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284 – The epidemiology of congenital syphilis in São Paulo: a public health problem with neurological sequelae

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Introduction: Syphilis is an infectious disease that has vertical transmission. If left untreated, it can lead to severe neurological problems, seizures, cerebral palsy, mental retardation, and learning problems in the baby.

Objective: To carry out epidemiological investigations to monitor cases of syphilis and treat infected pregnant women early.

Methods: This study statistically analyzes and describes the cases of congenital syphilis in São Paulo between 2010 and 2020 through data obtained by the Information System of Notifiable Diseases (SINAN).

Results: The total number of cases of syphilis during pregnancy was 81,401 and 30,665 (37.67%) evolved to congenital syphilis. From 2010 to 2020, the percentage change in the number of annual cases was 169.94%. Among the metropolitan regions, São Paulo had the highest incidence (55.67%). The mother diagnosis occurred during prenatal care in 59.59% of cases; At the time of delivery or curettage: 33.19%; after delivery: 4.67% Ignored/blank: 2.26%; Regarding skin color/ethnicity there is 18.36% (ignored in the research); White: 44.13%; Black: 4.26%; Yellow: 0.16%; Brown: 32.87%; Indigenous: 0.19%. Data on the mother's education show that 22.09% have incomplete elementary school; 10.93% completed elementary school; 13.19% have incomplete secondary education; 19.58% have completed secondary education; 1.06% have incomplete higher education and 1.18% have completed higher education; Ignored/blank: 30.79%. The data found that 17.34% did not realize prenatal care. Besides, 66.88% of partners did not realize treatment.

Conclusion: Cases of congenital syphilis are associated with lower maternal education, and white and brown skin color, thus being associated with greater social vulnerability. The growing increase in cases of congenital syphilis shows a public health problem and failures in prenatal care and treatment follow-up.

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290 – Chronic daily headache after COVID-19: a prospective cohort study

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Introduction: Headache may persist beyond the acute phase of coronavirus disease 2019 (COVID-19). Little is known about the frequency and impact of this persistent headache and the incidence of chronic daily headache (CDH).

Objectives: To evaluate the incidence, risk factors, characteristics and impact of CDH in patients with COVID-19.

Design: This is a prospective cohort study.

Methods: In the first stage, 288 patients were interviewed by telephone after the acute phase of COVID-19. In this phase, 199 patients who had headache were re-interviewed at least one year after COVID-19. Headache that persisted beyond the acute phase and had a frequency of 45 days or more in the first three months was considered as CDH.

Results: 123 patients were included, 56.1% were women, median age 50 (41; 58) years. Median time between the acute phase of COVID-19 and the interview: 804 (739; 892) days. Headache persisted beyond the acute phase COVID-19 in 52%. 20.3% had CDH. Those with previous headache and more severe headache in the acute phase had a higher risk of developing CDH. The group with CHD had more women, higher headache impact, higher frequency of headache in the first 3 months after COVID-19, more persistence of headache after 120 days of COVID-19, and less pulsatile headache when compared to those whose headache persisted.

Conclusion: The incidence of CDH after COVID-19 was 20.3%. This headache had a negative impact on the lives of these individuals. The presence of previous headache and higher intensity of headache were associated with a higher risk of CDH.

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294 – Clinical and epidemiological baseline characterization of dementia cases in a specialized center in Recife, Brazil

Ana Rosa Santana[1], Victor Ting Po Chy[1], Victor Mendes da Silva[1], Priscila Aparecida da Silva[1], Alberto Henrique Torres Trindade da Silva[1], Mariana Gonçalves Maciel Pinheiro[1], Victor de Moraes Chagas[1], Bruno Henrique Carneiro Costa[1], Luísa Couceiro de Albuquerque Macedo[1], João Pedro Matos de Santana[1], Breno José Alencar Pires Barbosa[1]

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Introduction: Population aging has been considered the main risk factor for the development of dementia globally. In low- and middle-income countries such as Brazil, the projections are for expressive numbers due to poor socioeconomic conditions. However, the epidemiological profile of dementias in Brazil is still little known due to the scarcity of studies, especially in the northeast of the country, where poverty and illiteracy are severe.

Design: Retrospective and descriptive study of data obtained from a comprehensive review of medical records.

Methods: Records of cases treated between 2018 and 2021 were examined, cases that did not have information in the medical record were excluded from the study.

Results: 128 medical records were analyzed, 109 of which met inclusion criteria. 57.8% of patients were male with a mean age of 65.3 years. 38.4% of the sample had less than 8 years of formal education and 15.1% were illiterate. Unskilled manual labor was the occupation type in 50% of the sample. The mean baseline Mini-Mental State Examination score was 16.88. Alzheimer's Disease (AD) was the most frequent diagnosis (37%) mostly in a moderate (22%) to advanced stage (20%) of dementia. 32% of all cases met criteria for mixed forms of Dementia. Frontotemporal Dementia (14%), Lewy Body's Disease (6%) and Pure Vascular Dementia (6%) were also reported. 69.7% of our sample underwent magnetic resonance imaging and 24% also had access to molecular imaging methods.

Conclusion: The degree of dementia of the examined cases caught our attention, most of them in a moderate or advanced stage, probably related to the time patients wait for care at a specialized center. Such bias may also explain the high frequency of non-AD pathologies in the sample, in contrast to data from the general population. With the limitation of its descriptive and retrospective design, the present study represents an important initiative towards characterizing data of Dementia in northeastern Brazil.

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295 – Allostatic load measures in elderly subjects with mild cognitive impairment and subjective cognitive decline: a cross-sectional analysis

Breno José Alencar Pires Barbosa[1], Maria Clara Ferreira Jesus[1], Maira Okada de Oliveira[1], Luciana Cassimiro[1], Isabella B. Avolio[1], Eduardo Sturzeneker Trés[1], Conrado Regis Borges[1], Thiago Bezerra Moraes Teixeira[1], Juliana Nery Souza-Talarico[1], Sonia Maria Dozzi Brucki[1]

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Introduction: Several studies suggest that chronic stress and persistently high levels of cortisol are related to cognitive dysfunction and neurodegeneration. 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.

Objective: The present study aimed to investigate the relationship between markers of chronic stress and cognitive status in BRAMS, making comparisons between participants classified as controls, subjective cognitive decline (SCD) and mild cognitive impairment (MCI)

Methods: Cross-sectional study with a subgroup analysis of the BRAMS cohort, in which individuals aged > 60 years had access to a comprehensive neuropsychological classification, clinical, imaging and laboratory measures. The allostatic load index (AL) was measured through anthropometric, endocrinological, cardiovascular, metabolic and inflammatory markers of chronic stress.

Results: 77 participants were evaluated, of which 65 met the inclusion criteria. The mean age was 70.2 (+6.01) years, with 72% being female and 11 years of schooling (interquartile range 7–15). 42 subjects were classified as MCI, 15 as SCD and 8 as controls. There was a significant difference between waist/hip ratio (0.94 in the MCI group versus 0,88 in the SCD group, P = 0.03) and in the measurement of the weighted AL index considering individuals in the extremes values of cortisol (36.9% in the MCI group versus 27.2% in the SCD group, P = 0.04). In the multivariate analysis, higher education was associated with reduced risk of MCI.

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 AL in the MCI group. Due to the cross-sectional nature of the study, the associations found do not allow inferring causality between the measures, which need further validation in the longitudinal follow-up of the participants.

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296 – The effects of early intervention on autism spectrum disorder prognosis: a narrative review

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Introduction: Autism Spectrum Disorder (ASD) is a multifactorial neurodevelopmental disorder that manifests itself in a classic triad that comprises communication, social interaction and the execution of stereotyped repetitive behaviors. The current estimate is that for every three detected cases of ASD, there are two undiagnosed cases that reach adulthood without adequate treatment.

Objectives: To investigate the impacts of early intervention (EI) on the prognosis of patients with ASD.

Methods: It's a narrative review of literature in the PubMed and ScienceDirect databases, using the descriptors: "Autism Spectrum Disorder", "early intervention" and "prognosis" registered in DeCS/MeSH, and using the Boolean operator AND. The inclusion criteria were: articles published in English, between 2013 and 2023. After analyzing titles, abstracts and full text, we selected 23 articles and included 17 additional studies.

Results: The average age for ASD diagnosis is around 4 to 5 years old, although the ideal age for diagnosis is around 2 years old. The use of specific screening and diagnostic methods enabled a reduction in the age at diagnosis by at least 2 years. Therapies inspired by Applied Behavior Analysis and the TEACCH model were the most effective for EI. Children diagnosed early, after intervention, demonstrated better cognition and language, in addition to a reduction in stereotyped behaviors and required less ongoing support at school than late-diagnosed children. Parental participation in therapies was significant for success.

Conclusion: EI positively influences the prognosis of ASD. The use of appropriate screening instruments and the training of professionals can favor early diagnosis and EI, especially in regions lacking specialists. Low sampling and the interference of external factors are relevant limitations of the studies found.

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302 – Evidence and affects in Duchenne muscular dystrophy in children and Golden Retriever dogs

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Introduction: Progressive muscular dystrophies differ in different ways due to their age of manifestation, the distribution of muscle weakness and the association of heart, central nervous system and peripheral nervous system. The most severe and common form of muscular dystrophies is Duchenne muscular dystrophy (DMD). Its involvement is 1/3500 male babies born alive and is attributed to 80% of cases of dystrophinopathies. The impairment of the dystrophin-glycoprotein complex in Becker and Duchenne dystrophies, in most congenital and girdle dystrophies, destruction of the sarcolemmal

muscle fiber occurs, releasing muscle serum enzymes. Its characteristics are defined by clinical, histological and electromyographic criteria. The symptoms result from the weakness of the skeletal, cardiac and visceral muscles; progressively worsen; histological changes include muscle degeneration and regeneration, without evidence of abnormal storage of any metabolic; the disease is heritable, even without familial reports.

Objectives: To compare the evidence and manifestations of DMD in humans in relation to Golden Retriever dogs with muscular dystrophy (GRMD), from breeders from Seattle-USA, named Canil GRMD — Brasil from the Department of Anatomy of the Faculty of Veterinary Medicine and Zootechnics of the Universidade de São Paulo.

Conclusion: The model that most resembles humans are dogs, in terms of clinical and pathological aspects of muscular dystrophy. Golden Retriever dogs (GRMD) are genetically homologous to humans, and their histological lesions are identical to patients with DMD. GRMD dogs have been characterized as an ideal model for studies of the pathological and genetic mechanisms of DMD, as well as therapeutic trials.

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305 – Eladocagene exuparvovec gene therapy improves motor development in patients with aromatic L-amino acid decarboxylase deficiency

Paul Wuh-Liang Hwu[1], Agathe Roubertie[1], Yin-Hsiu Chien[1], Ni-Chung Lee[1], Antonia Wang[1], Alexis Russell[1], Pedro Eugenio Pachelli[1], Luciana Giugliani[1], Andressa Federhen[1], Chun-Hwei Tai[1]
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Introduction: Aromatic L-amino acid decarboxylase (AADC) deficiency is caused by mutations in the DDC gene reducing AADC enzyme activity causing motor and neurodevelopmental impairments.

Objectives: Evaluate clinical outcomes in children with AADC treated with Eladocagene exuparvovec, recombinant adeno-associated viral vector serotype 2 carrying the coding sequence for AADC enzyme.

Methods: Therapy was infused bilaterally in the putamina of 30 patients aged 18–102 months receiving 1.8×10^{11} vg (n = 21) or 2.4×10^{11} vg (n = 9) followed for up to 120 months assessed using PDMS-2 key motor milestones including head control (partial or full), sitting (supported or independently), standing (with/away from support; up from cross-legged position), and walking (with/without assistance; 10 feet; taped line). Motor milestones were measured every 3 months for 1 year following gene therapy, then every 6–12 months for ≤ 120 months. Data extracted on January 4, 2022.

Results: At baseline, no patients had mastered head control or more advanced milestones. At year 1 of follow-up, patients were gaining the following skills (n): partial head control (26); full head control (15), sitting unassisted (7), supported standing (2). Progression of development was noted at years 5 and 10. By year 5 of follow-up, more advanced milestones were achieved (n): full head control (24), sitting unassisted (21) assisted walking (5), walking 10 feet (3), or walking upstairs (3). These abilities were maintained for as long as 10 years.

Conclusion: The data indicate that eladocagene exuparvovec can provide a durable, positive impact on motor development in patients with AADC deficiency.

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313 – Alexia without agraphia, case report

João Paulo Barile[1], Andreas Batista Schelp[1], Davi Vargas Freitas Teixeira[1], Sonia Maria Cesar de Azevedo Silva[1], Ana Luisa Rosas Sarmento[1]
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Introduction: Alexia Without Agrafia (AWA) is a syndrome in which the patient loses the ability to read while maintaining the ability to write. It's described in strokes in the territory of the left posterior cerebral artery (PCA) and is usually accompanied by right homonymous hemianopia (HH) or color anomy.

Case presentation: Male, 66 years old, complete higher education, right-handed, woke up two days ago with difficulty orienting himself, bumping into objects, visual difficulty in right hemifields. Neurological examination: preserved naming (when presented through sensory means other than visual), fluency, comprehension and writing, but inability to read, anomie for colors, HH on the right (R). CT Skull: hypoattenuating at occipital-temporal region left (L), in addition areas of encephalomalacia in the R occipital-temporal. Electrocardiogram: atrial fibrillation. US Doppler Carotid: no significant stenoses. Magnetic Resonance Imaging (MRI) Skull and Angio-MRI arterial phase: recent ischemia in the L occipital lobe and in L temporal lobe, involvement of splenium of the corpus callosum (CC), diffusion restriction and hypersignal in T2 and FLAIR (Fluid-Attenuated Inversion Recovery); previous ischemic lesion in the R temporal-occipital; hypoflow of bilateral PCA distal branches. Echocardiogram: enlarged L atrium. CT Skull 11 days after ictus with stability. Hospital discharge with Apixaban 5 mg every 12 hours, return to the neurology clinic.

Discussion: Lesion in the L occipito-temporal cortex with involvement of the splenium of CC leads to a disconnection syndrome called AWA. The CC has fibers that connect the two cerebral hemispheres. The occipital lobe and splenium are supplied by the PCA. In addition, PCA infarction L leads to HH on the R, thus, visual information (letters) interpreted in the R visual cortex (visual field L), explaining why the patient can see the letters but not read them.

Conclusion: Strokes are one of the main causes of morbidity. In the topography of the left PCA, we observed AWA.

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315 – What people diagnosed with multiple sclerosis in Brazil feel and how it affects their quality of life and employment status

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Introduction: Multiple sclerosis (MS) affects between 20 and 40 years of age, the peak of their productive life, and can cause permanent neurological disability, which can exert influence on their quality of life, income maintenance and employment relationship.

Objectives: To describe symptoms prevalence in a national wide sample of patients with MS and analyze their impact on quality of life, employment status and use of social benefits.
Methods: We performed a cross sectional, online self-reported survey, concerning demographic, clinical data (using validated scales), employment status and use of social benefits. The study was approved by the University's Ethics Committee and carried through an electronic survey sent to the patients affiliated to Amigos Múltiplos pela Esclerose.
Results: 466 patients answered the survey. Median age of onset, current age and disease duration were 30.2, 40.8 and 10.5 years; median PDDS (Patient Determined Disease Steps) was 2.5, which indicates minor to moderate disability; median MS Impact Scale and MS Walking Scale were 31 and 33%, which denotes minor to moderate quality of life and mobility compromise; 43% of them suffered fatigue and 53% reported not sleeping well. Unemployed patients presented a longer time from symptom onset to diagnosis and higher disability when compared to employed ones. Furthermore, half of the unemployed patients are receiving some social benefit, while only 6.8% of the employed patients.
Conclusion: This study presents symptom prevalence in a national sample of patients with MS and discloses that those with a diagnosis delay and more disability have higher percentage of unemployment and use of social benefits. Strategies to improve earlier diagnosis and better treatment plans can not only reduce patient disability but possibly increase employment retention and lesser use of social benefits.

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317 – Tolosa Hunt syndrome, case report

João Paulo Barile[1], Amanda Freitas Alves[1], Fernanda Maria Gonçalves de Sousa Moura[1], Sonia Maria Cesar de Azevedo Silva[1], Roberta Arb Saba Rodrigues Pinto[1]
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Introduction: Tolosa Hunt syndrome (THS) is a rare condition, incidence of 1/1.000.000 case per year, characterized by unilateral painful ophthalmoparesis caused by idiopathic inflammation in the cavernous sinus. The oculomotor nerve is most commonly involved (80%), followed by abducens nerve (70%), ophthalmic branch of trigeminal nerve (30%), trochlear nerve (29%).
Case presentation: Male, 77 years old, admitted with an acute moderate-intensity orbitofrontal headache on the left, envolving with palpebral ptosis of the left eye. Neurological examination: complete palpebral ptosis on the left and ophthalmoplegia of the entire ipsilateral extrinsic ocular musculature. A complete investigation was carried out: metabolic, rheumatological, serological tests without significant alterations and study of the cerebrospinal fluid with mild hyperproteinorachia, without pleocytosis. Magnetic resonance imaging (MRI) of the skull showed thickening of the cavernous sinus on the left, with contrast enhancement; Angio-MRI of the Skull and Neck without alterations. Therefore, THS was diagnosed and treatment with Methylprednisolone 1 g for five days, with complete improvement of headache and partial improvement of ophthalmoparesis. The patient was discharged with 60 mg of prednisone orally with instructions for gradual weaning off, return to the neurology outpatient clinic.
Discussion: THS diagnosis is based on the International Classification of Headache Disorders: unilateral periorbital headache; granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit on cranial MRI; paralysis of one or more of the oculomotor nerves; the headache must precede the ophthalmoparesis by up to two weeks or appear concomitantly. The exclusion of secondary causes is essential. Treatment of choice is corticosteroids, improvement of headache in the first days, and of ophthalmoparesis in 2–8 weeks.
Conclusion: Unilateral headache with ipsilateral ophthalmoparesis should raise the suspicion of THS.

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318 – Cadasil, atypical and familial presentation – family case report

Renan Iegoroff[1], Rafael Herlan Terceros Vaca[1], Gustavo Araújo Pinheiro[1], Alvaro Marcelo Huchani Huanca[1], Matheus Henrique de Souza Coradini[1], Leonardo Mariano Inácio Medeiros[1]
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Introduction: Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL), is a non-atherosclerotic, non-amyloid, hereditary cerebral disease of small vessels and capillaries caused by mutations in the NOTCH-3 gene located on chromosome 19. The presence of granular osmophilic material (GOM) deposition in the smooth muscle cells of vessel walls is the pathological hallmark of arteriopathy in CADASIL. GOM deposits in the basal lamina of smooth muscle of small vessels are pathognomonic for CADASIL. The presence of GOM in capillary blood vessels of the skin and muscle in biopsy and genetic studies (NOTCH-3 analysis) plays a key diagnostic role. Biopsy tests have high specificity (up to 100%) and low sensitivity (less than 50%). The NOTCH-3 test has been proposed as the primary diagnostic approach, allowing detection of 90% of affected individuals. CADASIL has an estimated prevalence between 2 and 5 in 100,000, and the phenotypic study demonstrates different clinical symptoms in the course of the disease within the same family. The average age of onset of clinical symptoms varies between 48.3 years in men and 52.2 years in women. Characteristic symptoms of migraine, stroke or TIA (transient ischemic attack), behavioral changes, early and progressive cognitive changes associated with leukoencephalopathy on imaging studies. The large association of symptoms often causes the diagnosis of CADASIL to be delayed. In this case, we had an association of gait ataxia within the framework of motor alterations, demonstrating the wide range of symptomatology of the pathology. This case report presents a familial course that started outside the most prevalent age group in the studies described and with an atypical presentation in an affected generation.
Case reports: Case 01: woman, 68 years old, started progressively forgetting to perform household activities after the age of fifty, associated with primarily generalized myoclonic epileptic seizures, evolving rapidly within five years to walking apraxia with the use of a wheelchair and tonic-tonic epileptic seizures. bilateral clonic disorders, comprehension aphasia and bradypsychism. Relatives report previous migraine without chronic aura and REM (rapid eye movement sleep) sleep behavior disorder (RMSD). On neurological examination, severe ataxia with bilateral dysdiadochokinesia associated with bilateral hypometric index-index. Bilateral ROT 4+/4+ with bilateral Hoffman and Babinski signs. MMSS and MMII with FGM 4/5 proximal and distal. Case 02: woman, 36 years old, pastry chef, had episodes of forgetting about everyday activities of her work, progressive in the last three years (cake recipes, budget accounts, orders placed) associated with confusion for spatial location on the way home/work, evolving to apraxia in writing letters and words and difficulty with calculations associated with monoparesis of the right lower limb for twelve months with progression to paresis of the lower limbs after six months and evolution to paresthesia of the upper limbs for three months. Associated with the condition, he has migraine without chronic aura and RMSD. The neurological examination showed Mini-Mental State Examination 22/30 (expected score of 29), list of animals in one minute: 09 animals; list of words starting with "F": 03 words; clock test: 2/4; difficulty with calculations and digital agnosia with

right/left apraxia; Upper limbs: eutrophic, FMG 4/5, bilateral distal; FMG 5/5 bilateral proximal; Lower limbs: eutrophic, bilateral FMG 4/5 distal and proximal with positive Mingazini; atypical gait with evidenced weakness in heel, toe and tandem gait; Bilateral dysdiadochokinesia with eumetric, slowed index-index; ROT 4+/4+ in the right side with positive Hoffman and Babinski signs. MRI Brain (17/09/2020): extensive area of hypersignal on T2 and FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) involving the periventricular white matter in all lobes without atrophic or expansive effect; Case 03: woman, 45 years old, started behavioral arrest epileptic seizures at the age of thirty-two, progressing to focal dysperceptive seizures with progression to bilateral tonic clonic seizures after eight months and multiple episodes of anterograde amnesia, presenting forgetfulness related to everyday work activities (exchanged worksheets , payments, calculation errors and budgets); associated with the condition presented migraine without chronic aura and RMSD. Genetic Test (04/06/2017): Heterozygous alteration in exon 8 of the NOTHC – 3 gene.

Discussion: CADASIL presents a rare cause of cognitive decline and is often overlooked in diagnosis, except in cases of high clinical suspicion in a familial course. Access to imaging tests becomes fundamental for the diagnostic segment and the primordial genetic test for etiological elucidation and family planning, in the report described the family presentation with the same course of satellite symptoms (migraine without aura, and RMSD) associated with cognitive alteration with anticipation of age of onset are hallmarks of clinical thinking. Cases described in the literature show that the clinical symptomatology is not necessarily related to the level of brain injury observed in the imaging exam, which could be explained by personal factors and which exon is affected. The NOTCH-3 gene has 24 exons, in which the literature reports exon 4 as the most common mutation, followed by 3, 5 and 6, mainly in the Caucasian population. In an Asian population, the most affected exon is 4 and 11, which is also found in Italian descendants. The mutation in exon 8, described in the clinical case, is found in a population of Portuguese origin, being the second most common mutation in this nationality, behind the mutation in exon 4. Brain MRI studies have tried to elucidate the most affected brain regions, aiming to trace a line of evolution. Involvement of the temporal lobe, external capsule and corpus callosum are described as probable markers of CADASIL, and can be used as an aid in the diagnosis due to its specificity of 86% and sensitivity of 89%. Studies also show that the frontoparietal area has frequent findings of hyperintensity (100%), followed by the temporal lobe (83%), less frequently affecting the brainstem, occipital lobe and cerebellum. Despite the great advances in the specialized literature, the causes of the important cognitive dysfunction presented in the course of the evolution of CADASIL remain unclear. However, studies have suggested that the process of cognitive decline is more related to the loss of cortico-subcortical connections than to brain atrophy itself, with these disconnections resulting from repeated transient ischemic accidents. Science has been looking for ways to change the prognosis of CADASIL, recent studies in gene therapy and neurogenetics show the importance of thinking about this pathology as a genetic disease of great importance to change the prognosis of this pathology.

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327 – Diagnostic yield of a NGS panel in Brazilian patients with sporadic peripheral neuropathy

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Introduction: The etiology of sporadic peripheral neuropathy (PN) remains unclear in a significant proportion of patients. In European and North American surveys, genetic causes have been increasingly recognized in these patients. Little is known about Latin American populations.

Objective: We assessed the diagnostic yield of a comprehensive next generation sequencing (NGS) panel in a cohort of Brazilian patients with PN without family history. Potential predictors of genetic etiology in this group were further explored.

Methods: We evaluated 41 consecutive adult patients regularly followed with PN and 1. Negative family history of PN or other neurological disorders and 2. Unclear etiology despite extensive laboratorial/neurophysiological (Nerve Conduction Study+Electromyography [NCS+EMG]) work-up. All patients then underwent genetic testing using a comprehensive (NGS) panel that included 72 genes known to cause PN. Only pathogenic or likely pathogenic variants according to American College of Medical Genetics and Genomics criteria were retrieved. For each subject, we recorded demographic, clinical and NCS+EMG data. Such variables were then compared between positive vs negative NGS subgroups using Fisher exact test ($P < 0.05$).

Results: Fifteen patients had diagnostic NGS results (8 men, median age = 43 years old), whereas 26 patients (15 men, median age = 47 years old) had negative or unconclusive results. Diagnostic yield of the PN panel was 15/41 = 36.5%. Seventeen distinct variants were found in 5 different genes; *PMP22* and *SH3TC2* were the most frequently identified. Earlier age of PN onset ($P = 0.013$), demyelinating pattern on NCS+EMG ($P = 0.021$) and presence of distal atrophy on clinical examination ($P= 0.043$) were associated with positive NGS results.

Conclusion: A significant proportion of Brazilian patients with sporadic PN has genetic etiology. NGS emerges as a diagnostically useful tool for idiopathic PN, particularly when there is earlier age at onset and demyelinating features.

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330 – Prognostic factors related to mortality in motor neuron disease at an outpatient care center in Salvador (BA)

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Introduction: Motor neuron disease is neurodegenerative and the understanding of its prognostic factors is crucial promoting better quality of life for patients. Epidemiological, clinical and nutritional factors have been described as influencing disease progression.

Objectives: To describe prognostic factors in patients with Motor Neuron Disease at a specialized outpacient care center in Bahia.

Methods: This is an observational, and cross-sectional study, carried out by collecting data from medical records of patients with diagnosis of amyotrophic lateral sclerosis (ALS), using forms to acess epidemiology data, clinical characteristics, and nutritional assessment in the beginning of treatment and the time of assessment. The time from the first symptoms until the application of palliative measures was also recorded.

