functional deterioration, disproportionately between 2019-2020; assessed by increasing the CDR from 1 to 2; increase in FAST 2 from 4,5 to 6,5 and in FAST 3 from 4,5 to 6; $p < 0.05$. The last two configured significant functional decline. There was an increase in the prevalence, over the four years, of the use of anticholinesterases, anti-NMDA and antipsychotics (36,5x74%; 20x46%; 19x45%, respectively) and their side effects. We also found a higher prevalence of urge-incontinence (1,4x38%) and of some neuropsychiatric symptoms, such as anxiety symptoms (35x58%) and insomnia (18x40,5%). Cognitive worsening was evidenced, disproportionately between 2019-2020; through the MMSE, SVF and CDT (15x11; 6x4,5; and 2x0, respectively; $p < 0,05$), the first of which configured significant cognitive decline. There was a significant increase in caregiver stress (11x74%). **Conclusion:** Patients with AD showed significant cognitive and functional decline after the beginning of the pandemic.

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TL 1105933

ACOMETIMENTO NEUROLÓGICO E COGNITIVO NA COVID-19: CARACTERIZAÇÃO CLÍNICA E EPIDEMIOLÓGICA

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Background: COVID-19 neurological manifestations were demonstrated during the pandemic, including cognitive impairment. **Objectives:** To determine the prevalence of cognitive and behavioral complaints (such as dementia, MCI or SCD) in a outpatient sample with recent SARS-COV2 infection. **Specific:** Evaluate the association of cognitive impairment with the presence of the polymorphism found in the APOE gene and with respiratory disease. **Methods:** Observational, longitudinal, prospective clinical study. **Inclusion criteria:** patients with confirmed Covid-19. Patients are evaluated in an outpatient clinic. They are evaluated through a standardized attendance record, with somatic and cognitive neurological assessment. Cognitive assessment involves the application of cognitive (ACER, MMSE and CDR), functional (Pfeffer) and psychiatric (GDS or Beck) screening instruments, in addition to subsequent extensive neuropsychological assessment. In addition, APOE polymorphism is analysed. **Preliminary Results:** To date, 225 patients were evaluated. The average age is 46.5 years, with 65.4% female, 79.16% with 8 or more years of schooling, in addition to 57.5% of the sample with cognitive complaints. **Conclusion:** The **Results:** so far in our study demonstrate that cognitive complaints are frequent in patients even in the chronic phase of the disease.

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TL 1105935

FRANK'S SIGN AS A PREDICTOR OF CARDIOVASCULAR DISEASE AND COGNITIVE IMPAIRMENT IN AN ELDERLY POPULATION: A CROSS-SECTIONAL STUDY

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Background: Frank's sign consists of a diagonal crease in the earlobe that extends from the tragus to the inferior-posterior edge of the auricle. First described as a predictor of cardiovascular diseases (CVDs), it was related to atherosclerosis, being later considered an independent cardiovascular risk factor and associated with cerebrovascular events and cognitive impairment. **Objective:** To assess the prevalence of Frank's sign in a general population aged 60 years or older regardless of the presence of cardiovascular risk factors and to evaluate the presence of this sign as a predictive factor of CVDs and cognitive impairment (CI). **Method:** Through a convenience sampling, we randomly recruited patients aged 60 years or older, not Hospitalized, from health services not related to cardiovascular affections from ABC Region and Itapeverica da Serra, in Greater São Paulo, who had their ears photographed in profile, responded to either the Mini Mental State Examination or the Montreal Cognitive Assessment depending on years of education, and a questionnaire with epidemiological and clinical variables. This study was conducted according to the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) Statement guidelines. **Results:** We evaluated 500 individuals, with 57% were women; 57% were white, 39% were black and 4% were asian; 55% had between 60 to 69 years; 64% had the sign; Frank's sign was associated with older people ($p=0.015$), white population ($p<0.001$), women ($p<0.01$), dyslipidemia ($p=0.044$) and had a tendency of association with hypertension ($p=0.058$), while there was no association with CI neither cardiovascular events as myocardial infarction or stroke. **Conclusion:** Frank's sign prevalence is more frequently found in white elderly women and doesn't seem to be a good predictor of CVDs neither CI, however, it showed effect with cardiovascular risk factors. Prospective studies should be taken to assure whether this sign is a marker for CVDs in this population.

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TL 1105949

ANALYSIS OF THE EPIDEMIOLOGICAL PROFILE OF HOSPITALIZATIONS IN PUBLIC HEALTH SYSTEM OF BRAZIL FOR DEMENTIA BETWEEN 2019 AND 2021

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Background: Dementia is a clinical syndrome characterized by gradual cognitive decline, involving at least two cognitive domains, associated with impaired functionality. Dementias usually have a progressive course, which leads to disability in different timing and degrees, and is associated with infectious and vascular complications, which often lead these patients to Hospital services. **Objective:** To analyze the epidemiological profile of Hospitalizations for dementia in the SUS in Brazil between the years 2019 and 2021. **Methods:** The research is an observational and cross-sectional study of Hospitalizations for dementia in the SUS in the years 2019 to 2021. The collection was carried out in the DATASUS database, with investigation of the variables: region, age group, gender, number of Hospitalizations, total value, mortality rate and days of stay. **Results:** In the period, there were 7908 Hospitalizations with 56% of cases occurring in the Southeast. The largest shares of Hospitalizations were between 60 and 69 years (16.7%), 70 and 79 years (23%), and more than 80 years (26.2%). The number of Hospitalizations was highest in the first year, with 2963 cases, while it was lower in the following year with 2421 cases and 2524 in the third. The average values per Hospitalization were R\$5381, R\$6323, and R\$5847 in 2019, 2020, and 2021, respectively. Total costs were R\$46M, mostly represented by the 40 to 49 year old age group, whose expenses were R\$12.1M. The average mortality rate was 11.2% for the period, with higher rates in the Southeast. The group between 40 and 49 years had more days of Hospitalization in each year, with 26.7% of the total, followed by the group over 80 years, which had 16.8% of the days. **Conclusion:** From the analysis of the number and mortality rate of cases, it is suggested that the long-term complications of dementias, which culminate in the Hospitalization of these patients, are a major health problem, associated with high total costs and values per Hospitalization still higher than the average monthly income for the period, and further studies are needed to measure the impact of the condition in the country. Furthermore, the preponderant regions were the Southeast and South, despite the population of the latter being smaller than the Northeast in numbers. Interestingly, it was shown that the groups with more Hospitalizations did not correspond to higher total expenditures or length of stay, even when analyzing the year individually.

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TL 1105988

RECRUITMENT ANALYSIS BY MEDIA IN A STUDY OF SUBJECTIVE COGNITIVE DECLINE

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Background: Subjective Cognitive Decline (SCD) is conceptualized as cognitive complaints in cognitively unimpaired (CU) individuals and is associated with progressive cognitive impairments. Recruiting these individuals can be challenging, especially because of the low education levels and limited access to information by a portion of the Brazilian population. **Objective:** We aimed to analyze the initial recruitment process through media in an ongoing study about SCD at the Hospital de Clínicas de Porto Alegre (HCPA). **Methods:** We analyzed the recruitment process through electronic (HCPA website and Twitter) and written local media of the participants in the study "Subjective Cognitive Decline and Cognitive Reserve: a multimodal approach." This study evaluated CU individuals aged 65+ years-old, with educational levels < 5 or > 10 years and with cognitive complaints. Exclusion criteria were the presence of uncontrolled psychiatric or clinical illness, cerebrovascular disease, and the use of psychotropic drugs. The recruitment was carried out exclusively by phone calls in two phases. In the first one, the exclusion criteria were applied and, in the second, the following scales were performed: Modified Telephone Interview for Cognitive Status (TICS-M); Generalized Anxiety Disorder Assessment (GAD-2); Geriatric Depression Scale; and the Subjective Cognitive Decline Scale. **Results:** From February to May 2022, 169 individuals presented interest in participating in the study. Most were women (74%; n=125), with a median age of 71 years-old [interquartile range (IQR): 67-74], a median of 11 years of formal schooling (IQR: 8.5-16), and most lived in Porto Alegre (71.6%; n=121). Most of the exclusions in phone call 1 were due to clinical illness (14.8%; n=25), cerebrovascular disease (5.9%; n=10), and off-target education (4.7%; n=8). The majority (62.7%; n=106/169) went to phone call 2 and, at the moment, 78 individuals were assessed. Of these, most were excluded because presented a TICS-M score below cut-off (44.9%; n=35/78) and absence of consistent cognitive complaints (23%; n=18/78). **Conclusion:** Most individuals recruited by electronic and written media were women with a high educational level. The main causes of exclusion were cognitive decline and the absence of consistent cognitive complaints. This assessment reveals the difficulty in recruiting SCD individuals through media and indicates the need for alternative recruitment methods Neurologia Cognitiva e do Envelhecimento

TL 1106068

KNOWLEDGE AND ATTITUDES ABOUT DEMENTIA OF PRIMARY CARE PHYSICIANS OF RIO GRANDE DO SUL: PRELIMINARY ANALYSIS

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Background: Knowing the knowledge and attitudes about dementia of primary care physicians (PCP) in the state of Rio Grande do Sul (RS) is essential for the establishment of educational measures. **Objectives:** To assess knowledge attitudes about dementia among PCP in RS. **Methods:** A form was sent by e-mail to the physicians of primary care in RS. We collected sociodemographic data, volume of patients with dementia treated and perception of difficulties in caring for these patients. A previously validated questionnaire was sent: "Quiz on Knowledge and Attitudes in Dementia". In addition to the descriptive analysis and comparison of data between the health macro-regions of the state, we performed principal components analysis in the results of the attitude quiz. **Results:** From March to April 2022, 147 PCP (from 135 municipalities) responded to the form. About half were women (51%; n=75) and the median age (interquartile range - IQR) was 35 (30-42) years-old. Most worked in Family Health Strategy teams (93.9%) and the time as a physician was 7 (4-14) years. Most had training in Family and Community Medicine (40.8%) or had no specialization (40.1%). The Missioneira (Northwest) region (21.8%) was the one with the most responses and the South region (7.5%) had the least. Most physicians see less than 5 patients with suspected dementia per month (71.4%) and 27.2% said refer less than 5 patients/month to tertiary care. The main reasons for referral were difficulty in drug management (35.4%) and behavioral difficulties (29.9%). The Mini Mental State Examination was the most used test, both as a screening (65.3%) and in clinical suspicion (79.6%). Less than half of the physicians perform cognitive screening (47.6%), mainly because of the patient's complaint (66%) and the presence of a family history of dementia (38.8%). The mean percentage of correct answers in the knowledge quiz was 42.9%, with no difference between the regions of RS. In the attitude quiz, the principal component analysis identified 2 factors: 1) frankly positive attitudes towards patients with dementia and 2) speed in revealing the diagnosis, but with a greater tendency towards referral. **Conclusion:** Knowledge about dementia is low among PCP in RS; however, most have positive attitudes towards these patients. Increasing the sample size may provide information about differences between regions in the state.

Neurologia Cognitiva e do Envelhecimento

TL 1106075

INFLUENCE OF DEPRESSION, INDEPENDENCE IN ACTIVITIES OF DAILY LIVING AND CLINICAL VARIABLES ON THE QUALITY OF LIFE OF ELDERLY PEOPLE WITH TYPE 2 DIABETES MELLITUS

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Background: Considering the currently increasing number of elderly individuals in society, it is necessary to emphasize that these individuals deserve to grow older with autonomy and good quality of life (QoL). **Objective:** To identify the factors, especially modifiable ones, that affect the quality of life (QoL) of elderly individuals with type 2 diabetes mellitus (EIDM2). **Method:** A cross-sectional study evaluated EIDM2 with emphasis on QoL, sociodemographic and clinical data, including depression (geriatric depression scale 15), family functionality/dysfunction (APGAR questionnaire), activities of daily living (KATZ index). Multiple linear regression was performed to assess the impact of factors on the five domains of QoL (WHOQOL-BREF). **Results:** Two hundred and fifty-three EIDM2 were assessed for possible study enrollment. The mean age was of 69.04 (± 6.52) years; the mean diagnostic time of DM2 was 15.49 (± 9.86) years; the highest mean score (on WHOQOL-bref) was in "social relationships" domain of 70.00 (± 15.92); and the lowest mean score in was "physical" domain of 60.74 (± 16.92). 67.9% of the individuals were females; 88.9% of the elderly individuals considered religion to be important; 48.8% of them said that they were not financially self-sufficient; 72.2% had their self-perceived health status as NOT good; 82.5% received polypharmacy; 40.1% of them had high HbA1c levels; 86.9% of the elderly reported absent family dysfunctions; 71.4% showed absent dependence for activities of daily living; 71.4% of them had absent depression. Factors that positively affected QoL were: absent depression (in all five domains), absent family dysfunction (in 3 domains), and absent dependence on activities of daily living (ADL) (in 3 domains). **Conclusion:** The modifiable factors that influenced QoL of EIDM2 were depression and family functionality. Furthermore, to our knowledge, this is the first study to show the importance of family functionality in QoL of EIDM2. Although the most feasible approach to improve QoL is probably to intervene in the above mentioned modifiable factors, it needs to be assessed by other prospective studies.

Neurologia Cognitiva e do Envelhecimento

TL 1106081

COGNITIVE DECLINE AND POSSIBLE BIOMARKERS IN RHEUMATOID ARTHRITIS: A SYSTEMATIC REVIEW

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Background: Cognitive dysfunction is a frequent complaint in patients with autoimmune disease and associated with impaired quality of life and loss of functionality. During the last years, a growing number of studies have investigated the link between cognitive dysfunction and Rheumatoid Arthritis (RA), highlighting the potential pathogenic role of several clinical, psychological, and biological factors. **Objective:** Considering the scarce literature on neuropsychiatric manifestations in RA, as well as the lack of serological markers that can help in clinical practice, we aimed to investigate serological and CSF biomarkers in humans, and its association with cognitive dysfunction in patients with RA. **Methods:** We performed a systematic review using PRISMA protocol. A systematic search was conducted in the PubMed/Medline, Embase, LILACS, Scopus and Google Scholar databases from inception up to November 2021. The inclusion criteria for studies were defined based on the participants involved, type of exposure, type of comparison group, outcome of interest and study design. **Results:** Five original studies were included, which provided data from 428 participants. The studies included plasma proteins, auto-antibodies, cytokines, interleukins, neurotrophic factors and lymphocyte subpopulations. Among plasma proteins, SHH was increased and TTR was reduced in patients with mild cognitive impairment; Anti-MBP and Anti-MOG negatively correlated with memory, executive function and attention. S100 β negatively correlated with memory and executive functions; some lymphocyte subpopulations positively correlated with attention, memory, and executive functions. IL-2, IL-4, IL-6 and TNF

Neurologia Cognitiva e do Envelhecimento

TL 1106110

ANALYSIS OF THE DISTRIBUTION OF DRUGS FOR DEMENTIA DUE TO ALZHEIMER'S DISEASE IN THE STATE OF RIO GRANDE DO SUL

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Background: Drugs for dementia due to Alzheimer's disease (AD) have been part of the SUS Specialized Pharmaceutical Care Component (SPCC) since 2005 and are provided free of charge as long as the request for these drugs meets the criteria of the national guidelines. Assessing the profile of this distribution can reveal inequalities in the supply of these drugs and, indirectly, in the quality of care for Alzheimer's patients. **Objectives:** To evaluate the prescription pattern and the profile of patients who receive prescription of medication for AD in the state of RS. **Methods:** All approved requests for donepezil, rivastigmine, galantamine, and memantine for the SPCC in the state of RS between July and November 2021 were evaluated. Patients' sociodemographic data, scores on the Mini Mental State Examination (MMSE) and Clinical Dementia Rating Scale (CDR), type and dose of the prescribed medication, municipality and health macro-region of origin were obtained. In addition, the human development index (HDI) and the prescription index (patients/100,000 population) of each macro-region were associated. **Results:** In the second half of 2021, 2382 AD patients were receiving some specific AD medication in RS. Most were female (65.5%) and the median age (interquartile range - IQR) was 79 (73.5-84) years. Most patients had education less than 8 years (n=1266; 71.7%). The median MMSE score was 15 (12-18) and most had CDR 2 (47.5%) or 1 (34.8%). In most municipalities, 339 (68.2%), there were patients with some prescription. The macro-regions "Serra" (37/100,000) and Norte (33/100,000) had higher prescription indices and the Sul region had the lowest (7/100,000). There was a strong correlation between prescription index and HDI ($Rho=0.877$; $p<0.009$, Spearman). The most prescribed drugs were donepezil (n=1014; 42.6%) and rivastigmine (n=691; 29%), and 464 (19.5%) patients were using memantine association. The "Centro-Oeste" macro-region showed a higher frequency of patients with CDR=2 (61.2%) and a higher frequency of rivastigmine prescriptions (42.1%) compared to other regions. **Conclusion:** The analysis of the profile of patients with AD who receive medication for the disease in the state of RS reveals some inequality, with less developed regions showing lower frequency of prescription, and prescription for patients in more advanced stages.

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TL 1106144

SUBJECTIVE COGNITIVE DECLINE AND COGNITIVE RESERVE PROJECT: PROTOCOL AND PRELIMINARY RESULTS

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Background: Subjective Cognitive Decline (SCD) is a condition characterized by consistent cognitive complaint in cognitively unimpaired (CU) individuals and is associated with progressive cognitive impairment, especially in the context of Alzheimer's disease (AD). Cognitive Reserve is the capacity to maintain cognitive abilities despite pathological damage. Studying the relationship between these two concepts is fundamental to understand the early stages of AD. **Objectives:** We aimed to show the study protocol and the preliminary **Results:** of the study. **Methods:** CU individuals with > 65 years old with cognitive complaints and with less than 5 or more than 10 years of formal education were recruited. Exclusion criteria were the previous diagnosis of dementia, uncontrolled neuropsychiatric/clinical illness, or cerebrovascular disease. The assessment was performed exclusively by phone calls. It consisted of a brief cognitive (Modified Telephone Interview for Cognitive Status - TICS-M) and anxiety/depression screenings, in addition to the SCD-scale. Individuals recruited were then evaluated in person with an extensive cognitive battery. Blood, cerebrospinal fluid, and saliva were collected to measure AD biomarkers (?-amyloid-42, total/phosphorylated tau), APOE genotyping and magnetic resonance imaging were also performed. **Results:** Between March and April 2022, 11 patients were included (90.1%, n=10/11 were female). Their median interquartile range (IQR) age was 70.5 (68.3-75) years old, with a median of 16 years of formal education for those in the high education group and 4 years for those with low education. The median TICS-M and SCD scores at the recruitment stages were 25 (IQR 20.8-26.5) and 8.5 (IQR 7-9), respectively. The main SCD-plus criteria reported were age at onset of SCD ? 60 years old (100%, n=11/11) and concerns associated with SCD (72.7%, n=8/11). Clinical Dementia Rating (CDR) global score was 0 in most patients (81.8%, n=9/11) - only 2 scored 0.5 (18.2%). In the cognitive battery, deficits were observed mainly in the Digit Span Memory Test (45.5%, n=5/11) and in the Five Digit Test (up to 54.5%, n=6/11). **Conclusion:** Most of the patients included had the diagnosis of SCD, showing that the telephone screening might be effective. The expansion of the sample may help us better understand the impact of cognitive reserve on the early stages of AD.

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TL 1106197

IMPACT OF THE SARS-COV-2 PANDEMIC ON ALZHEIMER'S DISEASE PATIENTS: A RETROSPECTIVE AND EVOLUTIONARY ANALYSIS OF COGNITIVE AND FUNCTIONAL ASPECTS

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Background: Alzheimer's Disease (AD) is a neurodegenerative disorder that manifests as deterioration of multiple cognitive domains and impairment of activities of daily living, for example. Such patients are at greater risk not only of contracting COVID-19, but greater vulnerability to accelerated functional and cognitive decline. **Objective:** The objective was to evaluate an evolution of the cognitive and functional performance of AD patients from 2018 to 2022. **Methods:** A retrospective cohort of 74 AD patients treated at a tertiary Hospital in Fortaleza was carried out, from 2018 to February 2022. Cognitive assessments were carried out, longitudinally and retrospectively, with the Mini-Mental State Examination (MMSE), Semantic Verbal Fluency (SVF) and Clock Design Test (CDT); with the Clinical Dementia Rating Scale (CDR) and Functional Assessment Staging (FAST). We used MMSE, FAST 2 (1-17) and 3 (1-7) to define significant decline; the last two being, respectively, numerical encodings of the FAST categorical variables. The evolution of the data in relation to the beginning of the pandemic was recorded as: before (2018 to 2019), after (2020 to 2021/22) and during the transition to the beginning of the pandemic (2019 to 2020). The annual evolution was then compared: 2018 to 2019 versus 2020 to 2021/22; 2018 to 2019 versus 2020 to 2021/22; 2018 to 2019 versus 2020 to 2021/22. **Results:** We evidenced that in four years of follow-up there was a functional deterioration, disproportionately between 2019 and 2020; assessed by increasing the CDR from 1 (0,5-2) to 2 (1-3); increase in FAST 2 from 4,5 (3-6) to 6,5 (4-9) and in FAST 3 from 4,5 (3-6) to 6 (4-6); $p<0.05$. The last two configured significant functional decline. Cognitive worsening was evidenced, disproportionately between 2019 and 2020; through the MMSE, SVF and CDT scores (15 versus 11; 6 versus 4,5; and 2 versus 0, respectively; $p<0,05$), the first of which configured significant cognitive decline. **Conclusion:** Patients with AD showed significant cognitive and functional decline after the beginning of the pandemic. The follow-up of these patients is necessary, aiming to improve the long-term extension of the effects of the pandemic in this known vulnerable group.

Neurologia Cognitiva e do Envelhecimento

TL 1106216

IMPACT OF THE SARS-COV-2 PANDEMIC ON PATIENTS WITH ALZHEIMER'S DISEASE: A RETROSPECTIVE AND EVOLUTIONARY ANALYSIS OF THE DEGREE OF SATISFACTION WITH TELECARE

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Background: Telemedicine allows Alzheimer's Disease (AD) patients to overcome barriers of access to health care services and increases accessibility for people with cognitive and functional impairments. **Objective:** This study aims to evaluate the feasibility of telehealth activity in the follow-up of patients with AD, focusing on the level of satisfaction with telemedicine care. **Methods:** A retrospective cohort of 74 AD patients treated at a tertiary Hospital in Fortaleza was carried out, from 2018 to February 2022. Throughout the cohort of the present study, questions were asked about telemedicine satisfaction. **Results:** Telemedicine was adopted as a modality in consultations for many patients during the pandemic. Throughout the cohort of the present study, we observed that 35 patients were seen at some point (2020-2021/22) by telemedicine; and that 39 had exclusively face-to-face consultations. In between the 39 patients treated by telemedicine, 29 (83%) patients, caregivers or family members interviewed reported having enjoyed the service and that it was satisfactory to their demands. Regarding the scores for telemedicine consultations, 81% of the interviewees assigned a grade of 8 or higher; 29% rated the service with a grade 10 (ten), 23% with 9 (nine) and 29% with 8 (eight). However, 3% of respondents assigned grade 0 (zero), 3%, grade 4 (four), 9%, grade 5 (five) and 6%, grade 7 (seven). **Conclusion:** This telehealth intervention promoted high levels of satisfaction during the isolation of the pandemic.

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TL 1106219

IMPACT OF THE SARS-COV-2 PANDEMIC ON PATIENTS WITH ALZHEIMER'S DISEASE: A RETROSPECTIVE AND EVOLUTIONARY ANALYSIS OF NEUROPSYCHIATRIC ASPECTS

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Background: Alzheimer's Disease (AD) is a neurodegenerative disorder that manifests as a variety of neuropsychiatric symptoms. Such patients are at greater risk not only of contracting COVID-19, but of increased physical and psychological suffering from the pandemic. Several studies carried out during this period showed the negative biopsychosocial consequences of isolation measures. **Objective:** Evaluate neuropsychiatric conditions in AD outpatients in Tertiary Hospital during covid-19 pandemic from 2018 to 2022. **Methods:** A retrospective cohort of 74 AD patients treated at a tertiary Hospital in Fortaleza was carried out, from 2018 to February 2022. Assessments were performed that sought to identify and characterize neuropsychiatric symptoms. The evolution of the data in relation to the beginning of the pandemic was recorded as: before (2018 to 2019), after (2020 to 2021/22) and during the transition to the beginning of the pandemic (2019 to 2020). The annual evolution was then compared: 2018 to 2019 versus 2020 to 2021/22; 2018 to 2019 versus 2020 to 2021/22; and 2019 to 2020 versus 2020 to 2021/22. **Results:** We observed, when comparing 2018 with 2021/22, an increase in all neuropsychiatric symptoms evaluated. Hallucinations ranged from 14.9% to 35.1%; depressive symptoms from 20.3% to 37.8%; and anxious symptoms from 35.1% to 58.1%. The two most prevalent neuropsychiatric symptoms in 2021/22 were anxious (58.1%) and depressive (37.8%) symptoms. As for the sleep changes, we observed an increase, between 2018-2021/22, in insomnia (from 17.6% to 40.5%); and hypersomnolence (from 0% to 13.5%). **Conclusion:** The social isolation provided by the covid-19 pandemic led to the worsening of neuropsychiatric disorders.

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TL 1106241

COVID19 AND COGNITIVE IMPAIRMENT – A FOLLOW-UP STUDY OF A CLINICAL HOSPITAL IN SÃO PAULO

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Background: Post-COVID19 syndrome is a condition characterized by signs and symptoms that occur within 3 months of the onset of COVID19 and that last for at least 2 months. In the past 2 years, cognitive impairment has frequently been associated with COVID19 with descriptions of attentional, executive, memory, and language disorders. Many studies have assessed these cognitive disturbances using online and telephone tests, often in isolated interviews. Moreover, some morphological and metabolic changes have been described in MRI and FDG PET-CT, respectively. There is no formal treatment for this condition, and more advanced knowledge in this important research in this field is needed. **Objectives:** To characterize the cognitive profile of this population pre- and post-rehabilitation, including neuroimage patterns and the effect of cognitive rehabilitation. **Methods:** We selected patients less than 65 years of age through social media with a history of COVID19, with at least 3 or more months of symptom and no more than 6 months, no relevant structural brain lesions, no history of neurodegenerative disease, important cognitive issues or psychiatric conditions and currently living in the city of São Paulo or nearby regions. They were evaluated in person or by remote video call 3 times by a neurologist (0, 3, and 6 months) with specific cognitive battery tests and questionnaires and then by a neuropsychologist for evaluation and training. Finally, some patients underwent neuroimaging. **Results:** 39 patients performed the first baseline evaluation and 14 completed the 6-month follow-up, 34 completed or were in cognitive rehabilitation, and 19 patients with underwent functional and structural neuroimaging (FDG PET CT/MRI). This sample has a mean age of 46.2 (SD11.4) years, 53% of females, 13.4 (SD3.4) years of education, 18% of hypertension, 6.2% diabetes, 15.6% dyslipidemia, 15.1% stroke/TIA, and 25% obesity. 59% of the assessments were face-to-face, 41% by telemedicine. The cognitive evaluation depicted: 90% had memory complaints, MMSE of 26.6 (SD2.7), delayed recall of 9.2 (SD1.6) out 10; semantic verbal fluency of 16.5 (SD5.1), FAS fluency test 27 (SD12.2), clock drawing test of 8.8 (SD2.2), and Boston naming test of 13.1 (SD2.4) out 15. **Conclusion:** Post-COVID19 syndrome and cognitive impairment are relatively common, and we need prospective studies in our population and other low and middle-income countries.

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TL 1106259

THE ADDENBROOKE'S COGNITIVE EXAMINATION–REVISED (ACE-R) IN THE DIAGNOSIS OF ALZHEIMER'S DISEASE: A NEW WAY TO EXPLORE ITS DIAGNOSTIC PROPERTIES

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Background: The ACE-R is an accurate and brief cognitive battery for the detection of mild dementia, especially for the discrimination between Alzheimer's disease (AD) and frontotemporal dementia. The aim of this study was to develop a new logarithm based on discriminative items of the ACE-R combined with relevant demographic characteristics that may interfere on cognition. **Method:** The ACE-R was administered to 102 patients with mild dementia due to probable AD and 161 controls from two Brazilian research centers. All individuals were submitted to the Mattis Dementia Rating Scale (DRS) and the ACE-R. The performance of the patients was compared and analyzed. Mokken scaling analysis was applied to identify the latent trait on the AD Group. Multivariate logistic regression and ROC curve analysis were carried out. **Result:** Mean \pm SD total scores in the ACE-R were 70.2 ± 10.8 in AD and 85.1 ± 8.2 in controls. AD Mokken ACE-R (AMokACE-R) comprises 12 items measuring the same latent concept. Logistic regression with cross-validation pointed that AMokACE-R + Years of education + ACE-R items Orientation (time) and Memory (name and address recall) share importance as independent variables ($p < 0.05$). The proposed logarithm reached an area under the curve of 0.968, with 93% sensitivity, 94% specificity, 90% PPV and 95% NPV. **Conclusion:** The new logarithm using the ACE-R items achieved high diagnostic accuracy in identifying mild AD versus controls. Furthermore, the final ROC curves showed the superiority of the model proposed in relation to the analysis of the subscales individually, as the literature previously reported. Further analysis in larger samples, with biomarkers or pathological confirmation, are necessary to confirm these findings.

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TL 1106318

VALIDATION OF THE ALTERNATIVE VERSION OF THE BRAZILIAN MONTREAL COGNITIVE ASSESSMENT (MOCA-BR): PILOT STUDY

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Background: The current version of the Brazilian Montreal Cognitive Assessment (MoCA-BR) did not have a reliable cross-cultural adaptation to Brazilian Portuguese. In previous stages of this study, the Alternative Version of the MoCA-BR was developed, with changes in the sections: Memory and Delayed Recall, Language and Naming. **Objectives:** to verify the influence of cross-cultural adaptation on the performance of cognitive tools, and the accuracy of the Alternative Version of the MoCA-BR. **Methods:** a pilot, prospective, longitudinal and analytical study. Both versions of the test were applied in a randomized and cognitively healthy population, between 18 and 60 years, within a medium interval of 54,56 days between the questionnaires. **Results:** out of 104 participants, 70 were included (64.3% female, 40.2 years). The alternative version obtained superior performances in the naming domain ($p < 0.001$), and in the adapted sentence in the language domain ($p = 0.003$). There was no significant difference in the delayed recall domain. The alternative version showed good internal consistency, with a Cronbach's alpha of 0.75. The cut-off point suggested by the study is 27 points, with sensitivity and specificity of 91.3% and 79.2%, respectively. **Conclusion:** Cultural factors affect the accuracy of cognitive tests, and adaptation is essential for their use in different countries.

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TL 1106336

DEMENTIA SYNDROMES AS NEGLECTED DISEASES

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Background: The neglected diseases are classically infectious diseases. However, in the health care of elderly patients, we observe that cognition and functionality are not routinely evaluated. Aging brings with it an increased risk of cognitive decline, moreover, elderly and low-income populations in cities in the interior of northeastern Brazil would still be exposed to other risk factors such as low education, poor control of chronic diseases and poor diet in childhood. In this context, the absence of a routine assessment of this population could be reflected in a large number of patients with the disease and lack of diagnosis of adequate treatments. **Objective:** The objective of this study was to evaluate the proportion of elderly people who would have dementia, but without a diagnosis and previous treatment of the condition. **Methods:** Patients aged 60 years or older from the municipality of Pindoretama-CE were evaluated. Patients in the coverage area of 3 health units were invited to participate in the study. They were evaluated by a neurologist with expertise in cognitive neurology. This assessment consisted of a structured medical interview, neurological and cognitive examination, and functionality tests. After evaluation, the data were discussed with at least two neurologists, to define whether or not the case would be a dementia syndrome. **Results:** A total of 213 patients were evaluated; 42 patients were identified as having dementia syndrome. Of this total, only 2 patients had a previous medical diagnosis of dementia and some treatment directed to the condition. **Conclusion:** The low recognition and treatment of dementia syndromes shows a negligence of the health care system regarding this condition.

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TL 1106364

THE INTERFACE BETWEEN SOCIAL COGNITION AND NEUROPSYCHIATRIC SYMPTOMS IN MULTIPLE SCLEROSIS: A NEUROPSYCHOLOGICAL INVESTIGATION

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Background: Multiple sclerosis (MS) manifests with motor deficits and non-motor symptoms, such as cognitive decline. One of the cognitive domains affected in MS is social cognition (SC), which refers to cognitive processes related to the perception, understanding, and mental inference, and is essential to interpersonal exchanges. Studies of SC in MS are still limited and the influence of depression, fatigue, and apathy on SC performance is unclear. **Objective:** To investigate the profile of patients with relapsing-remitting MS (RR-MS) in SC tests, and to explore correlations of SC measures with depression, fatigue and apathy. **Methods:** This cross-sectional study included two groups: 20 patients with stable RR-MS (mean age = 40.15 ± 10.15 , mean schooling 15.35 ± 1.98 , 4 men/16 women, mean EDSS score = 2.5 ± 1.5) and 20 healthy controls (mean age = 40.2 ± 10.17 , mean schooling 16.3 ± 1.9 , 4 men/16 women). RR-MS patients had no relapses in the last 90 days and no other neurological diseases. All participants underwent the Beck Depression Inventory, Apathy Scale, Fatigue Scale (MFIS), Brief International Cognitive Assessment for MS (BICAMS), Hayling test, and semantic (animal) fluency. SC was assessed with the Reading the Mind in the Eyes test (RMET), and the mini-version of the Social Emotional Assessment (mini-SEA), which is composed by the facial emotion recognition test (FERT) and the modified Faux-Pas test (mFP), which assesses mentalizing. **Results:** Groups did not differ on age, sex and educational level. Compared to controls, RR-MS patients had lower performance on semantic fluency ($p < 0.014$) and on working memory ($p < 0.011$), but not on episodic memory. Patients had higher score on the Apathy Scale ($p < 0.004$), but not on depression scale. Regarding SC scores, patients underperformed on the FERT ($p < 0.024$) and in the mFP ($p < 0.016$), but not in the RMET. SC measures did not correlate with depression, apathy or fatigue. **Conclusion:** Patients with RR-MS present SC deficits, which seem independent of neuropsychiatric symptoms. More studies are warranted to investigate the interface between SC deficits and psychiatric symptoms in MS.

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TL 1106386

THE USE OF THE COGNITIVE CHANGE QUESTIONNAIRE BY COMMUNITY HEALTH WORKERS AS DEMENTIA SCREENING METHOD

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Background: Brazil has a public health system that uses family medicine as main strategy and the Community Health Workers play a very important role. The Cognitive Change Questionnaire (CCQ) was a tool developed in Brazil to evaluate cognitive changes in a simple way to allow a triage for dementia patients. **Objective:** To evaluate the applicability of the CCQ by Community Health Workers (CHW) as a screening tool for dementia in a community. **Method:** This study was carried out in Pindoretama-CE, a 20.000 habitants town in the proximities of Fortaleza. Community health workers were trained on basic dementia principles and how to apply the CCQ. Patients assisted by 3 health care units were invited to participate in the study. The CHW was applied by the CCQ to people aged 60 or over individuals during routine home visits. Questionnaires were evaluated afterwards by a neurologist with expertise in cognitive neurology. Patients were then recruited and accessed by the same neurologist using a structured medical interview, neurological and cognitive examination, and functionality tests. The batteries used were: mini mental state exam, Brief cognitive battery, Dementia Rating Scale, Pfeffer's Functional Activities Questionnaire, Geriatric Depression Scale 15-item version. After evaluation, data were discussed with at least two neurologists, to define whether or not the case would be classified as dementia syndrome. **Result:** A total of 213 patients were evaluated; 42 patients were identified as having dementia syndrome. Statistical analysis comparing the score obtained in the test with the probability of the patient having or not having dementia is still in progress. **Conclusion:** Application of the QCC by CWH is a possible screening tool for dementia in small communities. Additional data regarding accuracy are needed before this strategy can be used in clinical practice.

Neurologia Cognitiva e do Envelhecimento

TL 1106410

THE INTER-RELATION BETWEEN GUT MICROBIOTA AND NEURODEGENERATIVE DISORDERS IN ELDERLY

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Background: The gut microbiota has a central role in the regulation of immune, endocrine, and neural pathways. Recent studies suggest that gut microbiota dysbiosis has influence in the development of neurodegenerative and neuropsychiatric diseases. The alteration in the gut microbiota during the aging process associated with inflammation and microglia activation can boost the risk of neurodegenerative disorders, like Alzheimer's and Parkinson's disease. **Objectives:** Identify what the scientific literature says about the relation between the gut microbiota and neurodegenerative diseases in elderly. **Methods:** A systematic review was performed in PubMed and Scielo databases using the keywords ("Microbiota" AND "Neurodegeneration" AND "Aged"). Articles with free access in Portuguese or in English published between January/2018 and May/2022 were included in the study. Articles out of context, repeated and editorials were excluded. It was found 30 articles and analyzed 15. **Results:** It was analyzed 8 transverse studies, 6 systematic reviews and 1 observational study. 6 of them approached the relation between gut microbiota and Parkinson's Disease (PD), 5 presented the association among dysbiosis and Alzheimer's Disease (AD) and 4 discussed the influence of gut microbiota and neurological alterations in elderly. The AD studies showed that dysbiosis causes a hypersensitivity of neurons which can amplify the immune response and accelerate degeneration via inflammation and release of cytokines. Other studies investigated the benefits of probiotics and its contribution for the prevention of neurodegeneration; however, its response depends on dietary habits, comorbidities, probiotic dosage, composition, and intake frequency. Studies showed that elderly with DP and severe gastrointestinal symptoms due to dysbiosis suffered more cognitive impairment as alterations of dopamine and motricity. One study also implies that alterations in the oral microbiome have correlation with DP. This is relevant to future therapeutic approaches since its management is easier than genetics and gut microbiota. **Conclusion:** The findings suggested that dysbiosis can amplify inflammatory responses contributing to neurodegeneration. However, the microbiota-gut-brain axis is not fully comprehended yet. It is necessary to have more research about it in order to better understand the therapeutic use of prebiotics and probiotics, its prognostic and its possible use as prevention in the future.

Neurologia Cognitiva e do Envelhecimento

TL 1106451

COGNITIVE AND BEHAVIORAL CHANGES RELATED TO COVID-19 IN PATIENTS EVALUATED AT A REFERENCE CENTER IN S&ATILDE;O PAULO, BRAZIL

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Background: Central nervous system involvement associated with Coronavirus Disease 2019 (COVID-19) has been reported, including cognitive impairment, even in patients with mild COVID-19. **Objectives:** To assess cognitive decline related to the SARS-CoV-2 infection in patients with neurological disease after COVID-19. **Methods:** Longitudinal prospective study developed to compare the cognitive performance of patients after COVID-19 based on cognitive complaints. Patients aged between 18 and 65 years without previous neurological disease were included in the study and illiterate people were excluded. Beck's anxiety and depression questionnaires were applied, also the Addenbrooke's Cognitive Examination-Revised (ACE-R) and the Trail Making Test (TMT), parts A and B. **Results:** Sixty one patients were evaluated, 41 women (67.21%) and 20 men. The mean age of all patients was 46.36 years (standard deviation – SD 9.92) and median of 46 years. In relation to schooling, mean of 13.93 years (SD 4.06) and median of 16 years. Forty-four patients had cognitive complaint (72.13%), 21 (47.72%) of them the only complaint. Regarding the time between the first symptom of COVID-19 and the onset of the neurological symptom, mean of 39.52 days (SD 40.7) and median of 29 days. Among all patients, the mean score of Beck's anxiety and depression questionnaires was, respectively, 16.72 (SD 10.05) and 13.31 (SD 6.97), and median of 15 and 12, which indicates mild to moderate degrees of anxiety and depression. The results of ACE-R were altered in 41 patients (67.21%) – 30/44 (68.18%) patients with a cognitive complaint and 11/17 (61.11%) patients without this complaint ($p = 0.07798$). The score on the mini-mental state examination (MMSE) was also altered in 41 patients. In the TMT part A, score altered in 39 patients (63.93%) – 10 had lower average performance, 8 borderline and 21 severe performance. In the TMT part B, score altered in 43 patients (70.49%) – 4 had lower average, 3 borderline and 36 severe performance. No patient had superior performance on any part of the test. Two patients could not take the TMT's due to motor limitation because of motor sequelae of COVID-19. **Conclusion:** Cognitive complaint was not a predictor of cognitive decline, and the cognitive decline was observed in most patients, in the MMSE, ACE-R and TMT assessments cognitive decline.

Neurologia Cognitiva e do Envelhecimento

TL 1104772

NEUROBIOLOGY IN AUTISM SPECTRUM DISORDER (ASD)

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Background: Autism spectrum disorder (ASD) is a disorder of the neurodevelopment associated with communication deficits and repetitive behavior characterized by a large and atypical variation in the connectivity of cortical and subcortical brain ligation. **Objective:** Study the neurobiological changes found in the patients with ASD in childhood and adolescence through reports in the literature. **Method:** Systematic review of the literature on the neurobiological aspects of ASD. Pubmed was used as a database, with descriptors such as neurobiology, autism, genetics, neurochemistry, neurotransmitters, magnetic resonance imaging, neurophysiology. The inclusion criteria were bibliographic materials available in English and Portuguese. **Findings:** It were identified 649 article with the use of keyword listed above. After the initial analysis, the articles were selected. One of the articles studied 662 patients showing that differene areas of the brain have increased cortical thickness, with the region being the most affect upper temporal gyre. The second most affected area would be the middle occipital gyre. One study demonstrated increased connectivity in the sensitive thalamic area. Other text claims that the increase on cortical thickness may be related to the severity of the clinic of the case. Pathological changes are more frequent in non-treated, suggestion that treatment in childhood has a beneficial effect on the brain development. The treated individuals may present reversal of the changes showing gains in brain fuction. **Conclusion:** Neurobiological changes, especially structural anatomical changes reported in most of eht reviewed articles,done in studies with a large number of patients and in different multidisciplinary centers, point out lines of research and study for our environment. The need for early treatment/rehabilitation was highlighted, since there is evidence in the literature of good results, including reversal of anatomical changes, which otherwise consolidate in adulthood

Neurologia Infantil

TL 1105010

THE PAINFUL MISSION OF COMMUNICATING THE DIAGNOSIS OF SPINAL MUSCULAR ATROPHY TO CHILDREN

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Background: The trinomial relationship between the doctor-child-legal guardian is essential in the process of a disease communicating and its prognosis. **Objectives:** To analyze the communication of the diagnosis of spinal muscular atrophy (SMA) by this trinomial relationship. **Methods:** Relatives of SMA patients answered a questionnaire containing **Objective:** questions and the Event Impact Scale – Revised. **Results:** The sample consisted of 50 volunteers, 94% of whom were female. 48% reported psychological sequelae related to the moment of diagnosis, having a direct correlation when the communication was made by them. 22% have a high risk of post-traumatic stress, relating the feeling of unpreparedness in communicating with the child. **Conclusion:** It was identified that the failure in communication is the main factor for negative repercussions with self-report of psychological sequelae, in addition to the high risk for post-traumatic stress syndrome.

Neurologia Infantil

TL 1105164

AUDIOVISUAL OVERSTIMULATION PROMOTES ANXIOTIC-LIKE BEHAVIOR IN A SEX-DEPENDENT MANNER IN MICE

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Background: Excessive use of screens is known to be related to increased incidence of mental disorders in children and adolescents. It has been demonstrated that mice subjected to audiovisual overstimulation show behaviors compatible with Attention Deficit Hyperactivity Disorder. **Objectives:** The aim of this study is to analyze whether there is a relationship between the sex of the animal and the anxious behavior of animals subjected to audiovisual overstimulation. **Methods:** Swiss mice (20 – 30 g), male and female, 21 days old, were used. For 42 days, the animals were placed in a room containing sound from children's television programs at a volume maintained below 70 dB, the level used in acoustic stress models. The animals were divided into groups (n = 6 each) as follows: control males and control females – animals without stimulation, but taken daily from the maintenance vivarium and kept in a separate room for 2 or 6 h daily. Stimulated males and stimulated females – animals with daily stimulation for 2 or 6 h. After 10 days of rest, the animals were subjected to the elevated cross maze test. **Results:** There was no difference in the behavior of male and female animals stimulated for 2 h daily. However, females overstimulated for 6 h daily, stayed longer in the open arm (*p<0.05) and produced more feces pellets (**p<0.01) when compared to the non-stimulated group (naive). The overstimulated male animals showed increased locomotor activity characterized by more entries into the open arm (*p<0.05) compared to the non-stimulated group. **Conclusion:** The higher preference for the open arm suggests anxious behavior, inferring impulsivity content. The results obtained point out that overstimulated females seem to show a higher propensity to take risks.

Neurologia Infantil

TL 1105181

THE RELATIONSHIP BETWEEN THE NEUROPSYCHOMOTOR AND NEURODEVELOPMENTAL CHARACTERISTICS IN 18 TO 47 MONTH-OLD CHILDREN

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Background: the knowledge of the health professionals of neuropsychomotor and neurodevelopmental skills of children is essential for their follow-up. **Objective:** to investigate and associate the neuropsychomotor and neurodevelopmental skills of 18 to 47 month-old children. **Methods:** this research is a transversal study of the relationship between neuropsychomotor and neurodevelopmental characteristics. It included 30 parents whose children are typical and aged from 18 to 47 months-old followed by a Pediatrics Service. As materials, the NPDC (Neuropsychomotor Development Checklist) and the M-CHAT (Modified Checklist for Autism in Toddlers) were used and both are composed by dichotomous questions. The participants were divided into two groups. The first represents the children who presented risk on M-CHAT and the second those who did not present risk. Descriptive percentages and statistics analysis, Mann-Whitney Test (as statistical significance p<0.05), were used to analyze the results. **Results:** The neurodevelopmental risk group included six (20%) of the total number of participants (30). This group did not achieve the expected neurodevelopmental stages such as directing their gaze by 6 months-old (33%); crawling by 9 months-old (33%); inviting other children or adults to play by 18 months-old (50%); pointing named parts of their own body by 12 months-old (66%) and using phrases with two or three words (66%). Those two last skills were statistically significant (p=0.007 and p=0.048, respectively). On the other hand, the second group, which was not at neurodevelopmental risk, presented neuropsychomotor skills below expected. However, the aspects were different if compared to the risk group, such as sitting without support by 6 months-old (20%); speaking isolated words by 14 months-old (25%) and mispronouncing speech sounds (33%). This group without risk also had some unexpected similar characteristics. Four children (16%) had not crawled by 9 months-old; six (25%) had not invited other children or adults to play; almost eight (33%) of them mispronounced speech sounds; six (25%) had not understood simple stories and all of them had pointed named parts of their own body by 12 months-old. **Conclusion:** Both groups underperformed on neuropsychomotor tasks and the ability of pointing named parts of their own body by 12 months-old got an important statistical significance. Thus, the neurodevelopment and neuropsychomotor follow-up is essential for typical children.

Neurologia Infantil

TL 1105557

MOTION SICKNESS IN CHILDREN AND ADOLESCENTS WITH NEUROLOGIC COMORBIDITIES

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Background: Motion sickness or intolerance to movements is the occurrence of signs or symptoms of conflict in the sensory perception of stimuli from the proprioceptive and vestibular systems. **Results:** in a number of symptoms including nausea, vomiting, dizziness and headache. The literature on motion sickness in adults is extensive due to its medical mission for military and space travel. In contrast, the pediatric population has not been well studied. This is surprising given that the prevalence of motion sickness in children to be higher than in adults. The difficulty of studies in this area and a lack of understanding this illness's etiology and its comorbidities can impact the course of the disease. An example of this are some neurological diseases that can be etiologically correlated as an aggravating the development of motion sickness. **Objective:** To evaluate the occurrence and characteristics of motion sickness in neuropsychiatric patients with followed up in a university Hospital in northeastern of Brazil. **Method:** It is an observational, analytical, cross-sectional study. All sex patients, younger than 18-years-old, with a previous diagnosis of a neurological disease, regularly followed up at neuropsychiatric service were included. Pregnant women and patients with severe systemic diseases were not selected. Data were collected through digital forms to analyze: sociodemographic factors, predictable diagnoses, susceptible to motion sickness and presence of headache. The Motion sickness susceptibility questionnaire (MSSQ) was Applied. The study had ethics committee approval. **Results:** The preliminary sample contains 70 patients, 35% of these presented motion sickness; 34% of patients are within the autistic spectrum (ASD) and of these 15.4% had motion sickness; 12% of patients have a diagnosis of attention deficit hyperactivity disorder (ADHD), and 30% of these had motion sickness; 20% of the sample has epilepsy, of which 30% had motion sickness; From the sample, 15% of migraine patients and of these 55% have motion sickness. **Conclusion:** The preliminary findings of our research demonstrated a high prevalence of motion sickness in children and adolescents with neuropsychiatric disorders. Even children with behavioral disorders, motion sickness could be verified by the parents' perception through the specific questionnaire (MSSQ). Our study also corroborates the literature on associative aspects of motion sickness with a previous diagnosis of migraine.

Neurologia Infantil

TL 1105704

BENEFITS OF APPLYING MUSIC THERAPY TO CHILDREN WITH AUTISM SPECTRUM DISORDER: A SYSTEMATIC REVIEW

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Background: Music therapy is a therapeutic modality that consists of the integration between music and health with the **Objective:** of providing intellectual development, communication and well-being, assisting in socialization factors to improve the quality of life of the individual. Therefore, several studies have analyzed the use of music therapy as a therapeutic intervention in the treatment of children with Autism Spectrum Disorder (ASD), since in this clinical condition there is a loss in social interaction and communication. **Objective:** To identify the main benefits of music therapy in the treatment of children with autism. **Method:** This is a systematic review, which used the following descriptors: "music therapy", "child", "autism spectrum disorder" with search in the databases: Medline, EBSCO, LILACS and SciELO. The inclusion criteria were articles in Portuguese, English and Spanish; full articles with the theme in question; articles published and indexed in the last 5 years in the mentioned databases. The exclusion criteria were review articles and articles that were not related to the theme. **Results:** 21 references that met the criteria of this research were selected. Of these, 20 articles evidenced the value of music and its role as a therapeutic resource in children with ASD and 1 was indifferent to the results presented. Those that presented positive results evaluated the application of music therapy in the medium and long term and the benefits presented were the development of new social interactions, improvement in the behavioral pattern of isolation, and improvement in communication skills. The study that did not show significant results evaluated the social affect and the parents' perception of social response and concluded that there was no considerable improvement. **Conclusion:** Based on the analysis of the studies presented, it can be concluded that the application of music therapy acts positively in the areas of socialization, communication, and language. Playing with instruments is a way to interact with others and establish strong social bonds, being one of the most important aspects of this practice. The benefits are not immediate, but in the long term they promote emotional satisfaction, break the patterns of social isolation and facilitate self-expression.

Neurologia Infantil

TL 1105809

CLINICAL-EPIDEMIOLOGICAL PROFILE OF EPILEPSY IN PATIENTS FOLLOWED-UP SINCE CHILDHOOD IN A UNIVERSITY HOSPITAL

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Background: Epilepsy is a neurological condition with high prevalence in neuropsychiatric care, requiring lifelong drug treatment in most cases, in addition to multidisciplinary team support. The individual with epilepsy may present neurobiological consequences such as cognitive, behavioral, and social disorders, causing great impact on the patient's quality of life. **Objectives:** To describe the clinical and epidemiological characteristics and the profile of drug use of epileptic patients followed up as outpatients in a specialized care service. **Methods:** Retrospective Cross-sectional study, with consultation of outpatient records of child and adolescent population with epilepsy in a university Hospital in Northeastern Brazil. Relative and absolute frequency distribution tables were used for descriptive data analysis. The Chi-square test for independence and Fisher's exact test were applied to analyze the existence of association between variables. The significance level was 5% ($p < 0.05$), using IBM SPSS 20.0 software. **Results:** The medical records of 282 patients with epilepsy (143 males and 139 females), aged 2 to 19 years, were analyzed. About the age of onset of epilepsy, the most prevalent age group was those older than 11 years (31.2%), followed by those younger than 2 years (30.9%), schoolchildren between 6 and 10 years represented 23.4% of the sample, and preschoolers between 3 and 5 years corresponded to 14.5%. As for the clinical features of epilepsy, 77.7% had focal epileptic seizures, 11.3% had indeterminate seizures, and 11.3% had generalized seizures. The recorded electroencephalogram of these patients showed alterations in 68% of the sample, with focal alterations being the most observed finding (28.4%). Some comorbidities investigated were migraine (34%) with and without aura, tension-type headache (2%), behavior disorders (20.9%), and sleep disorders (8.5%). Regarding antiepileptic drugs use, 73.3% were monotherapy, while 23.3% were taking two drugs, and only 3.5% were taking three drugs; and no significant difference was observed between males and females for these variables cited. **Conclusion:** As a chronic neurological condition, age of onset of epilepsy is more prevalent in teenagers and infants, with a higher incidence of focal seizures. We also observed that migraine is the main comorbidity diagnosed in these patients. Even in a referral center for epilepsy care, a large proportion of patients are well controlled with monotherapy.

Neurologia Infantil

TL 1105814

DETERMINANT VARIABLES OF THE ABSENCE OF VERBAL LANGUAGE IN PATIENTS WITH AUTISTIC SPECTRUM DISORDER

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Background: Autistic spectrum disorder (ASD) is a neurological disorder caused by the interaction of genetic and environmental factors. The individual presents atypical evolution in developmental milestones, behavioral changes and difficulty in communication and social interaction. Thus, it is noticeable, with about 5% to 50% of cases, that the first skill to present delay is language, becoming a predictor of ASD. **Objective:** To evaluate the determinant variables for the absence of verbal language at 3 years of age, and to describe their neuropsychomotor development. **Methods:** This is a documentary, observational, cross-sectional, retrospective, descriptive research with a quantitative approach. The following were analyzed: medical records collected in a protocol form from patients diagnosed with ASD, according to the DSM-V criteria, from a specialized institution in Teresina-PI, in the period between 2014 and 2019, after approval by the local Research Ethics Committee (CEP). The demographic variables used were age and gender, the clinical variables were neo and perinatal antecedents, neuropsychomotor developmental milestones, associated neuropsychiatric pathologies and disorders, and medications in use. The Denver Scale was used to assess neuropsychomotor milestones, data was organized using Microsoft Office Excel, and statistical analysis using the Biostatic 5.0. **Results:** In the present study, 90 medical records of patients diagnosed with Autism Spectrum Disorder (ASD) were analyzed, with a mean age of 11.65 years, of which 20 (22.22%) were female, while 70 (77, 78%) were male. It was possible to verify in this analysis, in isolation, that the delay in verbal language was more prevalent in those who were born by cesarean section ($p=0.00018$). **Conclusion:** The determining variable for the absence of verbal language in patients with ASD at 3 years of age was the mode of delivery, especially when correlated alone, demonstrating a greater tendency to language delay in those who were born by cesarean section, even in the absence of complications.

Neurologia Infantil

TL 1105823

EVEROLIMUS IN THE TREATMENT OF REFRACTORY EPILEPSY IN TUBEROUS SCLEROSIS COMPLEX: A CASE SERIES

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Case Presentation: Patient 1: A female infant was diagnosed with tuberous sclerosis complex (TSC) at 7 months of age after initiating generalized seizures associated with developmental delay and stereotyped behaviors. EEG showed generalized and focal epileptiform potentials and ictal activity from epileptic spasms. Brain MRI identified cortical dysplasias and subependymal nodules. The echocardiogram showed 2 cardiac rhabdomyomas and the exome revealed a deletion of 18 base pairs in exon 41 of the TSC2 gene in heterozygosity (chr16: g.2088293_2088310del; depth: 133x). First-line treatment for epilepsy in TSC was started with vigabatrin, but after an initial improvement period, seizures intensified, and social interaction worsened. Treatment with everolimus was then started, and the patient evolved with a substantial reduction of seizures, improved social and visual interaction and began to walk. Today, almost two years after starting treatment, the patient is in use of everolimus and oxcarbazepine and has good seizure control, with age appropriate development and without severe everolimus side effects. Patient 2: A 3-month-old male infant presented with clonic seizures in both eyelids, left hemiface and left arm. Brain MRI showed subependymal nodules in both lateral ventricles and cortical tubers in the middle gyri of both frontal lobes. EEG revealed epileptiform potentials in the left occipital region. A heterozygous missense variant was identified in the TSC2 gene (TSC Complex Subunit 2, OMIM*191092). Treatment was started with vigabatrin. Due to inefficient seizure control, levetiracetam was associated, followed by the introduction of oxcarbamazepine. However, the patient continued presenting 2 seizures weekly. After 5 months without adequate symptom control, everolimus was introduced, and seizure remission was achieved. Currently, about ten months after everolimus introduction, the patient remains seizure free and has adequate development for his age. **Conclusion:** TSC is a multisystem genetic disease in which epilepsy is a frequent manifestation and is often difficult to control. Everolimus is an mTOR inhibitor drug, with evidence suggesting benefits of its use in treating epilepsy in TSC. Inadequate control of seizures appears to play a crucial role in the cognitive and behavioral decline in patients with TSC, reiterating the importance of early diagnosis and treatment to improve the neurocognitive prognosis in these patients.

Neurologia Infantil

TL 1105826

ASSOCIATION BETWEEN SELF-MUTILATION AND CONSTIPATION IN PATIENTS WITH AUTISM SPECTRUM DISORDER

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Background: Self-injurious behavior (SIB) is defined as any intentional action that generates direct aggression to one's own body without conscious intention of suicide or exposure, as in the case of tattoos and piercings. SIB is considered a psychiatric disorder that is common in children with Autism Spectrum Disorder (ASD). In addition, gastrointestinal symptoms, such as functional constipation (FBC), are also associated with mental and behavioral health in children with ASD. **Objectives:** To investigate the existence of an association between CIF and SIB in a patient with ASD. **Methods:** This was a transversal study of applied, observational and exploratory nature with a quantitative and descriptive approach. For this, we used a questionnaire applied to patients with ASD in a care and rehabilitation institution. Therefore, the inclusion criteria according to the DSM-V were used. In addition, the FASM scales and the Rome IV criteria were applied for the diagnosis of BIS and ICF, respectively, and Fisher's Exact Test was used to assess the association between these variables. **Results:** A total of 74 patients with ASD were obtained, with a predominance of men, with 59 cases (79.7%). Mean age was not statistically significant enough to be considered a predictor variable for SIB or CIF. As for the level of education of the responsible family member, there was a prevalence in patients whose parents had a higher level of education, the most prevalent being those with complete high school and incomplete college education, about 33.8% (25) of cases, followed by those with complete college education, with 29.7% (22). To clinically stratify ASD, the CARS was used, in which 70.3% of participants were classified as mild/intermediate. Additionally, the prevalence of ICF was 37.8% and of SIB 37.8%, with no statistical association between them ($p=0.769$). **Conclusion:** Thus, it was noted that there was a high prevalence of SIB and ICF in the study sample. There was no association between CIF and SIB in patients with ASD.

Neurologia Infantil

TL 1105853

DENTAL APPROACH IN PATIENTS WITH MICROCEPHALY BY ZIKA VIRUS

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Background: Microcephaly is characterized by a neurological disease in which the child presents a decrease in brain diameter, causing motor sequelae, speech delay, behavioral disorders and craniofacial development. These characteristics also affect the child's oral health, because most of them present oral changes such as: periodontal diseases, dental caries, malocclusion, micrognathia, delayed dental eruption, dysphagia, bruxism and dental trauma that may have a direct and negative impact on their health. **Objective:** In this sense, the present study has as its main objective to analyze the craniofacial and dental alterations in patients with microcephaly. **Method:** This is a cross-sectional observational study with patients with microcephaly treated in Paraíba, which used a structured questionnaire divided into two parts: the first with questions regarding socioeconomic data and the second regarding data related to the object of study. The data was collected through the Google form and then tabulated by Excel. **Results:** 48 children were attended, 54% female and 46% male. 47% of the public examined were between 5 and 6 years old, with a family income of one to two minimum wages, making up 79% of the sample. All participants had significant neurological alterations, presenting seizures, spasms, motor deficits, and cognitive deficits. Regarding craniofacial alterations, the following were identified: lack of lip seal, tongue projection, and palate alterations. Regarding oral health, 49% visited the dentist once a month, and the reason for the visit was the need for treatment and follow-up. 90% use the public service, 57% report difficulty in finding a specialized dentist, 91% of caregivers reported difficulty in performing oral hygiene, for reasons of limited mouth opening, reflux, agitation and aggressiveness, 88% use a fluoride toothbrush and toothpaste for hygiene. Of the total sample, 32% of patients had no need for treatment and 68% had some need for treatment such as: prophylaxis, tartar removal, fillings and extractions. **Conclusion:** This work provided a better understanding of craniofacial and dental changes in patients with microcephaly, a topic little explored in research, in order to collaborate to improve the provision of care for these patients and promote quality of life.

Neurologia Infantil

TL 1105860

WEST SYNDROME: A LITERARY REVIEW

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Background: West Syndrome (WS) was first described by William James West (1793–1848) in 1841 and classically corresponds to an epileptic syndrome characterized by repetitive spasms and a pattern of hypsarrhythmia that result in a delay in neuropsychomotor development. This pathology has been progressively studied and can often be named as infantile spasm, based on the most relevant initial clinical event. Although its causes are unknown, in recent years new advances in this disorder have been made, and point to predisposing factors for the disease: hypoglycemia, lack of oxygen to the brain during childbirth, brain malformation, prematurity, sepsis, stroke, infections and genetic. **Objective:** To systematically demonstrate the West Syndrome questioning its complexities and peculiarities and what makes it a rare and complex syndrome. **Methods:** This is a systematic literature review based on the extraction of data in indexes such as PUBMED and MEDLINE. Publications from 2018 to 2022, in English, were considered, with the following descriptors: West syndrome, Neurologia, Epilepsy, Infantile spasms. **Results:** Overreliance on the term hypsarrhythmia is particularly problematic, since many children who experience infantile spasms will not have this classic pattern. Presence or absence on the pre-treatment EEG has no impact on treatment response, risk of subsequent seizures, or later developmental outcome. The hypsarrhythmia requirement in the criteria for West syndrome is unsupported by documentary evidence and is overly restrictive. Thus, almost all babies with the infantile spasms type of seizure also have West syndrome. **Conclusion:** It can be concluded that many advances have been made, however there are still many issues to be resolved about SW, which suggests steps forward. As a result, the diagnosis of West syndrome should not be made in a restrictive manner and even though the defining triad of the disease can be considered, the absence of a criterion should not exclude the diagnosis, so it should not interfere in the planned treatment for the disease.

Neurologia Infantil

TL 1105916

FETAL ALCOHOL SYNDROME (FAS): CONSEQUENCES FOR THE FETUS AND IMPAIRMENTS IN CHILD DEVELOPMENT

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Background: Alcohol is the most consumed psychoactive drug worldwide. Its metabolism happens more slowly in women, taking into account their body structure, increasing the risk of pathologies associated with its consumption, even if it is moderate. However, the demand for alcohol by women has become an issue of public health concern, especially when it occurs during pregnancy, causing harmful effects to the fetus during pregnancy, postpartum, and during the child's development, which is called FAS (Fetal Alcohol Syndrome). Children with FAS have characteristic facial alterations, prenatal and/or postnatal growth restriction, and structural and/or functional abnormalities of the central nervous system (CNS). They may present other congenital malformations, learning and memory difficulties, and functional, emotional, and behavioral problems. Currently, alcohol is considered the most common fetal teratogenic agent, but it is still neglected by many health professionals. **Objective:** This study aims to relate alcohol consumption during pregnancy with fetal neurological malformation, as well as to address ways to prevent this situation. **Methods:** This is a scientific study that allows a broad approach to fetal alcohol syndrome. The textual search was restricted to scientific publications from the SCIELO and PUBMED databases during the month of May of this year, using publications from the last 13 years with the following descriptors: Pregnant Women; Alcohol; Consequences for the fetus. Original articles indexed in the period between 2009 and 2021 were included and articles that do not have such descriptors and are not within the delimited period were excluded. **Results:** The harmful effects of alcohol consumption during pregnancy were confirmed and identified, especially regarding its consequences for the fetus and impairments in child development throughout life. In addition, ways to prevent this worrying issue for public health were addressed. **Conclusion:** Alcohol consumption by pregnant women can lead to irreversible neurological consequences for the fetus, postpartum, and throughout the child's development. This psychoactive drug is considered the most common fetal teratogenic agent, becoming a major public health problem. However, this situation is preventable if the woman abstains from drinking alcohol before conception and during pregnancy. In addition, they need to be routinely screened by health professionals for alcohol use during prenatal care.

Neurologia Infantil

TL 1106047

AUTISM AND COGNITIVE IMPAIRMENT IN DYSTROPHIC AND STRUCTURAL MYOPATHIES: ARE WE MISSING OUT?

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Background: Myopathies represent a wide spectrum of heterogeneous diseases characterized mainly by the abnormal structure and functioning of skeletal muscle. Due to the pathogenesis of myopathies being diverse and several systems being affected, neurodevelopmental disorders may also occur. However, amongst the genetic myopathies, intellectual disability (ID) and autism spectrum disorder (ASD) has been mostly described in Duchenne Muscular Dystrophy, Alpha-dystroglycanopathies and Myotonic Dystrophy, but not in other structural and dystrophic myopathies. **Objective:** Describe three patients with dystrophic and structural myopathies with ID and ASD. **Methods:** Description of three patients with ACTA1-myopathy and LAMA2-congenital muscular dystrophy with ID and ASD and literature review. **Results:** Patient 1 was a 7 years old male with hypotonia, proximal muscle weakness, myopathic face and muscle biopsy with nemaline rods, who presented with hemizygous pathogenic variant in ACTA1 (p.Gly57Arg). He was non-verbal, had stereotyped movements, poorly functioning play, poor social interaction and didn't comprehend simple commands. Patient 2 was an 8 years old boy with congenital hypotonia, with maximum motor ability to sit without support, that presented with myopathic face and multiple joint contractures. He had two pathogenic variants in LAMA2 (p.Tyr121* and Leu1581Profs*5), and spoke only few words, poorly functioning play, poor social interaction and only comprehended simple commands. Patient 3 was an eight years old girl, who presented with congenital hypotonia, muscle proximal weakness, but was able to walk unassisted, she also presented scoliosis. She had two pathogenic variants in LAMA2 (P.Cys1079Arg and exons 2-9 deletion). She spoke just simple phrases, had poorly functioning play, poor social interaction, wasn't able to read or write and had stereotyped movements. Both patients with LAMA2-CMD had white matter hyper-signal in T2 and FLAIR at Brain MRI, but none of them presented cortical malformation or epilepsy. All three patients scored for ASD in the Child Autism Rating Scale (CARS) and presented signs of ID. **Conclusion:** Given the gap in the literature regarding cognitive defects within the context of myopathies, we have presented patients with genetical myopathies associated ASD and ID and provided insight for clinicians handling these patients. As it may appear in a greater proportion of patients, early screening and multidisciplinary programs is necessary

Neurologia Infantil

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IMPACT OF THE COVID-19 PANDEMIC IN CHILDREN WITH AUTISTIC SPECTRUM DISORDER

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Background: Autistic Spectrum Disorder (ASD) comprises difficulties in social interaction and communication, as well as restrictive patterns and repetitive behavior. In the current pandemic, usual care, routine and activities directed to children with ASD changed due to preventive measures such as social distancing. With that, these children and their caregivers went through great stress, reflecting on different aspects of their lives. Goal: To assess the impact of the COVID-19 pandemic on children with ASD and their caregivers. **Methods:** A review was made utilizing the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) of published studies that assess changes in the well-being of children with ASD and their families during the Pandemic around the world and in Brazil, ranking each article according their Risk of Bias, upon which only the "Good" and "Regular" articles were utilized. **Results:** 161 articles were found upon initial search, of which 17 were selected. Amid the findings, there was an increase in symptoms related to ASD, greater difficulty in managing emotions, and the absence of school support was directly associated with an increase in behavioral problems. There was also a reduction in physical activities, increased screen time and changes in sleep habits. In most cases, the main caregiver was female, and among caregivers occurred an increase in their burden and their fears surrounding the pandemic, along with stress and mental health detriments, such as an increase in anxiety, depression and less time for self care. **Conclusion:** The pandemic can enhance inequalities for children in vulnerable groups, so it is essential to support the well-being of ASD children and their families as we emerge in this new way of living, focusing on making policies and approaches with individualized multidisciplinary planning, psychological resources and increased services for families in order to facilitate reintegration and ensure the development of the children. It is also important to make further studies to assess the effects of the pandemic.

Neurologia Infantil

TL 1106138

DESCRIPTIVE ANALYSIS OF THE CORRELATION BETWEEN THE PRENATAL AND THE OCCURRENCE OF ANOMALIES IN THE NERVOUS SYSTEM IN THE PERIOD OF 20 YEARS

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Background: Congenital anomalies are the fourth leading cause of neonatal death and 21% of these malformations are related to the nervous system. In this scenario, despite all advances in obstetrics and improvements in prenatal care for pregnant women, the prevention of mortality in neonates due to defects in the neurological tract remains a challenge to be overcome. **Objective:** Analyze the proper performance of prenatal care, correlated to the occurrence of deaths from neurological defects in neonates in Brazil from 1999 to 2019. **Methods:** Descriptive analysis, obtained from the DATASUS platform, of the frequency of prenatal care, the frequency of births with congenital anomalies and how many of these disorders are related to defects in the nervous system and mortality rate in children under 1 year old due to neurological diseases. **Results:** From 1999 to 2019, in Brazil, 702 neonates died due to defects in the nervous system and 3,443,087 women did not perform prenatal care or did it inappropriately. In addition, 30,734 children, whose mother didn't perform prenatal care, or it was performed inappropriately, were born with a certain congenital anomaly, which represents 21% more than those who had congenital disorders, but prenatal care was being performed. Furthermore, of a total of 170,392 cases of births with anomalies, 32% were characterized by neurological defects. **Conclusion:** It is possible to correlate the inadequate performance of prenatal and the occurrence of congenital defects, such as disorders in the nervous system, because, with the medical attendance, the pregnant woman can be treated for a certain pathologies and can be orientated to have some precautions, such as administrate folate (vitamin B9), which prevents neural tube malformations. This guidance is essential to avoid harmful consequences to the fetus and to prevent the occurrence of premature birth, which can impair the maturational processes of the brain, leading to anatomical changes, functional deficits, that can perpetuate into adolescence and adulthood, or death. In addition, in case the fetus is already with the disturb, the prenatal allows to early diagnosis and treat the condition, increasing the survival percentage. In resume, performing prenatal care correctly, is essential for the prevention or the successful treatment of defects in the nervous system in neonates and for the reduction of mortality in children under 1 year old.

Neurologia Infantil

TL 1106207

NEONATAL NEAR-MISS OF NEONATES BORN WITH CONGENITAL HYDROCEPHALUS IN BRAZIL: ANALYSIS OF A DECADE

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Congenital hydrocephalus (CH) is the variety of hydrocephalus present at birth and is related to impairment in the cognitive and motor development of the children affected. In the context of this disease, neonatal near-miss (NNM) is a tool to access the profile of these newborns and evaluate the care offered to them. This study aims to describe and analyze the neonatal near-misses among neonates born with congenital hydrocephalus (ICD-10 Q03) in Brazil from 2011 to 2020. Data from the Brazil Live Birth Information System (Sistema de Informações sobre Nascidos Vivos/SINASC) were used. NNM was defined by having one or more of very low weight at birth (<1500g), asphyxia at 5 minutes from birth (APGAR at fifth-minute score <7), or less than 32 gestational weeks at birth. A descriptive analysis of the data was made and the trends in both near-miss numbers and their ratio to neonatal mortality were evidenced from 2011 to 2020. This trend included a regression model to describe the behavior of the data along the time. In parallel, independent T-tests were used to compare the NNM-related variables between the group of neonates with CH and the live births in general. From 2011 to 2020, a total of 2550 NNM cases were observed among the 7699 newborns with CH, resulting in a NNM rate of 331 cases per 1000 live births. 2013 was the year with the highest NNM rate with a total of 380 NNM per 1000LB with CH, followed by 2012 (328 per 1000LB) and 2015 (335 per 1000LB). During the period, the NNMR trends presented two different behaviors, with an annual percent change (APC) of +1,84% from 2011 to 2013, and with an APC of -2,18% from 2013 to 2020, resulting in a total reduction of 7,85% in the ten years. Comparing the CH group and the general neonates, the first group presented a higher NNM rate ($p < 0,001$; 95% CI 279-317). Also, there was a higher NNM to mortality ratio ($p < 0,05$; 95% CI 0,10-0,62) in the neonates with CH, indicating a better success of care for the CH near-misses than for general near-misses, hence there are more cases of NNM per overall deaths. **Conclusion:** This study applied the neonatal near-miss concept to analyze the live births with congenital hydrocephalus in Brazil, presenting a change of behavior in trends in the last 7 years. A comparative analysis showed that CH neonates present with more cases of NNM, but at the same time present with a higher rate of NNM to mortality, indicating a recovery of the newborns from the conditions of birth.

Neurologia Infantil

TL 1106390

EPIDEMIOLOGICAL PROFILE OF DRUGS USED IN PEDIATRIC NEUROLOGY CARE OF PATIENTS WITH AUTISTIC SPECTRUM DISORDER (ASD) AT A REFERENCE HOSPITAL IN BRASÍLIA

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Background: Autism Spectrum Disorder (ASD) is a term used to describe a range of neurodevelopmental disorders characterized by persistent deficits in communication and social interaction and by the presence of repetitive or restricted behaviors and/or interests. Its prevalence was estimated, in 2012, between 1 and 5%, in developed countries, and it is increasing in recent years. Due to its impact on patients' quality of life, it is crucial to apply an appropriate and individualized treatment that encompasses behavioral and pharmacological interventions (which do not treat ASD itself, but possible related symptoms, such as agitation and irritability). **Objective:** To characterize the pharmacological treatment profile used by the pediatric neurology service of the Reference Hospital for patients with ASD. **Method:** A retrospective, descriptive, cross-sectional cohort study, using data collected from the medical records of patients diagnosed with ASD followed at the Pediatric Neurology service of the Reference Hospital. **Results:** The medical records of 109 patients with ASD were evaluated, among which there was 1 loss of follow-up. Of the patients evaluated, 19.2% were using antidepressants ($n=21$) and 45.8% were using antipsychotics ($n=50$). Among antidepressants, the most used was fluoxetine, used by 52.3% ($n=11$) of patients using antidepressants, followed by sertraline (47%). Regarding antipsychotics, risperidone was the most used, representing 74% of this group ($n=37$), followed by pericyazine (14%), aripiprazole (8%) and haldol (6%). 8 additional patients that had used risperidone in the past were identified, but had quit due to lack of response to the medication or side effects. Other medications such as cannabidiol, methylphenidate and anti-epileptics were being used by 19 patients. **Conclusion:** Pharmacological interventions in autism are reserved for particular situations in ASD and should have their use adjusted to the needs of each patient. The main medications used in the service analyzed were antipsychotics, especially risperidone, the first drug approved by the Food and Drug Administration (FDA) to treat irritability related to autism, followed by antidepressants. Other pharmacological treatments were implemented to control conditions such as agitation and insomnia, especially after a lack of response to the most commonly used medications, as a way of individualizing treatment.

Neurologia Infantil

TL 1106392

ASPECTS OF THE QUALITY OF LIFE OF FAMILIES CARING FOR PATIENTS WITH REFRACTORY EPILEPSY

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Background: Adaptive family resources such as communication, dominance, social support and financial well-being are associated with good social adaptability in patients with epilepsy. However, to date few studies were carried out to assess the impact of the presence of a patient on the functioning of the family. In the case of refractory epilepsies, the impact is even more significant, it is a stressful process which requires emotional mastery, physical integrity, as well as time, dedication, and financial resources. **Objective:** To assess the quality of life of family members who care for children with refractory epilepsy. In addition, to present the characteristics of the family's social, economic and emotional conditions, limitations, compromised by refractory epilepsy, evaluating the expectations, anxieties, and fears of the parents in relation to the risks of Hospitalization, death and regarding the academic and professional future of the child. **Method:** Descriptive study with a qualitative approach carried out remotely with the caregivers of children/adolescents with refractory epilepsy, who are followed up in a specialized outpatient clinic of a University Hospital in the Federal District. For data collection, a semi-structured form was used and an individual interview was carried out. Data collection was carried out through telephone contact. **Results:** Four families participated in the study, aged between 20 and 42 years. In this study, it was found that the children and adolescents participating in this study perform most of their daily activities with the help of their family members, thus requiring greater support and dedication from their caregivers. In addition, it was revealed that family members feel partially satisfied with their quality of life, this is due to several factors such as: lack of help from others to manage the child/adolescent, insufficient family income, collection, debt, overload, family conflicts, impact on marital life, feelings of guilt and unemployment, which cause anxiety, hopelessness and depression in family members. **Conclusion:** Caring for people with chronic neurological disorders has a negative impact on the family structure, causing psychological stress due to the need for adaptations that are not always achieved. In this way, care must be analyzed, not only from the perspective of the sick child, but extended to the entire family nucleus.

Neurologia Infantil

TL 1106400

MUSCLE ULTRASOUND AS A TOOL FOR RESPIRATORY ASSESSMENT IN PATIENTS WITH LAMA2-MD

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Background: LAMA2-muscular dystrophy (LAMA2-MD) is an autosomal recessive disease, and the most common form of congenital muscular dystrophy (CMD). Most of the patients develop a form of disease characterized by inability to achieve walking capacity, multiple joint deformities, respiratory insufficiency, and some degree of dysphagia. However, there is a gravity spectrum. There are still no adequate biomarkers to assess disease progression, and muscle ultrasound (US) can be a useful tool and also complement the assessment of respiratory and swallowing function. **Objective:** Evaluate, through muscular US, the function of the respiratory muscles, tongue muscles and correlate them with respiratory function, degree of dysphagia, disease severity and age. **Methods:** 10 patients with genetically confirmed LAMA2-MD were divided according to motor severity and evaluated. US of tongue, respiratory and paravertebral muscles were made. For muscles comparable to bone echo, the 4-point Heckmatt scale was used, for the others the classifications were hypoechoic, slightly hyperechoic, or very hyperechoic. Patients underwent respiratory function assessment and underwent neuromuscular disease swallowing status scale (NdSSS). **Results:** Two patients had severe presentation and were not able to sit without support. They presented genioglossus and genioglossus muscles very hyperechoic, had level 3 NdSSS dysphagia and required gastrostomy. They both had very affected external oblique, internal oblique and transverse muscles, but presented normal diaphragm, with normal thickening. They had altered polysomnography. 4 patients had classic disease presentation, with maximum motor ability to sit without support, and were under 6 years old. They presented genioglossus and genioglossus muscles slightly hyperechoic, affected external oblique muscle with normal internal oblique and transverse muscles and normal diaphragm, with normal thickening. They all had total lung capacity (TLC) above 50% and level 7 NdSSS. Other 3 patients had classic disease presentation but were older than twelve years old, who presented genioglossus and genioglossus muscles highly hyperechoic, affected external oblique, internal oblique and transverse muscles, with normal diaphragm, with normal thickening. They all had TLC below 35% and level 7 NdSSS. **Conclusion:** Muscle US can be used as a tool to evaluate disease progression and contribute to the assessment of respiratory function and dysphagia in LAMA2-MD.

Neurologia Infantil

TL 1106418

KINSBOURNE SYNDROME: A CASE PRESENTATION

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Case Presentation: Male infant, 1 year old and 10 months, third child of non-consanguineous parents, 2 healthy older sisters and mother with 2 abortions, presenting with aggressive behavior 3 days after immunization with inactivated polio vaccine and Hepatitis B vaccine in December 2021. Progression to ataxic gait and dysphagia after 15 days, with myoclonic movements of the upper limbs and opsochonus after 30 days. Admitted to the emergency department in January 2022, submitted pelvis, abdomen and chest CT, Cerebrospinal fluid and brain MRI without alterations. Underwent 5 days of pulse therapy with methylprednisolone at 30mg/kg/day, with no change. Then performed IVIG at 2g/kg in 5 days, ceasing opsochonus and improving gait. Myoclonus was controlled by Clonazepam. Discharged after 14 days but did not maintain the corticosteroid therapy at home. May 2022, he presented recurrence of opsochonus and worsening of ataxic gait. Admission exam: Regular general condition; irritated; unintelligible speech; trophism, tone and strength preserved. Exacerbated deep reflexes (grade 3), globally. Exalted axial face reflexes. Did not load weight on the lower limbs, making gait assessment difficult. Coordination with dysmetria and global decomposition. A new course of IVIG was performed, with no change to the clinical presentation. Then, a new course of pulse therapy with methylprednisolone was realized with global improvement, except for gait ataxia, in a lesser degree. A new screening for occult neuroblastoma was performed, with normal CT of the pelvis, chest and abdomen. **Discussion:** Kinsbourne syndrome is a rare inflammatory pathology, with no clear etiology. Up to 50% of cases are paraneoplastic, with neuroblastomas being identified in the majority of them. Despite the extensive search for neuroblastoma at this patient, no tumor processes were identified. He is still awaiting to perform full-body scintigraphy, in order to complete this search. **Conclusion:** Early recognition of Kinsbourne syndrome enables early treatment, leading to faster clinical improvement, with less cognitive sequelae. There is no consensus as to which immunomodulatory treatment to use, which varies according to the response of each patient. The absence of neuroblastoma at the time of the clinical occurrence of the syndrome does not definitively rule out its presence, requiring a new periodic evaluation.

Neurologia Infantil

TL 1106511

ANGELMAN SYNDROME: THE SEARCH FOR BRAIN KNOWLEDGE

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Background: Angelman Syndrome (AS) is a neurodevelopmental disorder, considered incurable, characterized by ataxia, intellectual disability, speech impairment, seizures, autistic behavior, hyperactivity and happy behavior, caused by the loss of function of the UBE3A maternally inherited region of the chromosome 15q11-13 and diagnosis is usually made between 1 and 4 years of age. For a consensus criterion for diagnosing AS, the clinical features of individuals are divided by their percentage of frequencies in the syndrome: consistent features, frequent features, associated features. **Objective:** To carry out a survey of current research on the definition of the profile of cases related to Angelman syndrome. **Methods:** This is a bibliographic review study, extracted from the PUBMED and The Febs Journal database, using an understanding of the brain areas affected in AS. **Results:** Identified by noted physicians and researchers, Angelman Syndrome is understood to be caused by the loss of the maternal copy of the UBE3A gene in the region of chromosome 15q11-13. To diagnose AS, the clinical characteristics of individuals are divided into percentage of frequencies in the syndrome. As consistent features (100%), all patients with AS have ataxia, functionally severe developmental delay, frequent laughter/smiling associated with hypermotor behavior, and speech impairment with no or minimal use of words, but greater use of non-verbal communication. In the frequent features, 80% of AS cases show a small head circumference associated with microcephaly, electroencephalogram abnormalities, and frequent and severe seizures during childhood that decrease with age but may persist into adulthood. Associated features in 20 to 80% of patients with AS include obesity, scoliosis, constipation, abnormal sleep cycles, wide mouth, squint, hypopigmented skin, attraction to water, and many others. Not all features need to be present for a diagnosis of AS, but it is mandatory that patients exhibit the four consistent features. **Conclusion:** It is believed that understanding about AS in the brain will only be achieved with a faithful recapitulation of brain development processes in emerging 3D models.

Neurologia Infantil

TL 1106515

BIOMARKERS OF BRAIN ELECTRICAL ACTIVITY IN INDIVIDUALS WITH AUTISTIC SPECTRUM DISORDER

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Autism Spectrum Disorder (ASD) is a neurodevelopmental disorder characterized by neurological disorders, which includes communication deficit, decreased social interaction, behavioral pattern, restricted and repetitive activities and interests. The diagnosis of ASD is clinical and based on the Diagnostic and Statistical Manual of Mental Disorders (DSM-5). This defines ASD as the entities Asperger's Syndrome, Autistic Disorder and Generalized Developmental Disorder not otherwise specified, which include Childhood Disintegrative Disorder and Rett's Syndrome. Due to the difficult diagnosis, numerous studies with electroencephalogram (EEG) exams in autistic individuals have shown interesting results in the search for a biomarker of this condition. In this work, carried out by analyzing the EEG record in autistic individuals (N = 22) in comparison to the control group (N = 8), it was found that in the TEA group there was a lower oscillation power of the low frequency waves (alpha, delta and theta) in relation to the control group, and in alpha and theta this finding occurred in the whole brain. Along with this, there was a slight predominance of the gamma oscillation power in the TEA group in relation to the healthy group, another fact was the inverse relationship between the oscillation power of the low frequency waves (alpha, theta and delta) and the power of range oscillation.

Neurologia Infantil

TL 1106538

EVALUATION OF CHILD DEVELOPMENT OF CHILDREN WITH MICROCEPHALY BY CONGENITAL ZIKA VIRUS SYNDROME

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Background: Microcephaly is a congenital malformation in which there is inadequate brain development. Its causes are multiple, among them: the exposure of the mother to the Zika virus during pregnancy. Children may have severe neurological impairment with the presence of brain abnormalities evidenced by intracranial calcifications related to cell death, ocular abnormalities, particularly microphthalmia, sleep disturbances, excessive irritability to touch, and epilepsy. In view of the important neuro-repercussions, children need to be monitored and evaluated regarding their development. **Objective:** To evaluate the child development of children with congenital microcephaly caused by the Zika virus. **Method:** A cross-sectional observational study was carried out, assessing child development using the Ages & Stages Questionnaires 3rd Edition (ASQ3 - BR) instrument. This study was approved by the ethics committee of the Federal University of Paraíba (CAAE: 42852621.0.0000.5188) and conducted according to the criteria of Resolution No. 466/2012 of the National Health Council. Nineteen children of both sexes were included in the study. **Results:** No child scored higher or close to the ASQ3 cut-off score. An important part of the sample showed severe impairment in all domains evaluated, scoring zero in the following skills: communication (84.16%), broad motor (73.64%), fine motor (89.42%), problem solving (84.16%) and personal skills and social (84.16%). **Conclusion:** The results of this study can provide evidence that alerts about the importance of continuous monitoring of these children by a multidisciplinary team, creating therapeutic proposals that stimulate the patient's development, reducing damage and promoting quality of life.

Neurologia Infantil

TL 1105459

PREOPERATIVE ENDOVASCULAR EMBOLIZATION OF GLOMUS JUGULARE TUMORS: A RETROSPECTIVE CASE SERIES OF 22 EMBOLIZATIONS IN 20 PATIENTS

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Background: Glomus jugulare tumors are rare, highly vascularized though slow-growing skull base tumors that originate from paraganglion cells of the neural crest. The approach for these lesions remains controversial, neurosurgeons are often faced with challenges due to the rich vascularization of the lesions as well as to the intimate relationship with neural and vascular structures. To minimize the risks for the patient and ensure total resection, a combined approach is ideal. **Objective:** Evaluate the profiles and outcomes of patients submitted to preoperative jugular glomus embolization in a Brazilian tertiary care Hospital. **Methods:** The present study is a retrospective analysis of a series of paraganglioma embolizations between January 2008 and December 2019. The embolization was performed through super selective catheterization of feeder arteries between 3 to 5 days prior the surgical procedure; feeders from the external carotid artery and internal carotid artery were embolized with gelatin foam, polyvinyl alcohol foam, or polyvinyl alcohol particles, embolic spheres, Bead Block, and ethylene-vinyl alcohol copolymer, or EVOH. The aim of the treatment was radical and complete tumor removal in one surgical procedure with preservation of the cranial nerves. **Results:** In the period studied 22 paraganglioma embolizations were performed in 20 patients, 11 of them had already been submitted to previous neurosurgical procedures. There were 11 women and 9 men, with a mean age of 51.25 years old. All patients underwent both embolization and surgery afterwards. Hearing loss was the most common symptom (50%), while 40% of patients had tinnitus, 30% had dysphagia, 25% had facial paralysis, 20% had hoarseness, and 10% had diplopia. The most common vessel involved was the ascending pharyngeal artery (90%), followed by the posterior auricular artery (15%), the internal maxillary artery or the occipital artery (10% each), and the superficial temporal or the lingual arteries (6% each). The embolization was deemed total in 13 procedures, partial in 4 procedures, and was not reported in the remaining 5. Only one patient had involvement of the internal carotid artery. No complications from embolization were recorded. **Conclusion:** Glomus jugulare tumors are complex lesions that demand a multidisciplinary approach to ensure satisfactory resection with preservation of lower cranial nerves. Preoperative embolization is essential to ensure one-stage operation.

Neurologia Intervencionista

TL 1105466

FLOW DIVERTER ENDOLUMINAL DEVICE (FRED) FOR THE TREATMENT OF INTRACRANIAL ANEURYSMS: 6 YEARS CLINICAL RESULTS

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Background: Flow diverter are one of several models of metallic stents used in intracranial aneurysms (IAs) embolization. They show an advantage over endosaccular coiling since they treat the weakened arterial wall by providing a scaffold for new endo-thelial cell growth to occur. The FRED (Flow Diverter Endoluminal Device) is a model of flow diverter that stands out for being a two-layer braided spiral that helps to reconstruct the blood vessel wall. **Objective:** Report the experience of the use of the FRED dispositive in the endovascular treatment of IAs and evaluate the safety and efficacy of this intervention. **Methods:** This retrospective and single-center observational study evaluates the use of FRED or FRED Jr in the endovascular treatment of IAs from July 2016 to April 2022. Patients included were older than 18 years, and treated with a FRED or FRED Jr dispositive; and were excluded if the implant did not occur for some complication or if they didn't perform a control cerebral angiography after the procedure. Dual antiplatelet therapy (acetylsalicylic acid and clopidogrel) was managed 5 days before and 6 months after the procedure and acetylsalicylic acid was used for another 6 months. Data were collected from the medical records: sex and age, number and location of IAs, complications, morbidity and level of IA occlusion - following the O'Kelly Marotta (OKM) classification scale. All intraprocedural complications were reported. The follow-up of patients was carried out 6 and/or 12 months after the procedure with a digital subtraction angiography. **Results:** Between July 2016 and April 2022, 191 patients were screened, which 31 met exclusion criteria. 135 patients were female. The mean age was 53 years. A total of 198 IAs were treated - 178 of them in the internal carotid artery, mainly in the ophthalmic segment. Following the OKM scale for occlusion of IAs, 61% fulfilled category D; while C, B and A were observed in, respectively, 11%, 7% and 6%. 28 patients had not undergone control angiography at the time of the study, but were not excluded because there was still time for follow-up. The morbidity rate was 16% (mRS = 1, mainly headache). No deaths were reported. **Conclusion:** IAs are more common in females after 50 years old. In this study, the occlusion rate was seen in 70% of the cases, a value that can still increase as follow-ups are concluded. Thus, high efficacy in the use of FRED in the endovascular treatment of IAs is suggested.