Results: 27 patients were enrolled in the study, 51.8% men and 41.8% women, with an average age of 55±11.1 years at diagnosis. No patients were diagnosed with frontotemporal dementia (FTD) and 3.7% had family history of ALS. Weight loss was observed in 95.8% of patients. Among the elderly population, 9% were classified as low weight at the first evaluation,

and 81% were classified as such during data collection. Among the adult population, 8.3% were classified as malnourished at their first evaluation, and 27% during data collection. About 81% of the population used some palliative measure. The time between symptoms these measures were of 32 ± 18.6 months for gastrostomy, 38 ± 41.1 months for AMBU use and 49 ± 43.9 months for BiPAP use.

Conclusion: A similar distribution of prognostic factors compared with literature was observed, with some particularities such as earlier age of onset, low prevalence of familial ALS and absence of FTD. Significant weight loss was observed, especially in those with bulbar ALS and in the elderly population, which is associated with a poor prognosis. Time from symptoms to palliative measures varied, but was shorter on average in patients with the bulbar form of the disease.

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331 – In vivo assessment of striatal compartments in patients with idiopathic upper limb dystonia

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Introduction: The striatum is an essential hub in the motor system associated with dystonia and other movement disorders. The function of the striosomes and matrix in motor control is not clear. A recently developed method using diffusion tensor imaging (DTI) enables us to distinguish compartments of the striatum, namely matrixes-like and striosomes-like voxels.

Objectives: To access striatal matrix and striosome compartments in patients with idiopathic upper limb dystonia using diffusion tensor imaging.

Methods: We analyzed 3T magnetic resonance imaging (MRI) images from 26 patients with idiopathic upper limb dystonia aged 43.88 ± 11.32 years (standard deviation, SD; range 19–60) with a mean disease duration of 12.55 ± 10.25 years (SD; range 1–25) and healthy controls aged 39.42 ±1 1.42 years (SD; range 19–58). The striatum was parcellated by targeting cortical regions that favored striosomes and matrix-favoring areas. The bilateral striatum was assessed for changes in volume and mean fractional anisotropy value.

Results: Patients show significant reductions of left Matrix-like voxels volume relative to controls (P = 0.022), with a moderate effect size (Cohen's d = 0.640). No difference was observed in the right striatum compartments.

Conclusion: By parcellating the striatum into striosome and matrix-like voxels, we showed that patients with idiopathic dystonia have a reduced volume in matrix-like voxels in agreement with anatomopathological findings of some genetic types of dystonia. Even in non-degenerative dystonias, volume differences may reflect an imbalance between matrix and striosome signaling, ultimately favoring the direct pathway.

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334 – National survey on neurocritical care education in Brazil: preliminary data

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Introduction: Neurocritical care (NCC) education involves integrating knowledge and skills from various complex areas. While the United States of America and Europe regulate didactic core training for medical residencies and fellowships, the quantity and the quality of such training in Brazil remain unclear.

Objectives: To understand how NCC training is currently performed in Neurology, Neurosurgery and Intensive Care residencies in Brazil.

Methods: A virtual survey was developed with 27 questions about the existence, duration, teaching method, quality of supervision and skills required in NCC training during medical residency. The planned sample was 390 answers.

Results: 120 participants from 41 Institutions and 8 Brazilian states replied. Median age was 36 years (26–76 years). About 66% were neurologists and 25% intensivists. Of them, 57% had training in NCC, 65% as mandatory rotation, 60% lasting more than 4 weeks, predominantly in the first 2 years of the course (42 and 45% respectively). Training happened in general Intensive Care Units (48%) with 5–20 beds (65%) under the supervision of intensivists (66%). The methodology most used was bedside discussion (98%). Among procedures skills, more than 80% reported having performed central venous catheter insertion, orotracheal intubation and neuroimaging discussion. The least performed skills were transcranial doppler (21%), electroencephalogram interpretation (20%), neurological multimodal monitoring (25%). It was highlighted that after training, few participants feel comfortable with the following diagnoses: control of intracranial hypertension (48%), spinal cord injury (39%), care for potential donors (42%), post cardiac arrest care (52%).

Conclusion: NCC education in Brazil is currently heterogeneous and at an early stage of development. Further studies are necessary to identify areas in need of improvement and promote advancements in the field.

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335 - Bilateral thalamic ischemic lesion mimicking bickerstaff brainstem encephalitis – a potential pitfall

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Introduction: Bickerstaff Brainstem Encephalitis (BBE) is a rare immunologic disorder that often occurs after an infection and is characterized by ataxia,

external ophthalmoplegia, and consciousness impairment. While it is a known differential diagnosis for posterior circulation syndromes, this case report highlights the potential pitfall of misdiagnosing a bilateral thalamic ischemic lesion as BBE.

Methods: Herein we report a single case based on a retrospective analysis of the patient's medical records.

Results: A 58-year-old man was admitted due to progressive incoordination, unsteadiness, and walking difficulty for seven days. His relatives reported drowsiness and horizontal bilateral diplopia three days before admission. His medical past was relevant for mild coronavirus disease 2019 (COVID-19) infection 40 days before neurological symptoms onset, arterial hypertension, obesity, and untreated obstructive sleep apnea. Neurological examination revealed global deep tendon areflexia, axial ataxia, right eye exotropia, and drowsiness. An initial computed tomography brain scan was normal, and cerebrospinal fluid analysis revealed cytoalbuminologic dissociation (cell count: 2.6/mcL and protein 0.82 g/L). A diagnosis of suspected BBE was made due to clinical and laboratory findings, and COVID-19 was identified as a possible trigger. Intravenous Human Immunoglobulin (0.4 g/kg.day) for five days was administered. The patient worsened despite the treatment. Alternative causes were therefore evaluated. Elevated troponin and echocardiogram were compatible with perimyocarditis. Brain magnetic resonance imaging revealed a bilateral thalamic lesion suggesting a ischemic lesion. Percheron artery infarct was suspected; however, digital angiography was not done due to the patient's deteriorating clinical condition.

Conclusion: BBE is a rare condition that may be mimicked by stroke, especially in the posterior circulation. As observed in this case, minor laboratory abnormalities could be a potential pitfall. The presence of risk factors for cerebrovascular disorders should be considered, even if the onset is gradual and subacute.

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339 – Stroke in brainstem topography after wasp sting: an unusual complication

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Introduction: Insects stings such as wasps and bees are common occurrences, especially in tropical countries, with local symptoms as the main manifestation, followed by rarer systemic complications such as severe anaphylactic reactions. Very uncommonly, patients may develop neurological manifestations such as stroke, which may occur by mechanisms that are still poorly studied, but probably linked to the inflammatory, thrombogenic and vasoactive venom's characteristics of these arthropods.

Objectives: Report the case of a patient with stroke as a complication after a wasp sting, developing symptoms of incomplete Wallenberg syndrome.

Methods: The information was obtained by the review of medical records and from the literature review about this topic.

Results: Through medical record review, we found the history of a previously independent adult patient for basic activities, who woke up with hypoesthesia and tingling sensation at the right limbs, after he having been stung by a wasp the day before. The patient presented recurrence of these deficits 3 times within 30 minutes, followed by a pulsatile left hemicranial headache. After that, he complained of vertigo, nausea and vomiting, as well as appendicular ataxia to the left, vertical nystagmus, dysarthria and hypophonia. The patient denied weakness and diplopia. In addition, conjunctival and palpebral edema, angioedema and infarcted lymph node next to the sting's location, were noted. During investigation, a diffusion-restricted lesion was detected at the left dorsolateral region of the medulla oblongata, as well as unaltered angiotomography. Finally, angiography showed maintained permeability of posterior-inferior cerebellar artery and the other arteries of the posterior circulation, with only a small parietal irregularity in V4 to the left.

Conclusion: Although rare, wasp sting is a possible cause of stroke due to thrombogenic and vasoactive mechanisms, and may be suspected when patients present with a temporally compatible history of sting and exclusion of other causes of stroke.

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341 – Mild cognitive impairment and early onset Parkinson's disease: an observational study

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Introduction: Early Onset Parkinson´s Disease (EOPD) is defined as an onset of motor symptoms of Parkinson´s disease (PD) after age 21 years and before 50 years. According to the Movement Disorder Society, PD Mild Cognitive Impairment (PD-MCI) can be defined as performing 1–2 standard deviations (SD) below appropriate norms in at least two neuropsychological tests within the commonly affected domains in PD (i.e., attention and working memory, language, executive function, visuospatial, and memory) in the absence of significant interference with functional independence. People with EOPD face unique challenges compared with those with late-onset PD since they potentially have more active roles in society and a longer time horizon of disease.

Objectives: We aimed to establish the PD-MCI frequency in EOPD patients and to define which subdomains were compromised in our patients.

Methods: 69 EOPD patients underwent an assessment including Mini-Mental State Examination, Frontal Assessment Battery, Clock Drawing Test, Hopkins Verbal Learning Test-Revised (HVLT-R), Trail Making Test A and B, Forward Digit Span and Backward Digit Span, Stroop Test, Beck Depressive Inventory, UPDRS-3, UPDRS-4, HY and UDysRS.

Results: The median (interquartile range [IQR]) age at PD onset was 43 years (23–49), the median (IQR) PD duration was 7 years (2–27), the median (IQR) LEDD was 800 mg (100–2430). The MCI frequency in this group was 40.6%. Patients were distributed in two groups (with and without MCI) and the scores between the groups had significant difference in most cognitive scales except HVLT-R.

Conclusion: Our study described a high frequency and a profile of MCI in EOPD patients (attention/working memory and executive function). We suggest that clinicians must search for neuropsychological impairments in EOPD in order to provide a more comprehensive treatment that includes multidisciplinary interventions.

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342 – Guillain-Barré syndrome associated with froin syndrome: case report

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Case report: A 31-year-old man was referred to our service after experiencing paresthesia, followed by distal weakness in both his feet and hands. These symptoms occurred 20 days after the administration of the fourth dose of the ChAdOx-1 COVID vaccine. A lumbar puncture was performed, which showed remarkably thick cerebrospinal fluid (CSF). Biochemical analysis of the CSF revealed 920 mg/dL of protein and a white cell count of 7.3/mm^3. An electromyography (EMG) was performed, which was compatible with the diagnosis of Guillain-Barré Syndrome (GBS). The patient was treated with intravenous immune globulin and showed clinical improvement. One month later, he experienced a recurrence of symptoms and underwent a new EMG, which again showed motor nerve conduction blocks. A new lumbar puncture was performed, and a thick fluid was obtained, with a protein concentration of 804 mg/dL. CSF flow blockage was suspected and the patient underwent a suboccipital puncture, which showed a more watery CSF, with a protein concentration of 227 mg/dL. A spinal magnetic resonance was performed, revealing an arachnoid web at the T3–T4 level. GBS relapse was presumed and the patient was treated with five sessions of therapeutic apheresis, which improved his strength and sensory symptoms.
Discussion: Blockage of CSF circulation at the spinal level can produce fluid with abnormally high levels of protein, which in this case was a confounding factor during the clinical investigation.
Conclusion: We report a case of concurrent GBS and spinal block, which led to abnormally high CSF protein levels during lumbar puncture, causing diagnostic ambiguity during the investigation.

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344 – Case report: ischemic stroke in a young woman

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Introduction: Ischemic stroke is responsible for about 62% of all stroke cases. The lifetime risk of having a stroke is approximately 25%. Worldwide, stroke is one of the leading causes of mortality and disability. The causes are diverse, and the etiology must always be investigated in order to initiate specific treatment and adequate prophylaxis.
Objectives: To report a case of ischemic stroke in a young woman after bronchoscopy with endotracheal tube replacement on an elective basis.
Methods: Information was obtained through clinical follow-up in a neurology ward.
Results: Patient, female, 27 years old, married, brown, with a history of polytrauma in 2015, after a bicycle accident, with the need for hospitalization in the intensive care unit, remaining intubated for nine days. Neurosurgical intervention was not necessary. The patient was discharged without any device, remaining functional, but two months later, she evolved with dyspnea, being diagnosed with tracheal stenosis, thus, a tracheostomy was necessary. In an outpatient routine, she was hospitalized for endotracheal tube replacement in October 2022. In the immediate postoperative period, she presented desaturation, requiring orotracheal intubation, sedation and breathing on mechanical ventilation. As she woke up weakly after ceasing sedation, a neurological investigation was initiated, being diagnosed with multiple areas of ischemic infarcts and identifying patent foramen ovale (PFO), moderate-grade paradoxical transseptal shunt as the most likely cause of the ischemic stroke.
Conclusion: Young patient with PFO submitted to mechanical ventilation, after an elective procedure, evolving with worsening of the right-to-left shunt, resulting in ischemic stroke with a high degree of disability.

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345 – Epidemiologic review: an analysis of the incidence of botulism in southeast Brazil from 2011-2021

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Introduction: Botulism is a rare life-threatening condition caused by Clostridium Botulinum bacteria toxins, which attack the various parts of the nervous system, like nerves, spinal cord and brain, causing symptoms, like dry mouth, facial weakness, trouble breathing and, in the worst cases, even paralysis. Being, for all the risks that this disease represents, a very important object of study and observation.
Objectives: Analyze the epidemiological profile of confirmed cases of botulism in the Southeast region of the country during the years 2011–2021.
Methods: This work was made using the data, from the Information System for Notifiable Diseases of the Information Department of the Unified Health System of Brazil, about the botulism. The data has been selected to represent only the 4 states, São Paulo (SP), Rio de Janeiro (RJ), Minas Gerais (MG) and Espírito Santo (ES), of the southeast region.
Results: The Brazil had 60 cases of botulism in the analyzed period, with 41.5% of them being in the Southeast region. SP had the most cases in the region with 56% of them, followed by MG with 24% and by RJ with 20%. No cases have been reported in ES. 92% of the reports were foodborne botulism showing an absolute prevalence of this spread form. With a deviation of ±1.35 in the botulism cases over the years. Among those diagnosed, 48% were women and 52% men. Still, 56% of the people were white.
Conclusion: With the data collected, it is possible to perceive a stable wave of incidence throughout the entire period, with major regional distortions. In addition, the overwhelming prevalence of the foodborne form of the disease is noticeable. however, due to the small number of cases, much of the data suffer from poor sampling and therefore may be biased, such as gender and race data, thus requiring more longitudinal studies to reach relevant numbers for this type of analysis.

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346 – Case report: stiff Person syndrome

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Introduction: The rigid person syndrome is rare, characterized by blockade of the GAD enzyme, responsible for inhibiting muscle contraction. Although it is not mandatory for the diagnosis, most patients are positive for Anti-GAD.
Objectives: To report a case of rigid person syndrome seen at Hospital Geral de Goiânia.
Methods: Information was obtained through clinical follow-up in a neurology ward and outpatient clinic.
Results: Patient, female, 32 years old, complaining of paresthesia ascending to upper limbs, worsening over a period of six months with paresis with gait impairment, increased tone and muscle spasms. Patient with multiple consultations with a neurologist and psychiatrist, using polypharmacy. During hospitalization, the patient reported severe pain in the limbs. On examination, she had grade 3 strength and quadrisegmental hypertonia with intermittent periods of generalized spasm, exacerbated during periods of greater anxiety. Hypothesis of rigid person syndrome was raised and anti-GAD 1,680 results were obtained. The patient responded well with high doses of diazepam, baclofen and pregabalin.
Conclusion: The case represents a definitive diagnosis of rigid person syndrome, a rare pathology that affects 1–2 patients per 1,000,000, mainly females. This study contributes to future research in this area since knowledge in this area is still scarce.

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347 – Literature review: the efficacy of mirror therapy in patients with phantom limb pain

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Introduction: Phantom Limb Pain (PLP) is a common symptom after an amputation, affecting more than 80% of the people who experience this procedure. The mirror therapy, based on a reflective illusion is a hopeful possible treatment for this condition, as it tricks the brain to believe that the amputated member is still there, allowing for pain management.
Objectives: The objective of this literature review is to search through clinical trials in order to collect data to relate the efficacy of Mirror Therapy in the treatment of the pain condition on PLP, establishing the possible reduction of the pain and the duration of the improvements caused by the procedure.
Methods: This review was made using the descriptors "Phantom Limb pain", "Mirror Therapy" and "Phantom Limb Syndrome" on the PubMed and New England Journal of Medicine advanced search mechanisms. The selected randomized Controlled trials totaled in 10, composed of articles from the last 16 years that fit the research's purpose.
Results: 9 out of the 10 trials used on this research showed improvement in the subjective pain of the patients on short term with the use of Mirror Therapy in both arms or legs. Some studies had even 100% of the patients with reductions on pain intensity, among a total pain reduction in the Visual Analogue Scale between 30% to 87% of improvement in all the trials. However, was not possible to determine the maintenance of this effect on a long term.
Conclusion: As the first experiments of mirror therapy demonstrate, the treatment seems to have good efficacy in the reduction of the pain on the PLP condition, but there is still a need for more research to determine the extent of the improvements on a longer schedule and it was not possible to create a consensus over the total efficacy of the procedure due to the different types of amputation and the low number of patients in each trial.

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350 – Cerebral watershed infarct as the first manifestation of systemic lupus erythematosus and antiphospholipid syndrome

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Introduction: Cerebral watershed infarcts are strokes that affect regions supplied by two main arterial territories. It is usually related to hemodynamic compromise or hypercoagulable state as noted in antiphospholipid syndrome. We present a case of a young patient, who developed this type of stroke as the initial manifestation of antiphospholipid syndrome (APS) and systemic lupus erythematosus (SLE).
Objectives: To report an unusual case of watershed zone stroke in the context of APS and SLE.
Case report: A previously healthy 15-year-old male patient presented fever and myalgia followed by dropped head and quadriparesis, which progressively worsened in the next five days. Initial neurological examination revealed quadriparesis, predominantly in the proximal muscles MRC grade II with hypertonia and pyramidal signs. Brain magnetic resonance imaging evidenced acute internal borderzone infarct in the territories of the anterior cerebral artery, and middle cerebral artery, bilaterally. Vessel wall imaging and cerebrospinal fluid showed no evidence of vasculitis. Computed tomography angiography revealed no large vessel occlusion, aneurysm or dissection, but showed right sigmoid sinus thrombosis. Laboratory tests revealed pancytopenia(hemoglobin 8,9; white blood cell 3510, 580 lymphocytes and 71.000 platelets) with positive direct Coombs, complement consumption, elevated erythrocyte sedimentation rate (47 mm/1st hour), positive anti-ds-DNA (>1:1280), anti-SSA and anti-SM-RNP. The Lupus anticoagulant, anti-beta2GPI and anticardiolipin antibodies were positive, filling laboratorial criteria for SLE and APS. Transthoracic and transesophagic echocardiography were unremarkable. The patient was treated with high-dose corticosteroids (1.5 g methylprednisolone in three days) and anticoagulants, presenting a progressive recovery of strength.
Conclusion: This case report reasures the important correlation between borderzone infarcts and hypercoagulable states. Antiphospholipid syndrome must be always remembered in these patients.

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351 – Atypical case of Wilson's disease presenting with unusual neuroimage features

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Introduction: Wilson's disease (WD) is a rare autosomal recessive disorder resulting in hepatic and neurological symptoms. Uncommon presentations are prone to diagnostic and treatment delay.
Objectives: Describe an atypical case of WD and improve clinicians awareness.
Case report: A 19 years-old male presented to us with seizures and upper limb tremor in the past three years, evolving to torsion-like movements in limbs and trunk. He was unable to open his eyes and had severe dysphagia. There was no consanguinity, perinatal complications or neurodevelopmental delay. Examination revealed generalized dystonia, blepharospasm, retrocollis, dysarthria, trismus, right hemiparesis and marked muscle wasting; there was a gray halo around both irises. The patient was bedridden and severely dependent. Magnetic resonance imaging (MRI) showed T2/FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) hyperintense lesions in the left hemisphere, basal ganglia and cerebellar peduncles. Laboratory workup revealed low ceruloplasmin levels (< 8 mg/dL; range: 16–45), serum copper of 44.2 mcg/dL (range: 70–140). A diffuse increased echogenicity was described in liver ultrasound. The patient had no clinical or laboratorial evidence of liver damage. Due to presumed Kayser-Fleisher rings, movement disorders and MRI findings, WD was suspected. Genetic test revealed two variants in the ATB7B gene, probably a compound heterozygosity. Having a final Leipzig score of 7, WD diagnosis was established.
Conclusion: WD may present with cortico-subcortical involvement. Hepatic subclinical involvement may delay WD diagnosis. It is important to have a high index of suspicion for WD in any movement disorder in young people. In order to prevent treatment delay and severe neurologic sequel, neurologists should be aware about atypical clinical and radiological presentations of this treatable disease.

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353 – Disease associated with anti-MOG antibody: a case report

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Introduction: Anti-MOG Antibody Associated Disease (MOGAD) is an autoimmune neurological condition with antibodies directed against the myelin glycoprotein of oligodendrocytes, with a monophasic or relapsing course. Symptoms include optic neuritis (ON), transverse myelitis, cerebral, brainstem, and cerebellar dysfunction. The diagnosis is based on the clinical picture and complementary exams, mainly the anti-MOG antibody. Treatment involves, in the acute phase, pulse with corticosteroids, plasmapheresis and, especially in children, immunoglobulin, and in the chronic phase (recurrent course), immunosuppressants, immunobiologicals and, especially in children, monthly immunoglobulin.
Objectives: To report a case of MOGAD.
Methods: Review of medical records and brief literature review.
Results: A 16-year-old male patient started with headache and fever, and the next day, low visual acuity (LVA) in the right eye (RE) with gradual worsening, and after two weeks, amaurosis in the RE and LVA in the left eye (LE). He received methylprednisolone pulse for five days, with improvement. After three weeks, he presented a new bilateral LVA, with amaurosis in the LE. Opted for new pulse therapy. Relevant findings included cerebrospinal fluid (CSF) with 15 cells, absence of oligoclonal bands in blood and CSF, neuraxial magnetic resonance Imaging with extensive bilateral ON affecting up to the optic chiasm, with gadolinium enhancement, and perineuritis. After results of non-reactive antiaquaporin 4 and reactive serum antiMOG, the diagnosis of MOGAD was confirmed, prednisone was maintained and azathioprine was prescribed. There was complete visual recovery afterwards.
Conclusion: This case sheds light on the diagnosis of an uncommon demyelinating disease that is MOGAD, however, a condition that should be remembered as one of the differential diagnoses of ON, especially in patients with atypical ON and in children.

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356 – Vertebral arterial dissection in a young patient, case report

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Introduction: Cervical artery dissection accounts for 1–2% of all ischemic strokes and is a common cause of stroke in young people, in which it represents 10–25%. Some studies have reported an increased risk of stroke in patients who present with a dissection, with most occurring soon after the onset of symptoms.
Objectives: To report a case of Vertebral Arterial Dissection in a young patient after Direct Trauma, treated at the Hospital Geral de Goiânia.
Methods: Information was obtained through clinical follow-up in a Neurology ward and outpatient clinic.
Results: Patient, female, 45 years old, reports direct trauma in the right cervical, frontal and left occipital regions. After one day, he suddenly presented right hemiparesis, right peripheral facial paresis, diplopia, vertigo and nausea, dysarthria. On examination, right horizontal gaze palsy, diplopia, right peripheral facial paresis, right curtain sign, tongue deviation to the left. Complete hemiparesis provided grade 3 on the right, tetrasegmental hyperreflexia, Trommer, Hoffman and Babinski on the right. Evidenced on Brain Magnetic Resonance Imaging recent ischemia in the base and pontine tegment lower right and higher left. Cervical Magnetic Resonance Angiography with signs of dissection of segment V3 of the Right Vertebral Artery. Antiplatelets therapy was proposed, evolving with good recovery of the deficits.
Conclusion: The literature shows that patients with arterial dissection have head or neck pain, stroke and Horner's syndrome. Vertebral artery dissection is believed to be a multifactorial process, intrinsic factors present in the setting of an exacerbating extrinsic factor (low-speed car accident, direct trauma, heavy lifting, or a rotational sports injury). As for therapy, more research is needed, there is no evidence of superiority of anticoagulation or antiplatelets therapy.

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357 – Pseudotumoral demyelinating lesion in a teenager, case report

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Introduction: Pseudotumor lesions are an atypical presentation of demyelinating diseases, commonly associated with Multiple Sclerosis (MS) of different varieties (Balo, Schilder and Marburg) and occasionally linked to other demyelinating processes, such as MOGAD, ADEM and NMOSD. Estimated prevalence of three cases per million per inhabitant and occurrence of 1.4–8.2% in patients with MS.
Objectives: To report a case of Pseudotumoral Demyelinating Lesion (PDL) in a teenager, attended at the General Hospital of Goiânia.
Methods: Information obtained through clinical follow-up in a Neurology ward.
Results: Male, 14 years old, onset of seizures 40 days ago. Ten days ago, he presented left crural monoparesis. On examination, hemiparesis of left crural predominance, left hyperreflexia, patellar and plantar clonus, left hemihypoesthesia. Normal cerebrospinal fluid (CSF), negative oligoclonal bands and oncotic cytology. Brain magnetic resonance imaging (MRI) showed an expansive cortical and subcortical lesion in the superior frontal gyrus and paracentral lobe on the right, cingulate gyrus and trunk of the corpus callosum on the right, with hypersignal on T2/FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) and focal areas of gadolinium enhancement, predominantly subcortical. Spectroscopy increased Choline and reduced peaks of N-Acetyl Aspartate. The perfusion study showed cold perfusion. Proposed therapy with high doses of intravenous corticosteroids for five days, with improvement of the deficit. A control MRI showing partial regression of the lesion after therapy, therefore the first hypothesis is a PDL.
Conclusion: PDL is a diagnostic challenge, as they mimic tumors and abscesses. Diagnosis based on the combination of clinical evolution, neuroimaging, CSF and brain biopsy, when appropriate. Initial treatment with high doses of intravenous corticosteroids, and most patients has good response. This case highlights pseudotumoral demyelinating diseases in teenage.