Neurologia Intervencionista

TL 1105222

MONONEUROPATHY MULTIPLEX: A 100-CASE SERIES

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Background: Mononeuropathy multiplex is a group of uncommon disorders. Definition is involvement of two or more noncontiguous peripheral nerves. National studies are needed to understand local epidemiology, as numerous causes have been identified. **Methods:** We retrospectively reviewed data of patients with an electrodiagnostic diagnosis of mononeuropathy multiplex. Etiology was divided as systemic vasculitis, isolated peripheral nerve vasculitis, chronic inflammatory neuropathy, infection (including leprosy), genetic, neoplastic disorders, and undetermined. Electrodiagnostic pattern was classified as axonal, demyelinating, intermediated (mixed), and associated pattern (clearly demyelinating lesions in some nerves and axonal lesion in others). When available, tissue biopsy was also reviewed. **Results:** A total of 100 patients were included - mean age was 47.4 years, 48 were male. Most frequent etiology was systemic vasculitis with 51 patients. Among these, lupus was the most common diagnosis, but rarer vasculitis that are seldom associated with mononeuropathy multiplex were also found. Isolated peripheral vasculitis was seen in three patients. Leprosy was the most frequent infection with 28 patients, only two presenting with pure neural leprosy. Chronic inflammatory neuropathy was represented by three cases of multifocal CIDP and 03 cases of multifocal motor neuropathy. Genetic was represented by two cases of HNPP. EDX was consistent with an axonal pattern in 70 cases, demyelinating in 12, intermediated in 13, associated in five. Entrapment neuropathies pointed towards an infectious etiology. A total of 17 nerve biopsies, 44 skin biopsies, three salivary gland biopsies, and seven renal biopsies were available to evaluation. Histology strongly suggested vasculitis in 33 biopsies, and 19 biopsies were diagnostic of leprosy. **Conclusion:** Facing a mononeuropathy multiplex, systemic vasculitis is the most frequent diagnosis. Very rare vasculitis types should also be considered. In our population, leprosy should always be a concern in asymmetric neuropathy. Although a diagnostic challenge, an etiology can be clearly identified in majority of patients. Non neural tissue biopsies can be very helpful with much less distress.

Neuropatias Periféricas

TL 1105278

FOLLOW-UP OF INDIVIDUALS WITH HEREDITARY TRANSTHYRETIN AMYLOIDOSIS

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Background: Hereditary transthyretin amyloidosis is caused by a genetic mutation that leads to deposition of the malformed transthyretin protein in various organs and tissues. Symptoms include peripheral neuropathy, cardiac, renal, ocular, gastrointestinal involvement and carpal tunnel syndrome. Autonomic function is often undervalued and is often compromised from the onset of symptoms resulting in cardiac conduction defects, orthostatic hypotension, urinary retention, sweating abnormalities, and sexual dysfunction. Genetic testing in pre-symptomatic individuals is now widely available and may provide a proper follow-up. In clinical suspicion and altered complementary exams, the diagnosis must be confirmed through tissue biopsy showing the deposit of amyloid material. **Objectives:** to make an early diagnosis of amyloidosis in asymptomatic carriers **Methods:** Asymptomatic individuals with hereditary amyloidosis from the Neuromuscular Diseases of Neurology outpatient clinic of the Pedro Ernesto University Hospital in Rio de Janeiro are included in the study. A semi-annual follow-up is carried out and individuals are periodically submitted to neurological and neurophysiological examination, evaluation of small fiber neuropathy (through QST (quantitative sensory testing), sympathetic-reflex response, dysautonomia questionnaire, heart rate variability) and cardiologic evaluation. **Results:** 10 individuals are being followed up and two individuals have developed signs and symptoms of the disease, confirming the deposit of amyloid material through salivary gland biopsy. A 32-year-old woman with dysautonomia and bilateral carpal tunnel syndrome has the Phe84Leu mutation and the index case was her father, who had a late diagnosis and died due to complications from the disease. A 22-year-old woman with Val50Met mutation presented small fiber neuropathy, with abolition of the reflex sympathetic response in the 4 limbs and QST abnormalities when compared to a healthy control matched by sex and age. The index case was his maternal aunt who died at age 38 due to dysautonomia. **Conclusion:** hereditary amyloidosis can be difficult to diagnose, since its clinical manifestations are quite heterogeneous. An early diagnosis is essential for an effective treatment. Monitoring asymptomatic individuals is very important for early detection of the disease.

Neuropatias Periféricas

TL 1105307

ULTRASOUND ABNORMALITIES OF THE MEDIAN NERVE IN LEPROSY NEUROPATHY AND CARPAL TUNNEL SYNDROME: HOW TO DIFFERENTIATE THESE CONDITIONS?

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Background: Leprosy is a chronic infectious disease caused by *Mycobacterium leprae* which primarily involves the nerves and the skin. Its clinical forms are defined by the host immune response and bacillary load, resulting in a wide clinical spectrum. Currently, nerve assessment in leprosy relies mainly on clinical evaluation and nerve conduction studies, but although neurophysiology provides detailed information about the dysfunction of affected nerves, it does not reveal anatomic changes, such as thickening and fascicular pattern changes. High-resolution ultrasonography (US) is an additional technique that permits the examination of multiple nerve trunks over a long course in a few minutes and is considered an accessible and reasonably precise method. **Objective:** This study aims to compare the sites of median nerve enlargement in patients with leprosy and carpal tunnel syndrome (CTS). **Methods:** During the year 2021, were included in this study 15 leprosy patients and 14 patients diagnosed with CTS, totaling 27 nerves in each group. All patients underwent a multisegmental US session of the median nerves, performed by an expert radiologist. The median nerve was scanned at the carpal tunnel (Mt) and 4 cm above the carpal tunnel (Mpt). The measurements of the cross-sectional area (CSA) and morphological characteristics such as echogenicity and fascicular pattern were assessed at these two points. We also calculated the absolute difference between CSAs of the nerve in the Mt and Mpt (?Mtpt = Mt - Mpt). **Results:** CSA of the median nerve was increased in leprosy (13.4; ± 4.4) and CTS (12.4; ± 4.6) nerves at the Mt, but with no significant difference. 100% (27/27) of the nerves with leprosy showed some morphological abnormality at the Mt point and only 40.7% (11/27) of the CTS group ($p=0.0042$). CSA was higher among nerves with leprosy (17.0; ± 6.7) as compared to CTS (6.6; ± 1.3) at Mpt (< 0.0001). 100% (27/27) of the nerves with leprosy showed some morphological abnormality at the Mpt point and none of the CTS group. The ?Mtpt in leprosy nerves was -3.6 (± 6.5) and 5.7 (± 4.0) in the CTS ($p < 0.0001$). A measure of the ?Mtpt ? 2.2 indicates the diagnosis of CTS with a sensitivity of 100% and specificity of 96.3%. **Conclusion:** Median nerve enlargement in leprosy using US seems to be more proximal than in carpal tunnel syndrome (CTS), which might be an important discriminating sign.

Neuropatias Periféricas

TL 1105355

POEMS SYNDROME: A BRAZILIAN CASE SERIES

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Background: Polyneuropathy, organomegaly, endocrinopathy, monoclonal-protein and skin changes (POEMS) is a rare paraneoplastic syndrome. It may be difficult to diagnose, specialty in developing countries facing specialist's shortage. In this study we sought to describe clinical, laboratory and electrophysiological characteristics found in 19 patients diagnosed in a Brazilian referral center. **Objective:** to describe the clinical and laboratory characteristics of POEMS syndrome in a Brazilian tertiary center. **Methods:** POEMS diagnosis was based according to current diagnostic criteria. Clinical data was retrospectively collected. **Results:** Fourteen out of 19 patients were male (73.7%). The mean age at onset was 48.6 years old (range 38 to 66) and the mean time for diagnosis was 13.7 months (range 5 to 48). Nerve conduction studies from all patients revealed a sensitive-motor demyelinating neuropathy. Six patients underwent nerve biopsy, and none had inflammatory infiltrate. Papilledema was present in 7 patients (36.8%). All cases had albuminocytological dissociation. A lambda monoclonal component was detected in serum from all cases, but clonal plasma cells on bone marrow biopsy were found in only 8 cases. Bone lesions were found in 18 cases (94.7%). Sixteen (84%) had typical skin changes. Endocrinopathy was present in 17 cases (89.9%), lymphadenomegaly in 6 (31.5%), splenomegaly in 9 (47%), hepatomegaly in 6 (31.5%) and thrombocytosis in 13 (68.4%), respectively. Two cases had Castleman's disease. **Conclusion:** POEMS syndrome is a rare condition. Our data suggest that the prevalence of endocrinopathy and bone lesions in our cohort were more frequently found than expected, and may be related to a longer time of disease duration leading.

Neuropatias Periféricas

TL 1105406

AValiação DA FORÇA PROXIMAL, DA MARCHA, EQUILÍBRIO, DA FUNCIONALIDADE E DA QUALIDADE DE VIDA EM PACIENTES COM POLIRRADICULOPATIA DESMIELINIZANTE INFLAMATÓRIA CRÔNICA

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Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) is an acquired disorder of the Peripheral Nervous System (PNS) where the myelin sheath is affected. Its etiology is still unknown and in most cases its evolution is progressive and symmetrical, with proximal and distal involvement of the upper and lower limbs, causing changes in gait, balance, functionality and quality of life. Several assessment scales have been proposed aiming to improve patient follow-up and management of secondary complications. The aim of this study was to evaluate the impact of proximal lower limb strength on gait, balance, functionality and consequently in quality of life in patients with CIDP. Scales that aid in the clinical assessment of gait, balance, functionality, muscle function and quality of life were used, such as: Dynamic Gait Index (DGI), Performance Oriented Mobility Assessment (POMA); Modified Rankin Scale (mRS), Medical Research Council (MRC) and The Medical Outcomes Study 36-item Short-Form Health Survey (SF-36) respectively, all in their Brazilian versions. Seven patients participated in the research, most of them males (71.4%), mean age of 48 years (median 41 years). According to the MRC, the patients had proximal muscle weakness (hip flexors) ranging in score from 3 to 4 (mean 3.57) and distal muscle weakness ranging from 2 to 4 (mean 3.29). According to the DGI scale, all participants were at risk of falling with a score equal to or below 19 points (mean of 17.43). According to the POMA scale, one patient (14.2%) had a high risk of falls with a score of 19, five participants (71.6%) had a moderate risk of falls, with scores between 20 and 24, and only one participant (14.2%) had no risk of falls, with a score of 25. The average among individuals was 22.71 points on the POMA scale. It was observed that the physical aspects obtained the lowest score in the average of the population under study in relation to the components of the SF-36, impacting more significantly on the sample. It can be concluded from this study that proximal and distal paresis in the lower limbs in individuals diagnosed with typical CIDP affects normal gait and balance patterns, according to the DGI and POMA scales, and these scales can be a risk predictor of falls in this population. It was also observed that functionality in this population, despite being a chronic and progressive disease, is not severely affected, according to the mRS scale.

Neuropatias Periféricas

TL 1105509

A LONG-TERM FOLLOW-UP OF BRAZILIAN PATIENTS WITH TRANSTHYRETIN FAMILIAL AMYLOID POLYNEUROPATHY AFTER LIVER TRANSPLANT

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Background: Transthyretin familial amyloid polyneuropathy (ATTRv-pn) is an inherited neuropathy, with over 140 mutations identified in the transthyretin (TTR) gene. ATTRv-pn is a severe length-dependent sensorimotor/autonomic neuropathy, which if untreated, leads to death in approximately 10 years (y). The liver produces 98% of the TTR circulating in the blood, and it was thought that liver transplant (LT) would stop disease progression. However, subsequent studies showed that the **Results:** were not good in late-onset patients (? 50y), those who were malnourished, with advanced disease and/or had non-V50M mutations. It proved to be very effective in controlling the neuropathic manifestations, but less effective in controlling cardiac manifestations. **Objective:** To describe the long term follow up of a series of ATTRv Brazilian patients with polyneuropathy who underwent LT. **Method:** We have reviewed the medical records of all patients (8) with ATTRv-pn who underwent LT and were followed in our institution. The diagnosis was made by genetic testing. We have used the Composite Autonomic Symptom Score 31 (COMPASS 31) for quantitative measure of autonomic symptoms. **Results:** All patients had the ATTRv-pn V50M variant. The mean age at symptoms onset was 31,2y. All patients had neuropathic pain before LT, and 2 of them experienced complete pain relief after it. All patients have clinical dysautonomia and the mean of the last COMPASS 31 was 52,6. 50% have confirmed cardiomyopathy; 37,5% are still under investigation and 12,5% do not have it. 87,5% have some arrhythmia or conduction-system disease and 50% needed pacemaker implantation between 2002 and 2022. The mean time from symptoms onset to diagnosis was 2y and from symptoms onset to LT was 5,37y. 6 patients are fully ambulant – Coutinho stage (CS) 1 -, 1 patient needs one walking aid – CS 2 - and 1 patient became wheelchair-bound – CS 3 - in the last 2y. The mean time from symptoms onset to the last clinical evaluation (2022) was 18,2y (8-23y), and from LT to the last clinical evaluation (2022) was 13,5y (6-18y). **Conclusion:** Our patients have most of the factors for good prognosis cited in the literature, as V50M mutation, early-onset and early-stage disease when they underwent LT. The 20-y patient survival rate was 62,5%, higher than the average in the literature (55,3%). The mean time from symptoms onset to diagnosis was lower than it is in Brazil (5,9y).

Neuropatias Periféricas

TL 1105599

EPIDEMIOLOGICAL AND CLINICAL CHARACTERISTICS OF GUILLAIN-BARRÉ SYNDROME CASES TREATED AT A PUBLIC UNIVERSITY HOSPITAL IN BRAZIL BETWEEN 2014-2020

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Background: Guillain-Barré syndrome (GBS) is the most common acute inflammatory neuropathy, which causes sensitive and motor symptoms, and can also be associated with dysautonomia. In Brazil, there are few epidemiological and systematized regional studies about the syndrome. **Objective:** The aim of the present study is to evaluate the epidemiological, clinical, and neurophysiological aspects of patients with GBS, treated at a public university Hospital, between 2014 and 2020. **Methods:** It is a retrospective study made by the analysis of 64 medical charts of patients diagnosed with GBS, between January 2014 and December 2020. **Results:** The majority of the patients are men (70,3%), with an average age of 39,1/±23,3. 79,7% of the patients claim to have had an infection before the symptoms, especially gastrointestinal (51,0%), respiratory (25,5%), and arboviruses (13,7%). Only 3,1% had a recent vaccination. Among the clinical forms of GBS, the most common was the classic sensory-motor form (70,3%) followed by the pure motor form (23,4%). The pharyngeal-cervical-brachial (3,1%), paraparetic (1,6%), and Miller-Fisher (1,6%) forms were less commonly observed. Regarding the physical examination at admission, all cases presented muscle weakness. Areflexia or hyporeflexia was present in 89,1% of the patients. Other clinical findings observed: sensory ataxia (48,4%), pain (46,9%), hypopallesthesia (43,8%), hypoaesthesia (40,6%), paresthesia/dysesthesia (39,1%). 34,4% of the patients had involvement of some cranial nerve and 23,4% had facial paresis. Dysautonomic symptoms such as tachycardia, dyspnea, and urinary retention/intestinal constipation have been observed in 23,4%, 17,2%, and 9,4%, respectively. In 25,0% of the cases, there was an asymmetric distribution of sensory abnormalities and in the assessment of motor function, there was a proximal predominance in 84,4% and an asymmetric distribution in 40,6%. The AMAN and PIDA neurophysiological patterns were the most commonly observed, both present in 40,3% of patients who underwent electroneuromyography. The time between symptoms onset and admission was 14,2/±13,0 days, with a Hospital stay of 11,3/±11,4. 15,6% required ICU admission and 12,5% developed respiratory failure and required mechanical ventilation. **Conclusion:** In the context of public health, it is important to recognize the clinical and epidemiological aspects of the GBS cases, to elaborate more efficient diagnosis and treatment strategies.

Neuropatias Periféricas

TL 1105746

SEVERE CHRONIC RECURRENT DEMYELINATING POLYNEUROPATHY AFTER A SINGLE DOSE OF CHADOX1-S NCOV-19: A CASE PRESENTATION

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Case Presentation: Female, 33 years old, was vaccinated with a single dose of ChAdOx1-S nCOV-19 (Fiocruz-AstraZeneca) at 02/02/2022. She was admitted to an emergency unit on 02/26/2021 with complaints of paresthesia in palate and limbs. CSF analysis showed: glucose of 52 mg/dL; proteins 72 mg/dL; 1 cell/mm³; and C. jejuni IgG ELISA indeterminate levels (0.93). Electroneuromyography (ENMG) identified acute motor and sensory demyelinating polyneuropathy on all four limbs. She evolved within 2 days with distal flaccid tetraparesis, dysphonia, dysphagia and dyspnea. At this point, a diagnosis of Guillain-Barré Syndrome (GBS) was made. Intravenous human immunoglobulin (IVIG) was started 3 days after symptom onset. Five days after IVIG, she developed facial diplegia, worsening of tetraparesis, and dysautonomia. A new IVIG infusion was performed 6 days after the first one, with improvement of limb paresis. In March 2021 she was readmitted with worsening of the facial diplegia, and plasmapheresis was instituted with good discussion. In April 2021 she returned with worsening dysphonia and paresis, with weak positivity to c-ANCA. Thus, suspicion was changed to ANCA-related vasculitis associated with polyneuropathy. She was then treated with methylprednisolone and cyclophosphamide, evolving with worsening of clinical state with need of mechanical ventilation. A new plasmapheresis and rituximab were performed, with a good clinical response. She was readmitted in May 2021 with worsening weakness and treated with plasmapheresis and IVIG. Specific antibodies were negative for systemic vasculitis, and a diagnosis of recurrent chronic demyelinating polyneuropathy (CIDP) was made. It was opted for a monthly maintenance of IVIG from 08/2021 until the present time. Currently, the patient is stable with preserved muscle strength. **Final comments:** CIDP is characterized by autoimmune, inflammatory, demyelinating lesions affecting peripheral nerves and nerve roots, with a chronic or remitting/recurrent course of neurological disability and is treatable with immunomodulatory therapies. Neurological complications have been reported after COVID-19 vaccination and C. jejuni infection. Longitudinal follow-up confirms the diagnosis of CIDP; the causal relationship with COVID-19 vaccination is possible considering other similar case presentations on literature. The absence of diarrhea and the weak antibody reactivity against C. jejuni decrease the chance of this agent as etiology.

Neuropatias Periféricas

TL 1105799

GUILLAIN-BARRÉ SYNDROME AFTER COVID-19 VACCINATION: CASE SERIES

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Background: Guillain-Barré syndrome (GBS) is an acute or subacute immune-mediated polyradiculoneuropathy generally preceded by an infectious trigger. The first link between GBS and vaccines was an 8-fold increase in incidence of GBS in receivers of a Swine influenza vaccine in 1976. Recently, MMR, HPV, MCV4 and Influenza vaccines were pointed as possible triggers, but this association is questionable. During the coronavirus (COVID-19) pandemic, cases of GBS preceded by COVID-19 vaccination have been reported, especially associated with viral vector-based vaccines. **Objective:** To describe cases of GBS diagnosed within 6 weeks after COVID-19 vaccination during the mass vaccination period. **Methods:** We performed a retrospective review of all patients diagnosed with GBS within 6 weeks after receiving COVID-19 vaccination admitted at our service from January 19th, 2021 to August 22nd, 2021. Exclusion criteria was the presence of other probable triggers during this period, including documented COVID-19 infection. Collected data include gender, age, vaccine received, time between vaccination and symptoms onset (latency), neurological manifestations, cerebrospinal fluid analyses, electrophysiological study, treatment and outcome at Hospital discharge. **Results:** During the studied period, 7 patients met the inclusion criteria (6 men and 1 woman) with an average age of 53.4 years old. 6 received ChAdOx1-S vaccine (AstraZeneca) and 1 received Ad26.COV2.S vaccine (Janssen). Except for one patient, who developed symptoms 1 day after immunization, all other patients had a latency time between 12 and 23 days. We identify that 5 of 6 patients who underwent lumbar puncture had an albuminocytological dissociation. Among the 5 patients who underwent electrophysiological study, 3 met the criteria for the variant of AIDP, 1 met borderline criteria for AIDP and 1 had a pattern compatible with AMSAN. Clinical presentation was heterogeneous, requiring individualized treatment. The patients received intravenous immunoglobulin, plasma exchange or both. The 3 non-severe patients were discharged with mild to moderate paresis and among the 4 patients that required intensive care, one was discharged to rehabilitation with tetraparesis and 3 were transferred to other services. **Conclusion:** This study reported cases in which there was temporal association between COVID-19 vaccines and development of GBS. Further studies are required to establish if there is a causal relation.

Neuropatias Periféricas

TL 1105873

CORRELATION OF ELECTRONEUROMIOGRAPHIC FINDINGS IN LEPROSY, ACCORDING TO THEIR CLINICAL FORMS

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Background: Leprosy is the most frequent infectious cause of peripheral neuropathy in the world and can lead to sequelae and physical disabilities that perpetuate the prejudice and stigma linked to the condition, especially due to the delay in diagnosis. **Objective:** To describe the electroneuromyographic findings in patients with leprosy, according to their clinical forms. **Methods:** The study is a retrospective observational analysis of medical records of patients with leprosy, from a Tertiary Referral Center, between the years 2014 and 2021, to correlate their clinical, epidemiological, laboratory and electroneuromyographic findings. **Results:** Among the 513 medical records analyzed, 71.7% patients (368/513) had neural thickening, 73.3% (376/513) had sensory symptoms, 49.3% (253/513) had motor symptoms and 50.5% (259/513) had leprosy reaction, being 34.9% (179/513) type I and 15.6% (80/513) type II. Regarding electroneuromyographic characteristics, the most affected sensory nerve was the superficial fibular nerve (15.5%), while the most affected motor nerve was the ulnar (12.6%). Regarding neural involvement, 126 patients had normal electroneuromyography (ENMG) and, among the 387 with abnormalities in ENMG, 13.2% (51/387) had mononeuropathy (MN) and 86.8% (336/387) had multiple mononeuropathy (MMN). Among the patients with the indeterminate (I) form (4.9%), 8% had abnormal ENMG and none had sensory/motor axonal impairment. In the primary neural leprosy (PNL) (20.3%), 96.2% had abnormal ENMG, being 88% with MMN and 6.9 (± 5.3) abnormal nerve/patient. In the tuberculoid (T) form (1%), 80% had abnormal ENMG, 50% with MMN and 3.0 (± 2.1) abnormal nerve/patient. In the borderline-tuberculoid (BT) form (49.3%), 68% had altered ENMG, 80.8% had MMN and mean number of altered nerves was 5.9 (± 5.2). In the borderline-dimorphic (BD) form (6.2%), 78.1% had altered ENMG, being 96% with MMN and 8.7 (± 5.5) abnormal nerve/patient. In the dimorpho-virchowian (DV) form (7.2%), 91.9% had altered ENMG, all of whom had MMN, with 10.1 (± 4.1) abnormal nerve/patient. Among lepromatous (LL) form (11.1%), 98.2% had altered ENMG, with 98.2% MMN and 7.8 (± 4.3) abnormal nerve/patient. Axonal involvement was more frequent in the PNL, DT, DV and LL. **Conclusion:** Leprosy is a spectral disease, characterized by a balance between host immunity and bacillary load. Therefore, the impairment and neurophysiological characteristics are distinct and may vary according to the clinical form.

Neuropatias Periféricas

TL 1106004

PERFIL DOS PACIENTES PORTADORES DE NEUROPATIA PERIFÉRICA DO MEMBRO SUPERIOR ACOMPANHADOS EM UM HOSPITAL DA CIDADE DO RECIFE

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Background: The peripheral neuropathy of upper limb functional bring great damage, interfering with the quality of life and socioeconomic conditions of individuals. **Objective:** To observe the prevalence of peripheral nerve lesions of the upper limb in a public service of the city of Recife, nothing in the prevalence of the type of disease or type of trauma that affect those individuals more. **Methods:** A cross-sectional study of descriptive and analytical, with composite sample of 71 patients in the neurosurgery clinic of the Hospital of the Restoration-Recife. Taking as a criterion for inclusion, patients with traumatic peripheral nerve lesions and / or compressive upper limb, and exclusion of patients with obstetric brachial palsy, leprosy and peripheral polyneuropathy. **Results:** Average age of patients: 43, 5 ± 16, 0 years, the most common disease in women was a mononeuropathy of the median nerve in the wrist (91, 2%, n = 31), in males the traumatic brachial plexus injury, with the etiological mechanism motorcycle accident (59, 5%, n = 22). **Conclusion:** Was obtained knowledge of the prevalence of peripheral neuropathies in a reference service in the city of Recife, as an incentive for greater integration between the interdisciplinary team and greater involvement of the physiotherapist in the rehabilitation of these patients.

Neuropatias Periféricas

TL 1106097

EVALUATE, IDENTIFY, INTERVENE: PRACTICAL REFLECTIONS FOR THE IMPROVEMENT OF HEALTH INDICATORS IN THE TREATMENT OF GUILLAIN-BARRÉ SYNDROME

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Background: Guillain-Barré syndrome (GBS) affects about 100,000 new patients worldwide every year. Mortality from this disease can range from 3 to 10%, depending, in terms of modifiable factors, on access to adequate health care, which can be problematic in low income regions. **Objective:** To determine the mortality rate, clinical complications profile, length of Hospital stay, time to initiation of pharmacological treatment and presence of side effects in Barbacena's health care region (251630 population). **Results:** 33 patients were included in the analysis. The mortality rate due to GBS was 6%. The rate of clinical complications during Hospital stay was 48,5% (n=16), mainly infection/sepsis (n=8, 24,2%), cardiac arrhythmias (n=5, 15,2%), intestinal constipation (n=5, 15,2%) and decubitus ulcers (n=3, 9,1%). All patients with GBS were treated with immunoglobulin (IgIV). About 78,8% (n=26) started IgIV within 2 weeks of symptom onset, and 21,2% (n=7) between 2 and 4 weeks of disease presentation. There was no treatment-related fluctuation and no need for retreatment with IgIV in any patient. Only 6,1% (n=2) of patients had IgIV side effects, both of them had headache and one of them had also diaphoresis and sinus tachycardia. No patient was treated with plasmapheresis. Most patients were Hospitalized for up to 2 weeks (45,5%). A small proportion (12,1%) was Hospitalized for more than 30 days. **Conclusion:** Although the length of Hospital stay was considered adequate in most cases (up to 2 weeks) and access to first-line treatment (IgIV) was excellent, the rate of clinical complications during Hospital stay was high, specially infections/sepsis and arrhythmias, which points to the need of protocols for better prevention of nosocomial infection, frequent evaluation of the swallowing capacity to avoid bronchoaspiration pneumonia, rotation of decubitus to avoid ulcers, routine cardiac monitoring and prompt treatment of dysautonomic complications. Non-ischemic chest pain due to IgIV would be an uncommon side effect (0,1-0,9%) of the medication, according to manufacturer. But in this study sample, this symptom was present in 6,1% of patients. The authors had a good experience in the treatment of this side effect by reduction of IgIV flow to a minimum of 0,5ml/kg/hour.

Neuropatias Periféricas

TL 1106255

CLINICAL AND GENOTYPICAL SPECTRUM OF HEREDITARY MOTOR NEUROPATHIES

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Background: The hereditary motor neuropathies (HMN), also known as distal spinal muscular atrophies, are genetically diverse group of diseases with predominantly distal lower motor neuron symptoms. More than twenty genes have been identified, with up to 60% of patients achieving a definite genetic diagnosis. Clinically, the HMN lie on a clinical spectrum alongside axonal Charcot-Marie-Tooth disease and hereditary spastic paraplegias. The HMN can be further divided into different clinical phenotypes, such as classical distal HMN, scapuloperoneal HMN (SPHMN) and spinal muscular atrophy with lower extremity predominance (SMALED). **Objective:** In this ongoing study, we describe the clinical and genotypical findings of patients with HMN in a tertiary healthcare service. **Methods:** We included 22 patients with clinical and electrophysiological diagnosis of HMN. We performed whole exome sequencing on 8 patients and a genetic panel for hereditary neuropathies on 7 patients, and 6 others await genetic testing. Clinical evaluation included phenotype and other neurological findings, such as pyramidal signs. Variant classification was in accordance with ACMG criteria. **Results:** Of the 22 patients, 15 were males and 7 were females. Classical distal HMN was the most common phenotype, with 17 patients. Of those, one patient presented with dHMN and myokymia, with an inconclusive genetic panel that included HINT1 gene. Another patient had a dHMN with upper limb predominance (Type V), split hands and lower limb pyramidal signs, with a variant of uncertain significance (VUS) in the GARS gene. Other genes associated with dHMN were VCP, SETX and HSPB1. Three patients presented with a SPHMN, with distal lower limb weakness and prominent scapular winging, two with a pathogenic variant on TRPV4 gene and another with pyramidal signs and a probable pathogenic variant on BICD2 gene. Two patients had a SMALED phenotype, with probable pathogenic variants in DYNC1H1 in one patient and BICD2 in another. Of the 15 patients with genetic analysis, 11 had a conclusive genetic diagnosis. **Conclusion:** There are few epidemiological studies on HMN, and to our knowledge, no systematic study has been conducted in Brazil. Interestingly, we found biallelic variants in VCP gene causing distal HMN, as well as a patient with SPSMA with a BICD2 variant, a gene normally associated with SMALED phenotype. Our study demonstrates the diverse clinical picture of the HMN.

Neuropatias Periféricas

TL 1106452

TEMPORAL ASSOCIATION OF GUILLAIN-BARRÉ SYNDROME WITH COVID-19 VACCINATION

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Background: Guillain-Barré Syndrome (GBS) is an acquired autoimmune disorder in which nerves myelin sheath is lost, especially the peripheral ones, leading to varying degrees of progressive and ascending motor weakness. The trigger is usually associated with respiratory and gastrointestinal infections, and, rarely, immunization. **Objectives:** To report a possible association of Guillain-Barré Syndrome to COVID vaccination. **Methods:** Description of 7 cases with post-vaccinal acute polyradiculoneuropathy. **Results:** In our series of seven cases of post-vaccinal COVID GBS, we observed no gender disparities, 3 males and 4 females, in an age range of 18-50 years old. All vaccines were ChAdOx1-S[recombinant], with 6 patients presenting symptoms after first dose and 1 patient after second dose. One patient had had H1N1 vaccine a week before the sars-cov-2 vaccine first dose. Ascending paraparesia began 15 days post vaccine, evolving to tetraparesia in 6 patients. Two patients had respiratory insufficiency, needing mechanical ventilation. All patients had cellular-proteic dissociation in CSF analysis, with proteinorraquia ranging from 92mg/d to 578mg/dl. Electroneuromyography showed a demyelinating pattern in five cases, but 3 were associated with secondary axonal degeneration. Other two patients showed an axonal pattern. **Conclusion:** The described cases had a temporal association of GBS with ChAdOx1-S[recombinant] vaccination. In literature, the interval between trigger and symptoms can vary, with some studies reporting 6 weeks onset after exposure. In our series of cases, mean time of symptoms onset was after 15 days of vaccination. The massive global program of sars cov2 vaccine provoked a surge of neurological complications, including GBS cases, a phenomenon well described in literature. However, this association is rare and the vaccination remains the most effective method of COVID-19 prevention.

Neuropatias Periféricas

TL 1106477

EVALUATION OF QUALITY OF LIFE IN PATIENTS WITH FAMILIAL AMYLOID POLYNEUROPATHY

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Background: Amyloidosis are a group of diseases caused by deposition of amyloid fibrils. A subgroup of those pathologies is associated with a gene mutation in a protein called Transthyretin, which may cause a series of affections, including a neurological manifestation, called Familial Amyloid Polyneuropathy (FAP). FAP is a rare, progressive and incurable disease, capable of causing great impact in patients' quality of life. **Objectives:** To evaluate quality of life in patients with Familial Amyloid Polyneuropathy; to characterize the sample regarding biological and sociodemographic variables. **Methods:** This is an observational, transversal, analytical and descriptive study, based on primary data from patients with confirmed diagnosis of FAP, recruited by convenience at an ambulatory in Salvador, Bahia, and collected in the year of 2021. There were collected sociodemographic and biological variables and six scores generated by the Norfolk QOL-DN questionnaire. Quantitative data were analysed and expressed in means/medians, according to their distribution, as well as standard deviations/interquartile range; and qualitative data were analysed and expressed in relative and absolute values. **Results:** In the period between July/2021 and December/2021, there were collected data from 16 patients, with a mean age of 56,19 years ($\pm 15,94$ years) and 9 (56,3%) of them were male. From those, 6 (37,5%) have not declared their race; 5 (31,3%) were brown; 3 (18,8%) were black and 2 (12,5%) were white. The autonomic neuropathy score, large-fiber neuropathy score and total affected quality of life score had means, respectively, of 4,19 ($\pm 4,23$), 30 ($\pm 28,4$) and 64,81 ($\pm 61,32$). The sample was divided in two age groups, by the mean age (56,19), older or younger, and it was applied Student's t-test, which has demonstrated that the previously cited scores has a similar mean - total affected quality of life score ($t(11,2) = 0,651$; $p = 0,523$); autonomic neuropathy score ($t(10,4) = 0,296$; $p = 0,773$); large-fiber neuropathy score ($t(14) = 0,344$; $p = 0,736$). The small-fiber neuropathy score, the symptoms score and the daily-life activities score had medians, respectively, of 3 (interquartile range: 0;8,75), 10 (interquartile range: 4,25;22), 2 (interquartile range: 0;15,25). **Conclusion:** In our sample, all patients had quality of life prejudice. Despite of being a chronic and progressive disease, it was not found any difference at quality of life decrease with age progression.

Neuropatias Periféricas

TL 1106556

MORTALITY PROFILE OF PATIENTS WITH INFLAMMATORY POLYNEUROPATHY IN BRAZIL BETWEEN JANUARY 2010 AND DECEMBER 2020

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Background: Inflammatory polyneuropathy is clinically marked by progressive symptoms of weakness affecting the distal and proximal segments with consequent loss of functional capacity. In addition, these patients develop global loss of reflexes and sensitivity, especially vibratory and positional. Due to its autoimmune characteristic that generates an inflammatory response on the neurons, the treatment is performed with immunoglobulin, immunosuppression, and glucocorticoids, generating stability and even remission. However, as the disease progresses, the patient may develop respiratory failure, a major cause of death in this etiology even with therapeutic progress. **Objective:** Describe the epidemiological profile of mortality for Inflammatory polyneuropathy. **Methods:** This descriptive study was carried out using data from the Departamento de Informática do Sistema Único de Saúde (DATASUS), in which the Inflammatory Polyneuropathy mortality profile between January 2010 and December 2020 was evaluated. The mortality during this period was analyzed by education, age group, month of death, federation region, ethnicity, and gender. In the description of these categorical variables, their relative frequencies (%) will be calculated. Since this is a public domain database, submission to the Research Ethics Committee was not necessary. **Results:** The total number of deaths in the 10-year period was 2283 individuals. There was a predominance of deaths in the school years between 8 and 11 years (22.56%), in the months of January (9.15%) and November (9.15%), in the age group between 60 and 69 years (20.59%), male (60.7%) and white (54%). **Conclusion:** There is a congruence of the mortality profile with the epidemiology of the disease. However, even with the treatment already established and the possibility of remission, the number of deaths from inflammatory polyneuropathy is still high. The higher incidence of mortality in patients with less education and older age suggest the lack of access to treatment as one of the causes for this fact. Thus, new studies are needed to solidify the literature about the motivations for maintaining high levels of deaths in an already well known disease.

Neuropatias Periféricas

TL 1104844

THE EFFECTIVENESS OF THE NEUROMODULATION TECHNIQUE USE IN BRAZILIAN HOSPITALS: A SYSTEMATIC REVIEW

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Background: The effectiveness of the neuromodulation technique leaves doubts that prevent its proper use. The process consists of the use of stimuli, such as electrical or chemical, to trigger the central or peripheral nervous system, improving its function in the human body. Having its first reports only in the 20th century, the technique is recent, and for this reason its effectiveness is discussed by many in the scientific community, making it of little use in Brazilian Hospitals. **Objective:** To clarify the effectiveness of the Neuromodulation Technique in the human body through its use and results in Brazilian Hospitals. **Methods:** This systematic review used scientific articles containing the keywords: "Neuromodulation" and "Transcutaneous Electrical Nerve Stimulation". The search was made in the databases: PubMed, MedLine and ScienceDirect, in the month of May 2022. The review had, as inclusion criteria, publications in the last 5 years, in Portuguese and English, that directly addressed the theme. Articles with foreign analyses, articles with no direct relation to the theme, and systematic reviews were excluded. Case presentation and randomized clinical trials were evaluated, with their results being analyzed to determine the effectiveness of the technique. **Results:** 20 references met the criteria of this search. Transcutaneous Electrical Nerve Stimulation (TENS) showed potential for improvement in cognitive functions and in reducing symptoms such as depression, although research is needed on customized TENS protocols. Preliminarily, TENS was shown to be safe, even during pregnancy; however, in the case of treatments such as laryngeal diadochokinesis, the technique showed inefficiency. **Conclusion:** Neuromodulation shows efficiency in pathological treatment and in the promotion of natural characteristics such as vocal variety. It is noted, however, that in cases such as the treatment of laryngeal diadochokinesis, the use of the technique has not shown efficiency, and traditional treatment is preferable. Neuromodulation is a favorable option for several situations, but not all, being necessary to evaluate what is the expected result and its comparison with the result of classical treatments. Considering the low amount of studies in the area, new researches should be done to prove the efficacy of the neurological technique according to the sought result.

Neurorreabilitação

TL 1104936

MULTIPLE SCLEROSIS IN DETAIL: EBOOK FOR PATIENTS

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Background: Multiple Sclerosis (MS) is a chronic neuroinflammatory and neurodegenerative disease that is a leading cause of neurological disability in young adults. Educational materials are important for health promotion, increasing knowledge about the disease and improving quality of life. **Objective:** Analyze and improve the readability and adequacy of educational materials for patients and relatives with Multiple Sclerosis. **Methods:** Thirteen original educational materials in flyers format written by a multidisciplinary team over 37 years were reviewed by a neurologist. Three MS experts then compared the original and revised materials using the Suitability Assessment of Materials (SAM) instrument, a grading scale used to evaluate suitability of health information materials for patients. The SAM criteria were each given 0, 1, or 2 points based on adequacy of the materials to address each criterion, with 0 indicating a “not suitable” rating, 1 indicating an “adequate” rating, and 2 indicating a “superior” rating. The total SAM score for each flyer was calculated as the sum of earned points divided by the number of possible points. The final SAM score was reported as a percentage, with 70-100 % indicating superior material, 40-69 % adequate material, and 0-39 % unsuitable material. Afterwards, six MS patients and two family members were interviewed to discuss the revised materials, the feedback was analyzed qualitatively. Finally, the revised flyers were converted into an eBook. Pearson correlation was calculated to determine inter-observer correlation between SAM scores assigned by the three evaluators. Paired t test was used for analysis of SAM scores between original and revised patient education materials. **Results:** It was observed that no specialist evaluated the original or revised flyers as unsuitable; however, the revised flyers represent a significant improvement in total SAM score compared to the original flyers. Specifically, the mean (\pm standard deviation) SAM score improved from $60\pm 77\%$ (adequate) for the original versions ($n=13$) to $88\pm 74\%$ (superior) for the revised flyers ($n=13$) ($p<0.001$). **Conclusion:** The eBook was produced based on the needs of target audience of the educational action, in understandable language and presentation. The material can be considered as a facilitator or support for conceptual and perceptual change and enrichment of skills or attitudes.

Neurorreabilitação

TL 1104937

FATIGUE, BALANCE AND GAIT OF PEOPLE WITH MULTIPLE SCLEROSIS AFTER TREATMENT WITH TRANSCRANIAL MAGNETIC STIMULATION ASSOCIATED WITH PHYSICAL THERAPY: A PILOT STUDY

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Background: Multiple Sclerosis (MS) is a demyelinating autoimmune disease that causes CNS damage and disabling symptoms. Transcranial Magnetic Stimulation (TMS) has benefits in the rehabilitation process when combined with therapeutic interventions. **Objective:** To evaluate the influence of TMS combined with physiotherapy on the balance, gait and fatigue of people with MS. **Methods:** Participated of this study 20 people with MS, 14 women and 6 men, aged 33 to 68 years (SD 50,0) and Kurtze Expanded Disability Status Scale (EDSS) between 0 and 6,5. The study was done at Brazilian Association of Multiple Sclerosis. The protocol consisted of 10 TMS sessions and 6 physiotherapy sessions with a protocol for lower limb strength training and static and dynamic balance. Participants were randomized and divided into 2 groups: Group I) with real stimulus, so that 10 received the TMS stimulus (primary motor cortex (Cz): 50 pulses per time, 30 trains, 20seconds of interval, totaling 1500 pulses at 90% of resting motor threshold and left dorsolateral prefrontal cortex (F3): 10Hz, 50 pulses per train, 40 trains, 20 seconds interval, totaling 2000 pulses at 110% of resting threshold), and Group II) sham, that received the application without TMS stimulation (10 patients). All underwent physiotherapy. The Berg Balance Scale, Timed up and Go Test (TUG) and Modified Fatigue Impact Scale (MFIS) was applied before the combined interventions and after 60 days. **Results:** It was found that 70% of participants showed improvement in balance, 20% a decrease and 10% a plateau on Berg Balance Scale. At the Timed up and Go Test, it was found 70% of improvement and 30% of decrease in performance. At the fatigue evaluation with Modified Fatigue Impact Scale, in Cognitive domain, it was found 90% decrease in fatigue values and 10% increase. In the physical domain, it was analyzed that 65% had an improvement in the indexes, 25% a worsening and 10% remained the same. In the psychosocial domain, 70% showed a decrease in fatigue levels, 20% an increase and 10% remained unchanged. **Conclusion:** This preliminary study suggests that TMS associated with physiotherapy has great potential in the rehabilitation of balance, gait and fatigue on people with MS. The promising results encourage the acquisition of additional evidence.

Neurorreabilitação

TL 1104938

TRANSCRANIAL MAGNETIC STIMULATION FOR DEPRESSIVE SYMPTOMS IN PATIENTS WITH MULTIPLE SCLEROSIS

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Background: Multiple sclerosis (MS) is a chronic disease of the central nervous system that affects the brain and spinal cord. Up to 50% of MS patients experience depressive disorders. Transcranial Magnetic Stimulation (TMS) has been used for depression. **Objectives:** To evaluate the effect of TMS on depressive symptoms in patients with MS. **Methods:** A quantitative study was performed with 20 people diagnosed with multiple sclerosis (13 relapsing-remitting, 4 primary-progressive and 3 secondary-progressive), aged between 33 and 68 years (Mean = 50.0, SD = 10.6 years), 6 men (30%) and 14 women (70%), with EDSS score from 0 to 6.5 (Mean = 4.6, SD = 1.9), time of diagnostic between 1 and 24 years (Mean = 10.0, SD = 7.2 years). The protocol consisted of 10 interventions by TMS (primary motor cortex (Cz): 50 pulses per time, 30 trains, 20seconds of interval, totaling 1500 pulses at 90% of resting motor threshold and left dorsolateral prefrontal cortex (F3): 10Hz, 50 pulses per train, 40 trains, 20 seconds interval, totaling 2000 pulses at 110% of resting threshold) during 10 consecutive working days. For evaluation, an interview was conducted to collect data and applied the BDI (Beck Depressions Inventory) at the beginning and at the end of the stimulations. **Results:** It was observed that 30% of the participants presented depression. It was found that 65% of participants had improvement in symptoms of depression. However, among those who had depression, 83.3% had improved symptoms. **Conclusion:** Depression is a substantial health problem in need of novel and effective treatment strategies. TMS is a non-invasive and safe treatment. This preliminary study suggests that TMS has potential for treating depression in people with MS. The promising results encourage the acquisition of additional evidence.

Neurorreabilitação

TL 1105033

ACCESS TO REHABILITATION AFTER STROKE IN BRAZIL (AREA STUDY): PRELIMINARY ANALYSIS

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The objective of this study is to investigate the Access to Rehabilitation After discharge from public Hospitals in Brazil (AREa study), up to six months after stroke. This is a cross-sectional multicenter study setting in eighteen public health centers. We presented a preliminary analysis of the first 100 participants (convenience sample) from seven centers collected between January, 2020 and January, 2022. Subjects within six months to 1 year post-stroke answered a survey about access to rehabilitation during the first 6 months after discharge. The primary outcome is the percentage of patients who had access to any physical therapy session after Hospital discharge within the first six months after stroke onset. The secondary outcomes are referral to physical therapy after Hospital discharge; the percentages of patients who had access to multidisciplinary team after discharge; the percentage of patients who received health instructions until six months after stroke; patients' and families' preferences about the rehabilitation process. In this preliminary analysis, the median age was 63 years-old [54,5; 70] and 50% of the patients were women. They answered the survey with a median time of 8 months [7; 10] post-stroke and 92% had ischemic strokes. The median NIHSS was 2 [1; 4,25] and the Modified Rankin Scale (mRS) 2 [1; 3]. Most participants (57%) reported that they had no access to Physical Therapy within the first 6 months post-stroke. In addition, 97% did not have access to physiatrists, 77% to speech therapists, 93% to psychologist, 83% to occupational therapist and 80% to nutritionist. Sixty-four subjects related that they were referred to a rehabilitation center, and Physical Therapy (37%) was the most cited, with a wait time of four weeks for the first visit. These preliminary findings indicate a gap in rehabilitation after stroke but may have been influenced by the pandemic. The study is ongoing. *AREa study group: Suellen Mary Marinho dos Santos Andrade, Lenise Valler, Gustavo José Luvizutto, Maria Elisa Pimentel Piemonte, Renata Viana Brígido de Moura Jucá, Octávio Marques Pontes Neto, Beatriz Neves, Luana Miranda.

Neurorreabilitação

TL 1105036

SUPERVISED, HOME-BASED, REAL-TIME VIDEOCONFERENCING TELEREHABILITATION PRESERVES PERCEPTION OF SOME CLINICAL ASPECTS IN PEOPLE WITH PARKINSON'S DISEASE: A RETROSPECTIVE STUDY

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Background: Clinical worsening in motor symptoms and quality of life in people with Parkinson's disease (PD) has been observed during the COVID-19 pandemic. Real-time videoconferencing telerehabilitation (RTT) has been recommended to mitigate the self-reported clinical worsening in people with PD in a safe way, as people are able to access remote rehabilitation services in their own homes. However, if long-term RTT can mitigate self-reported clinical worsening in PD is still unclear. **Objective:** To compare retrospectively the effects of 10 months of supervised, home-based, RTT and usual care without exercise training (UC) on self-reported posture, walking, bradykinesia, and quality of life in individuals with PD. **Methods:** People with PD who performed either RTT (n=20) or UC (n=20) were retrospectively assessed before pandemic (February to March 2020) and during pandemic (February 2021). People with PD participated in 60 min, group (6-10 per group) exercise sessions 2-3 days per week for 10 months (April 2020 to January 2021). Exercises included: dual tasking, gait training, lunges, dance, balance training and functional skill training. Items from UPDRS-III (posture, walking, and bradykinesia) and PD Questionnaire (PDQ-39) scores were assessed before and during pandemic by blinded evaluators. Effect size (ES) and Confidence Interval (CI) were calculated for within-group (before vs. during) and between-groups (during) comparisons. **Results:** There were no between-group differences in any variable before pandemic. ES and CI for within-group comparison showed that RTT improves perception of bradykinesia, preserves perception of walking and quality of life and worsens perception of posture. In addition, UC worsens perception of all evaluated aspects. ES and CI for between-group comparison showed that RTT is more effective than UC in preserving perception of walking, bradykinesia, and quality of life. **Conclusion:** Supervised, long-term RTT at home improves perception in bradykinesia and preserves perception of walking and quality of life, although RTT worsens perception of posture, which is linked to fall in people with PD.

Neurorreabilitação

TL 1105089

ELECTROMYOGRAPHIC PROFILE OF THE OROPHARYNGEAL SWALLOWING DYNAMICS IN INDIVIDUALS WITH PARKINSON'S DISEASE

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Objective: to determine whether the electromyographic profile of the suprahyoid muscle group and of the masseter muscles are modified according to the oropharyngeal swallowing dynamics in individuals with Parkinson's disease (PD). **Methods:** 30 subjects were assessed using videofluoroscopic swallow study (VFSS) and surface electromyogram (EMG) of the masseter and suprahyoid muscle groups during the swallowing of different food consistencies, and volumes. **Results:** Subjects of both sexes, older than 50 years, presented a decrease in the electric potential (EP) of the evaluated muscles in all impaired biomechanics of the oral and pharyngeal stages, except for the laryngeal penetration, where the EP was increased during the ingestion of the liquid sample. Muscle EP was reduced more frequently during the ingestion of semi-liquid and thickened liquid. There was a strong positive correlation, statistically significant, between the duration of contraction of the evaluated muscles and the duration of the oral transit time; moderate positive correlation, statistically significant, between the duration of contraction of the suprahyoid muscle group and the pharyngeal transit time; weak positive correlation with no statistical significance between the duration of contraction of the muscle pairs evaluated and the duration of the pharyngeal stage. **Conclusion:** the EP of the masseter and suprahyoid muscle groups are modified according to the presence of oral stage and pharyngeal stage swallowing disorders of certain consistencies and volumes, in patients with PD. Furthermore, oral transit times, duration of the pharyngeal stage, and pharyngeal transition were correlated with the contraction duration of the masseter muscles and suprahyoid muscle group.

Neurorreabilitação

TL 1105095

ELECTROMIOGRAPHIC PROFILE OF MASSETER AND SUPRAHYOID MUSCLES IN PEOPLE WITH PARKINSON DISEASE

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Objective: to characterize the electromyographic findings of the masseter and suprahyoid muscle group in maximal voluntary isometric contraction (MVIC) and during swallowing of different food volumes and consistencies, in individuals with Parkinson's Disease (PD) and healthy ones, who composed the Group Comparison (GC). **Method:** thirty individuals with PD and thirty healthy ones were evaluated through the Surface Electromyography (EMG), where the mean values of the electrical potential (EP) of the masseter and suprahyoid muscle group were obtained in MVIC and during the swallowing function. **Results:** it was observed that the EP of the masseter and suprahyoid muscle group was significantly lower in the Parkinson's Disease Group (PDG) in MVIC, and during the swallowing of all consistencies and volumes evaluated. The duration of muscular contraction was higher in individuals with PD, but with no statistical significance. In addition, muscular EP tends to reduce and the duration of muscular contraction tends to increase, according to the Hoehn and Yahr stages. **Conclusion:** The EP of the masseter and suprahyoid muscle group is reduced in MVIC and during the swallowing function in individuals with PD

Neurorreabilitação

TL 1105177

COMPARISON BETWEEN EFFECTS OF REPETITIVE PERIPHERAL NERVE STIMULATION ON UPPER LIMB MOTOR PERFORMANCE IN THE SUBACUTE AND CHRONIC PHASES AFTER STROKE

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Background: Repetitive peripheral nerve stimulation (RPNS) is a promising neuromodulation intervention to improve upper limb motor performance in stroke. Stimulation intensities higher than the sensory threshold (suprasensory) were associated with benefits compared to stimulation intensities below the sensory threshold (subsensory) or no stimulation, in patients in the chronic phase after stroke. There is limited information about the effects of different intensities of RPNS, early after stroke. **Objective:** To compare the effects of suprasensory and subsensory RPNS of the median nerve, on performance of the paretic upper limb in the subacute (< 6 months) and chronic (> 6 months) phases after stroke. **Methods:** This is an ongoing, multicenter, randomized, parallel-group, proof-of-principle clinical trial with blinded assessment of outcomes. Patients are randomized to 2-hour suprasensory or subsensory RPNS in the subacute (Groupsuprasubac and Groupsubsubac) or chronic phases (Groupsuprachronic and Groupsubchronic) after stroke. In all groups, motor training is performed after RPNS. The primary outcome is the improvement in performance in the Jebsen-Taylor Test (JTT), assessed before and after RPNS, as well as after training. Descriptive analyses of the preliminary results are shown. Statistical tests to compare differences within and across groups will only be performed at the end of the study. **PRELIMINARY Results:** Until March 2022, 39 of 72 volunteers were included. At baseline, motor performance was similar in Groupsuprasubac (n=7) and Groupsubsubac (n=8), but worse in Groupsubchronic (n=7) than in Groupsuprachronic (n=13). Improvement in JTT scores occurred in 28.5% of the patients in Groupsuprasubac, 75% of the Groupsubsubac, 61.7% of the Groupsuprachronic, and 57.1% of the Groupsubchronic. After training, performance improved in all patients. Changes in performance after stimulation were (in seconds, average \pm standard deviation; positive values indicate better performance): -18.4 \pm 35.3, Groupsuprasubac; 8.5 \pm 8.5, Groupsubsubac; 4.8 \pm 11.3, Groupsuprachronic; 5.5 \pm 16, Groupsubchronic. Improvements in performance after training were: 3.5 \pm 24.5, Groupsuprasubac; 19.7 \pm 12.7, Groupsubsubac; 17.9 \pm 11.6, Groupsuprachronic; 23.7 \pm 16.0, Groupsubchronic. **Conclusion:** Although preliminary, these results suggest that suprasensory RPNS may be beneficial to patients in the chronic, but not in the subacute phase after stroke. The study is currently recruiting patients.

Neurorreabilitação

TL 1105245

PERCEPTION OF PHYSICAL THERAPISTS ON THE USE OF SELF-MANAGEMENT PROGRAMS FOR PHYSICAL ACTIVITY IN POST-STROKE PATIENTS

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Stroke represents the second worldwide leading cause of death and the third leading cause of physical disability. As a result of the long-term damage generated, the rehabilitation guidelines for stroke recommend that these patients be taught to self-manage their sequelae. Self-management is a comprehensive concept that can be used in the management of behavioral aspects such as physical activity. This study aimed to characterize the knowledge of Brazilian physical therapists about the application of self-management programs to increase physical activity in the routine of post-stroke patients. The sample consisted of physical therapists who had already treated stroke patients, who knew the concept of self-management and its applicability to increase physical activity in these patients. The answers were collected through an online questionnaire with closed alternatives. Five questions were answered using the Likert scale (1 to 5). The score of this scale was used to measure the importance attributed by respondents to questions related to the use of self-management. The analysis was carried out regarding the mean and standard deviation, to determine, respectively, the degree of importance and agreement between the professionals' opinions. Four questions were analyzed regarding the correlation between the choice of methods and the frequency of use of self-management. Of 56 respondents, 29 participants were within the criteria to compose the sample. Not knowing the concept of self-management was the reason for the elimination of 19 respondents. These professionals claimed to use self-management half of the time (44.83%) or almost always (51.72%). The main limiting criteria for self-management were cognitive impairment (96.55%) and physical activity tolerance (100%). About the resources to assess the patient during the physical activity program, the more technological means, such as the use of accelerometers, were less acceptable among professionals (17.24%) and fatigue was the most cited (100%). Thirteen minutes, three times a week and twelve weeks were the protocol indicated for the most participants on how to organize a self-management program. Brazilian physical therapists generally agree with the literature, except for some points of divergence about inclusion criteria in the program and the use of technologies for monitoring patients. These results can guide a future self-management protocol for physical activity in the post-stroke Brazilian population.

Neurorreabilitação

TL 1105267

PHARYNGEAL SWALLOWING RESPONSE IN INDIVIDUALS AFTER ISCHEMIC STROKE: A PILOT STUDY

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Background: The onset site of the Pharyngeal Swallowing Response (PSR) provides information about the sensory motor model of swallowing. Absence or longer duration of this process is common in the post-stroke population. **Objective:** To identify the brain injury and the location of the PSR in post-stroke individuals. **Methods:** Cross-sectional, prospective, quantitative study. Twelve patients with mean age 68.58 (± 10.07), 5 women and 7 men, were evaluated. Included subjects ? 60 years old; confirmed ischemic stroke, responsive to perform videofluoroscopic (VDF). Pathologies that could cause dysphagia were excluded. During VDF, swallowing of 10 ml of extremely thickened food consistency was observed. The VDF and stroke images were analyzed by two speech pathologists and two neurologists. Lesion location was subdivided into: tentorial/supratentorial; cortical/subcortical and/or deep; right and/or left sides. A 4-point scale was used to identify the location of the PSR: 0 – posterior mandibular angle; 1 – epiglottic vallecula 2 – hypopharynx 3 – piriform recesses. **Results:** Five patients with PSR in posterior mandibular angle, 3 (25.0%) supratentorial lesion; 1 (8.3%) supratentorial/infratentorial and 1 (8.3%) infratentorial. Five patients with PSR in epiglottic vallecula, 4 (33.3%) supratentorial lesion, 1 (8.3%) supratentorial/infratentorial. Two patients with PSR in piriform recesses, 1 (8.3%) supratentorial/infratentorial and 1 (8.3%) infratentorial. PSR and cortical/subcortical lesion: five patients had PSR in the posterior mandibular angle, 1 (8.3%) deep lesion; 2 (16.7%) cortical/deep; 1 (8.3%) cortical/subcortical and 1 (8.3%) cortical. Five patients had PSR in epiglottic vallecula, 1 (8.3%) deep lesion; 1 (8.3%) cortical/deep; 2 (16.7%) cortical/subcortical and 1 (8.3%) cortical. Two patients with PSR in piriform recesses, 1 (8.3%) deep lesion and 1 (8.3%) cortical. PSR and laterality of the lesion: five patients with LRI in the posterior mandibular angle, 3 (25.0%) lesion on the left; 1 (8.3%) right; 1 (8.3%) bilateral. Five patients with PSR in epiglottic vallecula, 1 (8.3%) lesion left and 4 (33.3%) right. The PSR in piriform recesses, 2 (16.7%) both with left lesion. **Conclusion:** This pilot study allowed us to identify the site of the PSR and the location of the encephalic lesion. A larger sample would be needed to understand the relationship between lesion location and its impact on swallowing physiology.

Neurorreabilitação

TL 1105279

TREATMENT OF UPPER LIMB PARESIS WITH REPETITIVE PERIPHERAL NERVE SENSORY STIMULATION AND MOTOR TRAINING: ONE MONTH POST-TREATMENT FOLLOW-UP

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Background: In a randomized, placebo-controlled, proof-of-principle clinical trial, active or sham repetitive peripheral sensory stimulation (RPSS) were delivered as add-on interventions to task-specific training (TST) and functional electrical stimulation (FES) to 38 subjects with moderate to severe motor upper limb impairment in the chronic phase after stroke, three times per week, for six weeks. Increases in grasp and pinch force were significantly greater in the active than in the sham group. The per-protocol subgroup (25/38; 66%) only included patients who complied with at least 80% of the interventions. Outcomes were assessed in 19/25 (24%) of the per-protocol subgroup, one month post-treatment. **Objective:** To investigate whether gains in pinch and grasp strength remained in subjects of the per-protocol subgroup, followed up to one month after the end of treatment. **Methods:** Ten subjects were in the active and nine in the sham group. Within-group changes in performance (post-treatment versus 1 month later) were compared with the Wilcoxon test. Between-group differences in changes in performance were compared with the Mann-Whitney test. **Results:** There were no statistically significant differences in characteristics or baseline performance between the two groups (Table 1). There were no significant within-group differences between performance after treatment and one month later (Table 2). There were no significant between-group differences in changes in performance after treatment, compared to one month later (grasp, $p=0.447$; pinch, $p=0.094$). **Conclusion:** Gains in performance observed after treatment were retained at 1 month in the active and sham groups. These results should be interpreted with caution due to the limited power of this subgroup analysis.

Neurorreabilitação

TL 1105391

EFFECTS OF THE PERSONAL STROKE PROGRAM ON FUNCTIONAL INDEPENDENCE AND RISK FACTOR SELF-MANAGEMENT

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The Personal Stroke program is based on the social cognition theory and aims to induce long term behavioral changes, proposing an intervention centered on the individual and on the community. The objective of this study was to analyze the effects of the Personal Stroke program on improving the level of functional independence and risk factor management in post-stroke individuals. This is a single-arm, non-randomized, uncontrolled clinical trial with a non-probabilistic sample. Individuals with a clinical diagnosis of stroke, up to 18 years-old and who had access to the internet were admitted to the study. Those who had severe cognitive impairment assessed by the Mini-Mental State Examination (MMSE), clinically important musculoskeletal or neurological conditions and who were bedridden were excluded. Individuals were evaluated at baseline (A1) and 6 months after (A2), using the following instruments: Functional Independence Measure (FIM), Modified Rankin Scale (mRS) and Stroke Riskometer. Data were analyzed using the mean, standard deviation (sd) and frequency. Seven individuals were included in the study, all female, literate, with a mean age of 38.8 years-old (9.1 sd), a single vascular event, with the most prevalent ischemic etiology (71.4%) and with a mean injury time of 30.2 months. The Riskometer results show a 1.6% reduction in the average 10 years risk of a new stroke after the intervention, (A1: 16 (0.64) and A2: 14.4 (4)), while the 5 years risk increased by 0.41% (A1: 10.59 (0.3) and A2: 11 (0.83)). The FIM scores had an increase of 12.2% in the average of the general score (A1: 104.5 (9.7) and A2: 116.7 (7)) and in the mRS was observed a reduction of 0.5 (A1: 2.6 (0.8) and A2: 2.1 (0.9)). The Personal Stroke program was not able to produce an important reduction in the self-management of risk factors but could promote better outcomes in the functional independence measures.

Neurorreabilitação

TL 1105465

WHICH ELECTRICAL MUSCLE STIMULATION INCREASES CYCLICAL PERFORMANCE IN HEMIPLEGIC PATIENTS? AN EXPERIMENTAL STUDY

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Background: FES-assisted cycling is an intervention used in the rehabilitation of post-stroke hemiplegic individuals. However, it is necessary to establish electrostimulation parameters for the optimal effect of the intervention. **Objective:** To investigate the different conditions of FES-assisted cycling in the motor and cardiorespiratory performance of hemiplegic individuals and to understand the perception, credibility, and expectation of participants in each condition. **Methods:** These are preliminary data of an ongoing experimental study. The volunteers with stroke were recruited from the local community and the measures of cycling were collected under four different experimental conditions applied in a randomized sequence on the same day. Three different conditions (C1, C2, and C3) used electrostimulation in the quadriceps and hamstrings muscles. The C1 added electrostimulation to the tibialis anterior while the C2 to the gastrocnemius. In these first two conditions, the pedal used enables ankle movement, while C3 blocks ankle movement. A fourth control setup (C4) was performed without any electrostimulation. For each condition, motor performance was measured by the mean rotation per minute (RPM), cardiorespiratory performance was assessed by mean heart rate (HR) and at the end, the participant completed the Credibility and Expectation Questionnaire (QCE). The individuals' perception was analyzed through the mode of the answers to each item of the QCE. Descriptive and inferential statistics were performed applying MANOVA. **Result:** Five individuals (59.4 ± 8.7 years old; 60% male; 50 weeks after an ischemic stroke) were evaluated. The prevalence of hemiplegia was in the left limb [60%] and the level of disability measured by the Modified Rankin Scale was rated as one individual with severe disability (Rankin 4), 3 with moderate disability (Rankin 3), and one with mild disability (Rankin 2). Related to the conditions, there was no difference between the four setups regarding RPM and HR measurements [Pillai's Trace=0.993; $F(6.32)=0.459$; $p=0.833$]. Nevertheless C1, C2 and C3 were considered more logical and promising by the participants and in addition, the C3 was considered the most comfortable. **Conclusion:** It was observed that, by the individual perception, the setup C3 has greater potential to improve motor and cardiorespiratory performance. However, these results should be interpreted with caution until the study is completed.

Neurorreabilitação

TL 1105692

BIOLOGICAL PERCEPTION OF HUMAN MOVEMENT IN INDIVIDUALS WITH PARKINSON'S DISEASE: A CROSS-SECTIONAL STUDY

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Parkinson's disease (PD) is a multifactorial neurological disorder, with progressive degeneration and impairment of voluntary motor control. Action-observation could be an effective way to learn specific motor skills, but in PD the execution of movement and biological perception of human movements could be affected by the difficulty to planning motor actions. The aim of this cross-sectional study was to evaluate the biological perception of human movement in individuals with PD (PDG, $n=5$), comparing it with a control group of healthy individuals (CG, $n=5$). Edinburgh Handedness Inventory (EHI) and Mini Mental State Examination (MMSE) were used to characterized both groups. PDG and CG were evaluated in a single session of 30 minutes by a Human Movement Perception Task, in which the primary outcome measure was the number of correct answers (NCA) in each movement perception task and the verbal reaction time (VRT). As a secondary outcome measure, the Hoehn and Yahr Scale (H&Y) and section 3 of the Unified Parkinson's Disease Assessment Scale (UPDRS) were used only in PDG. Statistical analysis was performed using the BioEstat 5.3, using Mann-Whitney test and Spearman correlation. No statistical difference ($p>0.05$) was found when comparing the NCA of each task among the groups. A negative correlation ($rs: -0.91$; $p=0.0304$) was found between the H&Y and NCA for kick, long jump, sit to stand and riding a bike tasks. Further studies should be carried out based on the H&Y classification and its impact on the perception of human movement in this population. This study showed that there were no impairments in the perception of human movement by individuals with Parkinson's Disease. This study is ongoing.

Neurorreabilitação

TL 1105763

CONVENTIONAL SPEECH THERAPY COMBINED WITH FUNCTIONAL ELECTRICAL STIMULATION IN PATIENTS WITH DYSPHAGIA AFTER ACUTE CVA

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Background: CVA is the main cause of neurogenic oropharyngeal dysphagia. Electrostimulation has been used as a therapeutic tool in these cases. However, few studies prove its efficacy. We evaluated the effect of functional electrostimulation complementing standard speech therapy in patients with dysphagia after stroke in a stroke unit. **Objectives:** To evaluate the effect of short-term functional electrostimulation in a CVA unit to complement standard speech therapy in dysphagic patients after acute CVA and to indicate the benefits of different dysphagia techniques rehabilitation as standard therapy and electrotherapy. **Methods:** It is a randomized clinical trial, divided into an intervention group (IG) ($n = 16$) and a control group (CG) ($n = 17$). Data collection occurred from July 2017 to July 2020. The study included patients with ischemic CVA who had oropharyngeal dysphagia and required enteral feeding. Individuals with degenerative neurological diseases, neoplasia, pacemaker, cochlear implant, febrile state, and pregnancy were excluded. Patients underwent video endoscopy of deglutition (VED), an initial evaluation of the epidemiological and socioeconomic profile, a dysphagia risk assessment protocol (DRAP), and a functional oral intake scale (FOIS). Informed consent was obtained, and all methods were performed under the relevant guidelines and regulations (Declaration of Helsinki). The study was approved under number 2,388,931 and previously registered at ClinicalTrials.gov (NCT03649295) and the Brazilian Registry of Clinical Trials (RBR-56QK5J). **Results:** There was a significant difference in FOIS scores at follow-up for both groups. Both groups also tended to have improved levels of dysphagia as measured by FEES, although they were not statistically significant. Improvement in feeding occurred in both groups. No significant differences were detected between the groups before and after intervention by DRAP scores. Electrical stimulation showed no additional benefits beyond standard therapy when comparing results between groups. **Conclusion:** Based on the present study results there was a significant difference in FOIS level improvements of the patients in both the intervention group and the placebo group, with no significant difference between the groups. Electrical stimulation protocol use for five days in the acute phase of ischemic CVA did not generate additional benefits beyond conventional therapy and the variables analyzed improved in both groups.

Neurorreabilitação

TL 1106008

DOES NON-INVASIVE BRAIN STIMULATION IMPROVE FUNCTIONAL LOCOMOTION IN PEOPLE WITH MULTIPLE SCLEROSIS? A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background: Multiple sclerosis (MS) is a neurodegenerative disease that is the leading cause of progressive functional impairments in working-age people. MS symptoms can vary from sensorimotor to cognitive dysfunction, affecting activities of daily living and affecting the ambulation. Treating the walking disabilities associated with MS can delay its dysfunction throughout the disease course. **Objective:** This review aimed to analyze the effects of non-invasive brain stimulation (NIBS) on functional locomotion in people with multiple sclerosis (PwMS). **Methods:** The search strategy was based on the PICO process (transcranial direct current stimulation; any control group: gait, walking, or ambulation). Online databases such as PubMed, CINAHL, Web of Science, Embase, PsycINFO, and Lilacs were searched up to January 2022. Two review authors independently assessed the risk of bias in the included studies, and we used the Grading of Recommendations Assessment, Development, and Evaluation methods to rate the certainty of the evidence for each included study. A meta-analysis was performed by pooling the appropriate data using RevMan version 5.3. **Results:** One study included a qualitative synthesis and three a meta-analysis. In the meta-analysis, an improvement in the stride length (mean of differences: 0.19; 95% CI: 0.07 to 0.31; I^2 77%; $p = 0.002$) was found in the experimental group. **Conclusion:** Transcranial direct current stimulation as a NIBS associated with aerobic and resistance exercise can improve stride length in PwMS.

Neurorreabilitação

TL 1106023

EFFECTS OF NON-INVASIVE BRAIN STIMULATION (NIBS) ON VESTIBULOPATHY DISORDERS: A SYSTEMATIC REVIEW

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Background: Vestibular dysfunction (VD) is a dysfunction which affects the vestibular system causing dizziness and tinnitus. The physical therapy treatment for VD is based on exercises that can suppress the symptoms, such as adaptation, habituation, and substitution exercises. **Objective:** New types of treatments have emerged, such as non-invasive brain stimulation (NIBS), to treat chronic VD. Considering that NIBS is a promising approach to reduce VD symptoms, this review was aimed to analyze the effects of NIBS in patients with VD. **Methods:** We adhered to the methods described in the Cochrane Handbook for Intervention Reviews. The eligibility criteria were as follows: (a) individuals with vestibulopathy having clinical and neuroimaging; (b) Interventions: non-invasive brain stimulation, this intervention comprised transcranial direct current stimulation (tDCS) and repetitive transcranial magnetic stimulation (rTMS); (c) Control: any comparison or sham; and (d) Outcomes: dizziness and balance. We included randomized controlled trials and non-randomized studies from July 2004 to February 2020. We searched the PubMed, CINAHL, Web of Science, Scopus, Cochrane, and Ovid databases. Two pairs of reviewers independently screened all titles and abstracts. Two authors assessed the risk of bias for each study using the criteria outlined in the Cochrane Handbook for Systematic Reviews of Interventions and used the Grading of Recommendations Assessment, Development and Evaluation (GRADE) Methods to rate the certainty of the evidence for each outcome. **Results:** We identified 136 studies and included two studies. Both studies used tDCS application. One study used anodal cerebellar tDCS or sham at 2 mA for 25 min associated with vestibular rehabilitation therapy (VRT), and the other study used anodal tDCS over the left dorsolateral prefrontal cortex (F3) for 25–30 min associated with VRT at home. Both studies showed clinical improvement in the Dizziness Handicap Inventory (DHI), State-Trait Anxiety Inventory (STAI), Activities-Specific Balance Confidence (ABC), and Self-Rating Depression Scale (SDS) scores. Both studies presented higher-quality evidence on the GRADE scale and a low risk of bias. **Conclusion:** Based on two studies, anodal tDCS over F3 or the cerebellum associated with VRT improved chronic vestibular symptoms.

Neurorreabilitação

TL 1106057

VESTIBULAR REHABILITATION ON POSTURAL BALANCE AND INSTABILITY IN PATIENTS WITH PARKINSON'S DISEASE: A SYSTEMATIC REVIEW

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Background: Instability and postural imbalance impair the quality of life of Parkinson's disease patients (PD). Previous studies already described vestibular system connections to these functions, suggesting involvement of vestibular nuclei, the vestibulo-espinal lateral system, and the participation of basal ganglia in the reception of vestibular stimuli. We hypothesize that vestibular rehabilitation may contribute to the treatment of PD through central neuroplasticity mechanisms such as adaptation, habituation, and substitution. **Objective:** Investigate the efficacy of vestibular rehabilitation in postural instability and balance in patients with PD, in comparison to other routine interventions or care, using randomised clinical trials. **Methods:** The electronic data bases used were PubMed/Medline, Embase, Scopus, and the Physical Therapy Evidence Database (PEDro). The system review was registered in PROSPERO with the ID CRD42020210185, in accordance with PRISMA recommendations. The first stage of data extraction was carried out independently by two revisors, with any discrepancies being resolved by a third. The studies included had no restrictions on publication dates or languages, and the most recent update occurred in October of 2021. **Results:** 109 of the 440 searches were duplicated, and 303 were removed after reading the title and summary, leaving 28 candidates for full text reading. Twenty of these were excluded due to discordance, and four were annotated by a third reviewer due to discrepancies, leaving four studies for systemic review. One study was conducted in Turkey, one in Ireland, and two in Italy; the Hoehn and Yahr scale ranged from 1 to 4, and the average PD duration ranged from 3.3 to 10.3 years. Interventions occurred in all studies in the clinical state "on." The frequency of interventions ranged from one to three times per week, for a total of six years and eight weeks, with each session lasting 40 to 60 minutes. The Berg Balance Scale was described in all the studies, the Activities-Specific Balance Confidence scale and the Dynamic Gait Index in two of them, and the others had only one description. The majority of the outcomes in the four systemic review studies showed statistically significant improvements in postural stability and balance after vestibular rehabilitation. **Conclusion:** Vestibular rehabilitation is another type of therapy that can help patients with PD improve their balance and stability.

Neurorreabilitação

TL 1106094

ADHERENCE TO MOTOR REHABILITATION CAN INFLUENCE THE MOTOR TRAJECTORIES OF SMA PATIENTS TREATED WITH NUSINERSEN

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Background: Patients with 5q-Spinal Muscular Atrophy (SMA) should always undergo treatment with physical therapists in the motor area, but in many cases this happens only after the **Background:** of drug treatment, but not every increase in the adherence of rehabilitation can be beneficial. **Objective:** To evaluate the effects of changing the frequency and increasing adherence of motor physical therapy before or after the background of nusinersen. **Methods:** Twenty-eight patients with SMA type 1, 2 and 3 were evaluated using the CHOP-INTEND (Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders) and HFMSE (Expanded Hammersmith Functional Motor Scale for SMA) motor function scales. A questionnaire was carried out to identify the frequency and adherence of motor rehabilitation before and after starting treatment with nusinersen, with a 12-month follow-up. **Discussion:** In this study it is possible to see that those patients who started physical therapy only after drug treatment with an exaggerated increase in rehabilitation time did not present motor gains, as well as those patients who reduced the time of physical therapy. The greatest gains in the scores on motor scales were presented by patients who had previously performed motor physical therapy for about 3 hours a week, and for those who performed at least 1 hour of motor physical therapy per week and increased by up to 2 hours on average. Disease modifying treatment will not achieve its best result if there is no adequate rehabilitation associated with medication. **Conclusion:** It is important to evaluate the frequency and adherence of baseline rehabilitation before starting drug treatment. Therefore, it is possible to plan a change in physical therapies frequency after treatment, thus avoiding fatigue and risk of motor loss.

Neurorreabilitação

TL 1106367

SYSTEMATIC REVIEW BETWEEN NEUROMODULATION, EXECUTIVE FUNCTIONS AND OBESITY: IS THERE A RELATIONSHIP?

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Background: The brain is more sensitive to oxidative stress than other organs. Oxidative stress and inflammatory processes can negatively affect cognitive functions. The cumulative effect can potentially impact deterioration, especially of executive functions. **Objective:** to verify the results: of neuromodulation on the performance of executive functions when applied to overweight or individuals with obesity. **Methods:** This is a systematic review article based on articles selected from PubMed, ScienceDirect, BIREME, and Web of Science databases. The search was performed using the following combination of descriptors: ("problem solving" OR "executive function" OR memory) AND (tDCS OR TMS) AND obesity. Only controlled and/or randomized clinical trials published in any language between 2011 and May 31st, 2022, were analyzed using the PICOS strategy. The study was registered in the International Prospective Register of Systematic Reviews (PROSPERO) under registration number CRD42.021.261.770. **Results:** Using the descriptors mentioned, 763 articles were identified in the databases defined for the search. After selecting and analyzing the works found, 08 articles were selected. The number of people included in the eight selected studies was 301 participants. The majority were female (235 participants), with an average of 30.1 participants per study. The global nutritional status varied between low weight and grade 3 obesity, by the selected studies of 28.1 kg/m². Regarding the type of stimulation modality, the proportion between non-invasive neuromodulation strategies as a form of treatment was 75% (n=6) with tDCS and 25% (n=2) with EMT. Regarding the relationship between age and performance of executive functions, a study with older participants demonstrated the go/no-go task and the reduction of food desire. Differently, in a survey with young patients, an increase in the consumption of calories and a worse performance in the flanker paradigm task were demonstrated. It should be noted that both used the same target (anodic at F3) but in different modalities: electrical and magnetic, respectively. Furthermore, it improved the performance of the executive functions of the participants, especially in the groups of subjects with overweight or obese. **Conclusion:** The use of non-invasive neuromodulation has consistently reduced food cravings, especially for sweet foods, and fast food, and at the same time improved executive functions.

Neurorreabilitação

TL 1106369

APPRAXI@: THE ARTIFICIAL INTELLIGENCE-BASED SPEECH TRAINING APP

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Background: Verbal apraxia is the inability to program verbal movements voluntarily. However, rehabilitation should aim at active learning. Many applications with auxiliary subjects in the health area are being increasingly explored. However, very few speeches and language rehabilitation exist, and they are in Other languages (English and Russian), whose phonology is entirely different. It should be noted that public access to speech rehabilitation and the number of professionals are restricted, and the therapeutic process is lengthy. **Objective:** Develop an application to assist in the speech rehabilitation of people with verbal apraxia. **Methods:** A team of speech therapists with experience in the rehabilitation of acquired neurological disorders created the stimulus network with varying degrees of complexity. This network of stimuli was computationally absorbed through Artificial Intelligence techniques for IOS® and Android® technologies. The program was developed for minimally literate individuals with minimally functioning hearing and vision, as it has written, acoustic and visual stimuli that must be repeated and their degree of accuracy of the articulatory precision captured. The program has four interaction modules: sustained vowels, phonemes, sequences, and phrases. Each one will have three levels of complexity. In each module, there will also be subgroups of skills worked on. Among the various modules, the 'report' stands out, which informs the professional quantitatively and qualitatively about the speaker's errors. The research was approved by the CEP/UFPI/CONEP under the opinion number 5,111,083 and is currently in the implementation stage. The registration filing with the National Institute of Industrial Property is still on request. **Results:** During the 13 months after the research was installed, the conceptual design was awarded two research funding notices (CATALISA-ICT/Sebrae® and IFES). For validation of the prototype, 127 adults native to the Portuguese language participated, distributed in the states of São Paulo, Rio de Janeiro, Maranhão, Ceará, and Piauí. In this second stage, people with apraxia are invited to participate. **Conclusion:** It is expected that daily speech training will reduce the number of sessions for monitoring the Speech-Language Pathologist and increase the turnover of patients who will be seen, allowing better access to health at the tertiary level (rehabilitation).

Neurorreabilitação

TL 1106435

EFFECT OF ROBOT-ASSISTED TRAINING ON UNILATERAL SPATIAL NEGLECT AFTER STROKE: SYSTEMATIC REVIEW AND META-ANALYSIS OF RANDOMIZED CONTROLLED TRIALS

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Background: Several studies have shown that robotic devices can effectively improve motor function in stroke patients through limb activation. However, the effects of robot-assisted therapy on perceptual deficits after stroke is unclear. **Objective:** This review aimed to evaluate the effectiveness of robotic limb activation in patients with unilateral spatial neglect (USN) after stroke. **Methods:** A literature search was performed using MEDLINE, EMBASE, CENTRAL, CINAHL, and LILACS databases without language restrictions. Randomized controlled trials (RCTs) and quasi-RCTs of robot-assisted therapy for USN after stroke were selected. Two reviewers independently assessed the risk of bias and certainty of the evidence of the included studies. **Results:** A total of 630 studies were identified, including five studies for qualitative synthesis and four meta-analyses. The results of RCTs comparing robotic limb activation with a control group suggested an improvement in the degree of USN measured by the line bisection test (standardized mean difference [SMD], -0.64; 95% confidence interval [CI], -1.13 to -0.15; $p = 0.01$). There were no differences between the groups in the motor-free visual perception test 3rd edition (SMD, 0.27; 95% CI, -0.25-0.79; $p = 0.31$), star cancellation test (SMD, 0.26; 95% CI, -0.42-0.94; $p = 0.54$), Albert's test (SMD, -0.67; 95% CI, -2.01-0.66; $p = 0.32$), and Catherine Bergego Scale (SMD, -0.81; 95% CI, -2.07-0.45; $p = 0.21$). **Conclusion:** The study demonstrated that limb activation through robotic therapy can improve midline perception. However, there was no impact on tasks assessing visual scanning, functionality, or activities of daily living.

Neuroreabilitação

TL 1106440

PHYSICAL ACTIVITY AT HOME IS ABLE TO REDUCE THE WORSENING IN QUALITY OF LIFE IN PARKINSON'S DISEASE DURING COVID-19 PANDEMIC

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Background: The physical distancing, due to COVID 19 pandemic, causes an abrupt interruption in physical activity. The health services in all the world had to face the challenge to offer remote health to promote physical activity at home to avoid the general worsening of disease impairments. However, the efficacy of remote care is little known yet. **Objective:** To compare the effects of physical activity (PA) at home with and without remote supervision on the motor and non-motor aspects of daily life experiences of people living with Parkinson's disease during the physical distancing due to COVID-19 pandemic. **Methods:** 478 individuals with PD mean age of 67.6 years (SD=9.5), in stage 1 – 3 of disease evolution according to Hoehn and Yahr classification, living in 14 different cities representative of 5 different socioeconomic regions, participated in this study. Participants were asked to answer, through telephone interviews, a questionnaire about: (1) general information; (2) socio-economic status; (3) information associated with PD; (4) quality of life; (5) health conditions associated with COVID-19; (6) emotional health condition; (7) level of physical activity; (8) remote health care; (9) cognitive health conditions and (10) motor and non-motor aspects of daily life experience. Participants were divided into three groups, according to their self-reported physical activity at home: no PA group (NPA) (150 participants), PA with no-remote supervision group (PANS) (225 participants) and PA with remote supervision group (PAS) (103 participants). Comparison in motor and non-motor self-reported worsening between two groups was analyzed by ANOVA one-way. **Results:** ANOVA showed a significant statistical difference between the groups for quality of life ($F_{2, 475}=3.57$, $p=.02$, $ES=0.78$), but not for motor and non-motor aspects of daily life experience. The Tukey post-hoc confirmed that NPA reported more severe worsening in quality of life than PANS and PAS ($p<.05$). **Conclusion:** The PA at home, regardless of remote supervision, reduced the self-reported worsening quality of life in people living with PD during the physical distancing. However, it is not able to alleviate the negative impact of interruption in health care on motor and non-motor alterations associated with PD.

Neuroreabilitação

TL 1106490

PHYSICAL ACTIVITY INTERRUPTION DURING COVID-19 PANDEMIC AGGRAVATES THE WORSENING IN QUALITY OF LIFE OF PEOPLE WITH PARKINSON'S DISEASE

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Background: Reduced physical activity can collaborate to accentuate the motor and non-motor changes experienced in daily life by people with PD. **Objective:** To investigate the impact of interruption in physical activities due to physical distancing during the COVID-19 pandemic on the motor and non-motor aspects of daily life experiences of people living with Parkinson's disease in Brazil. **Methods:** 400 individuals with confirmed PD diagnosis mean age of 67.2 years (SD=9.5), in stage 1 – 3 of disease evolution according to Hoehn and Yahr classification, living in 14 different cities representative of 5 different geo-economic regions, participated in the present study. After presenting the informed consent form, and expressing agreement to participate in the study, participants were asked to answer, through telephone interviews, a previously established questionnaire that investigates: (1) general information; (2) socio-economic status; (3) information associated with PD; (4) quality of life; (5) health conditions related to COVID-19; (6) emotional health condition; (7) fear of falling; (8) level of physical activity; (9) cognitive health conditions and (10) motor and non-motor aspects of daily life experience. The participants were divided into two groups, according to their self-reported physical activity profile: no-interruption in PA group (NIPA) with 236 participants, and interruption in PA group (IPA), with 164 participants. The comparison in motor and non-motor self-reported worsening between two groups was analyzed by ANOVA one-way. **Results:** The groups were comparable in clinical aspects. The ANOVA showed a significant statistical difference between the groups for the quality of life and motor aspects of daily life experience ($p<.05$), but not for motor aspects of daily life experience. The Tukey post-hoc confirmed that NIPA reported less severe worsening in the quality of life and motor aspects of daily life experience than IPA. **Conclusion:** The interruption in PA due to physical distancing during the COVID19 pandemic promoted more severe worsening and motor aspects and quality of life in people living with PD in Brazil. Future studies should investigate feasible and effective strategies to guarantee PA maintenance at home, probably by telehealth.

Neuroreabilitação

TL 1106503

PHYSICAL ACTIVITY REDUCES THE WORSENING IN MENTAL-HEALTH AND QUALITY-OF-LIFE IN PEOPLE LIVING WITH PARKINSON'S DISEASE DURING THE COVID-19 PANDEMIC

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Background: Anxiety and depression are common non-motor alterations in PD. Increased levels of stress during the COVID-19 pandemic may have several short-term as well as long-term adverse consequences for this population. Reduced physical activity during the pandemic may contribute to increased psychological stress, thereby further aggravates the worsening in quality of life. **Objective:** To investigate the impact of interruption in physical activities due to physical distancing during the COVID-19 pandemic on the mental health condition and quality of life in people living with Parkinson's disease. **Methods:** 463 individuals with confirmed PD diagnosis mean age of 67.2 years (SD=9.5), in stage 1 – 3 of disease evolution according to Hoehn and Yahr classification, living in 14 different cities representative of 5 different geo-economic regions, participated in the present study. After presenting the informed consent form, and expressing agreement to participate in the study, participants were asked to answer, through telephone interviews, a previously established questionnaire that investigates: (1) general information; (2) socio-economic status; (3) information associated with PD; (4) quality of life worsening; (5) health conditions related to COVID-19; (6) mental health worsening; (7) fear of falling; (8) level of physical activity; (9) cognitive health conditions and (10) motor and non-motor aspects of daily life experience. The participants were divided into two groups, according to their self-reported physical activity profile: PA group (PAG) with 327 participants, and no PA group (NPAG), with 136 participants. The comparison of the mental health worsening and quality of life worsening between two groups was analyzed by ANOVA one-way. **Results:** The groups were comparable in clinical aspects. The ANOVA showed a significant statistical difference between the groups for the mental health worsening; ($F_{1, 461}=5.95$, $p=.015$, $ES=.78$), and quality of life worsening ($F_{1, 461}=8.1$, $p=.004$, $ES=.81$). The Tukey post-hoc confirmed a more severe worsening in the mental health condition and quality of life for NPAG than PAG. Besides, the worsening in mental health and in quality of life was directly correlated ($R=.49$; $p<.0001$). **Conclusion:** The interruption in PA due to physical distancing during the COVID19 pandemic promoted more severe worsening in mental health conditions and quality of life in people living with PD.

Neurorreabilitação

TL 1106517

INFLUENCE OF NON-INVASIVE NEUROMODULATION ON FOOD INHIBITION, EXECUTIVE FUNCTIONS AND BIOCHEMICAL PARAMETERS OF INFLAMMATION AND OXIDATIVE STRESS

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Background: The various pathologies that occur with the metabolic disorder promote chronic inflammation, leading to impairment of the integrity of the blood-brain barrier, which may affect cognitive functions. **Objective:** To correlate markers of antioxidant activity, inflammation and executive functions with tDCS in healthcare professionals. **Methods:** randomized, parallel, double-blind clinical trial. The target population consisted of health professionals from the clinical staff of a university Hospital. The following instruments were applied: Intense Food Cravings – Trait and State questionnaires, Brazilian Craving-Related Food Inventory, Three-Factor Eating Questionnaire, digit span test; Stroop Test of Colors and Words, Victoria version; Trail Making Test). Determinations of thiobarbituric acid reactive species (TBARS) and lipid peroxidation biomarkers were also performed. **Results:** A statistically significant reduction was found in QDIC-T values ($p<0.0074^{**}$); FCI-Br ($p<0.0005^{**}$); Digit span (forward order: $p<0.007^{**}$; reverse order: $p<0.01^{**}$); TSCP (conditions 1: $p<0.003^{**}$; 2: $p<0.006^{**}$ and 3: $p<0.008^{**}$); Trail Making Test (A: 0.05* and B: 0.007**) in the tDCS group when compared to the sham group. In this research, we observed that the application of tDCS triggered statistically significant changes in: women with a mean age predominantly between 31 and 40 years; middle-class income; high level of education; overweight and without previous infection by Covid-19; regarding: traits of food cravings, especially for sweets, fatty foods and traditional meal, executive functions. **Conclusion:** The use of tDCS influences the reduction of food cravings and the reduction of reaction time and the processing of executive functions, although there are no significant changes in brain bioenergetics to the point of impacting the biochemical parameters of inflammation.