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358 – Results obtained with cranial nerve block in the treatment of migraine in patients assisted in a reference hospital

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Introduction: The performance of Cranial nerve blocks (CNBs) in migraine has been increasingly used. CNBs are generally safe and well-tolerated procedures that can be performed in either emergency or outpatient settings. There is currently no guideline standardizing CNBs in migraine. In clinical practice, as well as the few published studies, the results are encouraging, justifying further studies in the area.
Objectives: Evaluate the clinical response, safety and tolerability to the CNBs performed in patients with diagnostic criteria for migraine accompanying in the headache clinic in a reference hospital, and evaluate the response through objective scales (HIT 6 and EVA) evaluating comparatively pre and after application in a time interval of 60 days.
Methods: Cross-sectional and retrospective observational clinical study in a convenience sample, in which the response to the CNBs performed in patients with migraine diagnostic criteria who accompany the headache clinic in a reference hospital, through the application of objective scales (HIT 6 and EVA) comparatively evaluating pre and after application in a time interval of 60 days, in the period from January 2021 to July 2022.
Results: There were 15 patients included in the present study. The evaluation of the averages from EVA and HIT 6 pre and after application was statistically significant with $P < 0.001$. Only 6.66% (1) complained of some adverse effect such as initial insomnia. 93.33% (14) denied any post-application adverse effect. Unilateral and bilateral application, type of drug used in nerve block and prophylaxis concomitant also showed statistical relevance in the variation of the VAS scale.
Conclusion: Despite the limitations and the reduced sample size, restricting to a more conclusive interpretation, the findings point to a potential therapeutic benefit and for the safety of the CNBs, justifying the performance of more studies in the area in order to create a protocol for the practice of CNBs.

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359 – Neurocysticercosis with spinal cord involvement, case report

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Introduction: Cysticercosis is caused by the larval stage Taenia solium. Clinical manifestations are divided into neurocysticercosis (intra- or extraparenchymal) and extraneural cysticercosis. Despite being an endemic disease in Brazil, in only 1% of cases it occurs with spinal cord injury. The aim is to report a case of subarachnoid neurocysticercosis with spinal cord involvement in a patient at Hospital Geral de Goiânia. Information was obtained through clinical follow-up in a neurology ward.
Case report: Female, 59 years old, patient with initial stabbing back pain at rest, evolving with lower limb paresthesia and progressing to paraparesis and paraplegia with urinary and fecal retention. On examination, areflexia in the lower limbs, Babinski and alteration of proprioception and palesthesia. On thoracic spine magnetic resonance imaging, the presence of a signal suggesting a subarachnoid blood component in metabolization and loculation of an anterolateral cystic configuration on the right at T4-T5 level, compressing and displacing the thoracic spinal cord, in addition to an alteration in the distribution of the roots of the cauda equina adjacent to the medullary cone with post-contrast intradural radicular enhancement in the equine tail, suspicious findings for arachnoiditis. The patient was urgently referred for neurosurgery, which, during the procedure, showed the presence of cysticercus and also the anatomopathological examination.
Conclusion: Despite being an endemic disease in Brazil, extraparenchymal neurocysticercosis with spinal cord involvement is rare, and it is necessary to report it in order to include it in the differential diagnoses of spinal cord diseases.

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360 – Neurosarcoidosis with meningeal impairment, cranial nerves and peripheral neuropathy, case report

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Introduction: Sarcoidosis is an immune-mediated disease in which there is granulomatous inflammation of the affected organs. Neurosarcoidosis occurs in 5–10% of all patients with sarcoidosis. Treatment is based on expert opinion and there are no randomized clinical trials in this regard. The aim is to report a case of neurosarcoidosis diagnosed at Hospital Geral de Goiânia. Information was obtained through clinical follow-up in a neurology ward.
Case report: Female, 58 years old, reports that three years ago she had paresthesia evolving with tetraparesis, rotatory vertigo, holocranial headache radiating to the neck and worsening with decubitus, hypoacusis, blurred vision and episodes of horizontal diplopia. On examination, saccadic fragmentation, mild anisocoria, fundoscopy bilateral papillary blurring and reduced vessel caliber, hypotonia and hypotrophy, grade 4 tetraparesis, brachial hyporeflexia, crural areflexia, gait ataxia, heel-knee dysmetria. During hospitalization, she developed polyuria and severe hyponatremia, water restriction and fludrocortisone were proposed, followed by prednisone. Brain Magnetic Resonance Imaging raised the hypothesis of carcinomatosis or an infectious process of the meninges. Screening for neoplasms was negative. Performed a meningeal biopsy suggestive of amyloidosis. However, evolution was not compatible with amyloidosis. Reviewed medical records, found axillary lymph node enlargement on chest computed tomography, performed biopsy, with diagnosis of sarcoidosis.
Conclusion: Neurosarcoidosis is rare and its treatment is based on expert opinions. The case described represents Neurosarcoidosis with involvement of the meninges, cranial nerves and peripheral neuropathy, treated with corticosteroid therapy, with good therapeutic response. This study contributes to future research in this area, since knowledge related to sarcoidosis is still scarce and more research is needed.

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361 – Case report: amyloidosis

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Introduction: Amyloidosis is caused by the deposition of low molecular weight proteins in the extracellular space. Amyloid deposits can cause several clinical manifestations, according to the type, quantity and location, among them: renal and cardiac failure, neuropathy and joint manifestations. The definitive diagnosis is made by tissue biopsy, showing positivity for Congo red. Treatment is complex and depends on the type, but includes, for example, chemotherapy and hematopoietic cell transplantation. The aim is to report a case of amyloidosis in a patient at Hospital Geral de Goiânia. Information was obtained through clinical follow-up in a neurology ward.
Case report: Male patient, 40 years old, with visual clouding and progression to bilateral total amaurosis. During the initial neurological investigation, signs of optic neuritis were found and the patient underwent pulse therapy, with no improvement. When returning to the neurologist, an Hb of 5 was found, as well as a significant change in renal function, and he was referred to the General Hospital of Goiânia for investigation. The patient had bilateral amaurosis, mydriatic pupils with mild plegia, without other neurological alterations, however, with systolic murmur, in addition to polyarthritis. Echo: moderate increase in myocardial thickness. diastolic dysfunction of ve and type 1. Necessary to rule out deposit/infiltrative disease. Due to the persistence of renal failure and the need to maintain hemodialysis, a renal biopsy was performed, obtaining positivity for Congo red in the tubules and glomerular wall.
Conclusion: A previously healthy young patient with a subacute clinical picture was diagnosed with amyloidosis after a confirmatory biopsy. This report aims to contribute to future studies in this area because it is a disease that is still little known and needs further studies.

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368 – Use of free genetic screening methods in neurology outpatients in cuiaba: advantages and interpretation difficulties

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Introduction: Medical genetics is increasingly gaining in modern medicine, through panels that enable the screening and diagnosis of rare diseases, becoming an effective ally in determination of some pathologies. In 2021, Invitae provided of medical outpatient clinics with kits for collecting genetic material in order to assist the specialist in diagnosing unusual and difficult-to-recognize conditions. In Cuiabá, the residency in Neurology was chosen to host the use of these methods.
Objectives and methods: List the genetic tests collected through oral swabs and quantify the diagnoses made, in addition to the difficulty in determining them due to the technical language used.
Results: In 12 months, 61 samples were collected, however, only 50 were analyzed. 11 tests were excluded. The analysis identified 30 male, representing 60% of panels, the average age of total sample was 33.64 years (SD). Received results from 20 samples with negative results in search for pathogenic alleles, while 22 showed only uncertain variants, not being conclusive for any diagnosis, just implying reproductive risk. Total of exams analyzed, 8 samples (16%) were positive for pathogenic variants, confirming the diagnosis of rare diseases and enabling appropriate therapeutic measures, initiation of supportive therapies and familiar guidance. Through this method, two patients were diagnosed with Spinal Muscular Atrophy type III, 1 with Wilson's disease and two others with Niemann-Pick disease type C.
Conclusion: Project ended in mid-2022, with the completion of free access to diagnoses. We emphasize that in many cases, access to genetic panel was extremely relevant, helped in the diagnostic direction, making it possible to start therapies for some patients with potentially treatable diseases. However, we encountered some difficulties, mainly in the interpretation of results provided by laboratory, due to extremely technical language and the large amount of information grouped in text. Conclud that medical genetics is extremely important in assisting medicals in diagnosis of rare diseases. However, there is a need for greater access to geneticists as well as genetic tests available in Brazilian Unified Health System.

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377 – Neurocriptococcosis in an immunocompetent patient: a case report

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Introduction: Cryptococcosis is an infection acquired through the lungs, the form of central nervous system involvement is neurocriptococcosis. The diagnosis is the investigation of yeasts in the cerebrospinal fluid using china ink. Treatment is amphotericin B and fluconazole. The aim is to report a case of neurocriptococcosis in an immunocompetent patient. This is a descriptive study, through the analysis of the medical record.

Case report: Female, 23 years old, healthy, with severe headache for 15 days, with progressive worsening, neurological examination without changes, performed skull computed tomography (CT) scan, no changes, cerebrospinal fluid (CSF) examination showed nucleated cells 204, presence of yeast, Venereal Disease Research Laboratory (VDRL): non-reactive and culture of Cryptococcus gattii, positive China ink. Magnetic resonance imaging of the brain showed hypersignal on T2/FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) affecting cortical sulci, encephalic fissures, cerebellar foliae and pial surface of the brainstem and diffuse leptomeningeal impregnation, volumetric reduction of the supratentorial ventricular system, suggestive of cerebrospinal fluid hypotension, findings of leptomeningitis, related to cryptococcosis. Treatment with amphotericin B and fluconazole was initiated. Patient with onset of focal neurological deficits, amaurosis and dysarthria. She presented with refractory headache, requiring serial liquoric punctures for relief of intracranial hypertension. She evolved with instability and underwent a right ventriculoperitoneal shunt. Postoperatively, he presented left hemiparesis. Skull CT showed hematoma and emphysema of adjacent extracranial soft tissues; right frontoparietal brain lesion and edema of adjacent parenchyma. She evolved with clinical instability and died after 61 days of hospitalization.

Conclusion: A pattern of CSF hypotension was shown due to compression of the frontal horns of the lateral ventricles by the granulomas. There was a need for ventriculoperitoneal shunt, but patient had fatal complications.

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378 – Diagnostic challenge in a middle-aged woman with recurrent ischemic strokes: a case of primary central nervous system vasculitis

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Introduction: Primary vasculitides of the central nervous system (CNS) are rare vascular disorders whose pathophysiology involves inflammatory infiltration exclusively in the vessels of the CNS and clinically without constitutional symptoms. The aim is to report a case of primary vasculitis of the CNS. Case report and brief bibliographic review with Health Sciences Descriptors.

Case report: This case portrays a 46-year-old woman with a history of multiple ischemic strokes. She previously had systemic arterial hypertension, prediabetes, obesity, and smoking. During follow-up after the second ischemic event, a small patent foramen ovale was detected with a positive microbubble test and the use of edoxaban was chosen. Transthoracic echocardiogram and magnetic resonance angiography of the cerebral arteries were without relevant changes. However, she recurred with new neurological deficits three more times. Edoxaban was replaced by dabigatran. Cerebrospinal fluid analysis was done twice and both were normal. Cerebral angiography was performed at the time of the last stroke and revealed suggestive signs of vasculitis. The characteristics of this case that corroborate the hypothesis of angiitis, to the detriment of reversible cerebral vasoconstriction syndrome, are the patient's age, chronic headache, and focal symptoms not temporally associated with the headache. Secondary causes of vasculitis have been extensively investigated with negative results. Intravenous methylprednisolone pulse therapy was performed for five days. She evolved with progressive improvement of the motor deficit. On hospital discharge was prescribed prednisone and azathioprine.

Conclusion: This report reinforces the importance of investigating CNS vasculitis in patients with recurrent ischemic strokes.

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379 – Hypopituitarism versus spontaneous subarachnoid hemorrhage

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Introduction: Spontaneous subarachnoid hemorrhage (SAH) is a medical emergency characterized by bleeding in the subarachnoid space, of non-traumatic etiology. Studies demonstrate the relationship of HSA in the pathogenesis of hypopituitarism, which are important due to the need to develop new therapeutic and diagnostic tools.

Objectives: To highlight hormonal complications after SAH.

Methods: A literature review was carried out in the Scielo and PubMed databases, using the descriptors: "aneurysmal subarachnoid haemorrhage" "hormone deficiency" and "hypopituitarism", limited to articles in English, available in full and published in the last five years. Twenty-seven articles were found, of which eight entered the study.

Results: The study indicates that hypopituitarism after SAH is associated with an unfavorable neurological outcome. Pointing to changes in gonadotropins in men, with less hair on the face and body, gynecomastia and hypogonadism. Anemia due to decreased erythropoiesis associated with hypogonadism, growth hormone deficiency associated with decreased exercise tolerance, increased centripetal fat, and premature atherosclerosis. Hyperprolactinemia is common in this group of patients due to interference with dopaminergic inhibition. These hormonal dysfunctions trigger fatigue, headache, mood swings, depression, cognitive

impairment and inability to perform daily activities in these patients, often being confused with neurological sequelae resulting from the patient's cerebral hemorrhage.

Conclusion: The endocrinological control of patients with SAH is relevant, due to its uncertain immediate prognosis. Hypopituitarism is a relevant complication, which if identified early on, there is the possibility of hormone replacement and improvements in the patient's quality of life. Knowledge about the complications of SAH has the role of clarifying not only hypopituitarism, but also cognitive diseases, contributing to elucidate possible therapeutic targets, gaps to be investigated on the subject.

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380 – Patisiran global open-label extension study at 36 months: effect of long-term treatment on mortality and ambulatory function in patients with hattr amyloidosis with polyneuropathy

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Introduction: Hereditary transthyretin-mediated (hATTR) amyloidosis is a progressive, fatal disease. Without treatment, patients experience a median survival of 4.7 years from diagnosis. The effects of long-term treatment with patisiran are being assessed in the ongoing Global Open-Label Extension (OLE) study (NCT02510261).

Objectives: To describe 36-month mortality and ambulatory function data in patients from the Global OLE study.

Methods: Patients were analyzed based on their enrollment in the parent studies: APOLLO-placebo, APOLLO-patisiran, and Phase 2 OLE. Mortality was analyzed in patients who received ≥ 1 dose of patisiran in the Global OLE.

Results: At Global OLE baseline, the APOLLO-placebo group had more severe disease, reflecting disease progression while on placebo in the parent study. At Month 36, mortality was lower in patients who initiated treatment earlier (APOLLO-patisiran 13.5%; Phase 2 OLE 11.1%) than the APOLLO-placebo group (34.7%). In multivariate analysis, NT-proBNP >3.000 ng/L, NYHA > 1 and placebo assignment in the parent study were significant risk factors for mortality. Greater proportions of patients in the APOLLO-patisiran (55.5%) and Phase 2 OLE (80.0%) groups stabilized or improved in ambulation compared to the APOLLO-placebo group (42.9%).

Conclusion: At Month 36 of the Global OLE, survival and the therapeutic benefit of patisiran on ambulatory function were greater in patients who received patisiran earlier. These results highlight the substantial impact of earlier diagnosis and treatment in patients with hATTR amyloidosis with polyneuropathy.

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382 – The impact of caregiver interaction with preschool children exposed inappropriately to screens on neurolinguistic development: a literature review

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Introduction: The first four years of life are critical for neurodevelopment. The overuse of technology in these first years can impact social and linguistic skills.

Objective: This review investigates the role of social interaction and caregiver engagement in the neurodevelopment of babies and preschoolers with premature or excessive screen exposure.

Methods: Searches on digital databases (MEDLINE, Embase, Cochrane, and LILACS) were performed using the keywords "child", "preschool", "neurodevelopment", "child development", "screen time", "child behavior", "social interaction", "caregivers" and "parents". Studies were selected using PICO (population, intervention, control, and outcomes) criteria. After a double-blind screening of the 31 articles found, 6 studies were included.

Results: Up to 23,168 children aged four years or younger were examined in the articles analyzed. All studies showed worse language development with an exposure higher than one hour per day, some of them showing a dose-response relationship between longer screen time and outcome severity. Two studies emphasized that prematurely exposing children to screens before the age of two has a harmful effect. Three studies demonstrated positive results regarding caregiver co-viewing and orientation during the activity. The nature of the media, parents' perception, and exposure during family meals were ranked as possible factors that can influence language acquisition.

Conclusion: This review highlights the importance of caregiver engagement in promoting neurolinguistic development in babies and preschoolers. Screen exposure can harm language skills, but conscious consumption with guidance may mitigate these effects. Exposure before the age of 2 and excessive use (exceeding 1 hour per day) between ages 2–4 can cause the most harm. Therefore, caregiver supervision and interaction play a crucial role in fostering optimal neurodevelopment in this population.

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383 – Neuroprotection by memantine after compressive spinal root lesion

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Introduction: Compressive root lesions are characterized by changes in the spinal cord microenvironment, which include motoneuron chromatolysis and degeneration, chronic gliosis, and glutamatergic excitotoxicity. Since exces-

sive NMDAr stimulation by glutamate leads to neuronal degeneration, the use of NMDAr antagonists has been proposed as a promissing treatment central and peripheral nerve injuries.

Objective: The present study aimed to investigate the neuroprotective effects of memantine, following compressive spinal root axotomy.

Methods: Adult C57BL/6J mice were subjected to unilateral ventral root crush (VRC) and divided into four groups: VRC+Vehicle, VRC+Memantine 30 mg/kg, 45 mg/kg, and 60 mg/kg. The treatment was administered orally for 14 days, starting immediately after injury. Twenty-eight days after the lesion, lumbar intumescences were collected and processed for motoneuron counting (toluidine blue staining), together with astrogliosis and microglial reaction assessment (immunohistochemistry for GFAP and Iba-1, respectively). The protocols for animal use and handling were approved by the local ethical committee (CEUA/UNICAMP, protocol nº 5740-1).

Results: Memantine rescued spinal motoneurons at all the studied doses when compared with the vehicle counterpart, being the 45 mg/kg group the most effective ($P < 0.001$). Memantine also downregulated microglial reactions at the doses of 45 mg/kg and 60 mg/kg ($P < 0.01$, and $P < 0.05$, respectively). Astrogliosis also decreased in all treated groups as compared to the control ($P < 0.01$).

Conclusions: The memantine has a significant antiinflammatory effect on glial cells, coupled with neuroprotection of motoneurons, indicating a possible translation to the clinic.

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384 – Complications after deep brain stimulator implantation in parkinson's disease

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Introduction: Parkinson's disease (PD) is the second most common neurodegenerative disease, its prevalence increases exponentially with age, generating a series of physical, cognitive and emotional non-motor symptoms that disrupt functionality and interfere with quality of life. Surgical treatment with a deep brain stimulator (DBS) implant is an established method for treating these patients.

Objectives: To describe the complications in DBS surgery for Parkinson's.

Methods: A literature review was performed using the SciELO, MEDLINE/PubMed, Virtual Health Library and LILACS platforms, with the descriptors: "Parkinson", "Deep brain stimulation", "Subthalamic nucleus" and "surgery". and "complicações" in Portuguese and English, in the last five years. In the end, 11 articles entered the review.

Results: Motor impairment in activities of daily living has been described as post-surgical complications, even if it is minor compared to psychobehavioral manifestations. With regard to the most reported psychiatric symptoms, impulsivity, delirium, mania, depressive states, aggression, hallucinations and delusions, were the complications found after DBS implementation. Decline in verbal fluency and postoperative confusion were evidenced in several studies.

Conclusion: Deep brain stimulation for Parkinson's disease is an established advanced therapy that addresses motor symptoms and improves quality of life, however, it has been associated with numerous neuropsychiatric symptoms, these symptoms can be distressing, requiring intervention psychiatric. In short, the studies found in the selected articles indicate that it is necessary to direct and reprogram the devices to reduce the postoperative neuropsychiatric impairment, thus allowing the anticipation and prevention of these neuropsychiatric complications, improving the safety and tolerability of these patients.

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385 – Detection of autism in infants prior to 18 months of age

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Introduction: In 2021, the third edition of the Child Health Booklet was published in Brazil, containing a positive novelty: the M-CHAT-R/F checklist, focused on the detection of Autism Spectrum Disorder (ASD) in children aged 16 to 30 months. The Booklet recommends screening for autism at the 18-month checkup. However, younger children have shown typical symptoms, such as delayed language development, non-responsiveness and lack of eye contact. In this regard, it is important to detect the signs of ASD early to diagnose and intervene as soon as possible.

Objectives: To assess the feasibility of screening for autism in children under 18 months.

Methods: Search terms were: "screening", "autism" and "early detection". Open articles in English and Spanish from 2013 to 2023 were selected in the PubMed and CAPES databases. We identified seven community-based papers in our review.

Results: We found 9 tests able to early detect signs of autism: PREAUT (at 4 and/or 9 months), CIRTEA (0–12 months), FYI (6-16 months), ITC (6–24 months), ADOS-2 (1 year to adult), TASI (12–36 months), SACS-R (12, 18, and 24 months consultations), CSBS-DP-IT (6–24 months) and M-CHAT-R/F (16–30 months). In the selected articles with Sensitivity (Se) data available, most tests used on infants under 1 year showed low sensitivity. However, early and repeated screening with a combination of tools facilitates early diagnosis and increases mean sensitivity: PREAUT at 4 and 9 months and CHAT at 24 resulted in a Se of 73%; CIRTEA + CHAT in a Se of 100% and SACS-R + SACS- PR (the latter at 31–60 months) in a Se of 96%.

Conclusion: Despite the recommendation to screen for autism at 18 months, we found that screening can be done earlier, including in the first year of life. In addition, although mostly, a definitive diagnosis can't be obtained at such an age, it is already possible to identify signs of atypical neurodevelopment in infants, and therefore allow earlier intervention.

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386 – Brainstem cavernoma: a case report

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Introduction: Cavernous malformation is a congenital vascular disease, composed of adjacent wide sinusoidal vascular spaces, without interposition of the brain parenchyma. This pathology is still underdiagnosed and neglected, even causing neurological disorders and a high risk of mortality, it is understood that the study and treatment of these injuries, as well as the follow-up of cases, is extremely important.

Objective: To report a clinical case of a patient diagnosed with Brainstem Cav-

ernoma, addressing the clinical evolution, therapeutic follow-up, clinical and surgical findings, through anamnesis and data obtained from medical records.
Case report: Male patient, 51 years old, Caucasian, carpenter, without underlying pathologies, is seen at the Specialty Medical Outpatient Clinic with a complaint of hypoesthesia in the right hand associated with pain in the right upper limb on exertion for 3 years. The patient was admitted to receive emergency care, magnetic resonance imaging of the cervical spine and skull was requested. The analysis revealed an intraparenchymal hematoma in the left portion of the pons, measuring 1.5 by 1.2 cm, with hemoglobin degradation products inside, without presenting impregnation after the intravenous injection of the paramagnetic agent, determining an expansive effect characterized by compression of the portion anterior left of the fourth ventricle.
Conclusions: The diagnostic conclusion was brainstem cavernoma, so clinical support was provided and neurosurgery for resection of the cavernoma. The patient underwent an outpatient follow-up again, in which he reported a significant improvement in the ataxia, maintaining the use of a walker to walk, in follow-up with physiotherapy and speech therapy. He needs help with daily activities due to permanent sequelae, unable to carry out work activities. We corroborate the need for observational studies, systematic reviews and case reports of brainstem cavernomas so that the best actions are always taken, improving the treatment of this rare condition.

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388 – Tuberculosis meningoencephalitis in São Paulo from 2001 to 2022: an epidemiological study

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Introduction: Tuberculosis (TB) is the most prevalent mycobacteriosis in Brazil. Among its extrapulmonary manifestations, one of the most worrying is meningoencephalitis (ME), which is characterized by inflammation of the brain parenchyma and meninges due to infection by the TB bacillus, leading to clinical conditions such as headache, decreased consciousness, intracranial hypertension and coma.
Objectives: To describe the epidemiological profile of confirmed cases of ME due to TB in the state of São Paulo between the years 2001 to 2022.
Methods: This work is an epidemiological investigation study of confirmed cases of ME due to TB between the years 2001 to 2022. Data were collected from the Notifiable Diseases Information System of DATASUS. Data were organized and analyzed using the Jamovi® statistical software.
Results: 418,553 TB cases were confirmed in São Paulo during the period. In this group 5,129 (1.2% of TB cases) had a confirmed diagnosis of ME. An average of 233 (±36.8) cases occur in an upward trend until 2009, when there was an inflection and a downward trend until 2022. Among cases of ME due to TB, data indicates that 64.4% occurred in men, the most prevalent age group was 20 to 39 years old (45.7% of the total) and the most prevalent skin color was white(34.2% of the patients). Another important correlation was the incidence of human immunodeficiency virus (HIV), amounting up to 53.3% of patients with these concurrent diseases. In regards to the final outcomes among those diagnosed with ME due to TB, there were 1,828 deaths (35.6%), of which 17.8% due to TB, and 2,285 patients were cured (44.5%).
Conclusion: The results showed a dispersed annual average of cases with a downward trend from 2009 onwards. Regarding the epidemiological profile, there was a higher prevalence in men, residents of urban areas, young and with an important correlation with HIV. In conclusion, ME due to TB had a high mortality rate and a reasonable cure rate.

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390 – Huntington's disease mortality in Brazil from 2011 to 2020: an epidemiological study

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Introduction: Huntington's Disease (HD) is a dominant genetic neurodegenerative pathology, with full penetrance, characterized by the CAG expansion of the huntingtin gene on chromosome 4, leading to neuronal death due to protein accumulation, which generates clinical presentation of motor, cognitive and psychiatric symptoms, especially Huntington's chorea.
Objectives: To describe the epidemiological profile of mortality from HD in Brazil between the years 2011 to 2020.
Methods: This work is an epidemiological investigation study of HD mortality between the years 2011 to 2020. Data were collected from the Mortality Information System of DATASUS. Data were organized and analyzed using the Jamovi® statistical software.
Results: 1,049 deaths from HD were confirmed in the analyzed period. Regarding the temporal analysis of deaths from HD, an average of 105 (±33.1) annual cases. There was a predominance of deaths in the Southeast compared to other regions of the country, concentrating 505 (41% of the total). Among deaths from HD, there was a higher prevalence of female deaths, with 583 deaths (55.5% of total). Regarding age, there was a predominance of deaths in the age group from 50 to 59 years old, totaling 311 (29.6% of the total). In regards to education, the deaths of individuals with 8 to 11 years of schooling were more prevalent, totaling 251 (23.9% of the total). In respect of skin color, white individuals accounted for the largest share of deaths, accounting 735 (70.0% of the total).
Conclusion: The results found an annual average of deaths increasing and dispersed, which may reflect a greater sensitivity of diagnostic methods, requiring further studies to confirm this association. Regarding the epidemiological profile, there was a higher prevalence in women, from the Southeast, aged over 50 years and with more years of education.