Neurorreabilitação

TL 1104824

CLINICAL-EPIDEMIOLOGICAL PROFILE OF PATIENTS TREATED FOR SLEEP DISORDERS IN A SPECIALIZED OUTPATIENT CLINIC OF THE PUBLIC HEALTH SYSTEM

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Sleep is a special physiological state that occurs cyclically, and its disorders affect quality of life and lead to metabolic and cardiovascular problems. The third edition of the international classification of sleep disorders (ICSD-3) classifies them as insomnia, sleep-related breathing disorders, central disorders of hypersomnolence, circadian rhythm of sleep-wake disorders, parasomnias and sleep-related movement disorders. This study aimed to discover the clinical and epidemiological profile of patients treated in the specialized sleep disorders outpatient clinic of the public otolaryngology and cervical-facial surgery service in the city of Manaus, AM, Brazil, between January 2019 and May 2021. The data of the analyzed sample were obtained from 72 medical records, in which it was observed that sleep disorders had a higher prevalence in the female sex, and predominated in the age group of between 51-60 years. Regarding life habits, both sexes presented a link with alcohol consumption (32.26%), tobacco use (11.1%) and were overweight (73.69%). Regarding the use of medication, 44.68% of patients reported using some type of hypnotic drug. In the item comorbidities, systemic arterial hypertension (SAH) was noted in 44.62% of the subjects analyzed. The most prevalent diagnosis was obstructive sleep apnea syndrome (45.5%), which is consistent with the finding that 35.59% of the individuals had class IV Mallampati scoring. Statistically, none of these variables were shown to interfere with sleep quality when evaluated by means of the chi-square test. Finally, it was concluded that, in this specialized service, most of the consultations involve overweight women and that obstructive sleep apnea is the most prevalent disorder.

Neurosonologia

TL 1105921

ASSESSMENT OF CONCORDANCE BETWEEN ULTRA-PORTABLE ULTRASOUND EQUIPMENT AND A TRADITIONAL ONE FOR THE DIAGNOSIS OF PARKINSON'S DISEASE BY TRANSCRANIAL SONOGRAPHY

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Background: The diagnosis of Parkinson's disease (PD) is clinical. Transcranial sonography (TCS) is used to assist in the diagnosis of PD, revealing an increase in echogenicity of the substantia nigra (SN) with an accuracy of 80-90%. With the advance in technology, ultrasound (US) equipment is becoming ever more portable, allowing the use of this method even in places with difficult access to traditional systems. There are still no studies to determine the accuracy of TCS performed on ultra-portable (UP) equipment within the field of PD. **Objective:** To verify the accuracy of the Lumify (Philips Medical Systems) UP equipment by differentiating data from healthy control subjects and from patients with PD and assessing the concordance between measures obtained from this equipment and traditional equipment. **Methods:** In this cross-sectional study, TCS was performed on 129 subjects, between September 2019 and July 2021, separated into two groups, one with healthy controls and the other with PD in the Outpatient Clinics for PD at the Hospital Universitário Gaffrêe e Guinle da Universidade Federal do Estado do Rio de Janeiro / Empresa Brasileira de Serviços Hospitalares or in the Clinic for Movement Disorders at the Hospital Universitário Clementino Fraga Filho da Universidade Federal do Rio de Janeiro. The tests were performed by insonation of the temporal bone window (TW) of healthy individuals and patients with PD. The parameters obtained in accordance with protocols established were: assessment of the quality of the TW and the size of the third ventricle. In addition to the measurement of the mesencephalon area and the SN hyperechogenicity, which, in the UP equipment, were performed by automatic measurement of an ellipse and, in the traditional one, by manual delineation of its borders. **Results:** The accuracy of the UP equipment for the diagnosis of PD was 93%, with a sensitivity of 98.3% and specificity of 86.7%, with a cut-off point of the SN area $>0.20 \text{ cm}^2$. The highest concordance was the assessment of the quality of the TW (Kappa index of 100% for the right TW and 83% for the left). The SN measurements presented with higher indices of concordance; ICC= 90% for the right SN and 93% for the left. The highest concordance between the systems was the measurement of the third ventricle (ICC 96%). **Conclusion:** The UP ultrasound is an accurate tool for PD diagnosis, and could be a valuable, practical, fast, and great potential bedside method.

Neurossonologia

TL 1104955

EVALUATION OF SLEEP QUALITY IN HOSPITALIZED PATIENTS IN A HOSPITAL IN SOUTHERN SANTA CATARINA

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Background: Despite the importance of sleep for health and recovering of Hospitalized patients, the Hospitalization period requires changes in routines that can be harmful to the sleep quality of patients. **Objective:** This study aims to compare intra and out-of-Hospital sleep quality, indicating the associated factors with worse sleep quality in Hospitalized patients and their possible clinical repercussions. **Methods:** this is a cross-sectional study conducted with 252 patients from a tertiary Hospital. In addition to searching medical records and conducting interviews to identify clinical characteristics and factors that interfere with sleep quality, the Pittsburgh Sleep Quality Index (PSQI) was used to assess sleep quality in home and Hospital environment. **Results:** There was a worsening in the components of duration, efficiency and subjective quality of sleep during Hospitalization, in addition to greater use of sleep-inducing medications and more complaints of daytime dysfunction when compared to the same components of the PSQI before Hospitalization ($p<0.05$). There was also an average reduction of 57 minutes in total sleep time during Hospitalization (7h52min vs. 6h55min; $p<0.05$). The main disturbing factors reported by patients were associated with the Hospital environment, such as interventions by Hospital staff and noise. Worse in inpatients sleep quality was also associated with presence of hypertension, diabetes and difficulty sleeping before admission ($p<0.05$). There was no association of sleep quantity and quality with blood pressure and blood glucose levels. **Conclusion:** Sleep in Hospital environment was considerably worse than home environment and the sleep disturbing factors were predominantly associated with the Hospital environment.

Sono

TL 1105497

INSOMNIA: AN ANALYSIS OF ITS PATHOPHYSIOLOGY, CLINICAL TREATMENTS AND RELATED COMORBIDITIES

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Background: Insomnia is characterized by difficulty in the quality of sleep, whether to initiate or maintain sleep, which leads to substantial suffering and daytime impairment, being more common in elderly people. The pathophysiology of insomnia is associated with sleep-wake regulation through wakefulness promoters such as orexin, norepinephrine, and histamine and sleep-producing chemicals such as GABA, adenosine, melatonin, and prostaglandin. Insomnia, therefore, results from increased orexin-mediated neuronal firing and inhibition of sleep-promoting areas. In this way, the diagnosis and treatment of insomnia depends on a clinical history that analyzes precipitating factors, behavioral issues and environmental factors that can lead to poor sleep quality. **Objective:** To highlight the pathophysiology, associated comorbidities and therapeutic strategies for treating insomnia. **Methods:** This is a systematic literature review based on data extraction in indexes such as PUBMED and MEDLINE. Publications between 2018 and 2022, in English and with the following descriptors: Insomnia, Neurology, Treatment, were considered. **Results:** Insomnia disorder goes beyond an individual complaint of poor sleep or misperception of sleep status, and there is evidence that insomnia in adolescents can be associated with changes in several biosystems, including functional cortical alterations and systemic inflammation. Insomnia disorder may also be associated with depression, hypertension, type 2 diabetes, gastroesophageal reflux disease, and asthma. The treatment of insomnia symptoms can be done through cognitive behavioral therapies and through this already achieving very positive results, at the same time they can be done through drug therapy that acts on GABA-A receptors exerting sedative, anxiolytic, muscle relaxant and hypnotic effects. **Conclusion:** In this way, it is important that physicians recognize and administer the treatment of insomnia to enable the reduction of morbidity related to it, which can guarantee the return of the quality of life that is lost, reduce daytime fatigue and the predictive risks for other pathologies.

Sono

TL 1105587

SLEEP-WAKE CYCLE PATTERN IN BLIND PEOPLE LIVING IN LOW LATITUDE AREAS (NATAL-RN-BRAZIL)

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Background: It is present in the current literature that blind individuals have sleep disorders related to circadian rhythmicity. These studies have been guiding, on a global, scale therapeutic approaches aimed at the blind. However, such studies were carried out in high latitude regions, so it is necessary to evaluate the pattern of the sleep-wake cycle in blind people in low latitude areas, with little variation in the photoperiod. **Objective:** Characterize sleep and rhythm of rest activity in blind people living in a low latitude region (Natal/RN). **Methods:** Participated in the study 19 total blindness people, living in the city of Natal. The parameters of the sleep-wake cycle were characterized by the actimetry technique (Actruster – Condor Instruments) that was used for 20 days. The analyses were done in the ActStudio software and were quantified sleep parameters and non-parametric variables of the rhythm of activity and rest (IV = intraday variability; IS = interday stability; M10 = 10 hours of higher activity; L5 = five hours of lower activity and RA = relative amplitude). 3 participants were excluded due misuse of the actimeter. Project was approved by CEP – UFRN. **Results:** The blind volunteers in this study had a sleep duration of 377.7min (6.28 hours) \pm 42 min, a sleep latency of 6 \pm 11 min and an average sleep efficiency of 84.8% \pm 6.1%. We found that 68% of volunteers have a sleep time recommended by the National Sleep Foundation (NSF), 12.5% have adequate sleep and 18% have sleep deficit. The time to go to bed was 23:30 \pm 98 min and the time to get up was 7:24 \pm 102 min. For the non-parametric variables of the rhythm of activity and rest, an average of IV of 0.72 \pm 0.18 and the SI of 0.4 \pm 0.09 were found. This shows that the blind subjects presented a little fragmented sleep and low stability of the circadian rhythm. The average value of M10 was 124 \pm 46, starting at 08:11 min \pm 123 min and for L5 17.3 \pm 8, starting at 02:25 \pm 118 min. The AR value of 0.64 \pm 0.3. **Conclusion:** The majority (68%) of blinds in our study had a sleep duration and a sleep efficiency above 85%. The subjects expressed a nocturnal sleep pattern, as observed by L5 and M10 value and interval, signaling an absence of desynchronization and suggesting a greater influence of non-photoc factors on synchronization. Thus, further studies are needed to evaluate which and how these non-photoc factors are synchronizing the sleep-wake cycle.

Sono

TL 1105589

IMPACT OF THE COVID-19 PANDEMIC ON NONRESTORATIVE SLEEP PATTERN IN COLLEGE STUDENTS AT A PRIVATE UNIVERSITY IN RECIFE

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Background: Because of the peak incidence of the new coronavirus, the university routine was completely modified. Absence of group conviviality can result in psychosocial implications, impairing restful sleep and reflecting on poor academic development. Therefore, if sleep is not restorative, the difficulty maintenance of deep sleep and muscle relaxation is associated with a set of adverse symptoms, such as physical, mental and emotional disturbances, among these, mood swings, memory consolidation and learning. **Objectives:** To report the incidence of the impact of non-restorative sleep. To investigate comorbidities effects on sleep and learning consequences. **Methods:** Cross-sectional study, observational, descriptive, retrospective, epidemiological quantitative. It was carried out by interviewing 204 students from a private university in Recife, through a semi-structured questionnaire placed on the Google Forms platform. **Results:** The sample was 204 college students distributed across health (74%), humanities (22.5%) and exact (3.5%) areas. Significant changes in sleep quality after onset of COVID-19 pandemic was felt by 73.5%. Comparing results between before and during COVID-19 period and the pandemic period, 72.1% of the students interviewed began to experience anxiety symptoms. Furthermore, 64.9% experienced intensification of symptoms of anxiety. Beginning depressive symptoms after the pandemic in 45.6%, while 35.2% reported intensification of depressive symptoms. The anxiety and depressive symptoms were related as perception of the individual. Regarding waking up, 75% reported feeling fatigued and unwell. Moreover, regarding the sleep pattern interfering in daily functioning, 73% affirmed fatigue during the day; 60.3% had mood changes; 49.5% memory changes; 74.5% felt interference in concentration, and 53.4% in the learning capacity. The sleep quality affects 72.1% in the educational part; 40.2% in the social part; 64.2% in the emotional part; 62.7% in the physical part. **Conclusion:** Our study showed that there was an impairment in the quality of sleep in college students, which may have impacted on the emergence or worsening of anxiety symptoms, and consequently impacted their learning and social context. Understanding these consequences of sleep disturbances and quality of life in the pandemic period, we can emphasize the importance of install a specialized psychopedagogical care program by the university institution.

Sono

TL 1105613

WOULD HEMIFACIAL SPASM BE A MOTIVATING FACTOR FOR POOR SLEEP QUALITY?

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Background: Hemifacial spasm (HFS) is a movement disorder characterized by clonic spasms, rarely tonic, involving the muscles of facial mimicry, especially the orbicularis of the eye and mouth of one of the hemifaces, with episodes of blinking, eye closure and involuntary contractions of the mouth. HFS usually causes great aesthetic and functional discomfort in the affected muscles, compromising quality of life and mood. Sleep is a biological function in the maintenance of quality of life, and the existence of sleep complaints in patients with HFS and possible correlations between these disorders motivated this study. **Objectives:** To analyze the clinical-epidemiological characteristics of individuals with HFS; to assess sleep quality, insomnia symptoms and/or mood changes in these patients; to relate possible associations between the variables and sleep impairment. **Methods:** Observational cross-sectional study that analyzed demographic variables, mood, sleep quality, presence and severity of insomnia symptoms in patients with HFS, followed up at the university center in northeastern Brazil, using questionnaires. The validated instruments were applied: Pittsburgh Sleep Quality Index (PSQI), Insomnia Severity Index (IGI), Hospital Anxiety and Depression Scale (HAD) to assess mood (depression and/or anxiety). Statistical analysis evaluated the association between the variables and sleep (chi-square test). **Results:** 29 patients participated in the study, 21 women (72.4%) and 8 men (27.6%), aged between 46 and 82 years (mean 63.76). 86% of the sample (20 women and 5 men) had poor sleep quality by the PSQI, and 58.6% (16 women and 1 man) had insomnia by the IGI. Mood disorder (depression and/or anxiety) in 58.6% (15 women and 2 men). Statistical analysis revealed a significant association between sleep and the following variables: sex, age group, BMI and mood disorder. HFS duration, only showed statistical significance, on the IGI scale. The only factor that did not show statistical significance was the interval of botulinum toxin application. **Conclusion:** The study showed that patients with hemifacial spasm have a higher risk of impaired quality of life. Among all non-motor aspects, this study showed the prevalence of sleep disorder among patients with HFS, and so, places this disease as an additional risk in sleep impairment, but not isolated.

Sono

TL 1105862

EXCESSIVE DAYTIME SLEEPINESS: SYMPTOM OF NARCOLEPSY AND OBSTRUCTIVE SLEEP APNEA

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Background: Excessive daytime sleepiness (EDS) is defined as the difficulty in staying awake and alert during the main wakefulness shift. It is characterized by sudden attacks of sleep, even though there is an attempt to stay awake, and is related to the main symptoms of narcolepsy and Obstructive Sleep Apnea (OSA). Narcolepsy is a neurological disease of genetic, autoimmune origin and consists of five main symptoms: EDS, cataplexy, sleep paralysis, hypnotic hallucinations and disturbed night sleep. In contrast, OSA is a narrowing of the upper airways that can result in poor ventilation during sleep, and is characterized by EDS, nocturnal snoring, and nocturnal awakenings with interruption of breathing. **Objective:** To systematically analyze how excessive daytime sleepiness is a fundamental symptom for both diseases and demonstrate the importance of an effective diagnosis in the treatment of these pathologies. **Methods:** This is a systematic literature review based on data extraction in indexes such as PUBMED and MEDLINE. Publications between 2018 and 2022 were considered, in English and with the following descriptors: Excessive daytime sleepiness, Narcolepsy, Obstructive sleep apnea. **Results:** EDS can be found in patients who have both pathologies or who have only one of them, for this reason, adequate clinical characterization is the starting point for determining the diagnosis. The diagnosis of both disorders must use tools that analyze sleep quality, situations in which drowsiness is expressed and associated symptoms: snoring, hallucinations, cataplexy. In addition, polysomnography can be used to analyze the quality of sleep. Finally, in cases where there is evidence of both diseases simultaneously, it is important to be aware that the therapeutic options are based on the signs and symptoms that weigh more heavily on the therapeutic options. **Conclusion:** The proper diagnosis of the cause of EDS goes far beyond the return of the patient's quality of life and the guarantee of daily social activities. As a result, when there is a complaint of EDS, the follow-up and clinical analysis must be cautious so that the treatment can adequately identify the disorder that affects the patient and help him/her.

Sono

TL 1105892

A PROSPECTIVE ANALYSIS OF SLEEP AND MEMORY QUALITY AMONG MEDICAL STUDENTS AT AN AMAZONAS UNIVERSITY FROM THE GLYMPHATIC HYPOTHESIS

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Background: With the glymphatic system discovery, responsible for 'cleaning' toxic waste from nervous tissue during sleep, the emergence of neurodegenerative diseases related to memory from a dysfunction in this system has been discussed. **Objective:** The goal was to evaluate the correlation between sleep quality and memory in medical students at an Amazonas university during a semester, and to observe how other factors (like the pandemic) could interfere in this process. **Method:** This was a prospective observational study approved by the Ethics Committee in 2021 under statement of No. 4.978.487, with the exclusion criterion being that students under the age of 18 could not participate. It was conducted using the Mini-Sleep Questionnaire (MSQ) to assess sleep quality, Epworth Sleepiness Questionnaire to assess the degree of daytime sleepiness, Prospective and Retrospective Memory Questionnaire (PRMQ-10) to assess retrospective and prospective memory, with the following cut-off points 25, 10 and 20 respectively, and a self-reported sleep diary for two weeks. **Results:** A total of 132 academics were included, 77 (58,38%) female and 55 (41,67%) male, the means (standard deviations) obtained from the PRMQ-10, MSQ and Epworth tests were 26,44 (6,43); 32,34 (8,88) and 10,58 (4,14) respectively. From the sample, 59 students were able to complete the sleep diary during the requested time: the total sleep mean was 6,84 hours (0,78). A high degree of memory failure complaints were evidenced in 114 participants (86.36%), excessive daytime sleepiness in 74 (56.06%) and sleep alterations in 114 (86.36%), and significant correlations at $p < 0.05$ level between anxiety and depressed states at the time of data collection; anxiety and worsening memory failure complaints during the pandemic period and worsening sleep and memory complaints during the pandemic; between PRMQ-10 memory failure complaints and MSQ sleep changes, and between the amount of sleep and better results on the PRMQ-10. **Conclusion:** Sleeping changes, daytime sleepiness, and memory failure were significantly observed among the students, evidencing the direct correlation between sleep and memory, since studies show that neurotoxic proteins can be deposited in the nervous tissue even after a poor sleep day, thus being a possible explanation for the correlation found between the MSQ and the PRMQ-10, i.e., the association between important sleep changes with high complaints of memory failure.

Sono

TL 1105945

CENTRAL HYPERSOMNIA AND CHRONIC INSOMNIA: EXPANDING THE SPECTRUM OF SLEEP DISORDERS IN LONG COVID SYNDROME – A PROSPECTIVE COHORT STUDY

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Background: Long-onset COVID syndrome has been described in patients with COVID-19 infection with persistence of symptoms or development of sequelae beyond 4 weeks after the onset of acute symptoms, a medium- and long-term consequence of COVID-19. This syndrome can affect up to 32% of affected individuals. **Objective:** The present study aimed to characterize and evaluate the prevalence of sleep symptoms in patients with long COVID syndrome. **Methods:** A total of 207 patients with post-COVID symptoms were evaluated through clinical evaluation with a neurologist, cognitive and sleep test and specific exams in the subgroup complaining of excessive sleepiness (ES). **Results:** Among 189 patients included in the long COVID sample, 48 (25.3%) had sleep-related symptoms. Insomnia was reported by 42 patients (22.2%), and excessive sleepiness was reported by 6 patients (3.17%). Four patients with ES were evaluated with polysomnography and test, multiple sleep latencies test, and Actigraphic data. Of a total of 4 patients with ES, 3 fulfilled the clinical criteria for central hypersomnia, and one reported symptoms of ES that resolved spontaneously after 6 months without intervention. We observed a high prevalence of cognitive complaints in these patients. **Discussion:** This study demonstrated a high prevalence of sleep symptoms in a cohort of long-term COVID-19 patients with persistent sleep disorders up to 18 months after infection. Insomnia was the most frequent sleep disorder in these patients. To the best of our knowledge, we have reported the first cases of central hypersomnia after COVID. One of our patients fulfilled clinical and polysomnographic criteria for narcolepsy. All patients with ES responded to wakefulness-promoting agents. We have reported three patients with documented central hypersomnia after COVID-19. All 3 patients were young and had no sleep disorders prior to COVID infection, suggesting a strong association with SARS-CoV-2 infection. To our knowledge, this is the first report of this association in the literature. In addition to the possibility of direct viral infection, hypersomnia after COVID might be related to immune and inflammatory hyperactivation resulting in hypothalamic infiltration by CD4+ and CD8+ T cells leading to neuronal damage. **Conclusion:** Sleep disorders are common in patients with long COVID syndrome, particularly insomnia. Chronic insomnia and central hypersomnia may expand the spectrum of post-COVID sleep disturbances.

Sono

TL 1105948

CANNABIDIOL FOR RAPID EYE MOVEMENT SLEEP BEHAVIOR DISORDER

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Background: Cannabidiol (CBD) is one of the main non-psychoactive components of Cannabis sativa and may represent an alternative route for treating REM sleep behavior disorder (RBD). **Methods:** a phase II/III, double-blind, placebo-controlled clinical trial was conducted in 33 patients with RBD associated with Parkinson's disease (PD). Patients were randomized to CBD in doses of 75 to 300mg or matched capsules placebo and were followed up for 14 weeks. Primary outcomes were the frequency of nights with RBD, CGI-I, and CGI-S. **Results:** CBD showed no difference in relation to placebo for primary outcomes. Regarding secondary outcomes, we observed a significant improvement in average sleep satisfaction from the 4th to 8th week in the CBD versus placebo group with $p=0.049$ and $p=0.038$, respectively. **Conclusion:** CBD, as an adjunct therapy, showed no reduction in RBD manifestation. An improvement in sleep satisfaction with doses of 75-150mg has been noted.

Sono

TL 1106048

SLEEP HABITS IN THE SLEEPING HAMMOCK IN A TERTIARY HOSPITAL IN THE STATE OF CEARÁ

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Background: The hammock is an item that has followed the evolution of society, since the indigenous reality, and has spread to the other inhabitants, in a way that involves a complex diversity of values, tastes and everything else that forms the national identity (1). Several benefits of the hammock have already been reported in the literature (2). The type 1 polysomnography (PSG) test is the gold standard method for studying sleep-associated parameters (3). It is not known whether there is a difference between the sleep assessment data obtained using PSG type 1 performed in bed and in the hammock. **Objectives:** To analyze sleep habits in the hammock sleeping and perceptions of patients in a sleep service of a tertiary Hospital in the state of Ceará. **Methods:** We perform an observational cross-sectional study. The parameters of sleep preferences and habits were evaluated by a questionnaire applied to patients at the sleep service of a large tertiary Hospital in the city of Fortaleza-Ce. **Results:** The final sample consisted of 100 patients. There was a predominance of females (67%). The mean age of participants was 51 years. Most of them came from the capital (Fortaleza). 33% of participants prefer to sleep in a hammock for different reasons. The most cited reasons were: greater comfort, milder temperature, being able to swing. 73% of that who preferred hammock came from the interior of the state. 45% of the participants prefer to sleep in a diagonal position on the hammock. During childhood, 59% of the participants preferred to sleep in a hammock. 44% reported that at least one parent prefers to sleep in a hammock. 70% of the patients have children. Of these, 20% reported that their children sleeps in a hammock. The most common diagnosis was Obstructive Sleep Apnea (42%). 14% patients reported that snoring improves when they sleep in a hammock. 27% of patients reported having a greater feeling of rest when sleeping in a hammock. **Conclusion:** The habit of sleeping in a hammock is part of our people's culture. Performing the PSG type 1 test in a hammock can provide more reliable data on the sleep conditions of individuals who sleep in a hammock. There may be a difference between the PSG data performed on the bed and on the hammock. Larger studies comparing the performance of the exam in the two environments are necessary for a better analysis.

Sono

TL 1106078

THE RELATION BETWEEN THE CHIARI MALFORMATION AND SLEEP APNEA

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Background: Chiari malformation occurs when the skull base and the area of the upper part of the spine are not properly formed, causing an inappropriate bulge of the cerebellum through the foramen magnum. This syndrome, by affecting the central nervous system, allows the development of respiratory syndromes. The most noteworthy of these is sleep apnea, characterized by noises and interruptions in breathing during sleep, gradually decreasing blood oxygenation and, consequently, increasing blood pressure and decreasing cerebral blood flow. Therefore, laboratory tests, such as polysomnography, are necessary to confirm the diagnosis of sleep apnea and magnetic resonance imaging to diagnose the type 1 Chiari malformation, also requiring neurosurgery to treat the Arnold-Chiari syndrome. **Objectives:** The present study aims to analyze the relation between Arnold-Chiari Syndrome and the breathing disorder Sleep Apnea. **Methods:** It has been accomplished a bibliographical research indexed in the databases SciELO, PubMed and Google Acadêmico, of studies published in the years of 2010 to 2018, using the following descriptors: "Apnea", "Chiari" and "Sleep". **Results:** By the selection of three different articles it was possible to infer that Chiari Malformation type 1 (MCI) is a congenital condition in which the cerebellum enters the spinal canal because of a malformation of the skull. This syndrome is strongly linked to respiratory disorders, evidencing Central Sleep Apnea, because of the proximity of the respiratory structures of the brain and the structures involved in the Chiari Malformation. As a direct consequence, there is a possible lack of oxygenation in the brain and very poor sleep quality. **Conclusion:** It is evident, therefore, that polysomnography, a test realized to analyze the quality of sleep by measuring brain waves, breathing and the amount of oxygen in the blood, becomes an ally in the identification of Sleep Apnea. While MRI is essential for the identification of Chiari type 1 malformation. Both decompression surgery, for the treatment of the malformation, as well as servoventilation for the treatment of apnea, are indicated. Thereby, as a direct consequence, there is a reduction in the symptoms of sleep-disordered breathing.

Sono

TL 1106100

BLEPHAROSPASM PATIENTS AFTER BOTULINUM TOXIN – SLEEP APPROACH

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Background: Blepharospasm is a focal dystonia that affects the orbicularis oculi muscles. The interest in non-motor symptoms is due to their impact on quality of life. **OBJECTIVE:** We evaluated the frequency of sleep disorders and circadian rhythm in a sample of Brazilian blepharospasm patients. **Methods:** A total of 51 patients, who met the clinical criteria for blepharospasm, evaluated by two specialists in movement disorders, were recruited from the outpatient clinic for movement disorders of two reference centers in the city of São Paulo. The selected 13 patients were evaluated from 13 days before to 13 days after using botulinum toxin. They were interviewed, underwent physical examination and actigraphy, and completed sleep diaries. **Results:** After using botulinum toxin, the group that reported sleep improvement exhibited a 50% decrease in sleep latency. There was no change in restless leg syndrome or circadian rhythm. Patients who reported no sleep improvement after using botulinum toxin presented poorer synchronization of the light-dark cycle. **Conclusion:** Blepharospasm patients have poor sleep quality. About 50% of the patients had sleep improvement after using botulinum toxin. The synchronization of the light-dark cycle may influence this improvement.

Sono

TL 1106273

ARTIFICIAL INTELLIGENCE AS A MONITORING STRATEGY FOR INDIVIDUALS WITH OBSTRUCTIVE SLEEP APNEA TREATED WITH POSITIVE AIRWAY PRESSURE – A PROPOSAL TO THE PUBLIC HEALTH SYSTEM IN THE STATE OF CEARÁ

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Background: Obstructive sleep apnea (OSA) is characterized by recurrent episodes of partial or total upper airway obstruction, with interruptions of the sleep cycle, often associated with intermittent hypoxic/hypercapnic stress. The prevalence of OSA is high and is strongly associated with cerebrovascular and cardiovascular events. Its gold standard treatment is performed through the application of positive airway pressure. This treatment, when properly instituted, is cost-effective. In the State of Ceará, four sleep-specialized services assist and follow patients undergoing treatment with positive airway pressure. The positive pressure equipment used for the treatment is provided to patients through the State Health Department. **Objective:** To develop a system that is applied to the public health system (SUS – Sistema Único de Saúde) for the management and monitoring of positive airway pressure users who are followed up in specialized sleep services and, from this product, generate a bigdata that allows offering solutions in the sphere of management, clinical follow-up and science based on information generated by artificial intelligence models. **Methods:** The developed system uses cloud computing techniques to interact with the user through conventional browsers and the acquired data are stored in the cloud, forming a bigdata. Artificial intelligence techniques will be used to extract information that enhances the treatment of patients with OSA, both individually and collectively. **Results:** The platform is under construction, based on the research of the necessary variables and constructs. An entity-relationship diagram was produced and integrated into the construction process. The combined variables for the artificial intelligence strategy are in the adjustment phase. It is expected that the developed program will make it possible to expand the follow-up of patients followed up in specialized sleep services, enabling better communication between services from the public health system, as well as providing information on the effectiveness of therapy to the State.

Sono

TL 1106309

VALIDATION OF THE BRAZILIAN PORTUGUESE VERSION OF STANFORD CATAPLEXY QUESTIONNAIRE

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Background: Cataplexy is the most specific REM clinical manifestation of narcolepsy, and it is characterized by a sudden and involuntary episode of loss of muscle tone during wakefulness. Cataplexy cannot be easily recognized when clinical features are atypical or when the physician is unfamiliar with its characteristics. The unstructured clinical interview is the only diagnostic method most common, and the diagnosis by targeted questionnaire might help identify cataplexy. **Methods:** Stanford cataplexy questionnaire is a self-applied 51-question questionnaire. This validation consisted of an initial translation and back-translation from English into Brazilian Portuguese of the questionnaire followed by a pilot study including 10 participants for the cultural adaptation of the scale. The application of the final version to 155 different participants, with 35 narcoleptic patients, was made. **Results:** 155 consecutive patients, aged 18-85 years old, completed the questionnaire. Cultural adaptations were made to a better comprehension of patients. The Brazilian version of the Stanford cataplexy questionnaire showed similar results to those depicted in the original version. **Conclusion:** The Brazilian Portuguese version of the Stanford cataplexy questionnaire showed to be a valid tool for assessing the patients with suspicion of having cataplectic events and has a potential impact on clinical decisions.

Sono

TL 1106313

APPLICATION OF THE RAPID EYE MOVEMENT SLEEP BEHAVIOR DISORDER SCREENING QUESTIONNAIRE IN PATIENTS WITH NARCOLEPSY

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Background: REM sleep behavior disorder is characterized by the loss of muscle atony during REM sleep associated with abnormal behaviors, often with dream enactments, which can even cause bodily harm to the patient and their bed partner. It is considered a REM sleep parasomnia and can be idiopathic or associated with other diseases, including narcolepsy. When associated with narcolepsy, it usually presents an earlier onset and milder symptoms than when it is associated with other diseases, often going unnoticed by patients and health professionals. **Objective:** To evaluate signs and symptoms of REM sleep behavior disorder in patients with narcolepsy through the application of a questionnaire. **Methods:** In the study, the Rapid Eye Movement Sleep Behavior Disorder Screening Questionnaire was applied to patients with type 1 (N1) and type 2 (N2) narcolepsy. All patients evaluated had excessive daytime sleepiness and had a previous diagnosis of the disease through polysomnography and the multiple sleep latency test. Subjects were classified into N1 and N2 according to the result of the cerebrospinal fluid hypocretin dosage, which was performed in all patients studied. **Results:** 71 adult individuals were evaluated, 45 of whom had N1 and 26 had N2. The score considered positive for the questionnaire was ≥ 5 . The mean score was 8.276 (SD: 2.801) in patients with N1 and 6.368 (SD: 4.669) in patients with N2. It was observed that there was a higher score on the questionnaire in patients with lower levels of hypocretin, and in the N1 group, the score result was significantly positive ($p=0.00331$). **Conclusion:** It was observed that patients with narcolepsy have high scores when responding to the Rapid Eye Movement Sleep Behavior Disorder Screening Questionnaire. Despite being a known comorbidity in people with narcolepsy, this research shows the symptoms of REM sleep behavior disorder are more important than usually considered in clinical practice.

Sono

TL 1106316

BRAIN COMPLIANCE BEHAVIOR DURING SLEEP – INITIAL ANALYSIS OF A PILOT STUDY

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Background. Intracranial compliance represents the cranium's ability to tolerate an increase in volume without a corresponding increase in intracranial pressure. Waves pulses obtained by non-invasive method of monitoring intracranial pressure curves is a validated and more practical form to evaluate brain compliance. Nonetheless, little is known about the impact of brain compliance on sleep. During the wake state and REM-sleep period, the brain metabolism is high and also the need for blood. Once the intracranial pressure is associated to blood volume, we hypothesized that the compliance during wakefulness and REM-sleep is higher than in N2 and N3. Our **Objective:** is to evaluate the behavior of brain compliance during sleep stages and wakefulness, through pulses morphology (P1, P2, P3) of intracranial pressure and P2/P1 ratio. **Methods:** The assessment of brain compliance was performed using the non-invasive intracranial pressure (NICP) method through the Brain4care® device overnight simultaneously with the performance of type I polysomnography (Alice 6, Phillips Respironics®). We evaluated brain compliance data in the first 10 minutes of each of the following situations: wakefulness (V), sleep N2 (N2), sleep N3 (N3) and REM (REM). **Results:** The patient did not complain in using the Brain4Care device throughout the night. We obtained the following P2/P1 ratios: V= 1.29; N2=1.21; N3=1.17 and in REM=1.30. The P2/P1 ratio was higher in REM when compared to NREM sleep ($p > 0.001$); higher in REM when compared to N3 sleep ($p > 0.001$). The behavior of the P2/P1 ratio was similar when comparing wakefulness and REM. **Discussion:** The main finding of this pilot study was that the P2/P1 ratio increased during the night's sleep, being higher during wakefulness and REM sleep. We observed a decrease in brain compliance in REM sleep. Although still an isolated study, this is an interesting finding, as brain metabolism increases during REM sleep, equivalent to what is observed in wakefulness. However, brain compliance in REM sleep is quite low, similar to conditions leading to intracranial hypertension. One possible cause of the increase in the P2/P1 ratio may be related to the presence of many hypopneas that occurred during REM sleep. The evaluation of brain compliance behavior in patients with sleep-disordered breathing will be carried out in the future, which will allow us to better understand the impact of obstructive sleep apnea in that particular situation. A relevant aspect in this study is the potential use of the variable brain compliance in the polysomnography set up, adding information with impact on clinical outcomes involving OSAS. **Conclusion:** Brain compliance appears to be associated with sleep stages and is progressively reduced from wakefulness to N3 sleep. In this case brain compliance is no different in wakefulness and REM. The decrease in brain compliance in REM sleep may be associated with severe obstructive sleep apnea.

Sono

TL 1106330

BAD DREAMS IN NARCOLEPSY TYPE 1

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Background: Narcolepsy can present with different combinations of severity and frequency of sleep paralysis, sleep-related hallucinations, and cataplexy. Symptoms related to REM sleep can happen frequently, such as lucid dreams, vivid dreams, epic dreams, and nightmares. **Objective:** The objective is to highlight the increased frequency of bad dreams in patients with type 1 narcolepsy, their impact on sleep quality, and possible factors associated with this increased frequency. **Methods:** We evaluated 52 patients diagnosed with type 1 narcolepsy according to the ICSD3 diagnostic criteria. They underwent clinical evaluation and application of the Pittsburgh Sleep Quality Rating Scale, Epworth Sleepiness Scale, and Narcolepsy Severity Scale. In addition to clinical data, data from complementary diagnostic tests and medications in use were also recorded. **Results:** Mean age of patients was 37.62 ± 11.974 , with a minimum of 18 and a maximum of 70. Females corresponded for 59.6%. More than half of the patients, 55.8% had bad dreams more than once a week in the month of the interview, 15.4% had them less than once a week and 28.8% had none in the last month. There was no correlation between the frequency of symptoms related to bad dreams and the narcolepsy severity scale (person 13.785, $p = 0.130$). Regarding symptomatic scales, the prevalence of poor sleep was associated with the Pittsburgh sleep quality index score ($F 5.278$; $p = 0.003$), but without association with the Epworth sleepiness scale ($F 0.616$; $p = 0.608$). When correlating with the use of medications, there was no correlation between the use of stimulants (person 1.192; $p=0.755$) or anti-cataplectics (person 5.120; $p = 0.163$). There was also no individual difference between each type of stimulant in use (person 7.591; $p=0.576$). The use of different antiepileptic antidepressants correlated differently with the prevalence of bad dreams (person 25.919; $p=0.011$), with tricyclic showing a negative association with a higher frequency of bad dreams and selective serotonin reuptake inhibitors having a positive association with the absence of bad dreams. **Conclusion:** Oneiric symptoms are frequent in type 1 narcoleptic population. The use of antidepressants with an anti-cataplectic effect may help to control these symptoms.

Sono

TL 1106489

CONTINUOUS MONITORING OF BRAIN COMPLIANCE DURING SLEEP: EXPANDING THE SPECTRUM OF POLYSOMNOGRAPHIC VARIABLES

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Background: Based on the principle of detecting small deformations of the skull caused by changes in intracranial pressure, a non-invasive monitoring method was developed. The aim of our study was to evaluate the possibility of performing continuous monitoring of brain compliance throughout the night, through pulse morphology (P1, P2, P3) of intracranial pressure synchronized to the other physiological variables routinely addressed in a type I sleep study. **Methods:** After assembling the patient to perform the polysomnography, we positioned the device on her head. The patient was instructed to call us if there was any discomfort regarding the use of the device. The assessment of brain compliance was performed by the non-invasive intracranial pressure (NICP) method through the Brain4care® device throughout the night. Sleep assessment was performed using type I polysomnography (Alice 6 – Phillips Respironics® device). We used the AASM scoring rules to analyze the sleep study. We superimposed the graph representing the variation of the P2/P1 ratio with the hypnogram. **Results:** The patient remained overnight with the sensor on her head. It did not move at any time during the monitoring. The recording duration was 5h 44min 42s. The useful pulses for evaluation were in the line above 60%. Only 6 minutes of the exam could not be analyzed due to the presence of artifacts interfering with the P1 and P2 waves. The P2/P1 variation graph was adequately superimposed on the hypnogram. In the morning, the patient reported no complaints related to device use. The signals recorded by the polysomnography equipment were not disturbed by the Brain4Care device. **Results:** In this pilot study, we were able to demonstrate the feasibility of including a sensor aimed at assessing brain compliance and performing continuous monitoring of its variation during sleep. The data shown here are part of a large project that involves analyzing the impact of obstructive sleep apnea on brain compliance. The data plotted on the hypnogram allowed a visual analysis of the P2/P1 ratio curve, which behaved as expected, considering the data that we have already obtained in the different sleep stages of monitoring this patient. **Conclusion:** It was possible to monitor overnight brain compliance through the non-invasive device Brain4Care. The Brain4Care device did not determine interference in the signal captured for performing the polysomnography, preserving the quality of both assessments.

Sono

TL 1106495

BIOCHEMICAL ANALYSIS, BODY COMPOSITION AND SLEEP QUALITY IN HEALTH PROFESSIONALS: IS THERE A RELATIONSHIP?

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Background: Food cravings are regularly experienced by individuals with and without eating disorders, especially for palatable, highly processed foods and high levels of carbohydrates, refined sugars, and/or fat. The relationship between changes in nutritional status, cardiovascular, neurodegenerative, metabolic-inflammatory diseases, and increased oxidative stress are widely established; however, there is still little evidence for the analysis of sleep quality. **Objective:** To outline the profile of nutritional status, food desire, biochemical markers of inflammation, and sleep quality in health professionals at a university Hospital. **Methods:** This pilot study was cross-sectional, quantitative, and prospective. The target population consisted of health professionals from the Hospital clinical staff with no previous neurological complaints. Questionnaires were applied to measure the Intense Desires for Food – Trait and State questionnaires; inflammation was recorded with sociodemographic and body composition data from bioimpedance. The Pittsburgh Sleep Quality Index and the Epworth Sleepiness Scale were used for sleep assessment. **Results:** Statistically significant correlations were found between inflammation, sleep quality, phase angle, and body fat percentage. It is inferred that moderate food cravings are related to poor sleep quality, a biochemical profile compatible with inflammation in health professionals. **Conclusion:** Overweight or obesity, increased oxidative stress, poor sleep quality and excessive day sleeping are present in the profile of healthcare professionals, regardless of the work shift. To establish cause and effect, we suggest longitudinal studies assessing these relationships comparatively in the long term.

Sono

TL 1106541

THE IMPORTANCE OF SLEEP FOR SUCCESSFUL NEUROREHABILITATION AFTER STROKE

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Background: Proper sleep has important clinical implications for the learning and re-learning of movements and activities of daily life for optimal stroke rehabilitation. Both sleep and rehabilitation are critical for many activities that may need to be learned or re-learned following stroke, and for functionality in everyday life. In this context, sleep is essential in neurorehabilitation practice as it supports neuroplasticity, memory, and learning. **Objective:** We aimed to systematically review the literature to answer if sleep (including naps) matters as a contributing factor for neurorehabilitation in patients after stroke in adults. **Methods:** A systematic review was performed on clinical trials investigating the influence of sleep on post-stroke neurorehabilitation. The PRISMA guidelines were followed. Seven investigations were included in this review. Data were extracted and assessed for quality and risk of bias. We looked at any neurorehabilitation setting, compared sleep versus no sleep in rehabilitation, and explored these factors in stroke patients versus healthy individuals. **Results:** This systematic review found that published neurorehabilitation interventions included nighttime sleep and long naps (? 90mins after intervention) as the intervention improved motor consolidation in patients following stroke. Sleep should be encouraged in the treatment of stroke to maximize outcomes. After a stroke, sleep disorders of any kind (e.g., OSA, insomnia, circadian rhythm disorders) can negatively influence neuroplasticity recovery and physical rehabilitation. **Conclusion:** The available data suggest that sleep should be considered in the treatment plan for successfully targeted physiotherapy to optimize cognitive and motor learning. Sleep hygiene and therapies to improve sleep, both quality and quantity are imperative in stroke treatments as they modulate rehabilitation outcomes.

Sono

TL 1106578

RESTLESS LEG SYNDROME/WILLIS-EKBOM DISEASE (RLS/WED) AND QUALITY OF LIFE IN HEMODIALYSIS PATIENTS

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Background: Chronic kidney disease (CKD) is associated with significant morbidity and mortality and can negatively impact on many aspects of health and well-being. Restless Leg Syndrome/Willis-Ekbom disease (RLS/WED) is one of the most common and unrecognized sleep disorders in CKD patients on hemodialysis and is associated to worse quality of life. **Objective:** The current study aimed to investigate the prevalence of RLS/WED in patients undergoing hemodialysis, as well as the association with quality of life domains. **Methods:** A total of 236 patients undergoing hemodialysis from three local dialysis units were enrolled from February to June 2022. RLS/WED was identified using a single screening question ('When you try to relax in the evening or sleep at night, do you ever have unpleasant, restless feelings in your legs that can be relieved by walking or movement?') and chronic persistent form was defined as symptoms at least twice a week. Quality of life was evaluated using the the Kidney Disease QoL Short Form (KDQOL-SF) by face-to-face interview. **Results:** The prevalence of RLS/WED in hemodialysis patients was 30.1%. About 20.4% had chronic persistent form. Patients with RLS/WED, compared to non-RLS/WED patients, had significantly worse KDQOL-SF scores across multiple domains as sleep ($p=0.003$), quality of social interaction ($p=0.02$), cognitive function ($p=0.001$), burden of kidney disease ($p=0.025$), effects of kidney disease ($p=0.01$), symptoms ($p=0.01$), physical functioning ($p=0.01$), social function ($p=0.04$), and emotional role (0.02). **Conclusion:** Therefore, this study show that RLS/WED is a common disorder in hemodialysis patients and is related to worse perception of quality of life in several of its domains.