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392 – Phenytoin-induced Chorea: a case report

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A 67-years-old woman, with no significant medical history, presents herself at Hospital do Servidor Público do Estado de São Paulo after a seizure episode following two weeks of a moderate heat trauma at home. At initial evaluation, no neurological deficit was found, but computed tomography showed a mild subdural hemorrhage in the left frontal area that was promptly drained. The pa-

tient was discharged with oral phenytoin 100 mg each eight hours. After thirty days, the patient returns to the hospital reporting the progressive onset of involuntary abnormal movements of upper limbs and head, associated with gait difficulties. The neurological exam revealed ataxia of limbs and trunk, severe gait ataxia, in addition to choreic movements involving superior limbs, neck, orofacial muscles and pathological horizontal nystagmus. The work-up for acute cerebellar ataxia and chorea was performed resulting in normal metabolic and inflammatory blood tests, cerebrospinal fluid analysis, neoplastic screening and brain magnetic resonance imaging, the only exception was serum phenytoin level: 29.4 μg/ml (therapeutic range: 10–20 μg/ml). After an extensive evaluation and differential diagnosis exclusion, a slowly withdraw was conducted, leading to marked symptom's improvement. At discharge, the patient was completely asymptomatic with no other treatment. The phenytoin has an extensive well described list of neurological side effects, commonly ataxia, headache, dizziness and insomnia, but choreic movements are rarely reported. It is due to the disturbance of dopamine metabolism at basal ganglia, involving both direct and indirect pathways. The choreic symptoms have no dose dependent relation, affecting mostly infants, previously brain injured patients, or concomitant drugs that inhibit hepatic metabolism. Our case elucidates a rare cause of chorea induced by a common pharmacological treatment, which demonstrates the importance of searching for uncommon drugs side effects.

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394 – Hallucinations and psychosis in Guillain-Barré syndrome: case report

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Introduction: Guillain-Barré syndrome (GBS) is an acute inflammatory peripheral neuropathy that occurs after infection or immunization. Wernicke's encephalopathy (WE) is caused by thiamine deficiency that classically presents with altered mental status, ataxic gait and ophthalmoplegia.
Case report: Male, 64 years old, presented with diarrhea, vomiting and hyporexia. Three weeks later, he developed concomitant acute tetraparesis and cognitive impairment. On examination, he presented with persecutory delusions, hallucinations, tetraparesis with global areflexia. The cerebrospinal fluid analysis showed important albuminocytological dissociation. Treatment with intravenous immunoglobulin associated with thiamine was started. The electroneuromyography was compatible with demyelinating sensorimotor polyneuropathy. Brain Magnetic resonance imaging showed FLAIR (Fluid-Attenuated Inversion Recovery) hyperintensities in mamillary bodies and periaqueductal gray matter. The patient was diagnosed with GBS and WE. He was discharged after two weeks with complete resolution of the cognitive impairment and improvement of tetraparesis. Altered mental status in a patient with GBS is not common. There are some reports of patients who presented cognitive disturbances related to autonomic dysfunction and more severe cases, which developed a few days after the onset of the motor condition. The reported patient had a change in mental status concomitantly with a motor condition, reported prolonged gastrointestinal symptoms, in addition to having received intravenous glucose. WE may occur after a short period of thiamine absorption/intake deficiency and may be precipitated by glucose administration. The patient showed rapid cognitive improvement after thiamine supplementation, and had typical EW changes on the brain MRI confirming the diagnosis of WE.
Conclusions: We must be aware of changes in mental status as this may indicate thiamine deficiency. The case elucidates the importance of thiamine replacement in patients at risk of vitamin deficiency.

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399 – Reversible posterior encephalopathy syndrome as a rare complication of oxaliplatin chemotherapy: a case report

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Introduction: The reversible posterior encephalopathy syndrome corresponds to a clinical and radiological diagnosis marked by headache, altered mental status, reduced visual acuity and seizures, associated with vasogenic edema mainly in posterior white matter. It presents a large heterogeneity of etiologies, such as cytotoxic drugs, eclampsia, vasculitis, and nephropathies, with arterial hypertension being the most important. In that way, quick diagnostic investigation is necessary to provide a better prognosis.
Case report: Case report of a patient who was in adjuvance combined chemotherapy, wich includes oxaliplatin, after pancreatic tumor resection, and presented reversible posterior encephalopathy as a treatment complication. The article was developed from data collection through literature review on the subject, as well as analysis of the patient's medical records. We found the history of a patient with pancreatic ductal adenocarcinoma resection, who underwent adjuvant FOLFIRINOX combined chemotherapy, which contain oxaliplatin. After the first dose, the patient developed severe headache, mental confusion, left-sided motor deficit, and reentrant epileptic seizures, requiring orotracheal intubation due to a non-convulsive status epilepticus. Patient was investigated with brain resonance, wich showed cortical and subcortical hypersignal lesions in the bilateral parieto-occipital lobes, as well as in the thalamus and right insula, and normal cerebrospinal fluid at the first and seventh day. She was also investigated with a panel for autoimmune encephalitis antibodies, with a negative result. After withdrawal of chemotherapy, the patient evolved with a gradual improvement of symptoms and resolution of seizures within twenty days, and was referred for motor rehabilitation at discharge.
Conclusion: Early diagnosis and treatment of the etiology contribute to a better prognosis for patients with reversible posterior encephalopathy syndrome.

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401 – Perception of quality of life between different genders in patients with multiple sclerosis

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Introduction: Multiple sclerosis (MS) is a chronic, demyelinating, inflammatory and neurodegenerative disease of the central nervous system. Quality of life (QoL) is significantly impaired in patients with MS. Motor disability only partially explains the reduction in QoL, as symptoms such as depression, fatigue and mood disorders also exert influence. Several characteristics in a patient with MS have been associated with worse QoL, including advanced age, late diagnosis and progressive form of the disease.
Objective: We evaluated in this review possible impacts of gender on QoL.

Methods: The database PubMed was searched for studies indexed from the year 2000 with the following descriptors: multiple sclerosis, quality of life, gender, sex.

Results: The impact of the reduction in QoL is worse for men than for women with MS in relation to motor function, vitality, sociability, emotional well-being and mental health. Men seems to have a greater cognitive decline, with worsening performance in verbal memory and executive function. Interestingly, gender and marital status can influence social support. Women had more support networks as they reported better availability from friends than from their male caregivers. It was also noted that men without a spouse feel less social support. Regarding non-motor symptoms, women feel more pain and have a higher prevalence of depressive and/or anxiety disorders. However, regarding sphincter and sexual disorders, the impact on QoL is greater in men. Women with high motor disability seem to maintain psychological well-being better than male patients.

Conclusion: Men seeking help later may be a factor influencing the natural history of MS. The diagnosis of a chronic disease for men seems to be faced differently due to cultural factors. The different impact of disabilities between genders makes us understand that the management must be specific for each sex to better meet the needs of patients.

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402 – Juvenile-onset amyotrophic lateral sclerosis due to homozygous COQ7 variants

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Coenzyme Q10 (CoQ10) biosynthesis defects may lead to primary CoQ10 deficiency. This rare inherited metabolic disorder has been associated with a heterogeneous genetic basis, including rare pathogenic variants in the *COQ7* gene. CoQ10 deficiency leads to dysfunction of mitochondrial energy, resulting in several distinct neuromuscular phenotypes, such as myopathy, axonal and demyelinating chronic polyneuropathy and hereditary motor neuronopathy. Herein, we present the first clinical association of *COQ7* pathogenic variants with Juvenile Amyotrophic Lateral Sclerosis. A 38-year-old Brazilian man started at age 11 years with a slowly progressive history of distal lower extremity weakness that evolved to the proximal segment, distal extremities of upper limbs and speech disorders. His parents were first degree cousins. His father and brother had similar motor symptoms. Neurological examination disclosed dysarthria, lower limb and distal-dominant quadriparesis, global amyotrophy with dominant distal involvement, and brisk tendon reflexes in the lower limbs. Neuroimaging studies were unremarkable, as well as cerebrospinal fluid analysis. Needle electromyography disclosed diffuse chronic denervation involving cervical, thoracic, and lumbosacral myotomes, and acute denervation (positive sharp waves and fibrillation) in the four limbs; preserved sensory neuroconduction studies. Broad next-generation sequencing-based gene panel testing disclosed the homozygous pathogenic variant c.1A>G in the *COQ7* gene. This description expands the pathophysiological basis associated with JALS. *COQ7* variants should be included in the differential diagnosis of juvenile-onset motor neuron disease.

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403 – Subacute fever and torpor in a hematological patient revealing atypical CLIPPERS with supratentorial involvement: a case report

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Case report: A 62-year-old patient was transferred from another hospital with complaints of subacute torpor and fever two months after an allogeneic bone marrow transplant. He had the diagnosis of chronic myelomonocytic leukemia type 2 four years before the beginning of the neurological symptoms. The disease remained stable for two years after the diagnosis, but progressed and he was submitted to a bone marrow transplant. The complications during the procedure were a febrile neutropenia, that resolved with a cycle of antibiotics, and a reactivation of cytomegalovirus (CMV) infection (he had a high risk to reactivation due to the status of antibodies of the donator, that was negative, and he had positive antibodies). He remained stable and was discharged from the hospital 30 days after the transplant. Two months after the transplant, the fever and torpor began, and he went to the emergency department in his hometown. There, he was diagnosed with a positive coronavirus disease 2019 (COVID-19) infection and reactivation of cytomegalovirus, with prescription of endovenous ganciclovir, without clinical improvement. His clinical symptoms evolved to loss of balance and progressive torpor, and was transferred to our hospital for clinical evaluation. In the initial examination, he was febrile, torporous, he was not collaborative to the neurological examination due to the level of consciousness. He was hypotonic and had withdrawal from a painful stimulus on four limbs. His tendinous reflexes were hyperactive and symmetrical. Due to his level of consciousness, the cerebellar maneuvers were not testable. He was then submitted to orotracheal intubation due to neurological deterioration. In his complementary investigation, the serum polymerase chain reaction (PCR) for CMV was already negative and his PCR for COVID-19 was also negative. The electroencephalogram showed slow generalized periodic discharges. His computerized tomography showed no abnormalities, so he was submitted to a lumbar puncture that showed 8 cells, with 97% lymphocytes, 74 mg/dL of protein, 26 mg/dL of glucose with no neoplasic cells and a positive PCR for *Toxoplasma gondii*. The PCR for *mycobacterium tuberculosis*, cryptococcosis, and viruses were negative. After the results, we started sulfadiazine and pyrimethamine and he was submitted to a magnetic resonance imaging that showed multiple nodular foci of hypersignal in T2/FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) and diffusion abnormalities on the cortical surface of the cerebellar hemispheres, thalamus, basal ganglia and subcortical white matter of the cerebral hemispheres, being more evident in the cerebellum and basal ganglia. Besides the atypical radiological findings for neurotoxoplasmosis, his serologic testing on the serum showed an positive immunoglobulin M and immunoglobulin G for *Toxoplasma gondii*. He had no clinical improvement with the antibiotics, and so, ten days after he was submitted to a new magnetic resonance imaging showing increase in the size and number of multiple small focal lesions with hypersignal on T2/FLAIR in the brain parenchyma, highlighting confluent le-

sions in the basal ganglia and focal lesions in the cerebellar hemispheres, which show contrast uptake, as well as in the thalamus and midbrain. His neurological symptoms evolved and he showed a minimally conscious state, with spasticity in the four limbs. He was submitted to a new lumbar puncture that showed 1 cell, 78 mg/dL of protein and 50 mg/dl of glucose, an improvement, despite clinical and radiological worsening. Because of the clinical worsening, he was submitted to a brain biopsy. His cerebral biopsy was negative for neoplasic infiltration of the cerebral nervous system, viral infections, cryptococcus and toxoplasmosis. It showed an interstitial CD3 lymphocytic inflammation with vasculitis and frequent macrophages, very rare CD20 lymphocytes and p24 and SV-40 negative. His neuropathological criteria were compatible with CLIPPERS (chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids). The patient was submitted to five days of metilprednisolone and with oral prednisone after, with slow clinical improvement, but probably due to the lack of systemic clinical status of the patient and delayed diagnosis because of the atypical presentation, the patient died of pneumonia.

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404 – Modification of palmar grip strength induced by overflow contralateral to hip contraction

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Introduction: Neurofunctional rehabilitation often employs various approaches to enhance recruitment and muscle strength through overflow, a phenomenon that amplifies the motor response of homologous muscles in the contralateral segment. overflow has been well documented in the literature, particularly in techniques like FNP and cross-education. The phenomenon of overflow is known to facilitate muscle activity in muscles that are not actively stimulated, resulting in unintentional contractions during a resisted isometric activity.

Objectives: The objective of this study was to investigate the overflow mechanisms of non-homologous contralateral muscles of the lower limb during an instructed handgrip task.

Methods: A total of 90 individuals, with a motor preference for their right upper limb (RUL), were divided into control (CG) and resisted isometry group (RIG) and underwent a protocol involving three maximal hand grip contractions with their RUL and LUL. In assessment 2, the CG performed the same protocol as baseline, while the RIG group performed three maximal isometric contractions of left hip flexion with three maximal contractions of hand flexion with their RUL. statistical analysis was conducted by a multiple comparison analysis of variance with $P < 0.05$.

Results: Observing the data from evaluation 1, both the CG and RIG showed a reduction in strength between the third and first attempt for both the left and right upper limbs ($F = 1.07$, $F = 0.68$, respectively). In evaluation 2, the decrease occurred only for the CG in both upper limbs ($F = 0.15$, $F = 1.29$, respectively).

Conclusion: Our study revealed that maximal muscle strength was maintained in RIG individuals during assessment 2, indicating the existence of overflow when performing an isometric hip flexion task concomitantly with the palmar grip.

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405 – Safety and efficacy of inebilizumab in AQP4+ NMOSD participants with history of immunosuppression treatment prior to N-MOmentum study

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Introduction: Inebilizumab (INEB), an anti-CD19 B cell-depleting antibody, is approved for the treatment of neuromyelitis optica spectrum disorder (NMOSD) in adults seropositive for aquaporin-4 antibody ($AQP4^+$).

Objectives: To evaluate long-term outcomes of INEB treatment in $AQP4^+$ NMOSD patients (pts.) from the N-MOmentum trial with a history of immunosuppressive therapy (IST) as compared to those without.

Methods: N-Momentum (NCT02200770) was a 28-week randomized phase 2/3 trial of INEB versus placebo, with an optional Open-Label Extension (OLE) (> 2 years). In this analysis, $AQP4^+$ participants who received INEB (through the OLE) were grouped by no history of IST beyond treatment of acute NMOSD attacks (naïve), or prior azathioprine (AZA) and/or mycophenolate mofetil (MMF) therapy.

Results: Among pts. who received INEB during the study, 94 received prior AZA/MMF and 103 were immunosuppressant naïve. The annualized relapse rate (95% confidence interval [CI]) for pts. with prior AZA/MMF was 0.11 (0.07, 0.17), compared to 0.08 (0.05, 0.14) for naïve. The annualized NMOSD-related inpatient hospitalization rate (annualized rate [95% CI]) for prior AZA/MMF was 0.15 (0.08, 0.27), and 0.12 (0.06, 0.22) for naïve. The percentage of pts with ≥ 1 study drug-related treatment-emergent adverse event (TEAE) was 30.9% (29/94) in prior AZA/MMF and 47.6% (49/103) of naïve; 4.3% (4) of prior AZA/MMF and 5.8% (6) of immunosuppressant-naïve reported ≥ 1 study drug-related serious adverse event.

Conclusion: This post hoc analysis evaluating long-term outcomes of INEB in $AQP4^+$ NMOSD pts. treated with prior AZA/MMF therapy demonstrated a similar efficacy and safety profile as pts. without prior IST.

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410 – Hip strength change induced by overflow ipsilateral to handgrip

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Introduction: Neurofunctional rehabilitation often employs various approaches to enhance recruitment and muscle strength through overflow, a

phenomenon that amplifies the motor response of homologous muscles in the contralateral segment. Overflow has been well documented in the literature, particularly in techniques like FNP and cross-education.

Objectives: This study aimed to investigate the overflow mechanism in non-homologous and ipsilateral muscles during an instructed handgrip task.

Methods: We recruited 90 subjects of both genders between 18 and 30 years with right upper limb (RUL) motor preference. Each subject was seated with their lower limbs flexed at the hip, knee, and ankle to 90 degrees. We used a lower limb dynamometer to measure baseline strength by having the subjects perform three maximal left hip flexion contractions with 6-second contractions and 10-second intervals. For the second evaluation, the subjects performed three maximal handgrip contractions on a rigid dynamometer, followed by three maximal isometric contractions for left hip flexion (also with 6-second contractions and 10-second intervals). After a 10-minute rest period, we took new measurements of the maximum voluntary isometric contraction (MVIC) for both handgrip and hip flexion. We excluded subjects who were unable to produce 70% of their MVIC during testing. We conducted a statistical analysis using a multiple comparisons analysis of variance with a significance level of $P < 0.05$.

Results: We found a decrease in strength between males and females during T3 (post-test) ($P = 0.006$), and an increase in strength between T3 and BL3 in female subjects ($P = 0.045$).

Conclusion: Our study demonstrated an increase in hip flexor strength in women when subjected to a maximal isometric task with the ipsilateral upper limb.

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414 – Omics profile of iPSC-derived astrocytes from Progressive Supranuclear Palsy (PSP) patients

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Progressive Supranuclear Palsy (PSP) is a neurodegenerative tauopathy and, to date, the pathophysiological mechanisms in PSP that lead to Tau hyperphosphorylation and neurodegeneration are not clear. The development of a model using neural cell lines derived from patients has the potential to identify molecules and possible biomarkers. We developed a model of induced pluripotent stem cells iPSC-derived astrocytes to investigate the pathophysiology of PSP, particularly early events that might contribute to Tau hyperphosphorylation, applying an omics approach to detect differentially expressed genes, metabolites, and proteins, including those from the secretome. Skin fibroblasts from PSP patients and controls were reprogrammed to iPSCs, which were further differentiated into neuroprogenitor cells and astrocytes. In the 5th passage, astrocytes were harvested for total ribonucleic acid sequencing. Intracellular and secreted proteins were processed for proteomics experiments. Metabolomics profiling was obtained from supernatants only. We identified hundreds of differentially expressed genes in PSP. The main networks were related to cell cycle activation. Several proteins were found exclusively secreted by the PSP group. The cellular processes related to the cell cycle and mitotic proteins, chaperonins of the TriC/CCT pathway, and redox signaling are enriched in the secretome of the PSP group. Moreover, we found spatial segregation by PCA in the metabolomics data, indicating distinct sets of metabolites between PSP and control groups. Our iPSC-derived astrocyte model can provide distinct molecular signatures for PSP patients and it is useful to elucidate the initial stages of PSP pathogenesis.

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418 – Anti-hu neuropathy as a paraneoplastic syndrome: the pursuit for the primary site.

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A 53-year-old man with a history of smoking and recent weight loss presented with progressive weakness and difficulty walking. Physical exam evidenced dysmetria on finger-to-nose test, worsened with eyes closed, pseudoathetosis, absent deep tendon reflexes and a neuropathic pattern of hypoesthesia and hypopalesthesia. He was unable to stand due to the severity of the ataxia. Initial investigation was positive for syphilis, with no history of treatment, and cerebrospinal fluid analysis showed hyperproteinrachia with normal cell count. Despite adequate treatment with penicillin, symptoms continued to progress. Electroneuromyography was remarkable for axonal sensorimotor polyneuropathy with sensitive predominance. A paraneoplastic panel was ordered and evidenced positivity to anti-Hu. The patient was then treated with high doses of methylprednisolone and plasmapheresis, with no improvement. An extensive search for malignancies was performed, and enlarged hilar lymph nodes were found on computed tomography scan. Positron Emission Tomography-scan showed pulmonary and mediastinal hypermetabolism. Tissue samples were acquired with no significant findings and tuberculosis, fungal infections and inflammatory granulomatous diseases, among other conditions, were ruled out. Due to the high suspicion for malignancy, the biopsies were reviewed by a pulmonary pathology specialist, submitted to additional immunohistochemical staining and, at last, the diagnosis of small cell lung carcinoma was made. Chemotherapy was initiated shortly after.

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420 – Atypical white matter changes on Bickerstaff encephalitis: a case report

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Introduction: Bickerstaff encephalitis is a rare disease and considered a spectrum of the Guillain Barré syndrome. We present a case report of subacute on-

set of gait impairment, impaired consciousness, ophthalmoplegia, respiratory dysfunction and severe dysautonomia with atypical and diffuse brainstem and diencephalic white matter abnormalities, a positive seric GQ1B and neurological recovery after two cycles of immunoglobulin and plasmapheresis. The aim is to describe an atypical case of Bickerstaff encephalitis.

Case report: A 30-year-old man was admitted with a ten days history of vertigo, dysphagia, bilateral ptosis and tetraparesis with brachial predominance. His examination showed appendicular ataxia, tetraparesis, ophthalmoplegia and ptosis. Due to the clinical presentation, we started treatment with immunoglobulin but he evolved with need of mechanical ventilation and had two cardiac arrests due to severe dysautonomia. His lumbar puncture showed no albuminocytologic dissociation. His magnetic resonance imaging showed nonspecific foci signal hyperintensities on FLAIR (Fluid-Attenuated Inversion Recovery) in cerebellum, hippocampus and globus pallidus. His serum GQ1B testing was positive, confirming the initial clinical hypothesis of Bickerstaff encephalitis. He was submitted to plasmapheresis and a second cycle of immunoglobulin, with complete clinical recovery six months after.

Conclusion: We described a case of atypical Bickerstaff encephalitis, highlighting the importance of recognizing atypical clinical patterns for an appropriate treatment.

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421 – Ramsay Hunt syndrome and viral meningitis: an association underreported in the literature

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Introduction: Ramsay Hunt Syndrome, a rare clinical entity, is caused by the reactivation of the varicella zoster virus, which results in inflammation of the geniculate ganglion of the facial nerve. Characterized by the triad: ipsilateral facial paralysis, otalgia and herpetic vesicles in the external ear. Due to the broad symptomatology, which encompasses atypical symptoms and present in several other pathologies, it is often underdiagnosed.

Case report: A 62-year-old woman admitted to Hospital Evangélico de Londrina due to paresthesia in the left hemiface, associated with vertigo and severe headache. On physical examination, peripheral facial paralysis on the left and vesicles on the palate and left external ear were observed. AVC protocol exams were requested (normal) and liquor was collected (clear, colorless appearance, 92% lymphocytes). New serologies and magnetic resonance imaging of the skull with contrast were requested. Prescribed acyclovir, prednisone, eye drops and ocular occlusion. The patient evolved with maintenance of headache and otalgia. Prednisone was replaced by intravenous dexamethasone, gabapentin and topical lidocaine were added, and other medications were maintained. Non-reactive requested serologies. Analysis of the herpes simplex virus by polymerase chain reaction > 30. In view of the data, a diagnosis of viral meningitis due to herpes associated with Ramsay Hunt syndrome was diagnosed. Maintained treatment, patient evolved with improvement of the condition.

Conclusion: Based on the clinical correlation and unfavorable prognosis, the importance of early identification and treatment lies in order to reduce the time of active disease and neural damage, avoiding possible complications.

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422 – Recurrent cerebral intraparenchymal hemorrhage after ayahuasca consumption: a case report

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Case report: A 57-year-old woman was referred to our service after noticing difficulty in coordinating the movements of the right hand. The symptoms occurred after she consumed ayahuasca in a religious ceremony and persisted after the psychedelic effects weaned off. On the first evaluation, she had right hemiataxia and increased blood pressure (200/100 mmHg). The head computed tomography (CT) scan revealed a left basal ganglia hemorrhage. No abnormalities were found on CT angiography. The patient later reported that a few years earlier, she was diagnosed with a hemorrhagic stroke at another hospital, which also happened within a few hours of ayahuasca intake. However, she has not stopped using it monthly since then.

Discussion: Ayahuasca is a potent plant-based hallucinogenic brew used traditionally for spiritual and medicinal purposes. The substance responsible for its effects is Dimethyltryptamine (DMT), which is known to be a potent serotonin receptor agonist, especially the 5HT2A. This substance may induce an acute raise in blood pressure and consequently trigger a hemorrhagic stroke. To this date, there are no cases reported of hemorrhagic stroke after using this drug.

Conclusion: Although ayahuasca has been associated with spiritual and therapeutic benefits, its potential risks should not be ignored, including serious complications, such as hemorrhagic stroke.

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425 – Genetic and clinical features of congenital titinopathy: a single-center cohort

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Introduction: *TTN* has the longest coding sequence of human genes and encodes titin, a huge protein that serves as the scaffold for sarcomere assembly. Titinopathy encompasses a complex spectrum of phenotypes with involvement of skeletal and cardiac muscles, with recessive and dominant autosomal inheritance.

Objectives: In this work we present the clinical and genetic data of seven patients with autosomal recessive TTN-related congenital myopathy, aged between 9 months and 38 years (mean age 14.73 years).

Methods: Clinical, muscle histology and genetic data are presented. *TTN* gene were analyzed by next-generation sequencing (NGS).
Results: Thirteen different variants were found, all classified as null variants (nonsense, frameshift or canonical splice site). All variants were presumably compound heterozygous. Only one variant was recurrent (c.44815+1G>A. All patients had a history of neonatal hypotonia. The latest age for gait acquisition was 7 years old, and a 19-year-old patient never acquired the gait. Joint hypermobility was prominent in two patients. No patient had ophthalmoparesis. Baseline CK ranged from 75 to 4619 U/L. Minicores and mild dystrophic changes were the most prominent findings on muscle biopsies.
Conclusion: Congenital muscle titinopathy commonly presents with null biallelic variants. Patients who present with distal hyperelasticity, normal or slightly increased CK, and dystrophic findings on muscle biopsy may have an initial etiologic suspicion of COL6-related myopathy, a more common group of skeletal muscle disorders and important differential diagnosis of TTN myopathy.

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426 – Hemorrhagic demyelinating lesions and systemic thrombosis due to Varicella Zoster infection: a case report

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Introduction: The investigation of hemorrhagic demyelinating cerebral lesions is challenging with diverse etiologies. We report a case of varicella zoster virus (VZV) infection with prominent hemorrhagic demyelinating component and systemic thrombosis.
Objectives: Describe a case of hemorrhagic demyelinating lesions and systemic thrombosis secondary to varicella zoster central nervous system infection.
Case report: A 45-year-old man presented with a one-week history of left leg strength loss associated with pyramidal liberation on the left side. Brain MRI showed two expansive hemorrhagic lesions, in the frontoparietal lobe and in the cingulate gyrus region. There were T2/FLAIR hypersignal in the subcortical area of both cerebral hemispheres and in the cervical cord, suggestive of demyelination. His arterial magnetic resonance imaging with vessel wall imaging showed focal tapering on the pial branches of the middle cerebral artery, but his digital angiography showed no abnormalities. The lumbar puncture had 8 cells, (lymphocyte predominance), 52 proteins/dL, and 53 mg/dL of glucose. In two days, he developed a left side hemiplegia associated with seizures and pulmonary thromboembolism, demanding lung mechanical thrombectomy and anticoagulation. He had a positive lupus anticoagulant and a positive VZV IgG in the cerebrospinal fluid associated with positive oligoclonal bands. He received intravenous acyclovir with complete resolution of the symptoms.
Conclusion: The clinical manifestations of VZV infection are wide, and we should suspect of this infection in patients with central nervous system demyelinating lesions, even without other skin lesions.