Sono

TL 1106586

PROPOSED PROTOCOL FOR DISPENSING POSITIVE AIRWAY PRESSURE EQUIPMENT FOR USERS OF THE BRAZILIAN UNIFIED HEALTH SYSTEM IN THE STATE OF CEARÁ WITH SLEEP-DISORDERED BREATHING

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Objective: This paper aims to propose a protocol to assist and standardize the dispensing of positive pressure equipment for patients with sleep-disordered breathing (SDB) in the Brazilian Unified Health System (SUS) of the State of Ceará. **Methods:** Methodological study for the development of a protocol for the dispensing of positive airway pressure (PAP) equipment to be implemented into the SUS, meeting the demands of the State of Ceará. The protocol was developed from an integrative literature review and is grounded on the opinion of experts. The chosen strategy was to apply electronic surveys made available to the consulted experts on two occasions. **Results:** The experts have defined the recommended PAP equipment, as well as the best way to organize the dispensing of those devices by the SUS according to the local needs. **Discussion:** The creation of this protocol aims to decrease the time needed to make PAP available for patients with SDB, which would improve the treatment efficacy and reduce costs for the public sector.

Sono

TL 1106618

CHRONONUTRITION AND NARCOLEPSY: A BEHAVIORAL APPROACH TO SYMPTOM MANAGEMENT

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Background: Although still incipient, there is an increasing body of data suggesting association between circadian nutritional preferences and obesity. Some studies demonstrated that concomitance between food intake and elevated levels of melatonin in bloodstream would predispose the individual to a higher insulin resistance, higher glucose levels and an increase in body mass index (BMI). It is widely known that obesity is correlated to a higher risk of developing sleep disorders, such as obstructive sleep apnea (OSA), which increments excessive daytime sleepiness (EDS) and conversely decreases quality of life. Also, it is known that patients diagnosed with narcolepsy tend to present with obesity and a sleep phase delay. In addition, it has been demonstrated that patients with narcolepsy tend to have altered bone mineral metabolism. A study using an animal model showed that the use of modafinil, a common drug used in the management of EDS, is associated with a reduction in bone mass. According to another study, circadian disruption plus sleep restriction would also predispose to disruption in bone homeostasis. There are still no studies that investigate the association between circadian nutritional preferences and the elevated BMI observed in patients diagnosed with narcolepsy, nor the reduction of bone mass in this population, to the extent of our knowledge. This is the aim of our study. **Methods:** We have selected 33 patients diagnosed with type 1 and 18 patients diagnosed with type 2 narcolepsy, based on a hypocretin measurement in cerebrospinal fluid previously performed. We intend to use data from medical records to evaluate the times and nutritional values of the main courses and assess their correlation to anthropometric endpoints. Also, we intend to compare bone mass and nutritional intake of micronutrients between these patients, evaluating the potential of these measurements of becoming biomarkers of disease. **Conclusion:** Narcolepsy is commonly associated with an important decrease in quality of life, in part because of its morbidity with obesity and OSA. Defining a behavioral approach to the treatment of obesity, based on nutritional counseling, would optimize symptoms control in these patients. In addition, the definition of a decreased bone mass as a part of the pathophysiology of this disease – and a possible biomarker – would allow faster and cheaper diagnosis, also favoring a better management of bone homeostasis in clinical follow-up.

Sono

TL 1106638

CHRONOTYPE AND HEMODIALYSIS-RELATED SOCIAL JET LAG IN CHRONIC KIDNEY DISEASE PATIENTS

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Background: Chronodisruption refers to the chronic disruption of circadian rhythms leading to disease. Alterations of circadian rhythms have been observed in CKD patients and there is accumulating evidence that some of them may adversely affect health. Social jet lag is a term describing misalignment between social and biological times conventionally opposing work days to free days. CKD patients in hemodialysis are distributed into treatment shifts that configure a commitment that occurs three times a week. **Objective:** The current study aimed to investigate if there is a hemodialysis-related social Jet Lag, to assess chronotype in hemodialysis patients and if there is inconsistency between the chronotype and the dialysis shift. **Methods:** A total of 194 patients undergoing hemodialysis from three local dialysis units were enrolled from February to June 2022. Chronotype was estimated using Morning-Eveningness Questionnaire (MEQ) and a modified version of Munich Chronotype Questionnaire (MCQT) was used to estimate hemodialysis-related social Jet Lag. **Results:** Hemodialysis related jet lag was 48.1 ± 1.4 min (range 0 - 4: 57) and was inversely correlated to age ($r = -0.216$, $p = 0.01$). Chronotype was distributed as 2.1% moderately evening, 31.4% intermediate, 39.2% moderately morning and 27.3% definitely morning. Forty-four patients (22.7%) were in morning shift treatment, 96 (49.5%) in afternoon shift and 54 (27.8%) in night shift treatment. Inconsistency between shift treatment and chronotype was detected in 38 (19.6%) and was related to higher MEQ scores ($p < 0.001$). **Conclusion:** Therefore, this study seems to indicate that there may be a hemodialysis-related social jet lag, that morning chronotype is the most frequent in hemodialysis patients and higher score in MEQ is related to inconsistency between shift treatment and chronotype.

Sono

TL 1106646

ASSESSMENT OF SLEEP AND QUALITY OF LIFE IN SCHOOL CHILDREN

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Background: Sleep plays an important role for the construction of the development of children, children and adolescents, having a great influence on the development of this population, affective and social. **Objective:** The objective of this study was to evaluate sleep and quality of school life in a public school in the city of São Paulo. **Method:** Sleep assessment was performed using the Sleep Disturbance Scale for Children (SDSC) with 26 items, divided into 6 subscales: Disorders of initiating and Maintaining Sleep (DIMS), Sleep-disordered breathing (SDB), disorders of arousal (DA), Sleep-wake transition disorders (SWTD), Disorders of excessive somnolence (DOES) and sleep hyperhidrosis (SHY). The SDSC was answered by the parents. The quality of life assessment was performed using the Pediatric Quality of Life Inventory (PedsQL), by both children and parents. **Results:** The SDSC was answered by the parents of 22 5-year-old children, 16 boys (73%) and 6 girls (27%). The average score on SDSC was 45.59 ± 11.86 . The DIMS had a higher mean (13.27 ± 2.95) when compared to the other subscales (SDB: 4.72 ± 2.69 ; DA: 4.22 ± 1.59 ; SWTD: 10.72 ± 3.23 ; DOES: 8.54 ± 2.69 and SHY: 4.09 ± 2.31). Parents and 17 children, 13 boys (76%) and 4 girls (24%) responded to the PedsQL™. There was no difference between the total quality of life score between the children's self-report and the parents' perception (76.89 ± 16.30 Vs. 84.84 ± 7.78 ; $p = 0.051$). Only in the physical capacity domain of the PedsQL™ there was a difference between the children's self-report compared to the parents' perception (84.76 ± 13.98 Vs. 95.21 ± 4.56 ; $p < 0.001$). **Discussion:** We observed that the schoolchildren's sleep was compromised by the mean score on the SDSC. DIMS are frequent complaints, as it is a transition period in which children prepare to experience the school routine, presenting anxious behaviors and, consequently, impairing the quality of their sleep. Despite this finding, there was no impairment in the quality of life of the children evaluated. In the physical capacity domain, the children's self-report score was lower than the parents' score. Studies show that the information provided by parents and/or guardians is not always equivalent to that reported by children. **Conclusion:** The children evaluated showed impaired sleep, with DIMS being the most prevalent. The child's self-report about their health is equivalent to the perception of parents/guardians.

Sono

TL 1106647

IS RESTLESS LEGS SYNDROME IN PARKINSON DISEASE PATIENTS ASSOCIATED WITH ANY SPECIFIC FACTOR?

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Background: Restless legs syndrome or Willis-Ekbom disease is a disorder characterized by unpleasant sensations associated with the need to mobilize the lower limbs. In Parkinson disease patients, restless legs syndrome is associated with worse quality of life and excessive sleepiness. Regarding other factors, results of different studies are controversial. **Objective:** To determine the factors associated with the restless legs syndrome presence in Parkinson disease patients. **Methods:** A cross-sectional study was conducted in 88 consecutive Parkinson disease patients from the outpatient clinic for 21 months. Participants underwent a clinical interview, assessment based on standardized scales (Epworth Sleepiness Scale, Parkinson Disease Questionnaire - 39, Pittsburgh Sleep Quality Index, International Restless Legs Syndrome Study Group rating scale), and video-polysomnography. **Results:** Out of the 88 participants, 25 had restless legs syndrome. In the multivariate analysis, restless legs syndrome in Parkinson disease has been associated with the symptom of smell loss and quality of sleep and life. In the univariate analysis, restless legs syndrome in Parkinson disease has occurred more frequently in women with higher frequency of insomnia, constipation, and anosmia than in the group without restless legs syndrome. **Conclusion:** Restless legs syndrome is a prevalent condition in patients with Parkinson disease and is associated with specific characteristics in this group of patients.

Sono

TL 1106649

SLEEP DISORDERS IN CONGENITAL ZIKA VÍRUS SYNDROME

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Background: Congenital Zika virus syndrome (CZVS) sleep characteristics in patients are still poorly studied, with only one study in which the Brief Infant Sleep Questionnaire (BISQ) was carried out, that shows that 34.1% of CZVS were defined as poor sleepers. No previous study performed electrophysiological assessment of sleep. **Objective:** The aim of this study was to clinically characterize sleep disorders in a case series of CZVS patients, correlating these findings with disease features and polysomnographic (PSG) results. **Methods:** This is a non-randomized, cross-sectional study with consecutive inclusion of confirmed CZVS. The evaluation was performed with PSG and the characteristics of sleep were evaluated using Sleep Disturbance Scale for Children (SDSC) and BISQ. **Results:** Ten patients were evaluated, with a mean age of 4,6 years, seven male (7/10). Eighty percent of the patients present a SDSC score more than 39, therefore, demonstrate sleep disturbances and 20% clinically relevant. The BISQ sleep pattern analysis showed that 10% of children were defined as poor sleepers, 10% had more than 3 wakes per night and 80% had nocturnal wakefulness period more than 1 hour. Polysomnography evaluation: Two patients were not evaluated in the polysomnography due to technical difficulties (exam duration and was no signal capture in the nasal cannula). Mild sleep apnea was detected in 5 patients and moderate in 2 patients, of these 2 with central and obstructive apnea. Mild desaturation in 5 patients and severe in 1 patient. Increase in sleep latency was documented in 5 children, normal in 2 and reduction in 1. Presence of periodic movement of members, with a PLM index of 29.8/h, in one patient. All showed reduced REM sleep, which was not detected in 1 patient. **Conclusion:** The frequency of sleep disorders in the population of children with confirmed CZVS is very high, being important the evaluation through polysomnography in this population.

Sono

TL 1106653

TRANSLATION, TRANSCULTURAL ADAPTATION, AND VALIDATION OF THE BRAZILIAN PORTUGUESE VERSION OF THE OBSTRUCTIVE SLEEP APNEA KNOWLEDGE AND ATTITUDES (OSAKA) QUESTIONNAIRE

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The knowledge and attitude of physicians regarding the identification and treatment of patients with obstructive sleep apnea can be assessed through the OSAKA (Obstructive Sleep Apnea Knowledge and Attitudes) questionnaire. This questionnaire was developed in 2003, and the University of Washington holds the intellectual property of the instrument. In Brazil, the current literature does not present any validated instrument that specifically assesses the medical knowledge of OSA. **Objective:** This study aimed to analyze the psychometric properties of the translated and cross-culturally adapted version of the OSAKA (Obstructive Sleep Apnea Knowledge and Attitudes) questionnaire in the Brazilian Portuguese language. **Methods:** The OSAKA instrument was translated by two independent translators, and the back-translated conciliated version was presented and approved by Washington University, which holds the intellectual property for the OSAKA questionnaire. A total of 176 physicians from different specialties electronically completed the OSAKA instrument and the ASKME (Assessment of Sleep Knowledge in Medical Education) questionnaire, which was used as an auxiliary instrument to analyse the construct validity. **Results:** The items from the knowledge and attitudes domains presented acceptable internal consistency values, with McDonald's omega coefficients (?) of 0.70 and 0.73, respectively. The OSAKA questionnaire showed a moderate correlation with the ASKME instrument ($r = 0.60$, $p < 0.001$) and excellent retest reliability, with an intraclass correlation coefficient of 0.81. There were differences in knowledge between the medical specialties ($p < 0.001$). Regarding attitudes, most respondents considered obstructive sleep apnoea and its diagnosis to be important and felt confident in identifying it, but the same majority did not feel confident in treating the disease. **Conclusion:** The OSAKA instrument, as a translated and cross-culturally adapted Brazilian Portuguese version, presented psychometric properties with adequate reliability and validity.

Sono

TL 1106657

SLEEP MEDICINE ON INSTAGRAM: FASHION DANCES AND NONSENSE

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Background: With the advancement of social networks we have a new profession: the “digital influencer”. No formal training or specializations are required, just communication skills and understanding of the target audience. In the context of the digital world, it is very common for health professionals to abandon their technical behavior based upon science knowledge, adopting a common language and a banal attitude, associating themselves with their target audience. **Objective:** The aim of this study was to analyze the Instagram profiles of sleep medicine professionals. **Method:** We did an Instagram search using only the word “sleep”. We access the searched accounts and only analyzed professional profiles. **Results:** We found 78 professional profiles. Of these, 12 otolaryngologists, 12 physiotherapists, 10 sleep consultants, 9 psychologists, 8 neurologists, 5 pulmonologists, 3 psychiatrists, 3 pediatricians, 2 dentists, 2 sleep educators, 1 sleep biologist/consultant, 1 biomedical doctor, 1 cardiologist, 1 scientist, 1 doula, 1 nurse, 1 coach, 1 speech therapist, 1 mentor/sleep therapist, 1 multiprofessional group, 1 psychoanalyst and 1 sleep specialist. Fifty-eight profiles present a marketing pattern focused on their professional performance; 17 profiles focused on diseases; 13 advertise their courses and 3 profiles their mentorships, 1 profile a webinar and 1 profile a podcast. The most frequent publications addressed were: 42% sleep in general, 26% infant sleep, 13% apnea and CPAP, 8% insomnia and 5% snoring. Regarding the content of the publications, 44 profiles (56%) have an informative/technical pattern, 19 (24%) have an informative/sensitizing pattern and 20 profiles (26%) have a sensitizing content. Considering the quality of the posts, 68% have a homemade pattern and 33% have a professional pattern. **Conclusion:** Advertising in the area of sleep medicine involves a heterogeneous group of people and professionals. Most of them, without the proper formal training required by the area, in addition to offering treatments, offer courses always aiming at a good financial return. The way they present themselves is typical of current times when they are equal in expressions to young influencers, centered on their own opinions and not on traditional technical-scientific standards. In addition, they refer to themselves with a varied and unofficial titles and measure their value by the number of followers and interaction with the public.

Sono

TL 1104998

EFFECTS OF TELEHEALTH ON SLEEP AND PHYSICAL ACTIVITY LEVEL IN PATIENTS WITH PARKINSON'S DISEASE DURING THE PANDEMIC

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Background: Telemedicine allows Parkinson's Disease (PD) patients to overcome physical barriers of access to health care services and increases accessibility for people with mobility impairments. Studies have shown similar effectiveness of virtual consultations versus face-to-face appointments for the management of PD symptoms. **Objective:** This study aims to evaluate the feasibility of telehealth activity in the follow-up of patients with PD, focusing on its effects on sleep and physical activity level. **Methods:** This study was performed at a Neurology Outpatient Clinic in Fortaleza, from May 1st to December 31st, 2020. Inclusion criteria were PD diagnosis and consultation in the clinic in the last 12 months and exclusion criteria were non-adherence to consultations and lack of technology for communication. All participants signed a consent form. During the study, patients participated in a teleconsultation, answered to 3 questionnaires-Pittsburgh PSQI Quality Index; Epworth Sleepiness Scale; and International Physical Activity Questionnaire- Short Form (IPAQ-SF)- and received health education advice, such as on healthy eating, sleep hygiene, fall prevention and encouragement on doing physical activity at least 30 minutes 3 times per week. After 15 days, calls were made to reinforce the recommendations and, after 45 days, the 3 questionnaires were reapplied. **Results:** A total of 107 patients participated, 58.9% were male. The mean age was 65 and mean disease duration was 10 years. 8.6% of the patients were attending physical therapy sessions; 25.7% were engaged in physical activity at least 30 min 3 times per week; and 36.5% needed walking aids. Perceived functional decline and worsening parkinsonian symptoms during the pandemic were reported for 32.7% of the patients. Baseline IPAQ-SF classification was as follows: 41.4% sedentary; 10.3% insufficiently active A; 9.2% insufficiently active B; 34.5% active; and 4.6% very active. There were improvements in three PSQI components: perceived sleep duration, sleep disorders and daytime sleep disturbance with an average of 1.7, 2.3 and 1.7 respectively. As for the level of physical activity, there was an increase in the number of days of walking and the number of days of moderate activities for at least 10 continuous minutes. **Conclusion:** This telehealth intervention promoted positive effects on sleep and on physical activity level of PD patients and may have greater benefits in longitudinal application.

Transtornos do Movimento

TL 1105051

FACTORS THAT NEGATIVELY IMPACT THE SEXUAL FUNCTION OF MEN LIVING WITH PARKINSON'S DISEASE

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Background: Among the non-motor alteration associated with Parkinson's disease, sexual dysfunction is less discussed and, consequently, less known. Satisfactory sexual function has been associated with better quality of life even in elderly. Therefore, it is imperative to know which motor and non-motor alterations negatively affect sexual function in order to find new approaches to improve this vital function. **Objective:** To investigate the impact of motor, non-motor function, and relationship quality on sexual function in men living with PD. **Methods:** Thirty-three men with a confirmed diagnosis of Idiopathic PD for 6.69 years (SD 4.39); mean age of 54 years (SD=9.98), in stage 1 – 3 of disease evolution according to Hoehn and Yahr classification, without dementia, participated in the present study. After presenting the informed consent form and expressing agreement to participate in the study, participants were asked to answer, through telephone interviews, a previously established questionnaire that investigates: (1) general information; (2) socio-economic status; (3) information associated with PD; (4) mental and cognitive health; (5) motor and non-motor aspects of daily life experience assessed by sections I and II of MDS-UPDRS; (6) International Index of Erectile Function (IIEF), and Male Sexual Quotient (MSQ); (7) Dyadic Adjustment Scale (DAS). The statistical analysis tested the correlation between total scores in the IIEF, MSQ and age, disease evolution, I – II MDS-UPDRS scores and DAS. **Results:** There was no correlation between age, disease evolution, the severity of non-motor alterations, relationship quality, and sexual function. However, there was a statistically significant correlation between MDS-UPDRS section II and sexual function ($R=-.50$; $p<.01$). Significant correlations can be observed in multi-domains of sexual function. **Conclusion:** For men living with PD, the disability level expressed by motor experiences of daily living is the main factor for the quality of sexual function. Then, interventions to improve motor function and reduce disability in daily living activities may help the men living with PD reach or keep a satisfactory sexual function.

Transtornos do Movimento

TL 1105085

INDICATORS OF TELEMEDICINE FEASIBILITY IN PARKINSON'S DISEASE PATIENTS DURING THE COVID-19 PANDEMIC

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Background: During the COVID-19 pandemic, access to health services was limited by government measures to combat the spread of the virus. Consequently, care for other diseases was impaired, such as Parkinson's disease (PD). Telemedicine was adopted as a way to make health care viable in the face of the lockdown state. **Objective:** To assess feasibility indicators of a telehealth intervention for patients with PD, during the pandemic isolation period in 2020. **Methods:** The sample consisted of 107 patients from the Movement Disorders outpatient clinic of a tertiary Hospital from May 1 to December 31, 2020. Eligibility criteria were: diagnosis of PD, face-to-face consultation in the previous 12 months, access to necessary communication and caregiver availability in cases of dementia. If eligible, they were invited to attend a medical appointment on WhatsApp® video calls. Recruitment, attendance, technical problems and acceptance rates were calculated as indicators of feasibility. Acceptance was assessed by a questionnaire with 10 questions about: feasibility, sense of security, satisfaction, effectiveness and future use of the intervention, with a visual analogue scale. **Results:** We had 61.3% recruitment and 90.7% attendance rates. Physicians perceived technical difficulties in 11.2% of consultations. The low recruitment rate was mainly due to the high proportion of patients who changed their telephone number and could not be reached. The inability of outpatient clinics to obtain registration data, the unavailability of technology and the absence of a caregiver were other problems found. Acceptance, in general, was higher than 80%, with the exception of a question about effectiveness in improving the symptoms of the disease, which had an average rate of 75%, suggesting that a single consultation is insufficient to resolve all patient complaints, especially those that require a multidisciplinary approach. Participants were referred to rehabilitation services, but in most cases there was no attendance due to the few public rehabilitation centers available. **Conclusion:** This is probably the first study to assess the feasibility of telemedicine on WhatsApp® in the care of patients with PD in the Brazilian public health system. The results showed good acceptance and attendance rates for the strategy, but more studies with a control group and intervention effect outcomes are needed.

Transtornos do Movimento

TL 1105107

APPLICATION OF SARC-F TO SAROPENIA SCREENING IN PATIENTS WITH PARKINSON'S DISEASE IN A SPECIALIZED OUTPATIENT CLINIC IN FORTALEZA

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Background: Parkinson's disease (PD) is characterized by progressive and disabling neurodegeneration. Among the disorders present in PD, musculoskeletal alterations, such as sarcopenia, added to the decline in functional capacity, can favor fall events and reduced functionality and quality of life of PD patients. **Objective:** Describe data obtained in screening for sarcopenia in patients with PD. **Methods:** A descriptive observational study was carried out, applying the SARC-F Questionnaire in 125 patients with PD in a specialized outpatient clinic of a tertiary Hospital in Fortaleza. The questionnaire evaluates the patient's perception of their limitations in the following conditions: strength to lift and carry 4.5 kg; ability to walk around a room; get up from a chair; climb a flight of stairs and the number of falls in the last year. Results greater than or equal to 4 mean probable sarcopenia. Only patients with Hoehn & Yahr (HY) 1 to 3 were included and patients with any other disabling diseases were excluded. **Results:** A total of 125 patients were analyzed, 72 (57.6%) male, with a mean age of 66.16 years. Of the total, the vast majority (42.4%) had HY of 2.5. Furthermore, 64 (51.2%) scored SARC-F positive, of which 33 (51.6%) were female and 31 (48.4%) were male, and 61 (48.8%) scored negative, of which 20 (33.3%) were female and 40 (66.7%) were male. Moreover, among the patients with positive SARC-F, 19 (29.7%) used some walking aid and 45 (70.3%) did not. Among the patients with negative SARC-F, 7 (11.5%) used some walking aid and 54 (88.5%) did not. Finally, among the total number of patients, 52 (41.6%) practiced physical activity for at least 30 minutes 3 times a week, of which 16 (30.8%) had positive SARC-F and 36 (69.2%) negative, and 73 (58.4%) did not practice physical activity, of which 48 (65.8%) had positive SARC-F and 25 (34.2%) negative. **Conclusion:** There is still few data in the literature that discriminate regarding the difference of sarcopenia screening in patients with PD, making more studies necessary for an adequacy of this analysis for this profile of patients.

Transtornos do Movimento

TL 1105145

DESCRIPTION OF CLINICAL CONDITIONS, PHYSICAL ACTIVITY LEVEL, DAYTIME SLEEPINESS AND SLEEP QUALITY IN PATIENTS WITH PD DURING THE COVID-19 PANDEMIC

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Background: Many patients with Parkinson's disease (PD) had health care impaired due to physical barriers and sanitary conditions imposed by the Government to contain the virus spread. As a result, there were reports of worsening of motor and non-motor symptoms in these patients during the pandemic period. **Objective:** Describe clinical conditions, physical activity level, daytime sleepiness and sleep quality in PD outpatients Tertiary Hospital Movement Disorders during covid-19 pandemic. **Methods:** The study was carried out in the period from May 1st to December 31st, 2020. Eligibility criteria included diagnosis of PD and face-to-face attendance in the previous 12 months and the criteria for exclusion were non-adherence to consultations and lack of technology for communication, unavailability of the caregiver in case of patient with dementia. During the study, patients participated in a teleconsultation and responded to 3 questionnaires: Pittsburgh Sleep Quality Index (PSQI), Epworth Sleepiness Scale (ESS) and International Physical Activity Short Form Questionnaire (IPAQ-SF). **Results:** 107 patients participated, being 58.9% male, with a mean age of 65.7 (± 17.1) years and mean duration of disease of 10 (± 6.7) years. The most comorbidities prevalent were arterial hypertension (44.9%); depression (32.7%); Type 2 diabetes (24.3%); dyslipidemia (23.4%) and osteoporosis (15%). In the IPAQ-SF there was 41.4% sedentary; 10.3% insufficiently active-A; 9.2% insufficiently active-B; 34.5% active and 4.6% very active; that is, 51.7% of the sample with a significant reduction in physical activity. The PSQI and ESS results were 12.1 (± 3.6) and 11.2 (± 5.9), respectively, showing poor sleep quality and increased daytime sleepiness. **Conclusion:** During the pandemic, the level of physical activity was unsatisfactory, with a predominance of a sedentary lifestyle, as well as poor sleep quality and daytime sleepiness. It is not possible to infer whether the quarantine has aggravated pre-existing conditions or caused these problems, since it is a cross-sectional study, but the results are worrying because they represent a greater risk of sarcopenia, falls, functional impairment and worsening of quality of life.

Transtornos do Movimento

TL 1105236

ANALYSIS OF UPPER LIMB STRENGTH IN PATIENTS WITH PARKINSON'S DISEASE: A DESCRIPTIVE CROSS-SECTIONAL STUDY

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Background: Parkinson's disease (PD) is a neurodegenerative disorder caused by disruption of the dopaminergic system. The muscle weakness is one of the main complaints reported by patients with PD, given the impacts on day-to-day activities. The clinical diagnosis of PD is based primarily on motor features, that usually starts in the upper limbs. The involvement of handgrip strength can be considered as an intrinsic characteristic of PD, besides being correlated with the motor symptom severity of the disorder. **Objective:** Correlate upper limb strength to the disability in patients with PD. **Method:** Descriptive cross-sectional study with PD patients from the Movement Disorders outpatient clinic of a tertiary Hospital carried out from September/2020 to May/2022. Demographic, clinical data and upper limb strength measurements were collected using the SAEHAN handgrip test with dynamometer according to the Southampton protocol and the cutoff suggested by the Revised European Consensus on Sarcopenia (2019) of <27kg for men and <16kg for women as determinants of weakness. PD staging was assessed using the modified Hoehn&Yahr (HY) scale and functionality using the Schwab-England (SE) scale. Eligibility criteria were mild to moderate PD (HY 1-3), no other neurological diseases with motor impairment, no chronic diseases with acute instability and no health conditions that compromise mobility. **Results:** The sample consisted of 125 patients, of which 72 (57.6%) were men and 53 (42.4%), women; 56.8% had a family history of PD, 53.6% had dyskinesia, 43.2% reported FOG, 20.8% needed walking aids and 41.6% were physically active. The comorbidities were: SAH (47.2%), depression (17%), PLD (16%) and DM2 (11.2%). Twenty-five patients (20%) had reduced palm strength, with an average age of 64.4 years in the group with normal examination and 71.1 years in the group with alterations. Of these, 12 were women and 13, men; 12 patients (48%) had SAH, 2 (8%) DM2, 3 (12%) PLD, and 9 (36%) depression. In the group with weakness in the upper limbs, the prevalent HY was 3, in 13 patients (52%); 3 (12%) had HY=2 and 9 (36%) had HY=2.5. **Conclusion:** Study carried out with Brazilian elderly had 28.2% dynapenic, compared to the 20% recorded in this analysis in patients with PD and a mean age of 71.1 years. Despite the correlation between the decreased upper limb strength in PD and a greater impairment of manual dexterity, it is important that further studies are carried out in this area.

Transtornos do Movimento

TL 1105244

ANTHROPOMETRIC PROFILE OF PARKINSONIANS FOLLOWED UP AT A REFERENCE CENTER IN FORTALEZA – CE

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Background: Parkinson's disease (PD) is a progressive neurodegenerative disorder. PD is known as a movement disorder with three cardinal signs: tremor, rigidity and bradykinesia. Weight changes and decreased muscle strength are also common in patients with PD. Describing anthropometric measurements and Body Mass Index (BMI) in patients with Parkinson's disease can improve interventions aimed at this public. **Objective:** The aim is to describe the anthropometric measurements of patients with Parkinson's Disease in a reference center in Ceará, Brazil. **Method:** this study has a cross-sectional and descriptive design. A sample of 125 eligible participants were included in the study, after signing the consent form. Data collection took place at the Walter Cantídio University Hospital, in Fortaleza, Ceará, Brazil, from February 8, 2021 to April 18, 2022. The inclusion criteria were: having a diagnosis of Parkinson's Disease according to the English Brain Bank diagnostic criteria; Age >40 years; stages 1, 2, and 3 of the Hoehn and Yahr scale. Participants with dementia syndrome, according to MSD-V; chronic diseases with acute exacerbation; active cancer or infection were excluded. The participants' data were included in the Research Electronic Data Capture (REDCap) platform, in compliance with the data protection law. **Results:** 72 men (57.6%) and 53 women (42.4%) with Parkinson's disease made up the sample, with a mean age of 65 years. Most patients were married (69.6%), did not practice physical activity (58.4%), did not use alcohol (88%) and did not smoke (97.6%). Comorbidities included hypertension (47.2%), dyslipidemia (16%), depression (13.6%), diabetes (11.2%), osteoporosis (8%) and generalized anxiety disorder (7.2%). Regarding BMI, 51 patients were overweight (men, 31 [44.9%]; women, 20 [38.5%]), 40 were normal weight (men, 23 [33.3%]; women, 17 [32.7%]), 17 were underweight (men, 9 [13%]; women, 8 [15.4%]) and 13 were obese (men, 6 [8.7%] and women, 7 [13.5%]). BMI of four patients was absent. Calf circumference was altered (<31cm) in 25 patients (20%). The Waist/Hip Ratio was altered in 48 patients (men, 40 [56%]; women, 8 [15%]), with missing data in one participant. **Conclusion:** these measures may highlight the need for greater attention from the health system in relation to the treatment and monitoring of patients with Parkinson's disease to improve their quality of life.

Transtornos do Movimento

TL 1105410

PARKINSON'S DISEASE IS RELATED TO HIGH EXPOSURE TO HOUSEHOLD INSECTICIDES IN BELÉM, PARÁ

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Background: Parkinson's Disease (PD) is a neurodegenerative disease that does not have a known cause but is strongly affected by environmental factors for its evolution, such as the extended exposure to household insecticides (HI). The HI, such as the pyrethroids, are used in residences for insect control, especially mosquitos carrying arboviruses. **Objective:** To evaluate the association between exposure to HI and the development of PD in three Brazilian cities. **Methods:** This is a clinical, observational, and case-control study. Participants were enrolled from three cities: Belém/PA (BEL), Ribeirão Preto/SP (RP), and São Paulo/SP (SP), and were submitted to the survey "Latin American Research Consortium on the Genetics of Parkinson's Disease" (LARGE-PD). The statistical analysis was done with the Chi-square test and Mann-Whitney test. **Results:** 373 patients with PD (BEL: 134; RP: 150; SP: 89) and 215 controls (BEL: 94; RP: 97; SP: 24) were assessed. The median age of patients and controls was 59 and 53 years old, respectively. In BEL, the exposure to HI between patients (73.8%) and controls (70.2%) was more common than in RP and SP ($p < 0.01$; RP – patients: 48%; controls: 51.5%; SP – patients: 35.9%; controls: 37.5%), but there were no differences in the exposure to HI between patients and controls in any of the cities. In BEL, the majority of individuals had high exposure to HI (exposure > 30 days/year) (patients: 69.6%; controls: 42.4%; $p < 0.0001$) compared with the other cities (high exposure: RP – patients: 15.2%, controls: 10%, $p = 0.56$; SP – patients: 31.2%, controls: 3.3%, $p = 0.77$), and patients with PD had greater proportion of high exposure to HI, compared with controls ($p = 0.001$). The overall median exposure time was 15 years, with no differences between patients and controls. **Conclusion:** In BEL, an Amazon Brazilian city with major exposure to HI, there was an association between high exposure to HI and the development of PD. High exposure to HI in BEL might be related to the Amazon environment, which promotes higher insect proliferation. Despite the weakness of the study (selection bias, memory bias, small sample, retrospective study), these results provide support to new studies that explore the association between PD and HI, mainly in less developed areas of the world.

Transtornos do Movimento

TL 1105452

A STUDY OF THE ASSOCIATION BETWEEN RELIGIOSITY AND DEPRESSION IN PATIENTS WITH PARKINSON'S DISEASE

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Background: Neuropsychiatric comorbidities are common in Parkinson's Disease (PD) and impact patients' quality of life. In parallel, religiosity is associated with the improvement of depressive and anxious symptoms in the general population. However, in other countries, the rates of religiosity among people with PD reported significantly lower levels of religiousness than did age-matched controls. That possibly happens because, in addition to the motor impairment that limits the experience of religious activities, depression, the main neuropsychiatric comorbidity, may influence the religious practices and beliefs of these patients. **Objectives:** To assess the level of religiosity in patients with PD and verify an association with depression and quality of life indices. **Methods:** Patients with PD diagnosed according to the 2015 MDS criteria were included in this study. We excluded cases with the hypothesis of atypical parkinsonism. Religiosity was assessed using the Duke University Religiosity Index, with 3 components: organizational religiosity (OR), non-organizational religiosity (NOR) and intrinsic religiosity (IR). Beck Depression Index (BDI) and PDQ 39 evaluated depression and quality of life indices, respectively. The cutoff point for moderate depressive symptoms was BDI > 19. The Spearman test was used to assess the correlation between the variables. This study obtained approval from the local ethics committee approval. **Results:** Twenty-eight patients were included, among which 3 had moderate depressive symptoms. The correlation coefficient between OR/NOR with the other variables was 0.308 for BDI ($p = 0.111$); 0.218 for PDQ 39 ($p = 0.264$) and 0.243 for UPDRS III ($p = 0.212$). As for the relationship between RI and the other scales, the value found was -0.348 for BDI ($p = 0.069$); -0.443 for PDQ 39 ($p = 0.018$) and -0.407 for UPDRS III ($p = 0.032$). Furthermore, the relationships between PDQ 39 and IDB (0.513; $p = 0.005$) and PDQ 39 and UPDRS III (0.614; $p < 0.001$) were analyzed. **Conclusion:** Only IR showed a significant correlation with PDQ 39 and UPDRS III. Intrinsic religiosity was weakly associated with better levels of quality of life and less motor impairment. Although not significant, a trend towards fewer depressive symptoms was observed in patients with a higher degree of intrinsic religiosity. There was also a significant association between quality of life with a lower degree of depressive symptoms and a lower degree of motor impairment.

Transtornos do Movimento

TL 1105471

CLINICAL AND GENETIC ANALYSIS OF A POPULATION WITH HUNTINGTON'S DISEASE IN AMAZONAS

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Background: Huntington's disease (HD) is a hereditary neurodegenerative disorder. The epidemiology of HD is still lacking in information covering all regions of Brazil. The first publication, bringing genetic aspects of 33 patients from 16 families with HD in Amazonas (AM), was published by our research group in 2020 (BRITO, 2020). These initial data showed that the incidence of HD in BF and the specific clinical aspects of this population should be better studied and included in the Brazilian series. **Objectives:** To analyze HD's prevalence, phenotypic and genotypic profile in a reference center for movement disorders in the North region of Brazil. Identify possible HD transmission clusters due to geographic characteristics. Describe the phenotypic characteristics of the population and correlate them with the genotypic findings. **Method:** prospective observational study with analysis of patients seen at a referral center. Genetic testing for Huntington's disease and clinical follow-up of patients and family groups. **Results:** The results: presented here correspond to 55 patients with suspected HD, 38 of which were effectively genetically tested and included in this analysis. Distribution by sex was 50% (19 H and 19 F). The mean age of symptom onset was 33 years (7 to 62 years), with a mean size of the expansions found at 48 repetitions (39 to 84). There was a positive correlation ($r=0.8$) between the size of the expansions and the age onset of motor manifestations. Parkinsonism was a frequent manifestation (79%), moderate to severe in 65% of cases. Chorea was found in 60% of patients, moderate to severe in 50%. **Conclusion:** Although considered a rare disease, an initial survey detected 19 families with Huntington's disease in Amazonas, with great phenotypic and genotypic variability. The continuity of the study should bring comprehensive epidemiological data on the presence of clusters of the disease in the Brazilian Amazon region.

Transtornos do Movimento

TL 1105487

GENETIC ANALYSIS OF A COHORT OF IDIOPATHIC PARKINSON'S DISEASE IN AMAZONAS

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Background: In the last 15 years, studies on Idiopathic Parkinson's Disease (PD) have advanced considerably, gaining knowledge about the genetic forms. However, complete information on the impact of these mutations, especially LRRK2 and GBA, on the clinical aspects of these patients is still lacking. **Objective:** To determine the frequency of LRRK2 and GBA mutations in a population of PD patients in Amazonas and to determine the influence of mutations on the clinical course of patients. **Method:** A cross-sectional cohort study with a multicenter character was performed to analyze the genetic forms of ILD. Blood samples were collected from 57 patients and initially analyzed for mutations in the GBA and LRRK2 genes, and 68 other genes were subsequently analyzed if the first analysis was negative. The clinical data was evaluated, comparing the evolutionary aspects of the disease between carriers and non-carriers of mutations. **Results:** After analyzing 57 patients, 2 cases with a mutation in the GBA gene (3.5%) and 3 cases with LRRK2 mutation (5.2%) were found. For the analysis of clinical data, we had access to 31 patients, of which 2 were carriers of LRRK2 mutation and 1 of GBA mutation. Patients' ages ranged from 32 to 89 years (mean = 58.6 years). Age at onset of symptoms ranged from 27 to 81 years (mean = 51 years). Among patients with the LRRK2 mutation, the mean age at onset of symptoms was 56 years, and in the GBA mutation, it was 49 years. Regarding the cognitive pattern, 19 of the 28 patients without mutations had moderate to severe cognitive decline (2 to 4 in the UPDRS), less intense in LRRK2 carriers (one patient with a score of 1 and a patient with 3) and mild in the patient GBA carrier, with a score of 1. The presence of hallucinations was less frequent in carriers of mutations than in non-carriers, with 1 case in LRRK2 mutation, against 11 cases in non-carriers. Sleep disturbances were more frequent and intense in those without the mutation (26 cases), against none in GBA mutation and one moderate case in LRRK2. **Conclusion:** The genetic analysis of this population showed the presence of 5 mutation carriers in the 57 patients studied (8.7%). In the clinical analysis, patients without mutations showed greater intensity of cognitive complications, hallucinations, and sleep disorders. The small study sample limits the broadening of the results.

Transtornos do Movimento

TL 1105648

ASSESSMENT OF UPPER LIMBS IMPAIRMENTS IN PARKINSON'S DISEASE: A SYSTEMATIC REVIEW

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Background: Progressive reduction of upper limbs functional capacity is common in Parkinson's disease (PD). In addition, upper limb freezing (FOUL) episodes can be very disabling during daily life activities. Only a few tests for assessing upper limbs (UL) in PD are available. And a standard upper limbs functional capacity assessment in PD is unknown. **Objective:** To identify the frequent outcomes measurements to assess UL functional capacity in PD. **Methods:** We reviewed systematically the literature published from February 2010 to February 2021 according to PRISMA guidelines. We analyzed only published studies in English from a systematic review in PubMed, using the following keywords: "upper limbs" OR "upper extremity" and "functional capacity" and "Parkinson's disease"[All Fields]). [MeSH Terms]). This study was registered at PROSPERO CRD 42021254486. **Results:** We found initially a total of 408 studies, 396 in PubMed and 12 in other sources. According to the inclusion and exclusion criteria, 40 studies published in English were included in this review. All quantitative study designs, including systematic reviews published from February 2010 were also included in this review. Studies that don't mention PD and that don't present UL assessments were excluded. No studies were found to assess UL functional capacity in PD. The most common upper limbs outcome measures found in the studies were: (i) to assess severity and progression of PD motor symptoms (tremor and bradykinesia): UPDRS-III (n=17), MDS-UPDRS (n=14), NFOG-Q (n=6), Nine Hole Peg Test (n=5), MOCA (n=4), Purdue Pegboard Test (n=4), Fahn-Tolosa-Marin Tremor Rating Scale (n=2); (ii) to assess manual dexterity: Nine Hole Peg Test (n=5) and Purdue Pegboard Test (n=4); (iii) to provoke and assess FOUL Spiral test (n=2) Funnel test (n=2); (iv) Use of technology such as sensors (n=3), app or digital platform (n=3). **Discussion:** Currently, physiotherapy guidelines for PD provide no standard recommendations to assess UL functional capacity. Only a few studies have shown that it is possible to assess tremors, bradykinesia, and manual dexterity. There is a lack of a standard UL functional capacity assessment. FOUL can be objectively assessed by evaluating the spiral-drawing or funnel tasks, but only a few studies reported these assessments. **Conclusion:** Current evidence is insufficient to recommend standard and effective outcome measures to assess upper limb functional capacity in PD. Further research is needed.

Transtornos do Movimento

TL 1105682

SEXUAL FUNCTION IN WOMEN WITH PARKINSON'S DISEASE

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Background: Impairment of sexual function in Parkinson's Disease (PD) is common and problematic, with many of its physical and mental aspects interrupted throughout the disease. Among the non-motor changes associated with PD, sexual function is less discussed and, consequently, less known. Satisfactory sexual function has been associated with better quality of life. **Objective:** To assess sexual performance and the couple's level of adjustment in women with PD. **Methods:** Forty-seven women with a confirmed diagnosis of idiopathic PD and age ranging from 38 to 68 years, stage 1 – 3 of disease progression according to the Hoehn and Yahr classification, without dementia, participated in the study. After submitting the Free and Informed Consent Term and agreeing to participate in the study, the participants were asked to answer a previously established questionnaire through telephone contact. The following were investigated: (1) general information; (2) socioeconomic status; (3) information associated with the PD; (4) mental and cognitive health; (5) motor and non-motor aspects of daily life experience assessed by sections I and II of the MDS-UPDRS; (6) Sexual quotient – female version (QS-VF); (7) Dyadic Adjustment Scale (DAS). Statistical analysis tested the correlation between total sexual performance scores assessed by QS-VF and DAS. **Results:** There was a statistically significant correlation between sexual performance assessed by QS – VF and DAS mainly for values ($p < 0.00$), behavior ($p < 0.00$), tired for sex ($p < 0.00$) and happiness ($p < 0.01$). **Conclusion:** For women living with PD, the quality of the marital relationship is the main factor for the quality of sexual performance. Therefore, seeking strategies with the objective of intervening and helping couples to achieve or maintain a good marital relationship is of fundamental importance to improve the sexual function/sexual performance of this population.

Transtornos do Movimento

TL 1105731

THERE IS AN ASSOCIATION BETWEEN COGNITIVE DEFICIT AND RELIGIOSITY IN PATIENTS WITH PARKINSON'S DISEASE?

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Background: Cognitive decline makes up the natural history of Parkinson's Disease (PD), especially in advanced stages. In this context, religiosity may be related to the status of cognition and executive functions, such as cognitive flexibility, which are usually also impacted in PD. Thus, it is essential to investigate the degree of cognitive impairment and its possible associations with religiosity. Therefore, the literature confirms the applicability of the MOCA for cognitive assessment even in populations with lower levels of education. **Objectives:** Analyze the degree of cognitive impairment and verify its possible association with religiosity, quality of life and depressive symptoms in patients diagnosed with PD. **Methods:** The MOCA and the Duke University Religiosity Index (DUREL) were applied to patients diagnosed with PD according to the 2015 MDS criteria. Patients with a hypothesis of atypical parkinsonism were excluded. DUREL was evaluated according to its 3 components: organizational religiosity (OR), non-organizational religiosity (RNO) and intrinsic religiosity (IR). Diagnostic of dementia or mild cognitive impairment based on the total test score, according to cutoff points in the literature. In addition, the Beck Depression Inventory (BDI) and the PDQ 39 were applied to assess depression and the degree of quality of life in these patients, respectively. The Spearman test was used to assess the correlation between the variables. This study obtained approval from the local ethics committee approval. **Results:** Twenty-eight patients, aged between 48 and 76 years, were included. Among these, 10 were diagnosed with dementia ($MOCA < 17.5$) and 9 with mild cognitive impairment ($MOCA < 22.5$). Among patients with dementia, 50% did not have any level of education. The Spearman test was calculated and the correlation coefficient of the MOCA with the other evaluated scales was: DUREL – RO and RNO (0.347; $p=0.71$), RI (0.06; $p=0.764$), BDI (0.063; $p=0.748$), PDQ – 39 (-0.063; $p=0.749$) and UPDRS III (-0.241, $p=0.217$). **Conclusion:** Approximately 32% of the sample was classified as having a mild cognitive impairment and 36% as having dementia. No significant association was found between cognition, religiosity, quality of life or depression. There was also no significant association between cognition and motor impairment in these patients.