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428 – Screening of vulnerable population with persistent neurological symptoms of COVID-19

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Introduction: The coronavirus disease 2019 (COVID-19) pandemic significantly impacts public health, with over 677 million infected and 7 million deaths. In addition, survivors of the disease can develop post-COVID symptoms, including neurological changes, which have been neglected in primary healthcare. These changes can include memory difficulties, peripheral neuropathy, and dysautonomia.
Objectives: To screen the vulnerable population with post-COVID symptoms, in order to generate an epidemiological and severity perspective of cases and generate management and rehabilitation resource allocation prospects.
Methods: A cross-sectional prevalence study was conducted with a questionnaire answered in an interview at the Santa Marina Basic Health Unit, Praia Grande, to identify the presence of post-COVID symptoms in a vulnerable population. The collected data were analyzed to generate an epidemiological and severity perspective of cases.
Results: Of the 22 patients identified with an average age of 43 years, dyspnea was the most common symptom, followed by cough and headache. Of the respondents, 9% were admitted to the ICU and had a higher burden of prolonged symptoms, including dyspnea on minimal exertion and peripheral neuropathy. In addition, 20% of patients reported difficulty with short-term memory recall, and 13% developed some form of dysautonomia, such as sphincter hypertonia.
Conclusion: The results suggest that post-COVID neurological changes are prevalent and severe in some patients. However, these symptoms are still neglected in primary healthcare and do not receive the appropriate investigation and treatment, requiring greater allocation of resources for their control.

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431 – Wall-eyed bilateral internuclear ophthalmoplegia (WEBINO) secondary to demyelinating brainstem lesion

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We report the case of a healthy 25-year-old man presenting with sudden onset dizziness, strabismus, and cloudy vision that improved when he closed one of his eyes. He denied pain with eye movement or color desaturation, as well as history of recent immunization or febrile illness. He did not present any other neurologic symptom, but he affirmed having had a limited episode of a dis-

crete strabismus four months before. In his first assessment at the emergency room his neurologic examination revealed a 30° exotropia of the right eye on the primary gaze position along with adduction deficit and abduction nystagmus bilaterally on conjugated horizontal eye movement, characterizing an internuclear ophthalmoplegia on both eyes. He also presented with asymmetrical convergence deficit, with inability on completing adduction on his right eye. On the vertical upward gaze there was also a vertical nystagmus. Eye fundus examination did not show retinal and optic nerve alterations. Visual acuity was normal. This set of findings qualified a WEBINO (wall-eyed bilateral internuclear ophthalmoplegia). During investigation, lumbar puncture showed mild hyperproteinrachia, with absence of oligoclonal bands and normal CSF (cerebrospinal fluid GigG (immunoglobulin G) index. He was submitted to a course of pulse therapy with methylprednisolone. Neuroaxis magnetic resonance imaging evidenced a demyelinating periaqueductal lesion, involving medium longitudinal fasciculus, ponto-mesencephalic junction and mesencephalic tegmentum, without gadolinium enhancement. As he remained symptomatic, plasmapheresis was indicated, with complete remission of symptoms afterwards. Following his investigation the tests for both anti-aquaporin-4 and anti-MOG antibodies were negative, and until the conclusion of this report, a diagnosis of clinical isolated syndrome remained as the main hypothesis.

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432 – Elderly's thrombolysis in the real-world setting: a cohort study

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Introduction: The higher prevalence of stroke in the elderly population is increasingly recognized, but safety and efficacy of thrombolysis in this population are still debatable in clinical practice.

Methods: This is a retrospective cohort study of stroke patients admitted in a health center in Brazil. We investigated the effectiveness of thrombolysis in stroke patients aged above 80y. The primary outcome was Rankin at discharge and the secondary outcome was lethality.

Results: We studied a cohort of 185 stroke patients aged above 80. Mean age was 84.9 (standard deviation [SD] 4.1). The majority was female (121–65.4%). Mean initial National Institute of Health Stroke Scale (NIHSS) was 12.2 (SD 6.7). Most patients had arterial hypertension (83.2%) with equal proportion between sexes. Thrombolysis was performed in 20.8%. The proportion of risk factors was the same for those who received or not thrombolytic therapy. Age, Aspects score, and comorbidity index were similar for the groups, as was the proportion of female/male. The rate of in-hospital infection was insignificantly higher in the non-thrombolytic group (34.9% for non-thrombolytic group and 25% for the thrombolytic one; P = 0.285). NIHSS was significantly higher for the thrombolytic group (14.6 versus 10.9 points – P = 0.0007). The rate of case-fatality was 30.1% for the non-thrombolytic group and 19.3% for those treated with thrombolysis. A multivariate logistic regression analysis disclosed in-hospital infection as the main predictor of lethality (odds ratio [OR] 3.1; CI 1.33–7.15; P < 0.0001) for both groups. Predictors of a good outcome at discharge (Rankin score ≤ 2) included a lower initial NIHSS score and the treatment with thrombolysis (adjusted OR 5.3; CI 1.79–15.80).

Conclusion: Thrombolysis in stroke patients aged 80 years was safe and effective, disclosing the strongest predictor of good short-term outcome.

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434 – Intracranial hypertension as initial presentation of multiple myeloma in a young man

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Introduction: Multiple myeloma (MM) commonly appears after the age of 50, corresponding to 1% of all cancers. Symptoms such as renal failure and hypercalcemia are usually the initial manifestations of the disease. Involvement of the central nervous system (CNS) it´s rare and a marker of poor prognosis. We report a young patient opening the MM with an initial manifestation that is rare in the CNS literature.

Case report: Male, 38 years old, previously healthy, admitted for holocranial and progressive headache for 1 month and visual blurring and mental confusion started two days before admission. On examination we evidenced papilledema and negative myoclonus in the 4 limbs and neck. Brain computed tomography, arterial and venous angioMRI were normal. Cerebrospinal fluid puncture show increased opening pressure (32 cm H_2O) and biochemical analysis was normal. Laboratory tests showed 24-hour proteinuria in the nephrotic range (3 g), microcytic hypochromic anemia and reduced creatine clearance (48,8 mL/min — Chronic Kidney Disease Epidemiology Collaboration). Contrast brain and orbits magnetic resonance imaging (MRI) presented thickening of the meninges and bone infiltration, suggestive of lymphoproliferative or hematologic disease. MRI of the spine (cervical and thoracic), thorax and abdomen showed signs of diffuse bone infiltration. HMM was confirmed by electrophoresis of proteins with expressive peak of gamma globulin (43%, VR 11,1–18%). Dexamethasone (4 mg 6/6 hour for three days) was administered, with complete resolution of mental confusion and headache. The patient was followed up with hematology to start chemotherapy.

Conclusion: We report a rare case of MM that started with predominantly neurological manifestations which responded very well to corticosteroid therapy in low doses after few days. This case illustrates how intracranial hypertension in a young patient requires extensive investigation for possible rarer differential diagnoses when initial tests are normal.

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435 – Quickly progressive insanity in emergency

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JP, 42 years old, female, previously healthy, brought for neurological evaluation with behavioral changes since May 2022, at the time, presented delusions of grandeur, persecution mania, disorganized thinking, mental confusion, being admitted to a psychiatric inpatient unit for clinical care. She was diagnosed with bipolar mood disorder and started treatment. At the time of hospital discharge, the family was instructed to investigate the condition with a neurologist, because the patient was lethargic, not very communica-

tive, did not recognize family members, incontinent with the need to use diapers, rigid, accepting only creamy diet, anorexia and with symmetrical and gross hand tremor. In January 2023, he came to the hospital because, in addition to this clinical picture, he described a patient who started with a persistent, daily fever of 40 degrees in the week before the medical evaluation. She was emaciated, dehydrated, Glasgow 13, with myoclonus in all four limbs, drowsy with appendicular rigidity. She was hospitalized due to a condition suggestive of a rapidly progressive dementia, and underwent a series of tests, some of which included a tomography showing signs of dilation of the ventricular system without signs of hypertension, cerebrospinal fluid with increased cellularity and proteins, in addition to Venereal Disease Research Laboratory (VDRL) 1:1024, associated with a serum VDRL of 1:64. The others serologies were negative. Treatment was started with benzathine penicillin 2,400,000 IU per day for 21 days in hospital, adjustment of lithium and quetiapine and introduction of aripiprazole until adequate control of the associated psychiatric symptoms. In the following month, the patient was able to identify family members, form simple sentences and maintained a stable mood, although the other neurological symptoms remained, probably as definitive sequelae of neurosyphilis. Early diagnosis of possibly treatable dementia syndromes, such as neurosyphilis, is essential to minimize the risk of neurological sequelae.

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437 – Muscle ultrasound changes correlate with functional impairment in spinal muscular atrophy

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Introduction: We investigated ultrasound patterns of muscle involvement in different types of Spinal Muscular Atrophy (SMA) and their correlation with functional status to determine the pattern of muscle compromise in SMA patients and the potential role of ultrasound to evaluate disease progression.

Methods: We examined muscles (biceps brachii, rectus femoris, diaphragm, intercostals and thoracic multifidus) of 41 patients with SMA (types 1 to 4) and 46 healthy age, and sex-matched controls using B-mode ultrasound for grey scale analysis (GSA), area (biceps brachii and rectus femoris) and diaphragm thickening ratio. Functional scales were applied to patients only. We analyzed ultrasound abnormalities in specific clinical subtypes and correlated findings with functional status.

Results: Compared to controls, patients had reduced muscle area and increased mean GSA for all muscles ($P < 0.001$), with a stablished correlation between the increase in GSA and the severity of SMA for biceps brachii, rectus femoris and intercostals ($P = 0.03$, 0.01 and 0.004 respectively) when using Hammersmith Motor Functional Scale Expanded. Diaphragm thickening ratio was normal in the majority patients, and intercostal muscles had higher GSA than diaphragm, in relation to the controls.

Conclusion: Ultrasound is useful for quantifying muscular changes in SMA and correlates with functional status. Diaphragm thickening ratio can be normal even with severe compromise of respiratory muscles in quantitative analysis, and intercostal muscles were more affected than diaphragm.

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438 – Association of neutrophil/lymphocyte ratio with risk factors and severity in ischemic stroke

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Stroke is the second leading cause of death and the primary cause of disability in Brazil. Ischemia resulting from a stroke triggers an inflammatory response in the central nervous system, primarily mediated by neutrophils, lymphocytes, and macrophages. Recent research suggests that the neutrophil/lymphocyte ratio (NLR) in peripheral blood can serve as a reliable marker of systemic inflammation in various diseases. The present study aims to quantify the NLR in patients diagnosed with ischemic stroke, who received treatment at a specialized clinic of a university hospital situated in the central region of São Paulo. Additionally, the study aims to investigate if any correlation exists between NLR values and risk factors associated with ischemic stroke and its severity — assessed using the National Institutes of Health Stroke Scale (NIHSS). This retrospective study is based on medical records of patients aged over 18 years, who had ischemic stroke between 2017 and 2022. The collection of clinical data includes information on the type of ischemic stroke and the presence of risk factors. The quantification of NLR is performed by analyzing blood count data from the patients' medical records. Of the 194 stroke patients whose data were collected, 80.5% had NLR values exceeding 2, a value considered non-physiological by literature and preliminary data obtained from healthy individuals at the same university hospital. It was also observed that patients with a more severe condition, according to the NIHSS scale, had a higher NLR. However, no statistically significant correlation was found between NLR, types of stroke, and the presence of risk factors. In conclusion, stroke patients exhibit a higher neutrophil-lymphocyte ratio, which is associated with the severity of the condition but not with risk factors. NLR can be a prognostic marker of stroke.

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444 – Transient global amnesia after a phenol peeling

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Case report: A previously healthy 60-year-old woman presented to the dermatology clinic for a deep phenol peeling. A few minutes after the procedure, she had become confused and disoriented, so the neurology clinic was called. On the neurological assessment, the patient kept asking her sister where she was and how she got to the hospital. It was noted that the patient had deep anterograde amnesia and retrograde amnesia ranging from a few hours before the arrival at the hospital, with preserved episodic and semantic memories. The remainder of the neurologic exam was normal. She was submitted to basic laboratory evaluation, which showed no abnormalities. Brain magnetic resonance imagin showed a mild FLAIR (Fluid-Attenuated Inversion Recovery)

hyperintensity on the left medial temporal lobe, without signal alterations in the DWI (diffusion-weighted imaging) image. After about 4 hours of the onset, the disorientation had resolved and she was discharged after 24 hours of observation, asymptomatic.

Discussion: Phenol peeling is the most aggressive of all peels, and as this substance is absorbed by the skin, metabolized by the liver, and excreted by urine it is associated with systemic complications, such as cardiac arrhythmias and acute kidney injury. There are no previous reports of the occurrence of a transient global amnesia (TGA) episode after such a procedure, although it is known that it can be precipitated by physical exertion and psychological stress. The pathophysiology of a TGA episode is thought to be related to metabolic stress to the CA-1 subfield of the hippocampus. Although the association is unclear, we can make an assumption that phenol could have a disruptive metabolic effect on this particular brain area.

Conclusion: We report a case of TGA preceded by a deep phenol peeling. Although it is impossible to establish causality in this case, it is important that the neurologist be aware that certain medical conditions, procedures and substance administration can be associated with TGA.

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446 – Is it possible that the metamorphosis suffered by Gregor Samsa could have been a neurological syndrome known to Guillain Barre?

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In 1912, Franz Kafka gives life to the character Gregor Samsa, who undergoes a metamorphosis capable of transforming him into an insect. The metaphors used in the literature are widely applied as a way of rewriting a story in an attractive, creative and curious way, in this article, however, the possibility is raised that this narrative mentions a recurrent neurological pathology know as Guillain Barre Syndrome (GBS). The patology is characterized by an ascending, symmetrical polyradiculoneuropathy, which is often associated with viral prodromes or post-vaccination periods. In classic GBS, motor symptoms may be accompanied by associated sensory symptoms. During the text, it is possible to notice when Gregor says that "the lower part of his body was, perhaps at that moment, the most sensitive of all". One of the complications of GBS is characterized by weakness of the oropharyngeal and cervical muscles leading to hypophonic dysarthria as possibly mentioned in the book "an irrepressible and painful wheezing" and as the condition progress the realization that "certainly, they no longer understood their words". One of the most severe symptoms of Guillain Barré syndrome is the involvement of the ventilatory muscles. We correlated this possibility with the following text: "the shortness of breath was already beginning to manifest itself, since, even in the old days, I did not have a completely reliable lung". Diagnosis and correct treatment are essential to ensure lower morbidity and mortality in patients. The exposition of technical-scientific arguments associated with citations from the book as the basis of a literary clinical reasoning.

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448 – Cerebral venous thrombosis presenting as refractory status epilepticus on patient with complex arteriovenous fistula in a patient with PTEN mutation spectrum

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Introduction: The dural arteriovenous fistulas (dVAFs) are a subtype of brain arteriovenous malformations. Our case displayed an atypical case of complex dVAFs secondary to PTEN mutation spectrum manifesting with cerebral venous thrombosis, intense venous hypertension and status epilepticus.

Objetives/methods: Demonstrate that the clinical spectrum of PTEN mutation is heterogeneous and may involve patients without classic phenotypes presenting with atypical dVAFs.

Results: A 30-year-old famale patient was admitted with acute impairment of consciousness and refractory status epilepticus. He had a past history of complex dVAFs, submitted to multiple embolizations. Her admission MRI showed dVAFs in the transverse and sigmoid sinus associated with cerebral venous thrombosis of Rosenthal's basal vein causing venous congestion. She underwent four new embolizations, in addition to surgical disconnection of the venous drainage, but remained in a minimal state of consciousness. To elucidate the genetic mechanism behind the clinical phenotype, the exome showed a heterozygous mutation in PTEN c.932_933insA.

Conclusion: It is inferred that the coexistence of refractory and recurrent dVAFs is due to the existence of dormant channels between the dural arteries and the sinuses, which open when the sinus is occluded and venous pressure increases, or that venous hypertension causes hypoxia and production of angiogenic factors. In our patient, changes in angiogenic factors may be responsible for dVAFs, as in some patients with PTEN mutation. There are few case reports in the literature that show the correlation of patients with the PTEN genotype with arteriovenous fistulas without other systemic manifestations.

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451 – Tubular aggregate myopathy with rhabdomyolysis: an uncommon presentation in elderly male

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Introduction: 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. The aim to describe

an uncommon presentation with rhabdomyolysis overlapping with a tubular aggregate myopathy in elderly men.

Case: 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.

Results: 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 magnetic resonance imaging revealed a heterogeneous edema pattern, with predominant proximal impairment. No evidence of atrophy was detected. Statins were promptly withdrawn. After 5 to 7 days, creatine phosphokinase lowered to normal level and muscle biopsy confirmed tubular aggregates 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. This disease can present as a pseudo metabolic feature, without progressive weakness, mimicking the metabolic myopathies.

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452 – General knowledge of stroke in Aparecida city: reports from an academic expedition

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Introduction: Stroke is the second main cause of death and the leading cause of disability in Brazil. The incapacity to recognize the disease and its underlying risk factors can compromise its prevention and delay the search for a health professional.

Objectives: To analyze the general knowledge regarding recognizing and preventing stroke in a low-income community in Brazil.

Methods: This is a cross-sectional study conducted in the city of Aparecida in January 2023. Patients attending a program organized by students of "Faculdade de Ciências Médicas da Santa Casa de São Paulo" called "Programa de Expedições Científicas Assistenciais" were interviewed. The questions verified the understanding of stroke, the risk factors, and the capacity to recognize, prevent and manage it. An ethics committee previously approved this research, and a consent form was acquired. Patients younger than 18 years old or that did not sign the consent were excluded.

Results: From a total of 260 participants, 34.6% were men; the average age of 53,8 years; most had an incomplete lower secondary school (31.5%) level of education and self-declared as white (56.9%). Among the interviewees, 42.3% conceptualized stroke or the popular term "derrame", and 61.2% could not define either; 85.4% recognized at least one symptom, mostly paresthesia (26.8%). The most noted strategies for prevention were physical activities (33.8%), healthy eating (31.2%), and controlling blood pressure (21.9%). When witnessing a stroke, 54.6% would call the emergency service, and 0.76% would time the beginning of symptoms. In addition, 74.3% had an acquaintance who suffered from a stroke.

Conclusion: Most of the interviewees had little knowledge about stroke, which can lead to higher rates of preventable morbidity and mortality. Therefore, it is necessary to reinforce mechanisms for recognizing, preventing, and managing the disease.

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453 – Dync1h1 mutation in a patient with multiple neurological conditions

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Introduction: Mutations in the cytoplasmic dynein 1 heavy chain 1 gene (DYNC1H1) were first described in 2010 related to dominant lower extremity-predominant spinal muscular amyotrophy (SMA) 1 and after to Charcot-Marie-Tooth (CMT) disease, neurodevelopmental disorders, central nervous system malformations and hereditary spastic paraplegia. The aim this case report intends to show a recent known genetic mutation that can be related to two different conditions simultaneously — CMT and SMA.

Case report: F.R.S, male, 49 years old, noticed proximal left hemiparesis in his childhood; he started walking late and had a lot of falls as a child. Cognitive development was normal. The motor deficit progressed to a right hemiparesis at the age of 39 years old. Deficit progression was noticed because he had difficulty to go up and down stairs. Non-consanguineous parents, family history positive for physical disability of unknown cause. Neurological examination evidenced cavus foot, amyotrophy and tetraparesis with distal predominance of the lower limbs. Electroneuromyography was compatible with progressive proximal SMA. Thoracic and cervical magnetic resonance imaging showed syringomyelia and Chiari malformation (CM) type 1. Gene panel was compatible with DYNC1H mutation.

Discussion: It is well known DYNC1H1 mutation relation to CMT and SMA, but this case had some different conditions rarely described that also could be investigated to be related to this gene mutation as CM type 1 and syringomyelia.

Conclusion: The gene mutation identification is important to physician to promote recognition of multiple associated conditions. Until now, there is no treatment for the described conditions, but eventually, it could change and neurologists must be prepared to look for multiple diseases related to specific genetic condition.

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457 – Lambert-eaton myasthenic syndrome and anti-acetylcholine receptor antibody: myasthenia gravis overlap syndrome or false positive?

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Lambert-Eaton Myasthenic Syndrome (LEMS) is a rare neuromuscular presynaptic disorder that can be paraneoplastic or primary autoimmune. Clinical features include proximal weakness, autonomic dysfunction, and areflexia.

The pathophysiology is related to production of antibodies against voltage-gated calcium channels (VGCC), and consequent reduction in the release of acetylcholine (ACh). The diagnosis of LEMS is clinical, serological, and electrophysiological and Myasthenia Gravis (MG) is a differential diagnosis. The aim is to report a case of LEMS with negative anti-VGCC and positive anti-acetylcholine receptor antibody (Anti-AChR).

Case report: A 62-year-old female, former smoker, complained of weakness in the lower limbs for one year, initially intermittent, and later continuous, occasional diplopia, and dyspnea. Physical examination showed proximal weakness and mild asymmetric eyelid ptosis. Electroneuromyography showed presynaptic defect of the neuromuscular junction, with an increase of up to 163% after high rate nerve stimulation. Anti-AChR antibodies = 1.14 (Reference value < 0.4) and anti-VGCC = 35.8 (Reference value < 40). The DELTA-P (Dutch-English LEMS Tumor Association Prediction) Score was 2 and the screening tests for neoplasia at the time of diagnosis, and six months later were normal. The disease progressed for about a year until it stabilized with pyridostigmine and human immunoglobulin. The anti-AChR antibody is characteristic and a diagnostic criterion for MG. The positivity of this antibody in the case of LEMS is not usual. Such a positive result in this case should be interpreted as an overlap syndrome of MG and LEMS, with a differential diagnosis of false-positive serological result.

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458 – Analysis of the intestinal microbiota and its relationship with neuropathologies

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Introduction: The human intestinal microbiota corresponds to the ecosystem of colonizing microorganisms of the intestine that has an important role of protection to the organism. In addition, it has a direct relationship with the nervous system, known as the bowel-brain axis. Changes in the intestinal microbiota have been associated with several neuropathologies, and disbiosis repair has been shown to improve specific symptoms of some diseases.

Objectives: This study aims to analyze the neurological implications caused by intestinal microbiota in humans.

Methods: Review of integrative literature, consulted the Databases PubMed, SciELO and Google Academic. Chosen as descriptors (DeCS): "Microbiota", "Gastrointestinal Microbiome" and "Nervous System Diseases" separated by Boolean connectors, and articles in English and Portuguese.

Results: In this sense, among the therapeutic techniques that objectify to recolonize the "'sick" intestine, the use of probiotics and fecal microbiota transplantation stand out. Symbiotics, a combination of probiotics and prebiotics, proved beneficial for symptomatological manifestations of neuropsychic disorders such as depression and chronic stress.

Conclusion: Although some of the relationships of the intestinal-brain microbiota axis and changes in the intestinal microbiota, as well as the pathophysiology and benefits arising from its health, there is still a lack of studies to make consensus whether a change in the intestinal microbiome would be an epiphenomenon or the cause of neuropathologies in humans.

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461 – Gastrointestinal dysmotility associated with Parkinson's disease's mechanism

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Introduction: Parkinson's Disease (PD) is a condition of the brain that consiste of the death of dopaminergic neurons in the substantia nigra, therefore causing dyskinesias and dystonias. Besides the motor symptoms, the neurogastro motility is affected by the disease, since gastrointestinal dysfunction is a frequent and clinically relevant symptom of PD.

Objectives: To link the neural pathways and neurotransmitters that involve the neuroenteric system control and the PD's pathology.

Methods: A systematic literature review was performed based on data extraction through the advanced research engine from Pubmed. Publications with the descriptors "dysmotility" OR "gastro motility" AND "Parkinson" were selected.

Results: Through clinical and pre-clinical studies on PD, there has been hypothesized a gut-brain axis that is connected through hormones, neurotransmitters and dopamanergic inputs. This hypothesis is supported by evidence in the showing of accumulation of alpha-synuclein in the vagal system and Enteric Nervous System, the use of drugs such as peripheral dopaminergic blockers and serotonin for gastroparesis, the ghrelin effects on the central dopaminergic system through modulation of the mesencephalic dopaminergic signaling tested on rats, the gastrointestinal autonomic neuropathy detected in PD patients and the establishment of gut dysmotility before motor onset symptoms. Therefore, dysmotility isues such as delayed gastric emptying may not only be a symptom of PD, but also contrubute to the pathogenesis itself through impaired signaling.

Conclusion: The gut-brain axis can be not only a tool for PD diagnosis but also a treatment target to restrain the advance of the disease. Although many articles are related this subject, there is a lack of designed trials for atypical movement disorders. To explore the dysmotility in PD, there is a need for multi-modality standardized tests to evaluate severity and prevalence.

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462 – The epidemiology of accidental tetanus in Brazil

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Introduction: Tetanus is an acute intoxication caused by tetanospasmin, which binds irreversibly to spinal cord and brainstem receptors, being taken there through retrograde axonal transport, blocking inhibitory neurotransmission, promoting muscle contraction, spasms and a hypersympathetic state.

Methods: The work is a descriptive study that statistically analyzes and describes the cases of accidental tetanus in Brazil between 2012 and 2022 through data obtained by the Information System of Notifiable Diseases (SINAN).

Results: The total number of cases was 2.596 and 846 progressed to death. The annual media was 236 cases. From 2012 to 2022, the percentage change in the number of annual new cases was -38.55%, demonstrating a drop in the number of cases. Also, the percentage change in the lethality from 2012 to 2022 was -76.53%. Among the regions, Northeast had the highest prevence (31.9%). Regarding skin color/ethnicity is: White: 32.51%; Black: 8.35%; Brown: 52.54%. The most affected age group was 40–59 years old (n% = 39.21). Of the 846 reported deaths, 313/846 occurred in this age group. About sex, 2212 cases ocurred in men. Data on the education show that 129 were illiterate; 29.9% have an incomplete secondary education; 8.16% have completed secondary education; Ignored/blank: 47.45%.