Transtornos do Movimento

TL 1105870

ASSESSMENT OF QUALITY OF LIFE IN PATIENTS WITH PARKINSON'S DISEASE: A TERTIARY CARE CENTER EXPERIENCE IN NORTHEASTERN BRAZIL

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Background: Motor and non-motor symptoms in Parkinson's Disease (PD) usually progress and cause physical, mental and social impairment, affecting the individual's level of functionality and patients' quality of life (QoL). **Objective:** To evaluate and to describe QoL in PD patients. **Method:** Between May 2021 and January 2022, a cross-sectional study was done at a Movement Disorders clinic in Teresina/PI. QoL assessment was done using the Parkinson's Disease Questionnaire 39 (PDQ39) in Brazilian Portuguese version. We also analyzed medical records to obtain demographics, HOEHN and YAHN (HY) Scale scores and Levodopa Equivalent Daily Dose (LEDD). Patients were categorized according their age range. Data was analyzed with RStudio software. Regression test was performed with disease duration and PDQ-39 domains, considering $p < 0.05$ as significant. **Results:** During the period, data were collected from 93 patients, most of whom were male (63.4%) and aged between 65 and 74 years (41.9%). As for the duration of the disease, most had between 5 and 10 years of evolution (44.1%) and the LEDD was between 350 and 700 (33.3%). The mean LEDD of the general population of our patients was $677.37 (\pm 372.07)$. On the HY Scale, most were between stages 1 and 3 (91.4%). Regarding the PDQ-39, 4 out of the 8 domains had means above 50, and therefore, a worse perception of quality of life, namely Mobility ($71.93/\pm 30.24$), Body discomfort ($70.60/\pm 23.71$), Activity of daily living (ADL) ($62.95/\pm 29.24$) and Emotional well-being ($54.25/\pm 29.63$). The domain with the lowest score in the general population of this study was Social Support ($34.41/\pm 13.91$). When comparing the averages of men and women separately, the domains with the greatest scores were also Mobility, Body Discomfort, ADL and Emotional Well-being. However, in all these domains, women had greater scores when compared to men. In addition, after regression test, there was statistical significance in the Mobility ($p=0.023$), ADL ($p=0.016$) and Communication ($p=0.006$) domains in patients with more than 20 years of disease duration. **Conclusion:** It was noted that both motor and non-motor aspects may be related to a worse perception of QoL by these patients. In addition, other individual characteristics, such as sex, lead to different perspectives on quality of life. Thus, it is necessary to direct integrated care strategies aimed at the support and treatment of people with Parkinson's Disease, taking into account their individualities.

Transtornos do Movimento

TL 1105922

WHY SLOTHS ARE SO SLOTH? IS THE BRADYPUS A PHYSIOLOGICAL PARKINSONIAN ANIMAL MODEL?

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Background: There is no natural animal model for Parkinson's disease, only experimental animal models that do not help in understanding the pathophysiology and natural evolution of the disease. The *Bradypus Variegatus* (Common Sloth) is a tropical forest animal (atlantic forest) that has the unique behavior of slowness and bradykinesia being called a Sloth. To this day, it is not known why it was so slow (including study of physiology, pathology, hormone dosages, etc.). **Objective:** to describe and correlate the characteristics of motor behavior and physiology of the *Bradypus Variegatus* with Parkinson's disease, discussing the possibility of being a physiologically determined "Parkinsonian" animal model. **Method:** data collection based on a literature review on the study of the Common Sloth and its bradykinesia. Animal neuroanatomy, neurophysiology and animal behavior studies, with special attention to data related to Parkinson's disease. **Results:** *Bradypus* has several "Parkinsonian" features. The most common is bradykinesia. However, paradoxical kinesis is also exist (agile when threatened. It has quick strikes with its claw in threats and fights. Hypomimia with reduced blinking and facial expression, smell too. Predominance of flexor tone (flexor muscles are antigravity), with posture of flexor rigidity most of the time. Slow transit colon with a frequency of 1 evacuation per 10 days. Slow responses to painful stimulus. In summary, 1) marked bradykinesia 2) Paradoxical kinesis 3) very slow intestinal transit 4) Sialorrhea 5) facial hypomimia with reduced blinking 6) reduced sense of smell 7) predominance of postural flexor tone with clear flexor hypertonia are common signs of *Bradypus* and also to parkinsonian patients. These characteristics are useful in their animal lifestyle (they lives in trees) and this "hypokinetic-hypertonic extrapyramidal status" favors them to remain in the trees, even during sleep, without fatigue. **Conclusion:** This is the first description in the literature that correlates *Bradypus* with Parkinson's disease and places it as the first natural animal parkinson's model, not from a pathological point of view, but with a natural and successful "Parkinsonian status".

Transtornos do Movimento

TL 1106061

THE NEURAL BASIS OF FACIAL EMOTION RECOGNITION IN PROGRESSIVE SUPRANUCLEAR PALSY: A NEUROIMAGING STUDY

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Background: Deficits in facial emotion recognition have been described in Progressive Supranuclear Palsy (PSP). However, the neural basis of these deficits remains uncertain. **Objectives:** To investigate the correlations between the Facial Emotion Recognition Test (FERT) and measures of cortical and subcortical volume in patients with PSP. **Methods:** 21 patients with PSP (age = 68.0 ± 7.4 years) and 20 healthy controls (age = 67.5 ± 10.8 years) underwent cognitive assessment, including the FERT, which is composed of 35 images from Ekman's portfolio, with 7 different emotions (happiness, sadness, fear, disgust, surprise, anger, and neutral). All participants underwent brain MRI (3T). We conducted two neuroimaging analyses: 1) subcortical structures were segmented with Freesurfer software and volumes were extracted for analyses. We explored correlations (Spearman's test) between subcortical nuclei volumes (putamen, caudate, amygdala; corrected for total intracranial volume) and FERT scores in PSP patients; 2) voxel-based morphometry (VBM) was performed with FSL software, exploring correlations between gray matter density and FERT scores within the PSP group. Age, sex and disease duration were considered as covariates. A threshold of 50 contiguous voxels was adopted, corrected at family-wise error (FWE) at the $p < 0.05$ threshold, for all analyses. **Results:** PSP and controls did not differ on age and schooling. PSP patients performed worse than controls ($p < 0.001$) on the FERT (total score, disgust, anger, sadness and neutral). Compared to controls, PSP had lower volumes of brainstem ($p < 0.001$), caudate ($p < 0.001$), putamen ($p < 0.001$), and frontal atrophy. Within PSP patients, surprise score correlated to right ($p < 0.001$, $\rho = 0.72$) and left ($p < 0.003$, $\rho = 0.66$) amygdala; disgust correlated to right ($p < 0.02$, $\rho = 0.54$) and left ($p < 0.03$, $\rho = 0.52$) amygdala; neutral score correlated to right ($p < 0.01$, $\rho = 0.55$) and left ($p < 0.0001$, $\rho = 0.74$) putamen and right ($p < 0.008$, $\rho = 0.6$) and left ($p < 0.001$, $\rho = 0.7$) amygdala. VBM analyses yielded significant direct correlations between frontal pole and scores of disgust, sadness, anger, and surprise. Disgust also directly correlated with insula. **Conclusion:** Subcortical nuclei and fronto-insular regions play a role in the recognition of facial emotions in PSP, suggesting that a complex fronto-subcortical network participate in emotional processing in the disease.

Transtornos do Movimento

TL 1106129

THE EFFICACY OF DEEP BRAIN STIMULATION IN THE SUBTHALAMIC NUCLEUS IN ISOLATED DYSTONIA

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Background: DBS is a safe and effective therapy for patients with isolated (inherited or idiopathic) and medically refractory dystonia in both generalized and severe focal and segmental dystonia. Subthalamic nucleus DBS for dystonia publications has been limited to small series and case presentation. Therefore, the objective of this systematic review was to provide clinical evidence of the efficacy of DBS in the STN in isolated dystonia. **Methods:** For this article, we performed a comprehensive search in databases for articles published from 1987 to 2022. We included patients with primary dystonia, focal, segmental, or generalized, who underwent DBS placement in STN and had no prior brain lesioning or brain surgeries. Data collected included demographics, clinical features of dystonia, adverse events, stimulation parameters, and quality of life scales. **Results:** The literature search resulting in the identification of 20 relevant articles corresponding to 14 distinct studies and reporting 114 cases. There were 75 women (65,7%) and 39 men. The mean age of onset was 37.2 ± 19.1 years (\pm SD) and the duration of dystonia was 9.1 ± 8.8 years (\pm SD). They were 40 focal dystonia, 44 segmental dystonia, and 30 generalized dystonia. The mean follow-up time was 12.8 months (range 3-33 months). The BFMDRS movements and TWSTRS scores were significantly lower in post-operative values ($p < 0.0001$), with a mean decrease of 55.3% (CI 48.97-61.6). In total, 20 patients experience an improvement of more than 80% over the preoperative score. Subitem analysis revealed a 61.4% score for focal dystonia, 47.8% in segmental dystonia, and 57.5% in generalized dystonia. The BFMDRS disability scale scores had also improvement ($p < 0.0001$), with mean reduction of 43.9% ($p = 0.0057$). The quality of life was assessed in only five studies using SF-36 scores. The patients' quality of life was improved significantly ($p < 0.0001$) in every subitem of the scale. **Discussion:** This meta-analysis represents the current data on DBS in STN in primary dystonia. Our findings support sustained improvement in motor function, measured by absolute and percentage changes in the BFMDRS and TWSTRS scores from baseline. We also observed an improved ability to perform activities of daily living and quality of life access through BFMDRS disability and SF-36 score.

Transtornos do Movimento

TL 1106190

SOCIAL COGNITION IN PARKINSON'S DISEASE AND SUPRANUCLEAR PARALYSIS: A NEUROPSYCHOLOGICAL INVESTIGATION

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Background: Patients with Parkinson's disease (PD) and Progressive Supranuclear Palsy (PSP) have deficits in mentalizing and facial emotion recognition, two core functions of social cognition (SC). There is scarce data regarding the direct comparison between PD and PSP patients in SC measures. **Method:** We included three groups of participants: PD (n=18, mean age = 65.2±9.8 years, 9 men), PSP (n=20, mean age = 67.7±6.8 years, 8 men) and healthy controls (n=23, mean age = 64.7±10 years, 8 men). Groups were matched for age, sex and education. Participants underwent clinical and neuropsychological exams, including the Mini-Mental State Exam (MMSE), executive tests (Frontal Assessment Battery [FAB], spans, phonemic fluency, Hayling) and the mini-version of the Social and Emotional Assessment (Mini-SEA), which is composed of the facial emotion recognition test (FERT) and the modified faux-pas (mFP) test, which assesses mentalizing. Multivariate analyses considered executive performance and apathy as covariates. **Results:** Compared to controls, PD and PSP patients had lower cognitive efficiency (MMSE). PD and PSP groups did not differ on the MMSE. PSP had lower score than PD patients and controls in the FAB. PD patients scored lower than controls in the mFP, but not in the FERT. PSP patients performed worse than controls in the mFP and in the FERT. PD and PSP groups did not differ in the FERT, but PSP performed worse than PD in the mFP test. These differences were maintained when controlling for executive dysfunction and apathy. The FERT and the mFP distinguished PSP from PD patients with 89% and 67% accuracies, respectively. **Conclusion:** The assessment of SC may contribute to the differential diagnosis between PD and PSP.

Transtornos do Movimento

TL 1106217

THE EFFECTS OF HIGH-SPEED RESISTANCE TRAINING IN PARKINSON'S DISEASE PATIENTS ON MOBILITY: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background: The loss of strength in PD patients has an impact on their quality of life. Common activities like getting up a chair, as well as all daily life activities become difficult. Moreover, muscle weakness, postural deficits and decreased mobility have been related to increased risk of falls in PD patients. **Objective:** The aim of this systematic review and meta-analysis is to compare the efficacy of high-speed resistance training (HSRT) versus control or another form of physical activity intervention to improve mobility in Parkinson's Disease (PD) patients. **Data sources:** We performed a systematic literature search through electronic databases including MEDLINE via PubMed, LILACS via BVS, Embase via Elsevier and PEDro databases up to September 2021. **Methods:** We included randomized clinical trials with patients with PD, HSRT as the intervention, compared to a control group or another type of physical activity. Standardized mean difference (SMD) with 95% confidence interval (CI) were calculated based on the inverse of variance method. **Results:** The literature search identified a total of 7631 results among which 4 studies were selected for the meta-analysis presenting moderate to high methods quality. The meta-analysis showed a significant difference on mobility outcome in favor of HSRT compared to a control group (SMD = 0.42; 95%CI [0.00;0.83], p= 0.049). **Conclusion:** HSRT may improve mobility in PD patients when compared to a control or minimal intervention group. HSRT seems to be a good approach for resistance training to enhance mobility in PD individuals.

Transtornos do Movimento

TL 1106248

MOVEMENT DISORDERS AFTER ACUTE STROKE: A LARGE PROSPECTIVE COHORT STROKE STUDY

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Background: Post-stroke movement disorders (PSMD) can occur either acutely or in the first year following a stroke. Abnormal movements after a stroke may be either hypo or hyperkinetic, but involuntary hyperkinetic movements are more frequently observed during stroke follow-up. PSMD is uncommon, with a known prevalence of about 1% and an incidence of 0.08% per year, highlighting the rarity of PSMD. Most basal ganglia and thalamic lesions did not result in movement disorders, suggesting that stroke location may not predict PSMD. Also, PSMD may be associated with both ischemic and hemorrhagic strokes. Few studies concern PSMD prevalence in a designed prospective cohort. **Objective:** We aimed at stating the prevalence of acute post-stroke movement disorders (PSMD) in a large prospective cohort of acute stroke patients. **Methods:** We designed an open, prospective, one-year, single-center stroke cohort to follow all acute cerebrovascular disorders searching for any movement disorders during Hospitalization and after discharge. PSMD was the primary outcome, and death was the main competitive outcome. Our study included all acute stroke patients admitted to the Hospital from March 2021 to February 2022. Besides the demographic data, we captured clinical and neurological data: NIH stroke scale, modified Rankin scale, Glasgow coma scale, and abnormal involuntary movement scale. We also evaluated the first CT scan of all patients for stroke localization and type determinations. **Results:** We included 858 acute stroke patients. There were four acute PSMD patients in this cohort, yielding a prevalence of 0.5%. Table-1 discloses all demographic data. In all PSMD cases, the stroke was ischemic. Two patients revealed thalamic lesions (2.4% of thalamic stroke patients), and two had striatal ischemic strokes (2.1% of striatal stroke patients). Movement disorder occurred in the first 24h after ischemia in all cases. Concerning the abnormal movement phenotype, all were hyperkinetic. One case disclosed acute tremor after striatal ischemia, and in the other patients, dystonia was the prevalent movement disorder. **Conclusion:** To sum up, this large cohort confirms that PSMD in acute stroke is rare, with an established prevalence of 0.5% for all stroke events. PSMD was associated with striatal and thalamic ischemic stroke, and all cases presented acute hyperkinetic movement disorder.

Transtornos do Movimento

TL 1106331

INTERLEAVED STIMULATION FOR REFRACTORY TREMOR IN PD: A SERIES CASE

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Background: Interleaved stimulation (IS) in DBS provides individualized energy for two contacts alternately. Most documented indication concerns the management for levodopa induced dyskinesias. On the other hand, it is well acknowledged that refractory PD tremor responds better to high frequency DBS, although with adverse events on balance and speech. Animal studies demonstrate that coordinated reset stimulation of the subthalamic nucleus presents short and long-term improvement in the parkinsonian motor condition, with lower energy expenditure and lower incidence of complications. We hypothesized if IS, as a quasi-coordinated reset mode, would control tremor and avoid complications. **Objective:** To test IS would improve tremor without causing adverse effects. **Methods:** We evaluated 5 tremor-dominant Parkinson's disease patients, on medication and previously stimulated in a basic pattern, all at high voltage and frequency. All had DBS complication due to high total energy delivered, like balance and dysarthrophonia, and pain. We modified the stimulation to the interleaved pattern, using the contacts that the patients already had before. Data analyzed included Unified PD Rating Scale III and tremor score. **Results:** There were 4 male and 1 female, with mean age of 59. Mean baseline UPDRS was 60,6 and mean tremor score was 3,5. All 5 patients showed satisfactory improvement in parkinsonian symptoms and relief from stimulation-induced side effects after interleaved stimulation, established by a large effect size (Cohen d) in UPDRS III (d=3,71285) and tremor score (d=1,543766). All patients were free of complications after changing the stimulation mode to IS. **Discussion:** The use of the interleaved mode is known to improve dyskinesias, through stimulating pallid-fugal fibers in the Zona Incerta, and sensory-motor fibers in the laterodorsal zone of the subthalamic nucleus. Requires frequency reduction, so it would be a bad alternative for the tremor-dominant patient, as they tend respond to high frequencies. On the other hand, the high frequency leads to adverse effects such as hypophonia, dysarthrophonia and worsening of balance. **Conclusion:** With this study, we demonstrated a substantial improvement in the motor condition of these patients, in addition to an improvement in side effects.

Transtornos do Movimento

TL 1106333

CLINICAL ASPECTS OF SPINOCEREBELLAR ATAXIA IN THE STATE OF AMAZONAS

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Background: Spinocerebellar ataxias (SCAs) are a group of neurodegenerative hereditary diseases, with an autosomal dominant pattern. The prevalence of SCAs subtypes is variable among geographic regions and ethnicities. In Brazil, the occurrence of the main subtypes was studied mainly in the Southern and Southeastern regions. However, considering the ethnic diversity among Brazilian regions, those findings cannot be generalized. Thus, this study aimed to investigate and clinically characterize cases of SCAs in the state of Amazonas. **Method:** Research participants were recruited at the reference center for movement disorders in Manaus-AM. Genetic tests for SCAs 2, 3, 6, and 7 were performed. **Results:** Of 21 participants, nine were genetically confirmed: two families (N=5) were diagnosed with SCA 3; one family (N=3), with SCA 2; and one patient with SCA 7. In one of the SCA 3 families, symptoms were predominantly cerebellar: gait ataxia, dysarthria, hypotonia, and nystagmus. Pyramidal signs (hyperreflexia) and peripheral neuropathy were also observed. In the other SCA 3 family, cerebellar signs of gait ataxia and dysarthria were associated with peripheral neuropathy, cognitive dysfunctions (memory loss), and psychiatric disorders (irritability and depression). The SCA 2 family was clinically characterized by ataxia (more severe gait, but also appendicular and axial), movement decomposition, and dysarthria in association with slow saccades and mild bradykinesia. Lastly, the patient diagnosed with SCA 7 showed a condition of mild cerebellar signs (gait ataxia, hypermetria, and dysarthria) and predominant symptoms of loss of visual acuity, ophthalmoparesis, bilateral ptosis, and facial hypomimia. Pyramidal signs (hyperreflexia), distal atrophy, and tongue fasciculations were also observed. **Conclusion:** The research demonstrated the presence of families with SCA2, SCA3 and SCA7 in Amazonas. New data will be added with testing for SCA10. So far, we have been able to detect a low incidence of families with SCAs in the state of Amazonas, compared to other Brazilian studies.

Transtornos do Movimento

TL 1106417

ASSOCIATION BETWEEN BALANCE PERFORMANCE AND COGNITIVE IMPAIRMENT IN PEOPLE WITH PARKINSON'S DISEASE

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Background: People with Parkinson's Disease (PD) often use cognitive strategies to maintain balance and postural stability due to their deficits in terms of automaticity, among the motor symptoms presented in the disease, postural instability is one of the most disabling and seems to be related to cognitive deficits. **Aim:** In this study, we determined the cognitive factors that may be related to balance deficits in individuals with PD. **Methods:** A sample of 74 participants, recruited in a Parkinson's association in São Paulo, being 22 healthy elderly (CG) and 52 people with Parkinson's, subdivided into early (IPG) and moderate (MPG) Parkinson's was evaluated based on variables clinical and social, among the variables evaluated, the groups were homogeneous in terms of age (p=0.14), education (p=0.21). The groups were different in terms of motor presentation measured from the MDS-Unified Parkinson's Disease Rating Scale Part II and III. For balance evaluation, the Balance Evaluation Systems Test (BESTest) was used in its complete format, evaluating domains such as Biomechanics, Stability Limits, Postural Responses, Anticipatory Postural Adjustments, Sensory Orientation and Dynamic Balance during gait and cognitive effects and for the cognitive impairment, the Montreal Cognitive Assessment (MoCA), a brief screening instrument for mild cognitive impairment. It accesses different cognitive domains: attention and concentration, executive functions, memory, language, visual-constructive skills, conceptualization, calculation and orientation. This study was approved by a Local Ethics Committee (CAAE 67388816.2.0000.0065). **Results:** MoCA data, especially when comparing the MPG group with the others, were different due to the presence of some cognitive deficit with the progression of the disease (p=0.031). A positive correlation was demonstrated between the total values of MoCA and the Best TEST (p<0.009; r=-.3047), as in specific domains related to visuospatial memory and visuospatial orientation (p<0.001; r=-.3786), evaluated respectively. **Conclusion:** Visuospatial memory is important in everyday activities that require location of the individual in space, differentiation of shape and spatial location of objects. Previous studies indicate that visuospatial memory is related to gait rhythm and postural control in healthy elderly people and those with PD. Thus, deficits in visuospatial memory are related to a higher risk of falls in this group of people.

Transtornos do Movimento

TL 1106574

PREVALENCE OF PSYCHOSIS IN PARKINSON'S DISEASE – PRELIMINARY RESULTS OF A CROSS-SECTIONAL STUDY

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Background: Parkinson's disease (PD) is the second most common neurodegenerative disease. Its prevalence is close to 1% in the population over 60y and its incidence ranges from 8-18: 100,000 person-years. Its etiology is multifactorial, with the main risk factor being aging. Although known for its motor symptoms, it encompasses a variety of non-motor symptoms such as psychosis, whose pathophysiology involves a combination of endogenous (disease/patient) and exogenous (treatment) factors. There is still no direct association between dopaminergic treatment and psychosis, but it is believed that it may be triggered or "accelerated" by it. Its management involves the gradual withdrawal of contributing drugs and the background of antipsychotics if necessary. Estimates of the prevalence of PDP vary widely in the literature. **Objective:** To describe the prevalence of Parkinson's disease Psychosis (PDP) and investigate patients' sociodemographic and clinical profile. **Methods:** Ongoing descriptive, observational, cross-sectional study conducted in a Movement Disorders Outpatient Clinic in Salvador, Bahia. We included patients over 18y diagnosed with PD by the UK Brain Bank Criteria and excluded patients with psychiatric comorbidities. The study was approved by the local Ethics Committee. Data gathering involved questionnaires for clinical and sociodemographic evaluation. The NINDS/NIHM criteria, validated by the MDS, was used to diagnose PDP. The modified Hoehn & Yahr Scale (mH&Y) was used to stage the disease's progression and the MMSE for cognitive assessment. **Results:** Until now, the sample size has 23 patients. The median age was 63y (IQR 52-72), with 65.2% being male, 39.1% self-reported as mixed-race and 52.2% were married. Regarding education, 43.5% had completed high school, and the median MMSE was 28 (IQR 21-29). High Blood Pressure was the most common comorbidity (43.5%). The mean time of disease was 9.3±4.5 years and 34.8% had mH&Y of 2, while only 4.3% had mH&Y of 5. All patients were on levodopa therapy, 34.8% on dopamine agonist, 26.1% on amantadine, 21.7% on COMTi and 8.7% on MAO-Bi. The prevalence of psychosis in the sample studied is 8.7% (n=2). **Conclusion:** It is concluded that the prevalence of PDP in the studied sample so far is low, but corroborates with the current literature. We emphasize the importance of investigating non-motor symptoms in PD, often neglected, as evidenced by the low number of studies on PDP, especially in Brazil.

Transtornos do Movimento

TL 1106626

PARKINSONISM FOLLOWING COVID-19 IN A LONG-COVID COHORT – CASE SERIES REPORT

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Background: Even though SARS-CoV-2's role in causing Parkinson's disease (PD) has not yet been confirmed, recent studies describe a possible association between COVID-19 and movement disorders. SARS-CoV-2 invasion of vasculature in the brain may cause multiple outcomes through inflammatory responses that could lead to protein misfolding, a characteristic of neurodegenerative disorders. **Objective:** This study describes three patients who started parkinsonian symptoms after confirmed diagnosis of COVID-19 infection. **Methods:** We evaluated a prospective cohort study with patients followed in a post-COVID neurological outpatient clinic, recruited based on positive COVID-19 PCR and post-COVID neurological symptoms. The total sample consisted of 221 patients recruited for the study. Three had a diagnosis compatible with Parkinson's disease. **Results:** The first patient, a 44 years-old-female developed mild flu-like symptoms lasting 6 days. One month later, she describes an asymmetric rest tremor. Neurological exam shows facial hypomimia and cogwheel stiffness with bradykinesia in the left upper limb. The second patient, a 57 years-old-man, reported mild flu-like symptoms with anosmia lasting 7 days. The symptoms progressive after 10 days with difficult to count money at his work and difficult walking. On examination he had mild hyposmia and mild bradykinesia with stiffness in the left upper limb. The third patient, a 56 years-old-female had a history of antiphospholipid syndrome. She reported flu-like symptoms lasting 4 days. After 2 months started a rest tremor. Neurological exam disclosed mild cogwheel stiffness with bradykinesia. All the three patients were responsive to levodopa / benzeraside. **Discussion:** Since the beginning of the pandemic, several neurological manifestations have been described associated with COVID-19. Parkinsonism associated with viral infections has been described with other viruses. Historically, several cases of post-encephalitic parkinsonism were described after the Spanish flu in 1918. The cause of parkinsonism in the present cases has some hypotheses, among which direct viral infection with neurodegeneration, inflammatory mechanism, or multiple hits. **Conclusion:** Available data does not confirm a clear association between the Covid-19 infection and PD. Parkinsonian symptoms after COVID-19 infection seems to be a rare condition. We propose a possible association between Covid-19 and the development of PD.

Transtornos do Movimento

TL 1104816

PRELIMINAR STUDY FOR NEUROVISUAL ASSESMENT MODEL USING EYE TRACKING (NAMET) RELATED TO COGNITIVE FUNCTIONS FOR TBI PATIENTS

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Neuro Visual Assessment Model for visual and neuro cognitive rehabilitation following TBI can include Visual Acuity (VA), Visual Field, eye movements (Eye Tracking), Accommodation and Convergence. The main objective of the study is to present a standardized NAMET in patients who suffered traumatic brain injury (TBI) and are in the cognitive rehabilitation process. The main symptoms found were: Binocular Visual Deficit (80%) Photophobia (75%) Accommodation and Convergence Problems (70%) Diplopia (50%) Reading Deficit (50%) Oculomotor Deficits (80%) Visual Field (20%) NAMET detects alterations in saccadic eye movements, pursuits and fixation not visible in subjective tests without the use of the Eye Tracking Device. Some of the visual dysfunctions found could be carried out with the help of prisms, lenses and filters, improving the patients' time reading, oculomotricity, balance and posture performance. This improvement in the visual performance of post-TBI patients helps in the cognitive rehabilitation process with other professionals in a transdisciplinary team.

Traumatisismo cranioencefálico

TL 1105834

QUANTITATIVE ANALYSIS OF CONSERVATIVE TREATMENT FOR TRAUMATIC BRAIN INJURY IN BRAZIL

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Background: Traumatic brain injury (TBI) is a structural injury of the skull, brain or associated structures. Conservative treatment involves the use of drugs and a non-surgical approach. The present study compares the number of Hospitalizations, the total cost and the average length of stay between conservative treatments for mild, moderate and severe TBI in 10 years in Brazil through DATASUS. **Methods:** It is a cross-sectional epidemiological study with values obtained from the DATASUS database from December 2011 to November 2021. Data of conservative treatment of mild, moderate and severe traumatic brain injury were selected and grouped into Excel for the number of Hospitalizations, the average length of stay and total value. **Results:** There were 427,916 admissions for mild conservative treatment, 290,130 for moderate TBI and 112,141 for severe. As for the Hospital cost, conservative treatment for mild TBI had a total value of \$35,7340.07, moderate \$61,552,281, and severe \$64,395,813. The mean stay in the health care institution was 2.8 days for mild TBI, seven days for moderate TBI, and 11.5 days for severe TBI. **Conclusion:** Although conservative treatment for mild TBI was more frequent (51.5%), severe TBI had a higher Hospital cost (39.8%), justified by the higher Hospital stay and greater demand for supplies to maintain the clinical status. The mean length of Hospital stay and total expenses were increasing concerning the severity of the trauma.

Traumatismo cranioencefálico

TL 1105857

DECOMPRESSIVE CRANIECTOMY TECHNIQUES AND THEIR OUTCOMES IN REDUCING INTRACRANIAL HYPERTENSION

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Background: Decompressive craniectomy is a procedure that removes part of the skull and the underlying dura to reduce brain swelling-related intracranial pressure (ICP). The present study compares four DC techniques and their outcomes in ICP. **Methods:** We searched by PUBMED in November 2021, using the Mesh Terms "Decompressive Craniectomy AND Intracranial Pressure". Exclusion criteria were studies that did not mention positive and negative points of the surgery. Novel studies in English about decompressive craniectomy techniques published in the last ten years were inclusion criteria. **Results:** Classic DC is the most commonly performed operation for patients that require cerebral decompression after suffering a traumatic brain injury (TBI) or stroke. Window DC proved to be an effective alternative to classic DC in the process of high ICP after severe TBI, showing results in reducing ICP and improving ECG scores. In addition, it offers a better aesthetic result and prevents the skin flap from sinking into the incision cavity and a second bone replacement surgery. Bifrontal DC allow better visualization and exploration of the brain, a technique used in patients with a very high risk of mortality due to cerebral oedema secondary to trauma. Modified decompressive hemicraniectomy access the middle cerebral artery territory and, in general, the described technique allows lower postoperative risk. **Conclusion:** All of the techniques presented in this article reduce the ICP, but they have different esthetic and postoperative outcomes. The choice of the best procedure will depend on what causes intracranial hypertension and the patient condition.

Traumatismo cranioencefálico

TL 1106282

DIGITAL BIOMARKER-BASED INDIVIDUALIZED ASSESSMENT IN PROFESSIONAL BOXING AND MMA ATHLETES: A DIGITAL NEURO SIGNATURE IN ASSESSING SPORTS-RELATED TRAUMATIC BRAIN INJURY

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Background: Current computerized neuropsychological assessment (NPA) batteries for athletes at risk of sports concussion (SC) are inadequate proxies for traditional neuropsychological tests and have limited clinical utility in postconcussion assessments. Therefore, it becomes necessary research for new and supplementary methods of NPA on SC. An active digital biomarker-based tool is the Altoida Digital Neuro Signature (ADNS), an augmented-reality based test. A previous trial demonstrated 94% prognostic accuracy to measure dementia progression on patients with Mild Cognitive Disorder (MCI). **Objective:** Describe a case series of 15 professional fighters who underwent NPA by the ADNS and correlate worse performance with higher burden of subconcussive traumatic events. Study design: Case series; Level of evidence, 4. **Methods:** The study participants were 15 professional fighters who underwent NPA by ADNS. The mean age was 26.7 years. The higher number of head blows were estimated based on the longer exposure to sparring sessions and conventional training. The sparring hours practiced, measured in hours based on the last month, varied from 1 to 16 hours. The conventional training, measured in hours based on the last week, varied from 5 to 30 hours. Performance measures were computed using the assessment system software (ALTOIDA INC). All significant values reported for correlation between a higher burden of subconcussive events and the ADNS performance were obtained by the Spearman rank, which estimated the coefficient rho (?) of the respective analyzed measures. **Results:** Subjects performed a series of tests, determined by the ADNS. 8 athletes (53.3%) performed worse on tests that assessed planning skills and cognitive processing speed, despite not reporting complaints in the initial interview. Statistical analysis showed correlation between higher number of subconcussive events and poor performance on ADNS, mainly on tests that assessed cognitive processing speed (? = -.689, p < 0,01), and planning skills (? = -.644, p < 0,01). We also observed that 9 athletes (60%) met diagnostic criteria for diseases associated with sports practice. **Conclusion:** These results show linear correlation between higher exposure to subconcussive events and worse performance on ADNS. In summary, this study adds to the literature describing a new neurocognitive assessment method called Altoida DNS, on assessing cognitive decline at professional fighters.

Traumatismo cranioencefálico



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JUNIOR, Daniel Vicente de Siqueira Lima	TL 1106439, TL 1106633, TL 1106294, TL 1105453, TL 1106603, TL 1105778, TL 1106549	JÚNIOR, Luiz José Malta Gaia Ferreira	TL 1105536
JUNIOR, Euldes Mendes	TL 1104712	JÚNIOR, Luiz Severo Bem	TL 1105895, TL 1105182
JUNIOR, Fabio Antonio Serra de Lima	TL 1105848	JÚNIOR, Marcondes Cavalcante França	TL 1105564
JUNIOR, Hermes Castro Araujo	TL 1105657	JÚNIOR, Miguel Cândido da Costa	TL 1105902
JUNIOR, Ivo Knut Anders	TL 1105347	JÚNIOR, Mário Emílio Teixeira Dourado	TL 1106412, TL 1106062, TL 1106384, TL 1106426, TL 1106133
JUNIOR, Jorge Alberto Martins Pentiado	TL 1105133, TL 1105083, TL 1105366, TL 1105372	JÚNIOR, Mário Emílio Teixeira Dourado	TL 1106412, TL 1106062, TL 1106384, TL 1106426, TL 1106133
JUNIOR, Jose Wagner Leonel Tavares	TL 1106604	JÚNIOR, Mário Luciano Mélo Silva	TL 1106523
JUNIOR, José Wagner Leonel Tavares	TL 1105945	JÚNIOR, Pedro Helder de Oliveira	TL 1106581
JUNIOR, Laerte Honorato Borges	TL 1105601	JÚNIOR, Pedro José da Silva	TL 1106507
JUNIOR, Luiz Dalfior	TL 1106411, TL 1106424	JÚNIOR, Renato Campos Freire	TL 1105519
JUNIOR, Luiz Severo Bem	TL 1105857	JÚNIOR, Renato C F	TL 1106440, TL 1106490, TL 1106503
JUNIOR, Marcondes Cavalcante França	TL 1106363, TL 1105166, TL 1106408	JÚNIOR, Renato Pereira da Costa	TL 1106126
JUNIOR, Mario Emilio Teixeira Dourado	TL 1105320	JÚNIOR, Roberto Santos de Oliveira	TL 1106040
JUNIOR, Mario Luciano Melo Silva	TL 1106492	JÚNIOR, Sergio Roberto Pereira da Silva	TL 1105309
JUNIOR, Oscar Nelson Reimann	TL 1105123, TL 1105165, TL 1105329	JÚNIOR, Sérgio Roberto Pereira da Silva	TL 1105253
JUNIOR, Oscar Nelson Reimman	TL 1105116, TL 1105134	JÚNIOR, Tales de Oliveira	TL 1105445, TL 1105638, TL 1105656, TL 1105005
JUNIOR, Pedro Helder de Oliveira	TL 1106177, TL 1105405, TL 1106335, TL 1106010, TL 1105949	JÚNIOR, Tales de Oliveira	TL 1105445, TL 1105638, TL 1105656, TL 1105005
JUNIOR, Pedro Helder Oliveira	TL 1106125	JÚNIOR, Valdecir Boeno Spenazato	TL 1106203, TL 1106209
JUNIOR, Roberto Carlos Sousa Alves	TL 1106492	JÚNIOR, Wilson Alves de Oliveira	TL 1106208
JUNIOR, Sandro Adriano Souza Lima	TL 1105885, TL 1105471, TL 1105487	JÚNIOR, Wilson Marques	TL 1104755, TL 1105342, TL 1106404, TL 1105540, TL 1105547, TL 1105314, TL 1105307, TL 1105509
JUNIOR, Silas Fernandes Avelar	TL 1106155	JÚNIOR, Álvaro Alves de Sá	TL 1106214

KAIMEN-MACIEL, Damacio Ramon	TL 1105844, TL 1105795, TL 1105771, TL 1105831	LAZERA, Vitória de Mendes Sá	TL 1106442
KAMADA, Daniela Midori	TL 1105235	LEAL, André Giacomelli	TL 1105459, TL 1105466
KANAWA, Gleica Maria de Macena	TL 1105565	LEAL, Leonardo Barbosa	TL 1106531
KANEMATSU, Maiara	TL 1105031	LEAL, Lucas Luan Gonçalves Barros	TL 1105363, TL 1105373
KAUARK, Roberta Borges Gomes	TL 1105447, TL 1105665	LEAL, Patricia Provenzano	TL 1106306
KAY, Cláudia Suemi Kamoi	TL 1106172	LEAL, Patrick Ramon dos Santos	TL 1106545
KERCHNER, Geoffrey A	TL 1105660	LEAL, Paulo Roberto Lacerda	TL 1106587, TL 1106589, TL 1105142, TL 1105272, TL 1105907, TL 1105900, TL 1105136, TL 1105419, TL 1106264
KERSTING, Nathalia	TL 1104685	LEANDRO, Cícera Simplicio Machado	TL 1105066, TL 1105114
KIM, Ho Jin	TL 1105040	LEDA, Jóshua Matheus de Sousa	TL 1105639
KIM, Kevin	TL 1104957	LEDERMAN, Alex	TL 1106516
KIM, Lenise Jihe	TL 1106100	LEE, Annelise Akemi Higa	TL 1106516, TL 1106610, TL 1106576, TL 1106406, TL 1106637
KINJO, Juliana Yumi	TL 1105031	LEE, Stefanie Hitomi de Nishi	TL 1105031
KLEIN, Gregory	TL 1105660	LEITE, Amanda Nassur Corrêa	TL 1105347
KLOEPEL, Manuela Aparecida	TL 1105062	LEITE, Bruno de Magalhães Barbosa	TL 1106596, TL 1106613, TL 1106573
KNOCHENHAUER, André Enoch	TL 1105122, TL 1105124	LEITE, Camila Ferreira	TL 1106273, TL 1106586, TL 1106653
KNORST, Gabriel Rocha Santos	TL 1105689	LEITE, Christian Max Uchôa	TL 1104780, TL 1105914, TL 1106197, TL 1106216, TL 1106219
KOFF, Marco Antonio Eduardo	TL 1105820	LEITE, Elder Machado	TL 1105882, TL 1105899, TL 1106549
KOFF, Marco Antonio	TL 1105183, TL 1105407	LEITE, Filipe Augusto Silva	TL 1106479, TL 1106556
KOGA, César Minoru Toita	TL 1106480	LEITE, Gerlânia de Oliveira	TL 1105164
KOSMINSKY, Ellen	TL 1106239	LEITE, Gleif Farias	TL 1105361
KOUYOUMDJIAN, João Aris	TL 1105190, TL 1105176	LEITE, Heloisa Ferraz Troijo	TL 1106329
KOUYOUMDJIAN, João A	TL 1104679	LEITE, João Marcelo Borba	TL 1106329, TL 1106206
KOWACS, Fernando	TL 1105617	LEITE, Juliana Magalhães	TL 1105693, TL 1106294, TL 1106603, TL 1105778
KRONBAUER, Lorena Fernandes	TL 1105505, TL 1105606, TL 1105618, TL 1106283, TL 1105171, TL 1105541, TL 1105844, TL 1105538, TL 1105795, TL 1105367, TL 1105771, TL 1105831	LEITE, Luis Felipe Magalhães Serne	TL 1106382
KRONBAUER, Lorena Ferndandes	TL 1106123	LEITE, Marco Antonio Araujo	TL 1105878
KRUEGER, Mariana Braatz	TL 1106091, TL 1106649	LEITE, Marjorie Bindá	TL 1105393
KRUG, Bárbara	TL 1106110	LEITE, Nicolas Fonteles	TL 1106273
KULIKOWSKI, Leslie Domenici	TL 1106175, TL 1106189	LEITE, Patrícia Lúcia Silva Sampaio	TL 1105070
KUS, William Pegoraro	TL 1106522	LEITÃO, Antônio Miguel Furtado	TL 1105780
KUSTER, Gustavo	TL 1106287, TL 1106319, TL 1106139, TL 1106307, TL 1106360	LEITÃO, Déborah Araújo	TL 1105937
LACERDA, Danielle	TL 1106580	LEMOS, Ana Flavia Andrade	TL 1105157
LACERDA, Eliza Maria da Costa Brito	TL 1105812	LEMOS, Ana Flávia Andrade	TL 1106413, TL 1106448, TL 1105111
LACERDA, Rebeca Montenegro de	TL 1105148	LEMOS, Andreza Salvio	TL 1105570, TL 1105668, TL 1105757, TL 1105269, TL 1105607, TL 1105530, TL 1105782, TL 1105665
LACERDA, Rebeca Montenegro	TL 1105326, TL 1105475	LEMOS, Isadora Versiane	TL 1105897
LACERDA, Roberta Ismael	TL 1104860, TL 1105285, TL 1104874, TL 1105428	LEMOS, Isadora Versiani de	TL 1105869, TL 1105710
LACERDA, Saulo Pimenta	TL 1106085	LEMOS, Isadora Versiani	TL 1105462
LAIS, Lucia Leite	TL 1106062	LEMOS, Nilson Batista	TL 1105895, TL 1105182
LAMAS, Gustavo Manginelli	TL 1105750, TL 1105635, TL 1105863, TL 1105296, TL 1105166, TL 1105630	LEOCÁDIO, Jéssica de Cássia Marques	TL 1106124
LANCELOTTI, Carmen Lucia Penteado	TL 1105793	LEON, Luciane Almeida Amado	TL 1106168
LANDIM, João Igor Dantas	TL 1106105, TL 1106234, TL 1106244, TL 1106616, TL 1106453, TL 1106107, TL 1106173, TL 1106632, TL 1106537, TL 1106236	LEON, Soniza Vieira Alves	TL 1106311
LANDIM, João Igor	TL 1106429, TL 1106466	LEONI, Tauana Bernardes	TL 1104778
LANG, Michael Ricardo	TL 1105347, TL 1105349, TL 1105116, TL 1105123, TL 1105134, TL 1105165, TL 1105329	LEOTTI, Vanessa Bielefeldt	TL 1104685
LANGE, Marcos Christiano	TL 1106191, TL 1106480, TL 1105365, TL 1105684, TL 1105725, TL 1105096	LEPRE, Vanessa Bernardo Nunes	TL 1106329
LARA, Carolina Haveroth	TL 1106156	LESTINGI, Silvia Cordova	TL 1106100
LARCIPRETTI, Anna Laura Lima	TL 1105216, TL 1105138	LEÃO, Bianca Lopes Cavalcante	TL 1106564, TL 1106590
LAURA, Matilde	TL 1105714	LEÃO, Bruna Acioly	TL 1104833, TL 1105908, TL 1104978
LAURINI, Lia Leal	TL 1105732, TL 1105639, TL 1105061, TL 1105744, TL 1105870	LEÃO, Maria Eduarda de Souza Arêa	TL 1106631, TL 1105373, TL 1105653, TL 1105814, TL 1105826
LAVOR, Vytor Alves de	TL 1105225	LIEM, Assuncion Martinez	TL 1105570, TL 1105757, TL 1105269, TL 1105607
LAZEIRA, Victória de Menezes Sá	TL 1105259	LIEM, Asunción Martinez	TL 1105494
LAZERA, Victória de Menezes Sá	TL 1105812, TL 1105916, TL 1106078	LIMA, Aline Vieira Scarlatelli	TL 1104955
LAZERA, Vitoria de Menezes Sá	TL 1105753	LIMA, Ana Rosa Melo Correa	TL 1105102, TL 1106239, TL 1106365, TL 1105764, TL 1105815, TL 1106184, TL 1105631
		LIMA, Ana Rosa Melo Correia	TL 1106467
		LIMA, Arieny Karen Santos	TL 1105363, TL 1105548
		LIMA, Arissa Crispim Maximo	TL 1106555
		LIMA, Brenda Matheus Nogueira de	TL 1104998, TL 1105085