Conclusion: Despite the reduction in cases, a reflection of greater vaccination coverage, tetanus still has a high lethality. The increase in cases with aging, but still in working age can be explained by the reduction of reflexes, ability and visual acuity. The pattern of higher prevalence in men can be elucidate by the work distribution, in which they are the majority in professions with higher risk, such as agriculture and construction. Furthermore, there is a pattern of low education linked to a greater number of cases, which can be explained by the work pattern and low adherence to vaccination campaigns in these groups.

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463 – Clinical features of headaches in an urban Mennonite group in Southern Brazil

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Introduction: Genetic variants play a pathophysiological role in headaches, especially regarding migraine. The Mennonites are a group that originated during the religious reformation in Europe, during the Sixteenth Century and have been geographically and genetically isolated throughout their history, harboring a distinctive distribution of diseases.

Objectives: To determine the clinical features of headaches in a group with direct Mennonite ancestry (MG) contrasting with other urban community members, that share a similar environment (control group, CG).

Methods: Subjects with headaches were asked to complete a questionnaire covering: the type of headache, presence of aura, frequency and duration of attacks, pain location and severity, analgesic use, premonitory and postdromic manifestations, Depressive Thoughts Scale, Epworth Sleepiness Scale (ESS), General Anxiety Disorder-7, Patient Health Questionnaire-9 (PHQ-9), Migraine Disability Assessment, and Composite Autonomic System Score.

Results: 103 participants were included (CG: 45, MG: 58). Migraine was the most common form of headache (CG: 91.1%, MG: 81.0%, P = 0.172), followed by tension-type headache (CG: 8.9%, MG: 15.5%, P = 0.381). Aura was identified in 44.4% and 39.7% in the CG and MG, respectively (P = 0.689). The groups differed only concerning the frequency of retroorbital pain (CG: 55.6%, MG: 32.8%, P = 0.027), PHQ-9 (CG: median 7, range 0–22; MG: median 5, range 0-19; P = 0.031) and ESS (CG: median 0, range 0–270; MG: median 0, range 0–108; P = 0.048) scores.

Conclusion: There were no major differences in the clinical attributes of headaches between the MG and CG. However, the latter showed more diffuse pain, sleepiness and depressive symptoms. Specific genetic or epigenetic variants in Mennonite descendants might account for these differences.

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464 – Detectable cognitive consequences by eye movement in post-traumatic brain injury individuals: a systematic review

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Introduction: Traumatic Brain Injury (TBI) is a leading cause of morbidity and mortality worldwide. TBI patients often suffer permanent motor, cognitive, and other negative effects that impact their daily lives.

Objectives: This systematic review aims to measure and analyze the cognitive impacts of TBI using ocular tracking techniques.

Methods: This is a systematic review of articles published between 2011 and 2021 in Portuguese, Spanish, and English. The articles were found in the PubMed and LILACS databases using the following research descriptors: "eye tracking"; "brain concussion" and "traumatic brain injury". The articles were filtered using the Rayyan program, a double-blind process based on the criteria established in this work, to select eligible and non-eligible articles.

Results: A total of 71 articles were found, but after excluding duplicates and non-eligible articles using the Rayyan program, 58 articles were deemed suitable for our trial. After applying the blind on and off methods, only 27 articles matched the previously established criteria to initiate the official trial. Our analysis revealed that TBI is prevalent in male adults between the ages of 20 and 35 years old. Most of the selected articles reported measurable deficits found in moderate to severe TBI detected with eye-tracking technology. Abnormal ocular movements, such as saccadic, anti-saccadic, fixation, and others, were associated with cognitive impairment.

Conclusion: This review concludes that the eye-tracking technique is an efficient tool to measure cognitive impairment resulting from TBI. However, many articles presented a narrow sample range, and broader studies are needed before using the data to the general population, as stated as a goal in some articles.

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465 – Manometry viability for Parkinson's disease constipation assessment

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Introduction: Parkinson's Disease (PD) is a condition in wich the dopamine presence is lowered, causing motor and non-motor symptoms. Constipation of different severities is related to the pathogenesis of PD and is a common affection before the onset of disease. Despite these factors, there is a lack of specific diagnostic tools for the assessment of this symptom in PD, which difficults pathophysiological evaluation of constipation in those with PD or refractory constipation.

Objectives: Identify if the findings in anorectal manometry could determine

the existence of PD and be included in the examination process of patients with history of constipation.

Methods: A systematic literature review was performed through an advanced search on Pubmed engine, in which 21 articles with the descriptors "anorectal" AND "manometry" AND "Parkinson" were selected.

Results: Using anorectal manometry, it was detected that patients with PD constipation and functional constipation have dysmotility of the colon and rectum, but there are certain differences in segmental colonic transit time and rectal anal pressure between the groups. Compared with idiopathic constipation, anal sphincter pressures on voluntary squeeze were lower in the PD patients. Furthermore, defecatory dysfunction is found early in the course of the disease additionally to later stages.

Conclusion: The use of manometry allows quantitative analysis of anorectal dysfunctions and provides early recognition of pelvic floor dyssynergy and colorectal assessment in PD. Besides, the findings in PD constipation are distinguishable from the findings in other types of constipation and in conditions of normalcy. Therefore, anorectal manometry could be used in the examination of patients with history of constipation in order to facilitate early recognition. In order to incorporate this practice, there is a need for more studies comparing the findings in PD and in other types of constipation.

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466 – Arginase 1 deficiency: a differential for progressive ataxia and intellectual disability

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Introduction: 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. Spastic paraparesis with onset in early childhood is the most obvious sign of the disease but other symptoms include irritability, delayed growth and development, ataxia, recurrent vomiting, feeding/protein aversion, and anorexia. Brain imaging may reveal cerebral atrophy and cerebellar atrophy may also occur. The *ARG1* gene sits on chromosome 6 (6q23) and there are at least 43 potentially disease-causing variants in *ARG1*.

Case report: A 56-year-old female patient with no parental consanguinity and unknown perinatal history — she had no contact with her parents. Since childhood, she reports never liked meat and that she had a lack of appetite and frequent vomiting. She also reports irritability without triggers and difficulties at school — illiterate despite 4 years of schooling. She denied gait difficulties until it was around 50 years, when she presented with progressive imbalance and incoordination. She is currently unable to walk without bilateral support due to severe gait ataxia in addition to appendicular ataxia.

Results: Extensive laboratory metabolic investigation was performed, serologies, electroencephalogram and cerebrospinal fluid without alterations. Brain magnetic resonance imaging showed diffuse cortical and cerebellar atrophy. Whole-exome sequencing revealed a homozygous pathogenic variant mutation p.Arg308Gln in the *ARG1* gene.

Conclusion: In systematic reviews, spastic paraparesis is the hallmark of the disease caused by *ARG1* deficiency, however, there are descriptions of rare, predominantly ataxic cases. In basically everyone, intellectual impairment is expected. Therefore, *ARG1* deficiency is still under construction and can also be a differential diagnosis for ataxia and intellectual impairment.

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468 – The importance of non-pharmacological strategies and the management of depression in patients with multiple sclerosis: an integrative review

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Introduction: Multiple sclerosis (MS) is a chronic and demyelinating disease of the central nervous system. Among the symptoms that patients with MS may develop, depression stands out, which is associated with an important decline in quality of life. The presence of this symptom is associated with a worse prognosis. There is much support in the literature of the positive role that non-pharmacological measures play in controlling depression in patients with MS, with a range of options for the patient to choose from.

Objectives: To analyze studies that deal with non-pharmacological therapeutic strategies against depression in patients with MS.

Methods: This scientific article was written in the form of an integrative literature review, which used the databases of the Scielo and the PubMed as a source of extraction of publications. The keywords used were: "Multiple sclerosis", "Depression" and "Complementary Therapies". Among the inclusion criteria, articles were included between the years 2015 and 2020, published in English or Portuguese, available in full and in the form of meta-analyzes, systematic reviews and randomized clinical trials.

Results: The bibliographic survey carried out led to 15 scientific articles, all of which were randomized clinical trials and published in English. The articles reported that non-pharmacological interventions, such as Cognitive Behavioral Therapy, physical activity, yoga, Guided Imagery, mindfulness and diet, had positive effects on depressive symptoms in MS.

Conclusion: The survey of data for the construction of the scientific article led to the conclusion that non-pharmacological measures play an important role in the control of depressive episodes in patients with MS. The combination of drug therapy and non-pharmacological practice leads to better responses.

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469 – Pharyngeal-cervical-brachial variant of Guillain-Barre syndrome overlap with Bickerstaff brainstem encephalitis

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Guillain-Barré Syndrome (GBS) is a rare disease. Its classic presentation is an acute tetraparesis and absence or diminished tendon reflex. Clinical vari-

ants such as Pharyngeal-cervical-brachial (PCB) and Bickerstaff brainstem encephalitis (BBE) can occur. We report a case of PCB and BBE overlap, with electrophysiological pattern of acute motor sensory axonal neuropathy. A 36-year-old man with recent dengue virus infection, was evaluated in the emergency department, complaining of double vision and eyelid drop for one week. Neurological examination showed palpebral ptosis, dysarthria, cervical and upper limb weakness, dysphagia, and diplopia with absent deep tendon reflexes in the upper limbs. The cerebrospinal fluid analysis as well as head computed tomography were unremarkable. Electroneuromyography demonstrated axonal sensory-motor polyneuropathy. The patient was treated with Human immunoglobulin, plasmapheresis and ventilation support. There was gradual improvement after treatment, with complete recovery after 6 months. About 60% of GBS cases experienced previous infection preceding neurological manifestations in up to 8 weeks. Although rare, dengue virus is a well-known possible trigger of GBS. Variants may overlap due to the existence of a crossed response against the glycoside GQ1b, related to PCB e BBE, expressed in peripheral nerves, spinal cord, cranial nerves, and brainstem, which probably occurred in the reported case. It is important to recognize isolated or superimposed clinical variants of GBS and its differential diagnosis.

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470 – Phenotypic and neuroimage differences in Corticobasal syndrome from two clinical cases

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Corticobasal syndrome (CBS) is a neurodegenerative condition characterized by cognitive and motor symptoms and neurologic-functional progressive deterioration. The phenotypes can be associated with the underlying proteinopathies like Corticobasal Degeneration (CBD), Progressive Supranuclear Palsy or Alzheimer's Disease (AD). This work aimed to study, in two cases, the correlations between clinical phenotypes, neuroimage markers and the underlying pathology. Clinical and neuroimage data of two patients was revised. The patient 1, 62-year-old, female, incomplete elementary school, presented progressive non fluent aphasia (NFA) for two years and evolved with limb apraxia and extrapyramidal signs of the right upper limb. The Fluorodeoxyglucose Positron Emission Tomography (FDG-PET) revealed moderate hypometabolism of temporal lobes and middle frontal gyrus predominating on the left side. It fulfilled the criteria for CBS and suggested CBD. The patient 2, 57-year-old, female, graduated, presented NFA, hemineglígence, cognitive impairment, bradykinesia and myoclonus. PET-FDG revealed asymmetric hypometabolism in the superior and inferior parietal lobes, posterior cingulum gyrus and worse in precuneus. The investigation of cerebrospinal fluid revealed consumed amyloid beta and increased phosphorylated and total TAU. It fulfilled probable CBS and suggested AD. These cases demonstrate the role of the PET-FDG in CBS and reveal its possible metabolic signatures: when caused by AD, the hypometabolism predominates in the posterior temporoparietal areas, and when caused by tau pathology, in the thalamus and brainstem, mainly contralateral to the most affected side. CBS has been widely studied with relatively new methods, like the cerebral FDG-PET. Studies that deepen the phenotypic heterogeneity and biomarkers of CBS would be important to improve its classification, prognostic and treatment.

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474 – Neuromyelitis optica in childhood: a case report

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Introduction: Neuromyelitis optica (NMO) is a relatively rare inflammatory disease that primarily affects the spinal cord and optic nerves.
Objectives: To report an atypical case of NMO in the child, with brainstem syndrome.
Case report: A 12-year-old patient, 20 days after Pfizer vaccination for severe acute respiratory syndrome coronavirus-19 (SARS-COV-19), started with asymmetrical ascending flaccid paraparesis, with global arreflexia, ataxia, dysphagia and dysarthria. On admission, after 2 months, thought of acute polyradiculoneuritis, immunoglobulin was administered without improvement. However, the patient evolved with signs of pyramidal release, methylprednisolone was administered with a good outcome. Cerebrospinal fluid: cytology: 0/mm^3, glucose: 59/mm^3, protein: 34/mm^3. electroneuromyography: upper and lower limbs show signs of polyradiculoneuritis in the hyperacute phase, the nature cannot be classified as myelin or axonal; Cranial magnetic resonance imaging (MRI): signal alteration compromising supra and infratentorial structures, T2/FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) symmetrical hypersignal in the bulbopontine transition suspicious for an demyelinating process. Cervical/thoracic spine MRI: no alterations. Anti Aquaporin-4 IgG: reagent.
Discussion: Clinical features are frequently more diverse than just optic neuritis and transverse myelitis in children, making a diagnosis can be challenging. The case series of pediatric NMO have shown a female preponderance and almost all NMO-IgG positive, which had relapsing course predicted by seropositivity. The median age of children diagnosed with NMO ranges from 10 to 14 years. Coexisting autoimmune disorders were reported in 42%. Longer term immunotherapy reduce the chance of further potentially disabling attacks.
Conclusion: Diagnosis of NMO isn't easy, especially when patients are children, the disease is potentially severe and needs to be better understood and managed.

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479 – Resection of low-grade temporal gliomas and the improvement of convulsive seizures

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Introduction: Epilepsy is a neurological disorder marked by recurring seizures, and secondary epilepsy refers to seizures that are generated by an underlying medical condition or injury. Low-grade temporal gliomas (LGTGs) frequently have epileptogenic potential, causing seizures. Tumor resection is often the preferred treatment when timing and compatibility with the patient and tumor attributes are determined.
Objectives: To discuss the main factors in the medical literature relevant to the improvement of seizures by resection of LGTGs and their surgical features.

Methods: A literature review was performed within the PubMed database, using the keywords "low-grade gliomas", "low-grade tumors", "resection", "seizures" and "epilepsy". Publications from 2010 to 2023 were included.
Results: Studies point out that gross full extension resection of LGTGs to achieve seizure freedom results in superior positive outcomes when compared to partial resection for Engel class I patients. Of the patients who had a partial resection, memory deficits were frequent. In addition, recurring epilepsy related to low-grade tumors and the time span of epilepsy were reported to be higher in children than in adults. Postoperative outcomes of patients with mesial temporal lobe lesions outperformed those with lateral temporal tumors. The addition of hypocampectomy and/or corticectomy of the anterior temporal lobe further improved the seizure freedom rate when compared to gross total lesionectomy.
Conclusion: Gross total resection of LGTGs provides a more favorable outcome than partial resection. After surgery, the seizure freedom rate is high (> 70%), and resection type is a significant predictor of seizure recurrence. Subtotal resection has a lower seizure-free rate compared to total lesionectomy, with additional benefits seen from hypocampectomy and/or corticectomy of the anterior temporal lobe. Tumor pathology or laterality did not significantly predict seizure freedom.

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482 – Community-acquired pneumococcal meningoencephalitis associated with neurosyphilis in an immunocompetent patient: case report

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Case report: A 28-year-old man with no comorbidities was admitted to our institution with a history of sudden holocranial headache, associated with fever, lowered level of consciousness and meningismus with the need for orotracheal intubation. Computed tomography of the brain was normal and the cerebrospinal fluid (CSF) on 05/28/2022 was yellowish, cloudy, glucose 6.0 mg/dL, protein 752 mg/dL, cells 25,600 mm^3 (neutrophils 92%, lymphocytes 5%), red blood cells 258 mm^3, CSF Venereal Disease Research Laboratory (VDRL) 1/8, serum VDRL 1/32, treponemal test positive, human immunodeficiency virus (HIV) negative. Ceftriaxone, ampicillin, and acyclovir were empirically started. Pneumococcus was identified in the culture of CSF and blood cultures on admission and the antibiotic regimen was adequate, maintaining only ceftriaxone. Antibiotic therapy lasted 14 days, he was discharged after 16 days of hospitalization, for outpatient follow-up, with no neurological deficits. Control lumbar puncture on 12/23 revealed clear, colorless CSF, glucose 56 mg/dL, total protein 31.8 mg/dL, no cells or red blood cells, cultures negative.
Discussion: Streptococcus pneumoniae is the most common cause of meningitis in adults, in older adults and in the current era, neurosyphilis, is most frequently seen in persons with HIV. There are no similar cases described in the literature. Despite the effectiveness of current antibiotics in clearing bacteria from the CSF, bacterial meningitis continues to cause significant morbidity and mortality worldwide. We describe a rare case of an immunocompetent patient with community-acquired pneumococcal meningoencephalitis associated with neurosyphilis treated with ceftriaxone who did not present sequelae or need for retreatment.
Conclusion: It´s a rare cause of meningoencephalitis and has significant morbidity and mortality. More studies are needed regarding susceptibility to meningoencephalitis by multiple germs in immunocompetent patients.

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483 – Infantile vaccine and cancer-associated cerebral venous thrombosis: an unusual cause of excessive daytime sleepiness

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Case presentation: A 10-year-old male presented with a sudden onset of excessive daytime sleepiness (EDS) in the last 36 hours. Currently, the patient sleeps around 18 hours per day associated with headache and irritability. Patient received his second dose of Pfizer-BioNTech COVID-19 (coronavirus disease) vaccine. He has a past medical history of subtotal resection craniopharyngioma around one year ago. Neurology exam showed new deficits of motor aphasia and drowsy but aroused easily to voice. Axial FLAIR (Fluid-Attenuated Inversion Recovery) and T2-weighted magnetic resonance imaging (MRI) show symmetrical hyperintense lesions on the thalamus. Magnetic resonance venography shows a lack of flow in the vein of galen, straight, superior sagittal and bilateral transverse extending to the right sigmoid sinus. The findings above confirmed the diagnosis of cerebral venous thrombosis (CVT). Patient received enoxaparin for three months. Patients complete resolution of symptoms and a new MRI shows complete recanalization.
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 an extensive CVT in the bilateral thalamus. Bilateral lesions of the medial thalamus can cause hypersomnia and decrease consciousness. Infection, trauma, neurosurgery, malignancy and thyroid problems are risk factors of children CVT. Another risk factor to be considered is the COVID vaccination. In literature, there are more reports of thrombocytopenia and CVT is very rare. Although these factors are associated, they are not necessarily causality.
Conclusion: Clinicians must be aware of CVT as a differential diagnosis of EDS, especially in the presence of sudden EDS onset or neuropsychological signs.

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487 – Radiculopathy C8-T1 atypical initial presentation: a case report

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Introduction: Radiculopathy is a common condition whose symptoms can include pain, sensory change, and motor weakness owing to mechanical and chemical irritation of the spinal nerve root. The aim is to report atypical clinical presentation of radiculopathy secondary to neoplasia.
Case report: A 62-year-old male patient was admitted with pain in the right scapular region of onset 2 months ago and progressive worsening. On patient

admission, he presented isolated loss of strength in the right hand with force grade V in the proximal right upper limb and right dropped hand and living reflexes in the right upper limb. Thoracic/cervical spine magnetic resonance imaging with expansive formations in the thoracic bone marrow of neoplastic aspect, compressive fracture of the vertebral body of T1 with impairment of the thoracic cord, reduction of the vertebral canal at level C3-C4. Electroneuromyography: bilateral acute C8-T1 radiculopathy, worse right and chronic radiculopathy on the left. Subsequently, he evolved with brachial paraparesis, exalted global osteotendinous reflexes, and inexhaustible clonus in the lower limbs. C4-T5 posterior cervical arthrodesis was performed.
Discussion: Cervical radiculopathy may leave the clinician perplexed by lack of clinical-radiological correlation. Of special interest is the weakness of the intrinsic hand muscles without radiological evidence for C8 radiculopathy. Overlapping with the T1 root, the C8 root innervates the finger flexors and all the intrinsic hand muscles. C8 radiculopathy is characterized by radicular neck pain, hand weakness, and sensory deficit of the ulnar fingers and medial forearm.
Conclusion: The lack of clinical-radiological correlation should not mislead the clinician from the correct diagnosis, and should not delay the surgical decompression of the cord and the roots.

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490 – Atypical presentation of Stiff Person syndrome: case report

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Introduction: Stiff Person syndrome (SPS) is a rare, immune-mediated neurological disease related to several antibodies, the most obvious being Anti-glutamic acid decarboxilase (GAD). This case intends to show atypical clinical presentation of Stiff Person Syndrome, in which the patient started with ataxia and gait alteration, evolving with dimidiated spasms.
Case report: L.S.M, male, 58 years old, denied previous comorbidities, smoking and alcoholism. He woke up with vertigo, associated with dysarthria and ataxia in the left side of the body. Two weeks after the onset of symptoms, he evolved with gait alteration and began to have episodes of painful, intermittent muscle spasms in the left upper and lower limbs. He presented negative serology for human immunodeficiency virus, syphilis and viral hepatitis, in addition to non-reagent rheumatological markers. Magnetic resonance imaging revealed a focus of signal alteration affecting the corticosubcortical surface of the cingulate gyrus on the right side, possibly corresponding to an inflammatory/demyelinating process. Cerebrospinal fluid analysis showed the presence of oligoclonal bands and anti-GAD reagent, with non-reactive anti-aquaporin 4.
Discussion: The typical symptomatology of SPS involves the musculoskeletal component, representing up to close to 93% of the symptoms referred by the patients. Among other predominant symptoms, pain (82%) and bulbar dysfunction (47%) stand out. Some patients may have atypical symptoms, especially during the onset of symptoms. The case exposes the predominance of vertigo and ataxic symptoms, unusual for this stage of SPS, which may lead to difficulties in the investigation.
Conclusion: Recognizing the atypical patterns of clinical presentation is an important responsibility of the neurologist, since early diagnostic elucidation is essential for defining the patient's prognostic determination therapy.

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491 – Machine learning algorithms to predict delayed cerebral ischemia after subarachnoid hemorrhage: a systematic review and meta-analysis

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Introduction: Delayed cerebral ischemia (DCI) is a common and severe complication after subarachnoid hemorrhage (SAH). Logistic regression (LR) is the primary method to predict DCI, but it has low accuracy.
Objectives: This study assessed whether other machine learning models can predict DCI after SAH more accurately than conventional LR.
Methods: PubMed, Embase, and Web of Science were systematically searched for studies directly comparing LR and other ML algorithms to forecast DCI in patients with SAH. Our main outcome was the accuracy measurement, represented by sensitivity, specificity, and area under the receiver operating characteristic.
Results: In the six studies included, comprising 1,828 patients, about 28% (519) developed DCI. For LR models, the pooled sensitivity was 0.71 (95% confidence interval [CI] 0.57–0.84; $P < 0.01$) and the pooled specificity was 0.63 (95% CI 0.42–0.85; $P < 0.01$). For ML models, the pooled sensitivity was 0.74 (95% CI 0.61–0.86; $P < 0.01$) and the pooled specificity was 0.78 (95% CI 0.71–0.86; $P = 0.02$).
Conclusion: Our results suggest that ML algorithms performed better than conventional LR at predicting DCI.

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494 – Huntington Disease-Like 2: a case report

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Case presentation: Male, 56-year-old, previously epileptic started with involuntary movements in the right hand at 47 years old evolving to torso, incoordination, behavioral and cognition disturbs. Paternal grandmother, father and cousin with similar symptoms, in addition to four asymptomatic children. From the onset of symptoms, it progressively worsened presenting involuntary movements, hallucinations, aggressiveness and neck drop. During the neurological examination had frequent cervical falls, tremors at rest in the limbs, and mood swings. Cranial magnetic resonance imaging (MRI) was with atrophy of the caudate nucleus and putamen. Genetic test for Huntington's Disease without evidence of characteristic expansion of the disease and Huntington Like 2 Test (Junctophilin-3) compatible with characteristic expansion of the disease.
Discussion: Huntington's Disease Like-2 has an autosomal dominant character. A rare disease related to repetitive mutations of cytosine-thymine-

guanine in the Junctophilin-3 gene. Frequently among 29–41-year-olds and represented by progressive movement disorders, dementia and psychiatric alterations and survival rate of 10 to 20 years after the onset of symptoms. Atrophy of the caudate and cerebral cortex is identified in cranial MRI. Diagnosis' based on genetic testing and treatment is symptomatic in addition to genetic counseling.

Conclusion: In this case the patient presented progressive movement disorders in addition to psychiatric and cognitive alterations as other family members without previous diagnosis of Huntington disease-like 2. Due to the rarity of the disease beside the genetic chance of hereditary cases follow-up with a specialist for symptomatic family members had been requested in addition to genetic counseling since 50% of siblings are at risk of developing the disease.

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495 – A case of neurosarcoidosis presenting with multiple cranial neuropathies

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Case presentation: Female, 40-year-old, started in 2018 holocranial headache with progressive worsening. Between 2018-2020, systemic symptoms started: skin papules, dry cough and polyarthralgia, in addition to new neurological symptoms: bilateral hypoacusis, hyposmia, hypogeusia, tongue deviation, dysphagia and dysarthria. In 2021, she had rapidly progressive visual loss in the left eye, until being completely blind. The patient started a follow-up in the neurology department at Hospital Universitário-Universidade Federal de Juiz de Fora and was admitted to investigate the case. Brain Magnetic Resonance Imaging showed pachymeningeal enhancement and nodular thickening, suggestive of neurosarcoidosis (NS). The investigation revealed bilateral optic neuritis, pulmonary and cardiac involvements. Immunosuppressive therapy with oral corticosteroid was started, with symptomatic stability. A meningeal biopsy showed a focal granulomatous inflammatory process without necrosis, negative for amyloidosis and infectious causes, confirming the diagnosis. After the result, methylprednisolone pulse was performed, subsequently suspended due to side effects and lack of clinical response. For this reason, we started methotrexate, with resolution of the headache and stability of deficits. Recent neuroimaging showed no signs of disease activity.

Discussion: Sarcoidosis is an inflammatory systemic disease that can affect any organ, being the most usual the lungs, lymph nodes, eyes and skin. NS represents 5–10% of all cases. Cranial neuralgias are the most common manifestation. The diagnosis is defined by the clinical syndrome, imaging and histopathological findings, and exclusion of mimics. Treatment requires high doses of corticoids with good and fast improvement. In this patient, the diagnosis was reached after three years of presentation, leading to late treatment, and despite current stability, she had permanent neurological deficits.