LIMA, Caio Vinicius Barroso de	TL 1106104, TL 1105726	LINHARES, Kennedy Soares	TL 1105565, TL 1106343, TL 1105843
LIMA, Camilla Silva Dantas Mendes	TL 1105658	LINO, Angelina Maria Martins	TL 1105178, TL 1105222
LIMA, Carlos Frederico Leite Souza	TL 1105815	LINS, Alice Cavalcante Almeida	TL 1104833, TL 1105908, TL 1104978
LIMA, Clarice	TL 1105414	LINS, Cícero José Pacheco	TL 1105481
LIMA, Danielle Pessoa	TL 1104998, TL 1105085, TL 1105107, TL 1105145, TL 1105236, TL 1105244, TL 1106217	LINS, Jose Jefferson Silva Cavalcanti	TL 1105388
LIMA, Daniel Meohas de Souza	TL 1105696	LINS, Jose Jefferson Silva Cavancanti	TL 1105380
LIMA, Eduarda Rabêlo	TL 1105164	LINS, José Jefferson Silva Cavalcanti	TL 1105252
LIMA, Emmanuel Avelino de	TL 1105445, TL 1105638	LINS, João Pedro Sá	TL 1105784
LIMA, Ester Costa	TL 1105697	LINS, Lucas Vergne Cheade	TL 1106547
LIMA, Fabricio D de	TL 1105545	LINS, Otávio Gomes	TL 1106467
LIMA, Fabricio Oliveira	TL 1106229, TL 1106553, TL 1106570, TL 1106520	LINZMEIER, Pietra Cani	TL 1105537
LIMA, Fabricio Oliveira	TL 1106244	LIRA, Arthur Oliveira	TL 1105895, TL 1105182
LIMA, Fabrícia Fontes	TL 1105500	LIRA-BATISTA, MARTA MARIA da SILVA	TL 1106379, TL 1106367, TL 1106369, TL 1106517, TL 1106495
LIMA, Fabrício D. de	TL 1104731	LIRA-BATISTA, MARTA MARIA da SILVA	TL 1106379, TL 1106367, TL 1106369, TL 1106517, TL 1106495
LIMA, Fabrício Diniz de	TL 1104979	LIRA-BATISTA, MARTA MARIA da SILVA	TL 1106379, TL 1106367, TL 1106369, TL 1106517, TL 1106495
LIMA, Fabrício Oliveira	TL 1106245, TL 1106620	LISBOA, João Vitor Mortari	TL 1105157, TL 1106407, TL 1104943, TL 1105109, TL 1105110, TL 1105111, TL 1105144, TL 1105488
LIMA, Fernanda Sousa	TL 1105702, TL 1105580, TL 1105585, TL 1105590	LISBOA, João Vitor M	TL 1106279, TL 1106278
LIMA, Gabrielle Benevides	TL 1105455, TL 1106501, TL 1106632	LISE, Emerson Oliveira	TL 1104889, TL 1104824
LIMA, Gleydyson Wesley Freire	TL 1106367, TL 1106495	LIU, Jonathan Wei Ting Wen	TL 1105123
LIMA, Isabel Maria Oliveira Macedo	TL 1106621, TL 1106631, TL 1106598, TL 1106575	LOBATO, Enedina Maria Monteiro	TL 1106104
LIMA, Izadora Vieira Bolzani Lopes	TL 1106237	LOBATO, Lara Willers	TL 1105864
LIMA, Jessica Layanne de Sousa	TL 1105164	LOBO, Camila Caroso	TL 1105863, TL 1105166, TL 1105785
LIMA, Joana Clara Oliveira Macedo	TL 1106598, TL 1105909	LOBO, Camila C	TL 1105284, TL 1105288
LIMA, José Guilherme Martins	TL 1105705, TL 1105716, TL 1105566, TL 1105990	LOBO, Izabella Rocha	TL 1106209
LIMA, José Rosemberg Costa	TL 1105450, TL 1105461, TL 1105574, TL 1105836, TL 1106528, TL 1105277, TL 1105355	LOBO, Luiz Felipe Nogueira de Figueiredo	TL 1105853
LIMA, Karla Vanessa Morais	TL 1105812	LOCHMÜLLER, Hanns	TL 1104679
LIMA, Karlla Danielle Ferreira	TL 1106469	LOIOLA, Rodrigo Bindá de Magalhães	TL 1104824
LIMA, Karlla Danielle Ferreira	TL 1105306, TL 1105049, TL 1106270	LONDERO, Renata	TL 1106298, TL 1106301
LIMA, Lidiane Andrea Oliveira	TL 1105498, TL 1105508	LONGO, Alexandre Luiz	TL 1106063, TL 1105096
LIMA, Lidiane Andréa Oliveira	TL 1105594, TL 1105465	LONGO, José Eduardo Trentim	TL 1105157
LIMA, Ludmila Machado	TL 1106296, TL 1105646, TL 1105624, TL 1105625, TL 1105619, TL 1105464	LONGO, Maria Francisca Moro	TL 1104891
LIMA, Luiza Aguiar	TL 1105869, TL 1105897	LOPES, Alessandro Augusto Viana Oliveira	TL 1105630
LIMA, Luiz Fernando Melo	TL 1105227	LOPES, Alexandre Bussinger	TL 1106104
LIMA, Lázaro de	TL 1105531	LOPES, Daniel Thomas Pereira	TL 1105293, TL 1105739
LIMA, Manuela Cavalcante Coling	TL 1106608, TL 1106623	LOPES, Edson Gabriel de Lima	TL 1105148
LIMA, Marcelo Ataíde de	TL 1106559	LOPES, Fernanda Cristina Rueda	TL 1105782, TL 1106540
LIMA, Marco Antonio Dantas Salles	TL 1104809	LOPES, Janine Lemos Mel Lobo Jofili	TL 1105861, TL 1105870
LIMA, Marco Antonio Sales Dantas de	TL 1106506	LOPES, Laura Cardia Gomes	TL 1105157, TL 1104943, TL 1105109, TL 1105110, TL 1105111
LIMA, Maria Luíza Marinho de Sá de Paula	TL 1106390	LOPES, Maycon Melo	TL 1106203, TL 1106281, TL 1105630
LIMA, Micael Porto Portela	TL 1105848, TL 1105872, TL 1106222	LOPES, Pedro Luiz	TL 1106223
LIMA, Pedro Lucas Grangeiro de Sá Barreto	TL 1105316, TL 1105405, TL 1106581, TL 1105949	LOPES, Rafaela Albuquerque	TL 1105092
LIMA, Pedro Lucas Grangeiro Sá Barreto	TL 1106196, TL 1106207	LOPES, Renan Barros	TL 1105836, TL 1106337, TL 1105837
LIMA, Raynrich Kevin Assis	TL 1106452	LOPES, Sarah Cerillo	TL 1105321
LIMA, Rian Vilar	TL 1106262, TL 1105301, TL 1105318, TL 1106117, TL 1106339, TL 1106544, TL 1106608, TL 1106623	LOPES-CENDES, Iscia	TL 1105215, TL 1105276
LIMA, Sylvio Ricard Gonçalves de Souza	TL 1106162, TL 1106620, TL 1106280, TL 1106090, TL 1106359, TL 1106429, TL 1106466, TL 1106308	LORENA, Ivan Augusto	TL 1105588
LIMA, Sylvio Ricard Gonçalves Souza	TL 1106450	LORENZONI, Leonardo Afonso	TL 1105857
LIMA, Sylvio Ricardo Gonçalves de Souza	TL 1106105	LORENZONI, Paulo José	TL 1106172
LIMA-SILVA, Thais Bento	TL 1106259	LOURENÇO, Elora Sampaio	TL 1105619
LIMONTE, Fábio Henrique	TL 1104728	LOURENÇO, Galileu Chagas	TL 1106121
LIN, Katia	TL 1105409, TL 1105103, TL 1105122, TL 1105124, TL 1105062	LOZ, Sabrina Hafemann	TL 1106063
LIN, Kátia	TL 1105395	LOZ, Sabrina Hafemann	TL 1106063
LINDNER, Stephanie	TL 1105123, TL 1105329	LUCAS, Ana Beatriz da Costa	TL 1105066, TL 1105114
LINHARES, Ana Paula Costa	TL 1105812	LUCAS, Elizabeth Lemos Silveira	TL 1106551
		LUCATO, Leandro Tavares	TL 1106252, TL 1106140, TL 1105048
		LUCENA, Adson Freitas de	TL 1106162
		LUCENA, Paulo Antonio Farias de	TL 1105748, TL 1105453
		LUCENA, Paulo Antonio Farias	TL 1106603
		LUCENA, Paulo Antônio Farias de	TL 1105810, TL 1106439

LUCENA, Paulo Antônio Farias	TL 1106294	MAGALHÃES, João Eudes	TL 1106317, TL 1106612, TL 1105524, TL 1106398, TL 1106187, TL 1106239, TL 1106365, TL 1105764, TL 1105815, TL 1106184, TL 1106467, TL 1106210, TL 1105557, TL 1105809
LUEMBA, Martinho	TL 1105532		
LUNA, Débora Nalígia Moraes	TL 1105672		
LUNARDI, Mariana dos Santos	TL 1105103		
LUPPI, Andrea de Martino	TL 1105850, TL 1105307	MAGALHÃES, Juliana Silva Almeida	TL 1106572, TL 1106605, TL 1106630
LUSTOSA, Júlio Augusto	TL 1105895, TL 1105182	MAGALHÃES, Jurandy Júnior Ferraz de	TL 1106320, TL 1106559
LUVIZUTTO, Gustavo José	TL 1106008, TL 1106023, TL 1106435	MAGALHÃES, Lara Cordeiro	TL 1106572, TL 1106135, TL 1106134, TL 1106605, TL 1106118, TL 1106524, TL 1106556, TL 1106574
LUVIZUTTO, Gustavo	TL 1106348		
LÉGORA, Davi Bravo Huguinim	TL 1106533, TL 1106471, TL 1106268	MAGALHÃES, Luciana Zelante Ambiel	TL 1105353
LÉGORA, Sophia Bravo Huguinim	TL 1106533, TL 1106471	MAGALHÃES, Marilza Campos de	TL 1105712
LÓCIO, Aline Moreira	TL 1105847	MAGALHÃES, Paolla Giovanna Rossito de	TL 1105346
LÓCIO, Aline	TL 1105865	MAGALHÃES, Paolla Giovanna Rossito	TL 1105370
MACDONALD, Matthew L	TL 1105215	MAGALHÃES, Pedro Silva Correa	TL 1105096
MACEDO, Aline Barbosa	TL 1105240, TL 1105242	MAGALHÃES, Samir Camara	TL 1106649
MACEDO, Amanda Souza Chaves	TL 1105414	MAGALHÃES, Samir Câmara	TL 1106091, TL 1106196, TL 1106207, TL 1106273
MACEDO, Laiane de Alencar	TL 1106410	MAGESTE, Priscila dos Santos	TL 1105462, TL 1105710, TL 1105479
MACEDO, Lidiane Alves	TL 1105230	MAGESTE, Priscila Santos	TL 1105869, TL 1105897, TL 1105741, TL 1106300
MACEDO, Victor Felipe Bispo	TL 1106418	MAIA, Fernanda Martins	TL 1106105, TL 1106162, TL 1106233, TL 1106234, TL 1106244, TL 1106616, TL 1106218, TL 1106461, TL 1106453, TL 1106107, TL 1106090, TL 1106159, TL 1106173, TL 1106359, TL 1106429, TL 1106501, TL 1106170, TL 1106450, TL 1106466, TL 1106493, TL 1106632, TL 1106502, TL 1106308, TL 1106537, TL 1106236, TL 1106473, TL 1106434, TL 1106499, TL 1105019, TL 1106226, TL 1106336, TL 1106386, TL 1106452, TL 1105763
MACEDO, Victor Fellipe Bispo	TL 1105102, TL 1105789, TL 1106187, TL 1105764, TL 1105815, TL 1106184, TL 1105589, TL 1105613		
		MAIA, Fernanda	TL 1106606
MACEDO, Yuri Silva	TL 1105770	MAIA, Isaac Holanda Mendes	TL 1106263
MACHADO, Amanda Batista	TL 1105624	MAIA, Larissa Almeida	TL 1105465
MACHADO, Antônio Fernando	TL 1105177	MAIA, Matheus Gonçalves	TL 1106533, TL 1106471, TL 1106433, TL 1106268, TL 1106282
MACHADO, Carolina Carmona Pinheiro	TL 1106214		
MACHADO, Denise	TL 1105407	MAIA, Raphael de Paula Doyle	TL 1105716
MACHADO, Luis Felipe Dantas da Nóbrega	TL 1106588	MAIA, Renata M	TL 1104775
MACHADO, Marcela Alcântara	TL 1106189	MAIA, Tonyato Fernandes Ribeiro	TL 1105872
MACHADO, Marcelo Teixeira	TL 1105833	MAIA, Víctor Queirós Calheiros Campelo	TL 1106608, TL 1106623
MACHADO, Maria Iris de Moraes	TL 1105230	MAITO, Vitor Teixeira	TL 1105402
MACHADO, Maria Iris Moraes	TL 1105774	MAKAREM, Louise	TL 1105247
MACHADO, Michel Ferreira	TL 1105115, TL 1105552, TL 1106249	MALHEIROS, Júlia Evaristo Figueiredo	TL 1105441
MACHADO, Richard Simon	TL 1106293, TL 1106491	MALISKI, Jessica	TL 1106564
MACHADO, Richard Simon	TL 1106293, TL 1106491	MALTA, Crissia Pitanga	TL 1105696
MACHADO, Roberta Ismael Lacerda	TL 1104860, TL 1105285, TL 1105428	MALTA, Maisa Vieira da Silva	TL 1105428
MACHADO, Stefano	TL 1105881, TL 1105581, TL 1105891	MANFRINATI, Luís Eduardo d'Almeida	TL 1106282
MACHADO, Vitor Oliveira	TL 1106179, TL 1106142	MANGANELLI, Luciane Aparecida Gonçalves	TL 1105519, TL 1105413
MACIEL, Damacio Ramon Kaimen	TL 1105505, TL 1105606, TL 1105618, TL 1106283, TL 1105171, TL 1105541, TL 1105538, TL 1105367, TL 1106123	MANGAS, Gabriel de Moraes	TL 1106203, TL 1106214
		MANGOLIN, Alison	TL 1105808, TL 1105836, TL 1105604, TL 1105798, TL 1105399, TL 1105592
MACIEL, Ernane Pires	TL 1105702, TL 1105580, TL 1105585, TL 1105590		
MACIEL, Guilherme Pablo de Santana	TL 1106384	MANGOLIN, Alisson	TL 1105821
MACIEL, Nicoly	TL 1106654, TL 1106655	MANHAGUANHA, Gustavo	TL 1106156
MACIEL, Thiago de Oliveira	TL 1106061	MANIVA, Samia Jardelle Costa de Freitas	TL 1106261, TL 1106558
MACIEL, Victor Augusto Zanesi	TL 1105808, TL 1105574, TL 1105836, TL 1106528, TL 1105387, TL 1105604, TL 1105798, TL 1105781, TL 1105399, TL 1105592	MANSUR, Kalil Ibrahim	TL 1105189, TL 1105432
		MANSUR, Leticia Fêzer	TL 1105066, TL 1105114
MACÊDO, Luisa Couceiro de Albuquerque	TL 1105777, TL 1106397	MANTESE, Carlos Eduardo	TL 1104957
MACÊDO, Stella Maria	TL 1105318	MANTOVANI, Charles Maroly Lessa	TL 1106404
MACÊDO, Victor Fellipe Bispo	TL 1105524, TL 1106365, TL 1105743, TL 1106210, TL 1105557, TL 1105809	MANTOVANI, Gabriel Paulo	TL 1106298
		MANZOLI, Isabela Reis	TL 1104697
MADRUGA, Kauan Alves Sousa	TL 1105227	MANZOLINI, Matheus Fernando	TL 1106595
MAGALHAES, Daiane	TL 1106362	MARAFON, Hellen Camila	TL 1105403
MAGALHAES, Felipe Barbosa	TL 1106600	MARANGA, Carina	TL 1106511
MAGALHAES, Francisco Neuton de Oliveira	TL 1106555	MARANHÃO, Eduardo	TL 1106188
MAGALHAES, Samir Camara	TL 1106064	MARANHÃO, Vicente Albuquerque	TL 1105397, TL 1105402, TL 1105403
MAGALHAES, Sarah de Sousa	TL 1106280	MARAZZI, Thire Baggio Machado	TL 1105450, TL 1105574, TL 1106528
MAGALHÃES, Daiane	TL 1105346	MARCELINO, Paulo Filho Soares	TL 1105738, TL 1105299, TL 1105274, TL 1105791, TL 1105848,

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MARCELINO, Paulo	TL 1104712	MARTINEZ, Alberto Rolim Muro	TL 1105296, TL 1106408
MARCHI, Bruno de	TL 1105988	MARTINS, Ana Carolina	TL 1104685
MARCHI, Luan	TL 1106252	MARTINS, Beatriz Souza	TL 1105752
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NAKAO, Natalia de Castro Fim	TL 1106413	NETO, Franklin de Castro	TL 1106286
NAKAZATO, Paula Carolina Grande	TL 1105472, TL 1105131, TL 1105694	NETO, Gilberto José de Melo	TL 1105534, TL 1105814
NARIMATSU, Keila	TL 1106516, TL 1106610, TL 1106576	NETO, Haldson Cesar Barbosa	TL 1105189, TL 1105432
NASCIMENTO, Ana Dolores Firmino Santos do	TL 1106427, TL 1106496, TL 1105125, TL 1105512	NETO, Heitor Nunes de Oliveira Sento-Sé	TL 1105552, TL 1105293
NASCIMENTO, Arthur Costa	TL 1105881, TL 1105581, TL 1105891	NETO, Heitor Nunes Oliveira Sento-Sé	TL 1106459
NASCIMENTO, Bruna Essado	TL 1105528	NETO, Herval Ribeiro Soares	TL 1105738, TL 1105299, TL 1105274, TL 1105353, TL 1105822
NASCIMENTO, Denner Matheus Ribeiro	TL 1105783	NETO, Ismar Andrade da Silveira	TL 1105294
NASCIMENTO, Fabricio Porto do	TL 1105560	NETO, Jasson José Moscon	TL 1105168
NASCIMENTO, Filipe Barroso	TL 1105892	NETO, Joaquim Fachine de Alencar	TL 1105895, TL 1105182
NASCIMENTO, Isaíra A P S	TL 1106440, TL 1106490, TL 1106503	NETO, Jose Hortencio dos Santos	TL 1106453
NASCIMENTO, Jeddson Rêgo	TL 1106418	NETO, José Dias de Assis	TL 1105799
NASCIMENTO, Marcia Helena Cassago	TL 1106531	NETO, José Elias Makaron	TL 1106329, TL 1106206
NASCIMENTO, Monique Evelyn Mendonça do	TL 1106427, TL 1106496, TL 1105125, TL 1105512, TL 1106569	NETO, José Lopes Tabatinga	TL 1106185
NASCIMENTO, Osvaldo J. M.	TL 1104755	NETO, José Luis Lima	TL 1106305
NASCIMENTO, Osvaldo J M	TL 1106097	NETO, José Ribamar Pereira	TL 1106375, TL 1106390, TL 1106392
NASCIMENTO, Osvaldo José Moreira do	TL 1105869, TL 1105479, TL 1105782	NETO, João Batista Guerra Barretto	TL 1106131
NASCIMENTO, Osvaldo José Moreira	TL 1106095, TL 1105239, TL 1105406	NETO, João Henrique Orsi	TL 1105270
NASCIMENTO, Osvaldo	TL 1105824, TL 1105879, TL 1106300	NETO, Luis Bandeira Alves	TL 1105895, TL 1105182
NASCIMENTO, Oswaldo	TL 1106380	NETO, Manoel Alves Sobreira	TL 1106125, TL 1106177, TL 1106525, TL 1106567, TL 1106622, TL 1106414, TL 1106091, TL 1106581, TL 1106103, TL 1105945, TL 1106048, TL 1106586
NASCIMENTO, Priscila Alves	TL 1105031	NETO, Manoel Moreira	TL 1105320
NASCIMENTO, Rairis Barbosa	TL 1106631, TL 1105348, TL 1105423	NETO, Moacir Pereira Leite	TL 1105046, TL 1104966
NASCIMENTO, Simony Lira do	TL 1106057, TL 1106217	NETO, Nivaldo Alves Calado	TL 1106588
NASCIMENTO, Suzete Farias da Guarda	TL 1105177		
NAUFEL, Maria Fernanda Soares	TL 1106618		
NAUFEL, Maria Fernanda	TL 1106313		
NEDER, Luciano	TL 1104755		
NEIDORF, Leonardo	TL 1105165		

NETO, Octavio Marques Pontes	TL 1105338, TL 1105875	NUNES, Diego Eduardo	TL 1105414
NETO, Octávio Marques Pontes	TL 1105450, TL 1105461, TL 1106528, TL 1105798	NUNES, Glaucus Fernando Vieira	TL 1105759
NETO, Olavo Santa Cruz Valadares	TL 1105557	NUNES, Helio Rubens de Carvalho	TL 1106435
NETO, Otilio Paulo da Silva	TL 1106369	NUNES, João Victor da Costa	TL 1105445, TL 1105892
NETO, Otávio da Cunha Ferreira	TL 1105895, TL 1105182	NUNES, Julia de Melo	TL 1106419, TL 1106538
NETO, Paulo Barbosa Leite	TL 1105102, TL 1105789, TL 1105524, TL 1106187, TL 1106365, TL 1105764, TL 1105815, TL 1106184, TL 1106467, TL 1105743, TL 1106210, TL 1105557, TL 1105809, TL 1106418, TL 1105589, TL 1105613	NUNES, Juliana de Oliveira	TL 1105752
NETO, Paulo Roberto Matos	TL 1106589, TL 1105272, TL 1105419, TL 1106264	NUNES, Júlia de Melo	TL 1105853
NETO, Pedro Braga	TL 1106125, TL 1106177, TL 1106525, TL 1106539, TL 1106567, TL 1106414, TL 1106076, TL 1106192, TL 1106385, TL 1106286, TL 1106402, TL 1106641, TL 1106581, TL 1106103, TL 1106081, TL 1105763, TL 1105945, TL 1106048, TL 1104998, TL 1105085, TL 1105107, TL 1105145, TL 1105236, TL 1105244	NUNES, Maria Cristina de Alencar	TL 1105267
NETO, Raimundo Marcial Brito	TL 1105462, TL 1105897, TL 1105741, TL 1106300	NUNES, Patricia Pereira	TL 1104833, TL 1104832
NETO, Raimundo Marcial de Brito	TL 1105869, TL 1105854, TL 1105878, TL 1105710	NUNES, Patricia Pereira	TL 1105908, TL 1104978, TL 1106578
NETO, Túlio Maranhão	TL 1106337, TL 1105277, TL 1105717	NUNES, Rebeca Holanda	TL 1106215
NEVE, Anuja	TL 1105660	NUNES, Thabata Emanuelle Martins	TL 1106279, TL 1106278, TL 1106475
NEVES, Carolina Daher de Alencar	TL 1105889	NUNES, Thábita Emanuelle Martins	TL 1105507, TL 1105790, TL 1106199, TL 1106205, TL 1105185, TL 1106527
NEVES, Larissa Lorrhan Santos	TL 1106380	NUNES, Zuriel Rodrigues Seixas	TL 1105005
NEVES, Luciana Oliveira	TL 1106220, TL 1106488	NÓBREGA, Ivna Lacerda Pereira	TL 1106158
NEVES, Pedro Rodrigues	TL 1106298, TL 1106301, TL 1105820	NÓBREGA, Katia Cirilo Costa	TL 1105682
NEVES, Rafaela de Souza Dias	TL 1105575	NÓBREGA, Kátia Cirilo Costa	TL 1105051
NICARETTA, Denise Hack	TL 1105921	NÓBREGA, Paulo Ribeiro	TL 1106125, TL 1105082, TL 1106177, TL 1106525, TL 1106539, TL 1106567, TL 1106622, TL 1106431, TL 1106414, TL 1106223, TL 1105316, TL 1106196, TL 1106581, TL 1106103, TL 1106207
NICOLI, Fernanda Michely	TL 1105833	ODA, Elder Yanaze	TL 1105420
NIEDERAUER, Andressa Gomes	TL 1104970, TL 1106463	OGUSUKU, Caroline Suemi	TL 1105115, TL 1105340, TL 1106343
NIMER, Gabriela Flor	TL 1105462	OKADA, Mariana	TL 1106287, TL 1106319, TL 1106307
NISHIYAMA, Alzira Nobuko	TL 1105369	OKAMOTO, Erika	TL 1105036
NITRINI, Ricardo	TL 1104692, TL 1106140, TL 1106259	OLIVAL, Guilherme Sciascia	TL 1104936
NOBREGA, Ana Caline	TL 1105089, TL 1105095	OLIVEIRA, Acary Souza Bulle	TL 1104864, TL 1104860, TL 1104862, TL 1105212, TL 1105221, TL 1105226, TL 1105283, TL 1105285, TL 1105559, TL 1105602, TL 1105633, TL 1104874, TL 1104940, TL 1105428, TL 1105516, TL 1105560, TL 1106189
NOBREGA, Ivna Lacerda Pereira	TL 1106501	OLIVEIRA, Acary Souza Bulle	TL 1105536
NOBREGA, Paulo Ribeiro	TL 1106076, TL 1106656, TL 1105945	OLIVEIRA, Afonso Henrique Costa de	TL 1106544
NOGUEIRA, Bruno Miceli Gonzalez	TL 1106311	OLIVEIRA, Americo Danuzio Pereira	TL 1105252, TL 1105380, TL 1105388
NOGUEIRA, Carlos Bruno	TL 1105406	OLIVEIRA, Ana Beatriz Garcez de	TL 1105696
NOGUEIRA, Clebeson Azevêdo	TL 1105245, TL 1105391	OLIVEIRA, Ana Carolina Silveira	TL 1105892
NOGUEIRA, Guilherme Nobre	TL 1106185, TL 1105780	OLIVEIRA, Ana Gabriella Camelo	TL 1106523
NOGUEIRA, Gustavo Carvalho	TL 1106617	OLIVEIRA, Ana Helena Barbalho Bezerra de	TL 1104805
NOGUEIRA, Luciana Tolstenko	TL 1105870	OLIVEIRA, Ana Maria dos Santos	TL 1105684, TL 1105725
NOGUEIRA, Mateus Henrique	TL 1106353, TL 1106607	OLIVEIRA, Ana Paula Pessoa de	TL 1105563
NOGUEIRA, Matheus Henrique	TL 1106312	OLIVEIRA, Ana Vitória Costa de	TL 1106544
NOGUEIRA, Patrícia T. B.	TL 1104755	OLIVEIRA, Andrea Lira	TL 1104707
NOLETO, Gustavo Sousa	TL 1105732, TL 1105744, TL 1105861, TL 1105870	OLIVEIRA, Andre Dias	TL 1105194
NOLETO, Gustavo Sousa	TL 1105732, TL 1105744, TL 1105861, TL 1105870	OLIVEIRA, André Dias de	TL 1105409
NONES, Diogo Pasquali	TL 1105116, TL 1105123, TL 1105134, TL 1105165, TL 1105329	OLIVEIRA, Anna Melissa Noronha	TL 1105593
NOUGUEIRA, Mateus Henrique	TL 1106074	OLIVEIRA, Arlindo Américo de	TL 1105347, TL 1105349, TL 1105123, TL 1105329
NOVAES, Bernardo Caetano	TL 1105684, TL 1105725	OLIVEIRA, Augusto César Penalva de	TL 1106451
NOVAES, Maria Rita Carvalho Garbi	TL 1106075	OLIVEIRA, Beatriz Barrozo Gonzalez de	TL 1105961
NOVAIS, Aurea Maria Lago	TL 1106134	OLIVEIRA, Beatriz Barrozo Gonzalez	TL 1106442, TL 1106481, TL 1105753
NOVAIS, Rodrigo Nobre de	TL 1106120	OLIVEIRA, Bianca Etelvina dos Santos de	TL 1106588
NOVAK, Edison Matos	TL 1106191	OLIVEIRA, Bruna Andrade de	TL 1105525
NOVAK, Felipe Trevisan Matos	TL 1106441	OLIVEIRA, Camila Maria	TL 1104685
NOVIS, Luiz Eduardo	TL 1105507, TL 1105790, TL 1106279, TL 1106278	OLIVEIRA, Camilla Ribeiro de	TL 1105528, TL 1105576
NUCCI, Anamarli	TL 1106302, TL 1106363, TL 1105564, TL 1106352, TL 1104979	OLIVEIRA, Carlos Mauricio	TL 1104707, TL 1105892, TL 1105948, TL 1105487
		OLIVEIRA, Celina de	TL 1106306
		OLIVEIRA, Clara Maria Limaverde	TL 1105763
		OLIVEIRA, Clariana Nascimento de	TL 1105447, TL 1105747
		OLIVEIRA, Clarice Pereira Sales	TL 1105586, TL 1105599

OLIVEIRA, Daniel Alves de	TL 1106188, TL 1105734, TL 1105777, TL 1106423, TL 1105773, TL 1106397, TL 1106444, TL 1106465, TL 1105237, TL 1106548	OLIVEIRA, Luis Henrique Sunderhus de	TL 1105716, TL 1105566, TL 1105990, TL 1105799
OLIVEIRA, Daniel Sabino de	TL 1105253	OLIVEIRA, Luiza Barbosa de	TL 1105528, TL 1105576
OLIVEIRA, Danilo Nunes	TL 1106391, TL 1106262, TL 1106604, TL 1105933, TL 1105945, TL 1106626	OLIVEIRA, Luiz Fernando Rodrigues de	TL 1105115
OLIVEIRA, Dayana Dourado de	TL 1106481, TL 1105259, TL 1106078	OLIVEIRA, Luiz Fernando	TL 1106249
OLIVEIRA, Denise Nunes	TL 1105301, TL 1105318	OLIVEIRA, Maira Okada de	TL 1105056
OLIVEIRA, Diego Silveira	TL 1106555	OLIVEIRA, Marcella Cabral de	TL 1105692
OLIVEIRA, Déborah Castro Ferreira de	TL 1105359, TL 1105839, TL 1105920	OLIVEIRA, Marcos Castello Barbosa	TL 1105206
OLIVEIRA, Déborah Castro Ferreira	TL 1105769, TL 1106500	OLIVEIRA, Maria Clara Barbosa de	TL 1105155, TL 1105223, TL 1105224
OLIVEIRA, Déborah de Castro Ferreira de	TL 1106326	OLIVEIRA, Maria de	TL 1106606
OLIVEIRA, Elienay Cassio	TL 1105730	OLIVEIRA, Mariana Pacheco de	TL 1106491
OLIVEIRA, Enedina Maria Lobato	TL 1105006	OLIVEIRA, Marina Rossi	TL 1105235
OLIVEIRA, Enrico Pinheiro de	TL 1106526	OLIVEIRA, Matheus Franco Andrade	TL 1106523
OLIVEIRA, Enzo Derick Guterres	TL 1106442, TL 1105259	OLIVEIRA, Milena Fernandes de	TL 1106619, TL 1105819, TL 1106428, TL 1106611
OLIVEIRA, Ericles Willy Machado de	TL 1105474	OLIVEIRA, Murilo A	TL 1104775
OLIVEIRA, Fabio de Nazaré	TL 1105521	OLIVEIRA, Pablo Nascimento	TL 1105115, TL 1105552, TL 1106249
OLIVEIRA, Fabio Pascotto de	TL 1105088	OLIVEIRA, Paula Luanna Carvalho de	TL 1105763
OLIVEIRA, Fernanda Aquino de	TL 1105381	OLIVEIRA, Paulo Henrique Martinelli	TL 1105666, TL 1105686, TL 1105699, TL 1105264, TL 1105390, TL 1105691, TL 1105641
OLIVEIRA, Francisca Adna Almeida de	TL 1105301	OLIVEIRA, Pedro Antonio Medeiros de	TL 1106549
OLIVEIRA, Francisco Tomaz Meneses de	TL 1106516, TL 1106610	OLIVEIRA, Pedro Barbosa	TL 1105150, TL 1105428, TL 1105726
OLIVEIRA, Fábio de Nazaré	TL 1105151	OLIVEIRA, Pedro Dantas	TL 1106437
OLIVEIRA, Gabriela Sanford Guimarães Caminha de	TL 1106226	OLIVEIRA, Pietra Baggio Peixoto de	TL 1105787, TL 1105302, TL 1104977
OLIVEIRA, Gabriel Carvalho	TL 1106474, TL 1106557	OLIVEIRA, Pietra Baggio Peixoto	TL 1105305
OLIVEIRA, Gabriel Cipriano Feitosa	TL 1106621	OLIVEIRA, Ricardo Santos de	TL 1105133
OLIVEIRA, Gabriel Salim Saud de	TL 1106506	OLIVEIRA, Roberto Teodoro Gurgel de	TL 1106062, TL 1106133
OLIVEIRA, Giovana Andrade de	TL 1105842, TL 1105871, TL 1105583	OLIVEIRA, Rodrigo Almeida Magalhães	TL 1106428
OLIVEIRA, Giuliano da Paz	TL 1105567	OLIVEIRA, Samantha Lopes	TL 1106605, TL 1106630
OLIVEIRA, Guiomar Nascimento de	TL 1105034	OLIVEIRA, Sarah Almeida Sales de	TL 1106544
OLIVEIRA, Gustavo Mesquita de	TL 1106090	OLIVEIRA, Sebastiao Carlos de Sousa	TL 1106042
OLIVEIRA, Helvia Bertoldo de	TL 1105559	OLIVEIRA, Silmara Ferreira de	TL 1105732, TL 1105744
OLIVEIRA, Herisson Rodrigues de	TL 1106059, TL 1106419, TL 1105913, TL 1106562, TL 1105874, TL 1106571	OLIVEIRA, Steffany Protásio Moura de	TL 1105783
OLIVEIRA, Hyago Casimiro Mendes de	TL 1105168	OLIVEIRA, SUZANA NOVAIS TEIXEIRA	TL 1105084
OLIVEIRA, Hélvia Bertoldo de	TL 1105283, TL 1105633	OLIVEIRA, Thais Barbosa de	TL 1105819, TL 1106428, TL 1106611
OLIVEIRA, Igor de	TL 1106548	OLIVEIRA, Thales Junqueira	TL 1105510
OLIVEIRA, Isabelle Jacqueline Weber	TL 1106574	OLIVEIRA, Thalís Silva de	TL 1106298, TL 1106301
OLIVEIRA, Isabelle Jacqueline Weber	TL 1106574	OLIVEIRA, Thaís Barbosa de	TL 1106619, TL 1106591
OLIVEIRA, Isabel Lopes de	TL 1106343	OLIVEIRA, Vanessa Fernandes de	TL 1105763
OLIVEIRA, Isadora Castro Ferreira	TL 1105769, TL 1106500	OLIVEIRA, Vinicius Pafume de	TL 1105601
OLIVEIRA, Isadora de Castro Ferreira de	TL 1105359, TL 1105839, TL 1106326, TL 1105920, TL 1105909	OLIVEIRA, Viviane de Sousa	TL 1106410
OLIVEIRA, Isadora Mônica Ponte de	TL 1106222	OLIVEIRA, Yuri Almeida	TL 1106492
OLIVEIRA, Jessica Blanc Leite	TL 1106302	OLIVEIRA-FILHO, Jamarly	TL 1104775, TL 1105655, TL 1105672, TL 1106272, TL 1105665
OLIVEIRA, João Gabriel Mansano de	TL 1105377	OLIVÉ, Montse	TL 1106175
OLIVEIRA, João Gustavo dos Anjos Moraes	TL 1106477	ONO, Carla Rachel	TL 1104692
OLIVEIRA, Juliana Marília Pereira de	TL 1105551	ORGE, Carolina	TL 1105053, TL 1105627
OLIVEIRA, Juliana Martini	TL 1106135	ORIKASA, Guilherme Seiti	TL 1105556
OLIVEIRA, Juliana Martini	TL 1106135	ORRICO, Camila	TL 1105447, TL 1105053, TL 1105747
OLIVEIRA, Lais Machado de	TL 1105861	ORSOLIN, Priscila Capelari	TL 1105595
OLIVEIRA, Leandro Freitas	TL 1105872, TL 1106152	ORTEGA, Adriana Banzatto	TL 1106551
OLIVEIRA, Leandro	TL 1104712	ORTH, Luiza	TL 1105477, TL 1105302, TL 1104977, TL 1105776
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OLIVEIRA, Letícia Bezerra de	TL 1105704	OTTONI, Letícia Pires	TL 1105770
OLIVEIRA, Letícia Farias	TL 1105639	PACE, Filipe Tupinambá Di	TL 1105377, TL 1105076
OLIVEIRA, Lilian Vieira de Sousa	TL 1105528	PACHECO, Ana Beatriz Vieira	TL 1106067, TL 1106148
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OLIVEIRA, Lucas Casimiro de	TL 1105420	PACHECO, Glauber Mota	TL 1105601, TL 1105510
OLIVEIRA, Lucas de Souza	TL 1105325	PACHITO, Daniela	TL 1104957
OLIVEIRA, Luciana Mattos Barros	TL 1105665	PACÍFICO, Fernando Augusto	TL 1106617
		PACÍFICO, Fernando Augusto	TL 1106583
		PADILHA, Wilton Wilney Nascimento	TL 1105853
		PAES, Maria Alice da Silva	TL 1105921

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PAIVA, Anderson Rodrigues Brandão de	TL 1106085, TL 1106370	TL 1105236, TL 1105244
PAIVA, Eduardo dos Santos	TL 1105784	PELLEGRIN, Eduarda de TL 1105138
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PAIVA, Isadora	TL 1105847	PENA-PEREIRA, Marcio TL 1106647
PAIVA, Kettley Alves	TL 1106273	PENICHE, Paula da Cruz TL 1105508
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PAIXÃO, Bárbara Oliveira	TL 1105346	PEREIRA, Cristiana Borges TL 1105198, TL 1105292
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PALMINI, Andre	TL 1105407	PEREIRA, Felipe Salvagni TL 1106522
PAOLILO, Renata Barbosa	TL 1106432	PEREIRA, Flávio Faria Costa TL 1105196
PAPAIZ, Fabiano	TL 1106263	PEREIRA, Fábio Araujo TL 1105472
PARANHOS, Daniel Rocha	TL 1106135, TL 1106134, TL 1106118	PEREIRA, Fábio de Araújo TL 1105694
PARANHOS, Daniel Rocha	TL 1106135, TL 1106134, TL 1106118	PEREIRA, Ivanio Alves TL 1106198, TL 1106257, TL 1105208, TL 1105209
PARENTE, Ana Raquel Jucá	TL 1105455	PEREIRA, Jorge Fernando de Miranda TL 1105378
PARENTE, Enzo Lima Alcântara	TL 1105791	PEREIRA, Jéssica Gonçalves TL 1106168
PARMERA, Jacy Bezerra	TL 1104692	PEREIRA, Katriane Endiel TL 1105240, TL 1105242
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PATATT, Júlia	TL 1105026	PEREIRA, Valeria Coelho Santa Rita TL 1105570, TL 1105757, TL 1105269
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PAULA, Telio Diego Cantalice de	TL 1105379	PERES, Daniele TL 1105123
PAULA, Thomas Vieira de	TL 1106378, TL 1105724, TL 1105863, TL 1105296	PERES, Gabriel Hoher TL 1105349, TL 1105116
PAULA, Thomas Viera de	TL 1105635	PERES, Mario TL 1105218
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PAULO, Larissa Emile	TL 1106206	PERINI, Laís Bissoli TL 1105409, TL 1105395
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PB, Bianca Etelvina Santos de Oliveira. João Pessoa	TL 1106597, TL 1106458	PESSOA, Alberlucio Esquirio TL 1105807
PE, Wilson Alves de Oliveira Júnior. Recife	TL 1106208	PESSOA, Alberlúcio Esquirio TL 1105294
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PEDROSO, Jose L	TL 1105284	PESSOA, Fabricio Silva TL 1105812
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PEIXOTO, Juliana de Castro Naves	TL 1105689	PICCOLO, Ana Claudia TL 1105321, TL 1105235
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PEIXOTO, Maria Clara Borges Girão	TL 1106222	PIEMONTE, Maria Elisa Pimentel TL 1105051, TL 1105682, TL 1106417
PEIXOTO, Satiko Andrezza Takano	TL 1106241	PIEMONTE, Maria Elisa P TL 1106440, TL 1106490, TL 1106503
PEIXOTO, Vitória Maria Torres	TL 1105225, TL 1106177, TL 1106376, TL 1106381, TL 1106396, TL 1106525,	PIMENTA, Matheus Resende Costa TL 1105216
		PIMENTEL, Ana Luiza Utrine TL 1105897
		PIMENTEL, Angela V TL 1105948
		PIMENTEL, Leonardo Halley Carvalho TL 1105363, TL 1105373

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VERAS, Samuel Ranieri Oliveira	TL 1106105, TL 1106620, TL 1106431	VITORINO, Luciano Magalhaes	TL 1105474
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VERDE, Lucas Antonissen Lima	TL 1105419, TL 1106264	WANDALL, Guilherme	TL 1105349, TL 1105537
VERDI, Giulia Maria Ximenes	TL 1106487, TL 1105362, TL 1105277, TL 1105717	WANDERLEY, Bianca Gomes	TL 1105635
VERSIANI, Isadora	TL 1105462, TL 1105869, TL 1105741, TL 1105710, TL 1105879, TL 1106300	WANZILEU, Ingrid Demosthenes	TL 1105626, TL 1105609, TL 1105728, TL 1105535, TL 1105553, TL 1105749
VIANA, Eloisa Alves	TL 1105155, TL 1105223, TL 1105224	WASEM, Matheus Pedro	TL 1105624, TL 1105625
VIANA, Leda Maria Cavalcante	TL 1106222	WATANABE, Nathalia	TL 1105421, TL 1105703, TL 1105078, TL 1105859, TL 1105740
VIANA, Lorena da Silva	TL 1105511, TL 1105855, TL 1106141, TL 1106130, TL 1105801, TL 1106446	WAYSZCEYK, Sheila	TL 1105349, TL 1105537, TL 1105527, TL 1105533
VIANA, Lorena Silva	TL 1106143, TL 1106624	WEBER, Eduarda Kotlinsky	TL 1105542, TL 1105026, TL 1105621
VIANA, Natália Araújo Nascimento	TL 1106631	WEINSHENKER, Brian G	TL 1105040
VIANA, Patrick Silva	TL 1105818	WELKER, Carolina Flores	TL 1105702, TL 1105580, TL 1105585, TL 1105590
VIANA, Ramon Távora	TL 1105498, TL 1105508, TL 1105594, TL 1105465	WERLE, Taís Michele	TL 1105820, TL 1105925
VIANEZ, Talisia Nascimento	TL 1105816	WERNECK, Lineu César	TL 1106172
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VICTOR HUGO CASTRO SÁ2	TL 1105294	WESCHENFELDER, Cesar Augusto	TL 1106298, TL 1106301, TL 1106372
VIDAL, Mariana Alemeida	TL 1105630	WESTIN, Larissa Kozow	TL 1105406
VIDIGAL, Daniela Name Chaul	TL 1105601	WHITAKER, Maria Eduarda Dall'Oglio	TL 1105751
VIDIGAL, Giovanna Paula	TL 1104937, TL 1104938	WIECKIEWICZ, Mieszko	TL 1105627
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		XAVIER, Arthur	TL 1105824
		XAVIER, Fernando	TL 1105407
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XIMENES, Marcelo Tognato	TL 1106406	TL 1106411, TL 1106416, TL 1106424, TL 1106436,
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YASSUDA, Clarissa	TL 1105276	TL 1105222, TL 1106255, TL 1106094
YASSUDA, Mônica Sanches	TL 1106259	ZANOVELLO, Ana Beatriz Gonçalves
YASUDA, Clarissa Lin	TL 1105555, TL 1105724, TL 1106312, TL 1105610, TL 1106212, TL 1106345, TL 1106594, TL 1106074, TL 1106353, TL 1106607	TL 1105756, TL 1106599
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YOSHIDA, Daniela	TL 1104816	TL 1105207, TL 1105906
ZABETIAN, Cyrus P	TL 1105410	ZEIGELBOIM, Bianca Simone
ZAIDAN, Paloma Peter Travassos	TL 1105613	TL 1106564, TL 1106590
ZAIDEN, Júlia Suzana	TL 1105177	ZENATTI, Gabriel Angelo Garute
ZAMBON, Amélia Limongi	TL 1105321	TL 1105477, TL 1105751, TL 1105787, TL 1105302, TL 1105305, TL 1104977, TL 1105776
ZAMBON, Antonio Alberto	TL 1105176, TL 1105178	ZENGO, Lucas Victoy Guimarães
ZAMBON, Antônio Alberto	TL 1106231	TL 1105477, TL 1105751, TL 1105787, TL 1105302, TL 1105305, TL 1104977, TL 1105776
ZAMBON, Atonio Alberto	TL 1104679	ZETOLA, Viviane de Hiroki Flumignan
ZAMORA, Paulo Henrique Fabiano	TL 1104955	TL 1105192, TL 1106318
ZAMPOLLI, Tainá Montini	TL 1105189, TL 1105432	ZIMMER, Eduardo
ZANCANARO, Rodrigo Bianchi	TL 1105531	TL 1106144
ZANETTE, Fernando Netto	TL 1105194, TL 1105642	ZIRPOLI, Bianca Berardo Pessoa
ZANETTE, Fernando	TL 1105409, TL 1105395	TL 1106224, TL 1106592, TL 1106238, TL 1106131
ZANETTI, Luisa	TL 1104891	ZOTIN, Maria Clara Zanon
ZANINI, Marco Antonio	TL 1104886	TL 1105065, TL 1105362
ZANIRATI, Gabriele	TL 1105407	ZUARDI, Antônio W
ZANOTELI, Edmar	TL 1105158, TL 1105213, TL 1105306, TL 1105358, TL 1106092, TL 1106093, TL 1106274, TL 1106377,	TL 1105948
		ZUCHNER, Stephan
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		ZUMBANO, Giulia Matiusso
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		ZUNIGA, Rubén David dos Reis
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		ZÉTOLA, Viviane de HIROKI FLUMIGNAN
		TL 1106191, TL 1106480, TL 1105365
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