Conclusion: This case demonstrates the difficulty in diagnosing NS, for lacking specific diagnostic tests, and delaying the treatment can lead to irreversible neurological damage.

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499 – Idiopathic basal ganglia calcification and Hoarding disorder

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Introduction: Basal ganglia calcifications are associated with many neurological and metabolic disorders, being present also on asymptomatic patients. It may present in its primary form, including familial and sporadic cases. Its secondary form is associated especially to hypoparathyroidism but also associated to infections, toxic exposure, rheumatologic diseases, mitochondrial disorders. It has an heterogenous clinical presentation with movement disorders and neuropsychiatric symptoms.

Case presentation: A 66-year-old patient presented with a progressive hoarding disorder for the last six years. In the last 2 years started an aggressive behavior, loss of acquired skills, urinary incontinence, sleep-wake cicle disorder and one episode of focal seizure. Physical examination revealed bilateral asymmetrical tremor, bradykinesia and cogwheel rigidity. MoCA test was 23/30 for 12 years of schooling. Brain Computed Tomography showed calcifications in basal ganglia affecting predominantly pallidum e thalamus and cerebellar hemispheres. Brain Magnetic Resonance Imaging revealed hypointensites in the same regions and in nucleus caudate suggestive of calcification. Laboratory testing for endocrine and calcium metabolism was normal. No clinical signs of other disorders.

Discussion: We presented a case of probable Idiopathic Basal Ganglia Calcification initially treated as a hoarding disorder. The normal laboratory results, lack of other clinical signs and familial history suggests a primary sporadic form that might be due to de novo mutations or transmitted by asymptomatic parent. The most commonly mutations in SLC20A2, PDGFB and PDGFRB but genetic testing is commonly unavailable. Parkinsonism is the most common movement disorder and the neuropsychiatric features include cognitive impairment, psychotic and obsessive compulsive disorders.

Conclusion: This case demonstrates that attention is needed to the progression of psychiatric disorders suggesting some rare neurological disorders.

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502 – Cadasil syndrome: a case report

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Introduction: Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is a genetic disease with an autosomal dominant transmission due to pathogenic variants in the *NOTCH3* gene on chromosome 19. This condition causes angiopathy and is associated with high risks of strokes and vascular dementia in young adults. The present

case reports a 60-year-old woman with the diagnosis of this condition after moderate cognitive impairment and advanced microangiopathy.

Case report: KAO, 60-year-old, presented for evaluation after a 10-year moderate cognitive impairment, with short term memory loss without functional impairment at that point. She also referred multiple episodes of neurological deficits along the years, including vertigo and gait impairment. Neurological examination showed a wide-based gait, dismetria and disdiadococinesia and global hiperreflexia. She scored 15/30 on the Montreal Cognitive Assessment, with noted attentional deficits, memory loss and visuoespacial impairment. Family history was positive for her 62-year-old mother having history of stroke, followed by major cognitive impairment. Brain Magnetic Rrsonance Imaging showed severe white matter impairment with confluent hyperintensities — Fazekas 3, in addition to hipointensity in frontal lobes and in left side of cerebellum, suggesting hemosiderin deposition. Cardiological exams didn't show any other significant cardiovascular risk factors. The patient was submitted to genetic testing that confirmed an atypical heterozygous pathogenic variant in *NOTCH3*.

Conclusion: CADASIL is caused, in approximately 95% of cases, by point mutations in the *NOTCH3* gene (a subtype of transmembrane receptor that acts in signaling between neighboring cells, being located in vascular muscle cells). This gene is located on chromosome 19p13.12 (OMIM #125310). The prevalence is 2:100,000, but varies in different populations. Penetrance is believed to be 100%, but it is age dependent. The severity of symptoms and disease progression are diverse, with great intra and interfamilial phenotypic variability

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506 – Analysis on the university use of anxiolytics: a review

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Introduction: The use of anxiolytics is becoming increasingly popular in today's society, especially among academics. However, improper use can cause adverse effects and become a health problem.

Objectives: This is an integrative literature review whose objective is to evaluate the neurological side effect of anxiolytics among students.

Methods: This is a systematic review, consulted the Databases PubMed, SciELO and Google Scholar. Chosen as descriptors (DeCS): "Anti-Anxiety Agents", "Nervous System Diseases" and "Students" separated by Boolean connectors, and articles in English and Portuguese.

Results: These evidenced the exacerbated and unknown use of anxiolytics as a problem in the school environment, including cognitive ones. Moreover, it was also exposed that neurological effects, in addition to cognition, can range from changes in social relationships, such as behavioral (mood destabilization, convulsion, psychosis). In addition, given its potential for dependence, it was also evidenced that medical prescription is essential for both initiation and cessation of treatment.

Conclusion: The exacerbated and unknowing use of anxiolytics as a problem in the school environment, having the potential to negatively affect mental and physical health. It is possible to infer those problems such as the failure of communication between the health professional and the patient, abandonment of treatment and disobedience to medical guidance are still recurrent.

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507 – SUNFISH parts 1 and 2: 4-year efficacy and safety data of risdiplam in types 2 and 3 SMA

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Introduction: Spinal muscular atrophy (SMA) affects individuals with a broad age range and spectrum of disease severity. Risdiplam (EVRYSDI®) is a centrally and peripherally distributed, oral survival of motor neuron 2 (*SMN2*) pre-mRNA splicing modifier that has been approved in over 90 countries worldwide. SUNFISH (NCT02908685) is a multicenter, two-part, randomized, placebo-controlled, double-blind study in patients with types 2 and 3 SMA (inclusion criteria: aged 2–25 years at enrollment). Part 1 (n = 51) assessed the safety, tolerability and pharmacokinetics/pharmacodynamics of different risdiplam dose levels in patients with types 2 and 3 SMA (ambulant and non-ambulant). Part 2 (n = 180) assessed the efficacy and safety of the Part 1-selected dose of risdiplam versus placebo in type 2 and non-ambulant type 3 SMA. In Part 2, participants were treated with risdiplam or placebo for 12 months; participants then received risdiplam in a blinded manner until Month 24. At Month 24, patients were offered the opportunity to enter the open-label extension phase.

Objectives: To determine the efficacy and safety of risdiplam in patients with Types 2 and 3 SMA after 4 years (48 months) of treatment.

Results: The primary endpoint (Part 2) of change from baseline in the 32-item Motor Function Measure (MFM32) total score in patients treated with risdiplam (n=120) versus placebo (n = 60) was met at Month 12. These increases in motor function were sustained in the second and third year after risdiplam treatment, as measured by changes in the MFM32, Hammersmith Functional Motor Scale — Expanded, and Revised Upper Limb Module. At Month 36 (data-cut: 6 September 2021), there were no treatment-related safety findings leading to withdrawal from either SUNFISH Part 1 or 2. Here we present 4-year efficacy and safety data from SUNFISH.

Conclusion: SUNFISH is ongoing and will provide further long-term efficacy and safety data of risdiplam in a broad population of children, teenagers and adults with SMA.

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508 – Multiple cerebral ring enhancing lesions: an atypical finding of high-grade glioma

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Introduction: High-grade gliomas are primary neoplasms of the central nervous system and can have multiple clinical and neuroimaging presentations. An unusual radiologic image can lead to diagnostic difficulty, and cancer treatment delay. In rare cases, primary brain tumor can mimic multiple abscesses in magnetic resoanance imaging (MRI). The aim of this paper is to describe a diagnostic challenge in MRI imaging of brain tumors. Case Report based on a retrospective analysis of the medical records of the patient.

Case report: This case report describes a previously healthy 48-year-old male evaluated for a first episode of tonic-clonic seizure. Brain MRI showed multiple focal cerebral ring enhancing lesions, with centrally restricted diffusion and susceptibility-weighted imaging (SWI) demonstrating incomplete hypointense rims in the lesion margin. Infectious and neoplastic diseases were suspected, including brain abscess, primary brain tumors or metastases. The initial systemic investigation for infection and primary tumor was negative and brain biopsy showed nonspecific inflammation. Empirical antibiotic therapy was started, with no clinical response. To better elucidate the diagnosis, surgical resection of the lesions was undertaken. Histopathologic exam showed a high-grade glial tumor, with 20% Ki-67, ATRX and IDH1 mutation, which was compatible with Grade IV astrocytoma.
Conclusion: Multiple cerebral ring enhancing lesions can be an atypical presentation of high grade glioma and a diagnostic challenge. In this cases SWI can help differentiate from brain abscess, which presents complete and smooth hypointense rims.

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509 – Acute myeloradiculitis due to herpes simplex virus in an immunocompetent woman: early response to acyclovir and complete recovery after treatment

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Case presentation: A 42-year-old female presented a febrile syndrome associated with acute urinary retention and constipation one week after noticing perineal painful and hyperemic vesicular lesions. Outpatient work-up yielded positive herpes simplex virus serologies (immunoglobulin M and immunoglobulin G) and oral acyclovir was started. She arrived at our service with no spontaneous diuresis; in addition, there were perineal and lower gluteal hypoesthesia, lower limb hyperreflexia and Babinski sign in the right foot. Thoracic and lumbar magnetic resonance imaging (MRI) with contrast was normal, as well as brain and cervical images posteriorly performed. Cerebrospinal fluid analysis showed lymphomononuclear pleocytosis and hypoglycorrhachia. Intravenous acyclovir (10 mg/kg every eight hours) was started and continued for 14 days; she presented spontaneous diuresis 12 hours after the first dose and progressively decreased intermittent catheter output. Methylprednisolone pulse therapy for five days was associated at D4 of acyclovir. She was asymptomatic at discharge.
Discussion: Herpes simplex virus type 2 (HSV-2) myeloradiculitis is rare and often underdiagnosed. An attentive neurological exam is essential for suspicion. Edema and T2 weighted imaging hyperintensity are possible findings at MRI; however, no changes are observed in many cases. Currently, there is no robust evidence for treatment, usually based on intravenous acyclovir and/or corticosteroids in high doses. Our case had an early and remarkable response to acyclovir. Prognosis varies in the literature; recovery has been reported after acyclovir was started three months from onset.
Conclusion: It is important to be aware of HSV-2 myeloradiculitis as a cause of acute urinary retention, even in immunocompetent patients. This case adds to the discussion about the role of acyclovir, given the lack of evidence for best treatment and its relation to prognosis

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510 – Epidemiological profile of migraine admissions at the Brazilian Unified Health System (SUS) between 2012 and 2022: a decade of pain.

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Introduction: Migraine is a type of headache characterized by a pulsating pain or sensation, and the pathophysiology is still not fully understood. There are two types of migraines, with aura and without aura, the latter being the most frequent. World Health Organization data suggest that, globally, the estimated prevalence of any type of headache among adults can reach up to 50%, being the sixth highest worldwide cause of years lost due to disability. In Brazil, about 15.8% of the population suffers from migraine. Studies are needed to draw more attention to this disease that causes much morbidity and disabling.
Objectives: The aim of this study is to analyze the epidemiology of migraine admissions at the Brazilian Unified Health System (SUS) from 2012 to 2022.
Methods: This is a descriptive epidemiological study, in which migraine admissions data was collected using the SUS Hospital Information System database (DATASUS/TABNET), from 2012 to 2022, by region, using the term "*Enxaqueca e outras síndromes de algias cefálicos*". Literature review was conducted using PubMed and SciELO databases. Statistical analysis was performed with Kolmogorov-Smirnov and analysis of variance/Tukey tests, using the PRISM software.
Results: From 2012 and 2022, Brazil had 96,857 migraine admissions. The most prevalent region is the southeast of Brazil with 33,487 admissions ($P < 0.05$ when compared to north and midwest). The least prevalent region is the midwest, totalling 5,026 admissions. The year with most recorded migraines were 2019, with a total of 11,996 admissions, and 2012 the least prevalent, with 5,908 admissions. Of all admissions, 65% occurred in women, being the most prevalent age group from 30 to 39 years old.
Conclusion: We observed an increase in hospital admissions by migraines in the last decade, particularly between women and in the southeast region of Brazil.

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511 – Distinct patterns of cerebellar damage in sporadic and ATXN2-related amyotrophic lateral sclerosis

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Introduction: Neurodegeneration in amyotrophic lateral sclerosis (ALS) is not confined to the corticospinal tracts and motor neurons. Some studies have lately suggested that the cerebellum is also involved in the disease. Despite that, it is not yet clear whether cerebellar damage is similar in sporadic (sALS) and specific subtypes of familial ALS. This is particularly relevant for

ATXN2-related ALS, because this gene is also known to cause cerebellar ataxia (when fully expanded).

Objectives: This study aimed to characterize cerebellar damage *in vivo* in patients with sporadic ALS and *ATXN2*-ALS.

Methods: Ten patients with *ATXN2*-ALS, 10 with sALS and 10 controls underwent high resolution T1 magnetic resonance imaging (MRI). Afterwards, we computed the volumes of all cerebellar lobules using the fully automated ACAPULCO tool. Between-group comparisons were then performed for each individual lobule employing linear regression with multiple comparison adjustment. $P < 0.05$ were considered significant. Disease severity at the time of MRI acquisition was assessed with the ALSFRS scale.

Results: Mean age of sALS, *ATXN2*-ALS and control groups were 61.5 (±10.7), 62.5 (±14.4) and 61.5 (±13. 9), respectively. Both ALS cohorts had similar ALSFRS scores ($P = 0.46$). Sporadic ALS patients had volumetric reduction at the left VIIIB ($P = 0.02$), right VIIIB ($P = 0.002$), left X ($P = 0.027$) and right X ($P = 0.007$) lobules relative to controls; and volumetric reduction at the medullary body ($P = 0.025$) relative to *ATXN2*-ALS. No volumetric change was found between *ATXN2*-ALS and controls.

Conclusion: Segmental cerebellar atrophy was found in sALS, but not in *ATXN2*-ALS. This suggests that the mechanisms underlying both disorders may be different.

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512 – Metabolic syndrome in patients with Duchenne muscular dystrophy

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Introduction: Duchenne muscular dystrophy (DMD) is the most frequent muscle dystrophy in children. It is an X-linked condition caused by loss of function variants in the *DMD* gene. During disease course, affected patients lose lean body mass, become severely motor disabled and are often treated with long term steroids. All these factors pose them at increased risk for developing metabolic syndrome (MS), even as children. Surprisingly, there are few studies addressing this aspect in DMD cohorts, particularly in Brazil.

Objectives: To estimate the frequency and risk factors for DMD-related MS in a Brazilian cohort.

Methods: This is a cross-sectional study involving 33 patients with DMD regularly followed at Universidade Estadual de Campinas. All subjects underwent detailed clinical (blood pressure, body mass index, motor function) and laboratorial (serum glucose, glycated hemoglobin (HbA1c), cholesterol and triglyceride levels) evaluation. We also recorded demographic, genetic and steroid use (cumulative dose) data from each subject. MS was defined according to the international federation of diabetes criteria for children/teenagers.

Results: Mean age of patients was 10.54 years and there were 60% of wheelchair bound. Most of them had either deletions (n = 15) or duplications (n = 6). None of the patients fulfilled the entire set of criteria for MS. However, particular clinical features of the syndrome were frequently found: Obesity in 60%, Central obesity in 70.36% and increased blood pressure in 21.7%. Glucose and Hb1Ac were normal in all individuals, but increased cholesterol and/or triglyceride levels were noticed in 33.33% of the patients.

Conclusion: Metabolic abnormalities are frequent and should be actively investigated in DMD. The existing MS diagnostic criteria may not be adequate for these patients and contribute to underdiagnosis.

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513 – Subacute combined degeneration of the spinal cord in copper deficiency: case report

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Introduction: Copper deficiency is a rare disease that can present with a wide variety of symptoms, with myelopathy as the most common neurological presentation. The clinical features are characterized by sensory ataxia, spasticity and sensory-motor neuropathy. The risk factor that has the most impact is previous gastrointestinal surgery, but a large part of cases remain without a determined cause. The aim to present a case of combined subacute medullary degeneration secondary to copper deficiency with good response to treatment by reviewing the patient's medical record.

Case report: Male, 47-year-old, with no pathological history or previous surgeries. His symptoms started 12 days after coronavirus disease 2019 infection, as paresthesias in the lower limbs, urinary retention and progressive weakness. Imaging exams showed longitudinally extensive cervical and thoracic medullary injury compromising posterior and posterolateral regions, without contrast uptake. Complementary investigation demonstrated low serum copper. After oral and parenteral copper supplementation, the patient evolved with significant improvement. The etiological investigation of copper deficiency remains undetermined until the moment.

Discussion: Myelopathy secondary to copper deficiency is characterized by being clinically and radiologically indistinguishable from subacute combined spinal cord degeneration due to vitamin B12 deficiency. Diagnosis and treatment are often delayed. Neurological manifestations are only partially reversible with copper supplementation, demonstrating poor prognosis. In the case described, the important recovery evidenced after the treatment stands out. It is suggested that combined oral and parenteral therapy with a multidisciplinary approach, as motor physiotherapy, are related to better neurological recovery.

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514 – Case report: ultrasound examination in central retinal artery occlusion in a patient with infective endocarditis

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Introduction: Central retinal artery occlusion (CRAO) is an important cause of sudden monocular blindness. It can be divided into arteritic and non-arteritic forms, with non-arteritic being the most common, typically caused by emboli.

Ultrasound is useful to detect embolic CRAO, with retrobulbar spot sign commonly found in atherosclerotic etiology. Case report of a patient with CRAO secondary to infective endocarditis, obtained by medical records review.

Case report: A 65-year-old male patient presented to the emergency department with sudden monocular vision loss one month from admission with weight loss of 9 kg, fatigue, dyspnea and daily fever of 39 °C for three months. He had 20/20 vision acuity on the left eye and complete vision loss on the right eye. An afferent pupillary defect was present on the right eye. A holosystolic cardiac murmur was found, most prominent on the aortic site. An orbit ultrasound was performed, which showed a retrobulbar spot sign on the right eye, indicating a CRAO, with a high likelihood of embolic cause. A transesophageal echocardiogram showed double aortic lesion with predominant stenosis, with a 0.6 cm mobile filament on the anterior leaflet suggestive of vegetation. The diagnosis of infectious endocarditis was thus established by the modified Duke criteria and antibiotic treatment was started.

Conclusion: This case report highlights the utility of using ultrasound examination in investigating sudden visual loss, as it helps in determining an embolic cause when the retrobulbar spot sign is found, and excluding arteritic forms of CRAO. This report also shows endocarditis as an uncommon etiology for cardiac emboli to the central retinal artery for CRAO with retrobulbar spot sign in ultrasound evaluation.

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515 – Acute pregabalin cerebellar ataxia

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Female patient, 65-year-old, complaining of imbalance and difficulty walking for 15 days, associated with dizziness and diplopia, with worsening symptoms in the last four days. In recent use of pregabalin 75 mg daily to treat chronic pain, with dose increase in the last four days. On examination, he presented ataxic gait, drunken speech, pendular reflex, dysmetria and dysdiadochokinesia. We opted for the suspension of Pregabalin and complementary investigation for cerebellar ataxia with imaging examination, examination of the cerebrospinal fluid (CSF) and laboratory tests. Magnetic resonance imaging showed mild microangiopathy with, no other findings. Normal CSF examination, including search for oligoclonal bands. The search for rheumatological antibodies, tumor markers and serologies was negative. After the suspension of Pregabalin, the patient showed progressive improvement, with total reversal of symptoms. Acute cerebellar ataxia is defined as a syndrome that occurs in less than 72 hours in previously healthy individuals. It is clinically characterized by loss of balance and coordination. A heterogeneous group of conditions can cause the syndrome, including toxic causes. Several drugs cause ataxia, depending on the dose and duration of exposure. Although uncommon, anticonvulsants cause movement disorders, including pregabalin. Most adverse effects of this class of drugs occur within the first two weeks, ranging from mild to moderate. Movement disorders are rare, but when found they usually appear as ataxia or tremor. Acute cerebellar ataxia comprises a spectrum of neurological disorders in which ataxia is the main symptom. The heterogeneity of the etiologies of cerebellar ataxia is wide and requires hospitalization and extensive laboratory investigation. Among the causes, even if rare, is the use of pregabalin, widely used in clinical practice to treat neuropathic pain.

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516 – Acute multiple sclerosis after immunization with Pfizer-Biontech COVID-19 vaccine

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Introduction: Vaccination against SARS-CoV-2 (Severe Acute Respiratory Syndrome Coronavirus 2) is crucial to controlling the pandemic. Although there are not yet enough data on the coronavirus disease 2019 (COVID-19) and the risk of patients develop Multiple Sxlerosis, it is likely that older patients with higher disability and associated complications have a higher risk of complications from COVID-19 infections. Multiple sclerosis is defined as an inflammatory demyelinating disease characterized by the presence of episodes of neurological dysfunction in at least two areas of the central nervous system (brain, spinal cord and optic nerves) separated in time and space.

Case report: A 37-year-old male patient, took the first dose of the pfizer vaccine in July 2021, after three months in October 2021, he started symptoms of bilateral paresthesia in the lower limbs evolving with weakness and faecal incontinence, 21 days later he sought medical service and underwent magnetic resonance imaging of the skull with the finding of eight small oval foci (> 3 mm) of hypersignal in T2 and FLAIR (Fluid-Attenuated Inversion Recovery) in the white matter of the cerebral hemispheres, including periventricular regions. without signs of acute inflammation. cerebrospinal fluid puncture showed the presence of oligoclonal bands associated with an immunoglobulin G index of 1.006 mg/dL. Vaccinated patients can present a myriad of symptoms and autoimmune disorders have been correlated to it.

Conclusion: There is no cure for multiple sclerosis, the focus is the fast recovery from acute attacks reducing new relapses and slow progression of disease as well as brain atrophy and disability accumulation. To accomplish this purpose there are several disease modifying therapies for relapsing-remitting multiple sclerosis. Interferon beta medications are the most prescribed medications, with the mechanism of reducing inflammation and increasing nerve growth. Patients taking interferons need blood tests to monitor liver enzymes and experience side effects.

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517 – Miller Fisher syndrome secondary to Epstein-Baar virus mononucleosis

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Miller Fisher syndrome is a rare variant of Guillain-Barre syndrome and usually presents with at least two of the following features: ataxia, areflexia, and ophthalmoplegia. Male patient, 16 years old, without comorbidities. Odynophagia began, followed by diarrhea and asthenia. Prescribed antibiotic therapy for tonsillitis. After one week hospital admission with neurological examination with mild dysarthria, global grade 5 strength, bilateral naso-index dysmetria, walking gait, positive romberg and areflexia. Tomography and magnetic resonance imaging of normal. Liquor with protein cytological dissociation. Electro-

neuromyography with absence of H reflex bilaterally, suggestive of demyelinating neuropathy. Abdominal ultrasound with homogeneous splenomegaly. Cervical ultrasonography with prominent reactive looking lymphnodes. Epstein Barr virus Reactive IgM and non-reactive IGG. A diagnostic hypothesis of Miller Fischer syndrome secondary to mononucleosis was raised, despite a negative anti-GQ1B antibody. Immunoglobulin at a dose of 0.4 kg/kg/day was requested. Patient followed immunoglobulin infusion in intensive care unit bed. He was discharged from the hospital, maintaining only mild gait ataxia. Miller Fisher syndrome within the spectrum of Guillain-Barré syndrome occurs due to an aberrant acute autoimmune response to a previous infection, such as the Epstein-Barr virus. Approximately two-thirds of cases are preceded by symptoms of an upper respiratory tract infection or diarrhea. Miller Fisher syndrome is mainly associated with dysfunction of the cranial nerves, but as exemplified in our case, it can occur in its absence. Several studies have suggested that antibodies against gangliosides, anti-GQ1b, are a specific feature, but the absence of antibodies does not exclude the disease. Cerebrospinal fluid shows proteincytological dissociation. Electroneuromyography may show reduced or absent sensory responses. Imaging may be normal or show thickening and enhancement of the spinal nerve roots. Treatment is aimed at supportive care and the therapy of choice is IV immunoglobulin or plasmaphere. Assertive diagnosis associated with early treatment contributes to a favorable clinical response.

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518 – Genetic profiling of RYR1-related myopathy in a tertiary neuromuscular center

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Introduction: The *RYR1* gene encodes the skeletal muscle ryanodine channel, an ion channel responsible for regulating calcium release from the sarcoplasmic reticulum. Variants in *RYR1* are cause of a wide phenotype of myopathies, with autosomal dominant and recessive inheritance.

Objectives: In this work we present the clinical and genetic data of 43 patients with myopathies presumably related to the *RYR1* gene, carrying 47 different variants

Material and methods: Clinical and genetic data are presented. *RYR1* gene were analyzed by next-generation sequencing.

Results: Twenty patients have an autosomal dominant (AD) inheritance pattern, with 16 different variants observed. All AD variants were missense. The c.14582G>A, p.(Arg4861His) variant was the most recurrent in the AD group (three patients). Seventy-five percent of these variants were found in the channel and activation core region, especially in the transmembrane domain. In the presumed autosomal recessive inheritance (AR) pattern group, 23 patients were included, with 31 different variants described, the majority being missense variants (23). Eight were null variants (four splice site, two frameshift, one nonsense and one copy number variation). The c.7027G>A, p.(Gly2343Ser) variant was the most recurrent in the AR group (four patients).

Conclusion: Autosomal dominant forms of *RYR1*-related myopathy are related to missense variants usually located in the channel and activation core region. Missense variants are also the main type observed in autosomal recessive forms, but in this AR group, null variants are also found, with no specific cluster along the gene.

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519 – Analgesic potentiation in post-traumatic occipital neuralgia, a case report

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Context: Occipital neuralgia is characterized as uni or bilateral pain in the topography of the occipital nerve. The condition may be harrowing or acute and worsens with digital pressure on the back of the neck. The pain appears in the distribution of the greater (GON), minor (LON), and third (TON) occipital nerves. Besides that, it may radiate to the retro-orbital region and angle of the mandible, and it can be associated with paresthesias. A less common cause is direct head trauma, and it is believed that the mechanism for its development is the injury or entrapment of the nerve fibers due to post-traumatic fibrosis. The conservative treatment consists of antiepileptics, tricyclic antidepressants, or antipsychotics. In addition, it includes anesthetic blockade of the GON, LON, and TON guided by ultrasonography of the skull, which is an essential option for treatment and identifies the presence of entrapment of the quoted structures. We present the case of a 37-year-old man, a victim of physical aggression with traumatic brain injury, without previous comorbidities or use of daily medications, who developed occipital neuralgia after the event. Cranial computed tomography showed occipital depression in the topography of the GON. Due to the absence of neurological alterations, the medical team chose conservative treatment with phenytoin. As the pain persisted, the professionals performed a hydro section and suboccipital block with anesthesia, resulting in improvements.

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520 – Pseudochoreoathetosis in a patient with idiopathic sensitive ganglionopathy: a case report

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Sensitive ganglionopathy has been associated with systemic diseases. In general, this rare neurological syndrome predates the onset of systemic symptoms. It affects selectively large nerve fibers and, accordingly, is manifested with loss of protopathic touch, vibratory perception, and proprioception. We describe a case of pseudochoreoathetosis in an elderly female patient with idiopathic sensitive ganglionopathy, manisfested with unsteadiness and weight loss in the last six months. The patient had systemic arterial hypertension, and a history of tobacco and alcohol abuses. The neurological examination showed a mild cognitive impairment, preserved proximal appendicular strength and reduced distal strength, generalized areflexia, bilateral non-evoked plantar reflex, dysmetria of the four limbs with significant impairment of position perception (worsening with eyes closed), tactile, thermal and painful hypoesthesia on the upper limbs, in the trunk and cervical region, bilateral anesthesia and crural apalesthesia, and marked loss of proprioception. Involuntary movements were observed, predominantly distal to the limbs,

with sustained spontaneous bilateral extension of halluces, alongside slow, constant, low amplitude, serpentine, and involuntary movements of the toes, fingers and wrists, better defined as pseudochoreoathetosis due to severe loss of proprioception. A thourough diagnostic evaluation, performed by cranial and spinal magnetic resonance imagem, cerebrospinal fluid analysis, autoantibodies dosing, cyanocobalamin and folic acid dosing, infections diseases testing and neoplasic screening, yielded no positive results. The electroneuromyography of the four limbs showed marked axonal impairment, exclusively restricted to sensitive neurons. Accordingly, a diagnosis of sensory ataxia due to idiopathic ganglionopathy was established.

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522 – Case report: cryptococcosis and unexpected outcome

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Introduction: *Cryptococcus* meningoencephalitis is the most frequently encountered manifestation of cryptococcosis, a fungal infection often found in immunocompromised individuals. The purpose of this case report is to present an unexpected death of an immunocompetent patient with cryptococcosis, evidencing a fulminant course.
Case report: A 65-year-old male patient, previously immunocompetent, was admitted to our hospital mechanically ventilated after deep sedation due to agitation and altered mental status. Three days prior to admission, he had a throbbing headache with no response to analgesics. After normal laboratory tests, evaluation of the cerebrospinal fluid was performed with positive culture of Cryptococcus neoformans and a positive India ink. Moreover, cranial computed tomography had nodular calcification in the right frontal lobe and in the left thalamus. Therefore, treatment was started with amphotericin B plus fluconazole, furthermore, therapeutic lumbar drainage was repeated daily. After six days, the patient was extubated and confirmed that his headache had a one-year course, on that day his intracranial pressure was 13 cm. On the next day he was discharged from intensive care unit, and later on that day was found dead.
Conclusion: Cryptococcosis is a potentially lethal infection, often with a more severe clinical course in immunocompetent patients. Therefore, prompt diagnosis and treatment are needed to avoid a fulminant course.

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523 – Fibrocartilaginous embolism causing spinal cord infartion: case report

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A 41-year-old male presented with strength, sensitivity and sphincters deficits of acute evolution after a mild car accident, characterized by paraplegia and areflexia of the lower limbs, hypoesthesia with sensory level at T6 and preserved artresthesia. A major diagnostic evaluation was carried out with neuroxias imaging tests, extensive laboratory investigation and exclusion of occult neoplasia. Cerebrospinal fluid was normal. Magnetic resonance imaging of the thoracic spine revealed signal alteration between T4-T7 without enhancement after contrast injection. A short course of intravenous corticosteroid therapy was attempted without any clinical improvement. Due to the acute presentation, history of minimal trauma, absence of findings in laboratory investigations, no other risk factors, clinical and images compatible with anterior spinal cord infarction, Fibrocartilaginous Embolism (FCE) was defined as a probable etiology. FCE is a rare cause of spinal cord infarction and consists in the migration of fibrocartilaginous nucleus pulposus material through the nearby vasculature into one of the spinal cord vessels, usually at the thoracic level and affects the territory of the anterior spinal artery. It can be associated with minor trauma and other causes. The probable diagnosis of FCE is based on data from the clinical history, exclusion of other alternative diagnoses and neuroradiological findings, such as changes in the spinal cord signal in the vascular territory and degenerative changes in the vertebral disc. Definitive diagnosis is possible only with biopsy. Treatment is supportive with physical rehabilitation. Although rare, FCE should be considered as a differential diagnosis in cases of unexplained medullary infarction in young patients without cardiovascular risk factors. The case in question is important and exemplifies that a high diagnostic suspicion is necessary to elucidate these cases.

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525 – Staphylococcus aureus brain abscess due to chronic sinusopathy and mastoiditis: a case report

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Introduction: A brain abscess is an intraparenchymal collection of pus and a life-threatening infection. Despite the advancements in imaging and diagnostic techniques, observational studies suggest mortality rate still remains high. The clinical presentation usually includes fever, headache and focal neurologic deficits. About 90% result from pericranial infection such as sinusitis, mastoiditis and otitis media, and many are hematogenous borne such as bacterial endocarditis. *Bacteroides*, *Peptostreptococcus* and *Streptococcus* are the most pathogens identified in brain abscesses due to contiguous spread from pericranial infection, while *Peptostreptococcus* and *Streptococcus* are the ones in patients with cardiac origin. *Staphylococcus* is common in posttraumatic and postoperative cases. The treatment must be individualized according to the pathogen and the patient. Initial therapy should include broad spectrum antibiotics. Most pyogenic abscess also requires a surgical intervention. The aim is to describe a brain abscess case as a result of an untreated chronic sinusopathy and mastoiditis.
Case report: A 31-year-old man presented to the Emergency Department of our hospital with fever and progressive cognitive decline, which had been worsened the last five days. Associated with headache for over a year, that didn't get relieved with analgesics, chronic cold and right otorrhea. His past medical history included splenectomy as a kid with immunizations up to date and a chronic sinusopathy and mastoiditis without any treatment. His social history included chronic alcohol consumption and smoking. Admitted with normal ranges of vital signs and physical examination, except for his neuro-

logical examination which showed notable disorientation. Complete blood count and metabolic panel results were normal. Serologic testing for human immunodeficiency virus and hepatitis were negative. Head computed tomography scan revealed a large mass (64 x 37 mm) involving the left frontal lobe with edema and 9 mm midline shift. After admission, empirical parenteral antibiotics were started with ceftriaxone 2 g, oxacillin 2 g, metronidazole 500 mg and the patient underwent immediate neurosurgical drainage of the large abscess. The cerebrospinal fluid cultures were positive for *Staphycoccus aureus*. The patient had previous brain magnetic resonance imaging that indicated a chronic sinusopathy of all four paranasal sinuses and a right mastoiditis. Since the normality of blood exams and no better hypothesis to explain the abscess, the chronic sinusopathy had been considered the main cause for the patient's outcomes. The patient underwent a sinusectomy. After five days, the patient was discharged from the Intensive Care Unit due to better clinical evolution. The patient signed the informed and consent form to participate in this research.

Conclusion: As the brain abscess continues with a high mortality rate, it could be important to focus attention on recognizing the risk factors and early treat them, including sinusopathy and mastoiditis, in order to avoid severe complications such as brain abscess and eventually prevent the occurrence of critical outcomes.

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528 – Wernicke encephalopathy in a patient with uncontrollable vomiting. A case report

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Wernicke encephalopathy is a neurological disorder caused by lack of thiamine. The main symptoms include ophthalmoplegia, ataxia, and confusion. We described a case of an elderly woman who presented with sudden-onset vertigo, uncontrollable vomiting and blurred vision on the left eye. On the following days, she evolved with gait imbalance with a tendency to fall to the left side, fluctuating drowsiness and bilateral ophthalmoplegia. She had a previous history of systemic arterial hypertension, depressive disorder, gastritis and gastric cancer 15 years ago, treated with chemotherapy and radiotherapy. No history of tobacco or alcohol abuse. Neurological examination showed disorientation in time and space, drowsiness, gait ataxia and positive Romberg test. Furthermore, she had Babinski sign on the right toe. In addition, she had bilateral ophthalmoplegia, visual acuity in the right eye 20/40 and left eye 20/50. Serum tests were normal, including thyroid function, ions/metabolics, and viral serologies. Cerebrospinal fluid analysis showed six leukocytes (100% mononuclear) and 51 mg/dL of protein. A brain nuclear resonance imaging showed hypersignal on T2 and FLAIR (Fluid-Attenuated Inversion Recovery) sequences located in the medial portions of the thalamus, symmetrically and bilaterally, in the periaqueductal gray matter, around the fourth ventricle and in the cortico-subcortical transition of the post-central gyri, which in the context suggested Wernicke's encephalopathy. The treatment was performed with thiamine (300 mg/day), with significant neurological deficit recovery.

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529 – Quality indicators of a stroke unit in Curitiba (Brazil)

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Introduction: Stroke represents the second leading cause of death and disability in Latin America. Thus, there is a constant need to understand the profile of patients admitted for stroke, as well as to analyze the quality indicators of treatment in stroke centers.

Objectives: The aim of this study was to analyze the quality indicators of a stroke center and to determine the epidemiological profile of inpatients.

Methods: This is an observational cross-sectional study that included individuals admitted with stroke or transient ischaemic attack (TIA) in the stroke unit of Complexo Hospital de Clínicas da Universidade Federal do Paraná from November 2020 to October 2022. The variables and quality indicators were collected through the application of questionnaires in two moments, on admission and hospital discharge.

Results: Of the 407 patients included, 49.1% were men, with a mean age of 65.4 years (standard deviation ±15.3). Ischemic stroke was the most common type of stroke (84.5%). The majority of patients (62.6%) did not receive specific intervention, while 41.9% underwent intravenous thrombolysis, with a median door-to-needle time of 30 minutes. In 69.1% of the individuals, thrombolysis started in less than 60 minutes. All patients underwent neuroimaging, 90.9% carotid imaging and 92.6% had their cardiac rhythm rated by EKG or 24h-Holter. Dysphagia screening was applied in 62.4% of the cases and smoking cessation advice was provided in 50.5%. Of the patients with ischemic stroke and TIA, only 5 (1.3%) did not receive anticoagulant or antiplatelet agent for secondary prophylaxis and in 14 (3.75%) this data was unknown.

Conclusion: Intravenous thrombolysis is not performed in most hospitalized patients, however, when performed, it presents an adequate door-to-needle time. Most patients received secondary prophylaxis. Management of patients in a stroke unit allows for a thorough etiological investigation and optimization of secondary prevention and rehabilitation.

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530 – Non-motor symptoms and signs of Myotonic Dystrophy type 1

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Introduction: Myotonic Dystrophy Type 1 (DM1) is a genetic disease that presents neuromuscular manifestations and multisystemic clinical repercussions, such as cardiac and respiratory disorders, sleep disorders and impaired swallowing, among others. It is the most common muscular dystrophy in adults.

Objectives: To determine the epidemiological profile of patients with DM1 treated at the Neuromuscular Diseases Outpatient Clinic of the Complexo Hospital de Clínicas da Universidade Federal do Paraná (CHC-UFPR).

Methods: A total of 27 individuals diagnosed with DM1, assisted at the Neuromuscular Diseases Outpatient Clinic of the CHC-UFPR, were evaluated between May 2021 and March 2022. For this purpose, their medical records with the clinical data were analyzed.
Results: The sample consisted of 78% male subjects with mean age at onset of symptoms of 27.6 ± 10.8. The most frequent muscular manifestations were myotonia (100%), weakness of the distal muscles of the upper (96.3%) and lower (96.3%) limbs, myotonic facies (92.6%). The most common non-motor manifestations were excessive daytime sleepiness (74.1%), frontal baldness (66.7%), pharyngeal globus (62.9%), choking or coughing during and/or at the end of swallowing (62.9%), cataracts (59.2%), dysphagia (55.6%), chest pain (55.6%), cognitive impairment (44.4%), dyspnea (44.4%). Of the patients, 22.2% had a previous history of pneumonia.
Conclusion: The DM1 patients in this study presented an epidemiological profile consistent with that described in the literature. Non-motor manifestations are common and should be investigated, since complications such as bronchopneumonia are important causes of mortality in these patients and may negatively impact the quality of life. Therefore, DM1 patients require multidisciplinary monitoring and evaluation.

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531 – Atypical course of schistosomal myelitis: a diagnostic challenge

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Acute myelitis is a severe neurological complication of schistosomiasis which clinical features present as lower limb weakness, sensory disturbance and intestinal or bladder dysfunction. A 60-year-old male farmer presented with low back pain and progressive paresis of right lower limb that worsened to plegia over ten months. MRI demonstrated hypersignal lesions in the T2/FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) sequence in the ninth dorsal segment level. A Kato-Katz examination was negative for Schistosoma mansoni and a rectal biopsy confirmed an active infection. The patient received methylprednisolone (MTP) and a single oral dose of praziquantel (50 mg/kg), with partially improvement, being able to walk with assistance. Six months later, after coronavirus disease 2019 vaccine (Pfizer), he reported a worsening of neurological symptoms. New magnetic resonance imaging revealed no change compared to previous one. Aquaporin-4 antibodies were negative in serum, cerebrospinal fluid analysis showed 15 leukocytes (93% mononuclear), protein and glucose within the normal range. Autoimmune profile, serology for syphilis, hepatitis B, Hepatitis C, and human immunodeficiency virus were negative. The patient received a second pulse of MTP and repeat course of praziquantel, after which he improved his left lower limb weakness. Oral corticosteroid therapy was weaned. Nine months later, he experienced new exacerbation of crural paraparesis. Repeat cerebrospinal fluid analysis showed a normal pattern and new spine magnetic resonance imaging had no changes, but stools testing was positive for hookworm eggs, taenia sp and entamoeba coli in three different occasions. Therefore, he also received treatment with ivermectin and albendazole. The delay in diagnosis and treatment likely contributes to poor outcomes. As praziquantel is effective only on the mature adult worms but not on the larval form, a second dose should be considered in recurrent myelitis related to spinal neuroschistosomiasis.

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537 – Stiff Person syndrome and lumbosacral plexopathy post COVID-19

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Introduction: Coronavirus diasease 2019 (COVID-19) has been associated with inflammatory, ischemic and autoimmune neurological complications. We report a case of Stiff Person Syndrome (SPS) as an autoimmune neurological complication in COVID-19, an association still rare in the literature.
Case report: A 45-year-old woman, presented with back pain of strong intensity with irradiation to the left lower limb, associated with painful muscle contracture and weakness in the same limb with progressive worsening in two days. Neurological examination revealed strength grade II, hyporeflexia, tactile and vibratory hypoesthesia, and paresthesia of the left lower limb, with increased tonus and impossibility to bend the left knee. She presented with positive severe acute respiratory syndrome coronavirus 2 polymerase chain reaction 12 days before the onset of symptoms. cCerebrospinal fluid and lumbosacral magnetic resonance imaging without significant changes, left thigh MRI with nonspecific edema and electroneuromyography compatible with left lumbosacral plexopathy. Stiff Person syndrome and autoimmune lumbosacral plexopathy were hypothesized. The treatment was started with methylprednisolone 1 g/day for five days and immunoglobulin 0.4 g/kg/day for five days with partial improvement. Anti-GAD AC performed, non-reactive. Screening for neoplasms was negative. In evolution, the patient progressed with involvement of the right lower limb, trunk, and upper limbs, respectively, with a fluctuating course and partial and temporary response to human immunoglobulin infusion and methylprednisolone pulse therapy.
Conclusion: We report the associated time between SPS and COVID-19. SPS is a rare autoimmune disease related to anti-GAD AC and other antibodies. The temporal link with infectious agents that may trigger the autoimmune attack is uncommon in the literature, as the diagnosis is usually made years after the onset of symptoms. In this case, the diagnosis was made soon after infection, which allowed us to report another autoimmune neurological complication in COVID-19.

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538 – Cytotoxic lesion of the corpus callosum associated with Tumor Lysis Syndrome

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Case presentation: MCP, 39 years, female, with primary rectal cancer with lung metastases being treated with weekly cycles of chemotherapy, without other comorbidities. At the end of 46 hours of another cycle of this treat-

ment, patient presented an episode of weakness associated with release of the sphincters, without clonism. Physical examination with stable vital signs, closed eyelids, non-contacting and glasgow coma scale (GCS) 10. Laboratory tests with hypocalcemia, uremia, hyperphosphatemia, increased creatinine, hypomagnesemia, hyperuricemia. On the following day she underwent hemodialysis and evolves with improvement in the level of consciousness (GCS 14), communicative, mild confusion and anterograde amnesia. No alteration at neurological examination. cranioencephalic magnetic resonance imaging with contrast demonstrating signs of diffusion restriction in the splenium region of the corpus callosum. With this, it was possible to determine the diagnosis of cytotoxic lesions of the corpus callosum (CLOCCS) due to tumor lysis syndrome.

Discussion: CLOCCs are secondary lesions associated with varied etiologies, including drug therapy, malignancy, metabolic abnormalities (e.g. electrolyte imbalance) and infections. Cell-cytokine interactions lead to massively increased levels of cytokines and extracellular glutamate, resulting in dysfunction of callosal neurons and microglia, and cytotoxic edema develops as water becomes trapped whitin the cells. CLOCCs appears in magnetic resonance imaging as areas of increased signal intensity on FLAIR and low diffusion on diffusion weighted. This lesion is usually midline, reversible and symmetric.

Conclusion: It is important to know the radiographic features of CLOCCS and theirs known causes, so the source can be found and addressed and avoid misdiagnosis of primary colossal conditions.

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539 – Urinary retention as an initial manifestation of adrenomyeloneuropathy

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Patient RMS, male, 38-year-old, previously dyslipidemic, was admitted to the emergency complaining of severe abdominal pain in the lower abdomen, diaphoresis, and acute retention, after drinking alcohol (ingestion of six glasses of beer). After being submitted to a urinary catheter — with a flow of 2000 ml of urine-, the patient showed instantaneous improvement in pain complaints and diaphoresis, remaining asymptomatic afterwards. He reported a previous feeling of mild difficulty -eventual- to urinate in recent years. He denied constipation or associated erectile dysfunction. Upon neurological examination, the patient was alert, preserved mental state, oriented in time-space-person, mobilizing the four limbs with globally preserved strength (grade V), without gait, static balance or sensory dysfunction. Eumetric and eudiadochokinetic. With no alterations in the assessment of the cranial nerves, but with signs of bilateral pyramidal release — cutaneous-plantar extension, presence of hyperreflexia and clonus. On inspection, eutrophic patient, atypical facies, bald and presented hyperpigmentation of the skin. He denied any type of previous sphincter alteration, as well as any other previous neurological symptoms. When actively questioned about his personal history and life habits, the patient also reported a history of having discovered an adrenal insufficiency in routine exams years ago, and that after extensive investigation and treatment with corticosteroids for months, without much improvement (sic), it was decided to interrupt the follow-up. Regarding alcohol consumption, he said to consume only socially. He denied frequent abuse or use of other drugs (licit or illicit). As for the family history, he said that years ago a brother was diagnosed with Guillain-Barré syndrome, and he has a nephew of the maternal family with a genetic neurological disease. He mentioned that his grandmother underwent genetic investigation, and it was discovered that she had the same disease. He denies any related prior personal investigation. Patient underwent laboratory and imaging investigation. Laboratory tests, including partial urine, serology, inflammatory and rheumatological tests, tumor markers, serum vitamin levels, thyroid function, kidney and liver functions, that were normal. Ultrasound of the kidneys and urinary tract was unchanged. Abdominal tomography showed only a small fecaloma in the rectal ampulla. Magnetic resonance imaging of the brain showed the presence of a slight increase in the periventricular white matter signal in the posterior portions of the brain parenchyma, affecting the bilateral occipital and parietal lobes on brain resonance. No significant degenerative or other changes in the spinal cord were seen on full-spine MRI. The cerebrospinal fluid analysis resulted in parameters within normal limits. Due to the suspicion of hereditary neurological pathology with a pattern of inheritance suggestive of X-linked disease, associated with adrenal insufficiency, a peroxisomal panel was used, which showed increased concentrations of C24:0 and C26:0 concentrations, and in C24/C22 and C26/C22 ratios (very long chain fatty acids), which confirmed the diagnosis of adrenomyeloneuropathy.

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544 – Alzheimer's: what is the difference between or frontotemporal dementia

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Introduction: In the study of neurodegenerative diseases, Alzheimer's disease (AD) has been classically considered with a typical presentation of cognitive symptoms and neuroanatomical changes. However, there are clinical phenotypes of AD whose neurobiological bases are similar to frontotemporal dementia (FTD). In this sense, the heterogeneity of these pictures leads to inaccurate evaluation and diagnosis processes, due to the scant knowledge about their neurocognitive symptoms. The early stages of Alzheimer's-type dementia are classically characterized by memory impairment, whereas behavioral and personality changes appear in the early stages of FTD. However, in clinical practice, the differential diagnosis is difficult.

Objectives: The objective of this systematic review is to establish neuropsychological characteristics and similarities in patients with AD and FTD, identifying key elements for their differential diagnosis, through clinical and imaging exams, with the objective of enhancing the clinical management of the patient.

Methods: A bibliographic survey was carried out in SciELO and PubMed databases and indexers, using the terms "frontotemporal dementia", "alzheimer's" and "neuropsychology", in Portuguese and English. Six updated articles were then selected, considering their adequacy to the objective of the work.

Results: Evidence suggests that there are important differences in cognitive domains such as language (eg, verbal fluency), memory, social cognition, executive functioning, and behavior; These aspects should be considered fundamental in any process of neuropsychological evaluation and diagnosis.

Conclusion: There are linguistic aspects that promise to be powerful biomarkers for the differential diagnosis between AD and FTD, namely semantic and phonemic verbal fluency and semantic-grammatical alterations, which may be fundamental in differentiating the diseases leading to an adequate conduct for each patient.

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545 – Reversible Cerebral Vasoconstriction Syndrome trigged by an unusual trigger: a case report

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Introduction: Reversible Cerebral Vasoconstriction Syndrome (RCVS) is a rare condition of secondary headache to reversible multifocal narrowing of cerebral arteries, coursing with recurrent thunderclap-like pain, associated or not with focal neurological deficits and seizures. The mechanism is unknown, but an abnormality in control of cerebrovascular tonus triggered by a vasoconstrictor trigger is suggested. Diagnostic criteria is based on RCVS2, with high diagnostic accuracy. The aim is to report an unusual trigger cause for SVCR in a woman admitted in ER at Hospital Geral de Cuiabá.
Case: CMP, 43-year-old, admitted at the cardiology ER, with an implantable cardioverter-defibrillator (ICD) due to asymmetric septal hypertrophic cardiomyopathy, referring after 2 sequential shocks of ICD, started a sudden, intense holocranial headache, worst in her life, associated to nausea and vomiting, without focal neurological deficits. Brain computed tomography and angiotomography of cranial vessels was performed, showing a thin layer of cortical subarachnoid hemorrhage (SAH) in right postcentral gyrus region, without aneurysmal dilations or others. Opioid analgesia was performed and Nimodipine was started as prophylaxis for cerebral vasospasm. On next day, presented a new episode of pain with the same characteristics, a new neuroimaging was acquired, maintaining the characteristics described. A skull arterial magnetic resonance angiography was requested for a better evaluation, however, because of ICD, it was unable to perform. RCVS2 was calculated, totaling 10 points, with high accuracy, diagnosing the pathology. Event prophylaxis initiated with Verapamil and Amitriptyline to pain and mood control and, during hospitalization, presented just one new episode of pain after measurements until telemetry is performed.
Conclusion: SVCR is a rare entity and must be readily differentiated from malignant causes, such as aneurysmal SAH or others. Due to the recurrence of pain associated with the potential trigger and exclusion of other causes, diagnosis was established. RVCS2 scale was essential for the outcome in question. It´s being followed up at the headache and arrhythmology ambulatory, with a new imaging exam in three months, described in literature to verifying the reversal of initial findings.

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547 – Structural MRI analysis of basal ganglia volume and white matter tracts in upper limb dystonia

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Background: Dystonia is known as a network disorder. There is evidence of volumetric changes in structures associated with the traditional physiopathology, such as basal nuclei. One approach to studying the neural pathways is through tractography, which can provide insights into the structural connectivity of neural networks that may be disrupted in dystonia.
Objectives: To evaluate brain structural changes of motor networks and basal ganglia volume in dystonia.
Methods: Twenty-six patients with right upper limb dystonia and 29 healthy controls underwent 3T magnetic resonance imaging and evaluated in terms of DTI and T1 data. The XTRACT FSL tool was utilized to examine fractional anisotropy of the bilateral anterior thalamic radiation, superior thalamic radiation, superior longitudinal fasciculus, cortical spinal tract, middle cerebellar peduncle, forceps major and forceps minor. Using T1-weithed data, volunteers were also evaluated in terms of volumetric changes in bilateral Putamen, Caudate, Pallidum and Thalamus extracted using Freesurfer 7.0 volumetric segmentation. For group comparison, we conducted an analysis of covariance controlling for sex and estimated intracranial volume.
Results: For uncorrected p-values, patients with upper limb dystonia show diminished FA volume in the right corticospinal tract relative to controls ($P = 0.025$). Region of interest analysis of subcortical regions volume based on T1-weighted images shows that patients had diminished left caudate volume ($P = 0.031$) and right putamen ($P = 0.041$). However, using FDR multiple comparisons correction, no difference was observed between groups: right corticospinal tract ($P = 0.329$), left caudate volume ($P = 0.16$), right putamen volume ($P = 0.16$).
Conclusion: Our study could not replicate previous findings describing structural changes in dystonia. This could be to methodological differences, as well as the fact that we selected only patients with upper limb dystonia, as opposed to studies that included other types of dystonia. The neuroimaging analyses were conducted with the utmost rigor, utilizing the optimal preprocessing and statistical analysis methods. The nature and characteristics of structural alterations remain unclear and may vary depending on the subtype of dystonia. Therefore, additional structural studies and meta-analyses are warranted to advance our knowledge of this network disorder.

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548 – Multiple cranial couple syndrome secondary to neurosyphilis: case report

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Male, 62-year-old, with Type 2 Diabetes Mellitus, seeks care due to altered balance and worsening palpebral ptosis. The patient had had peripheral facial palsy on the left for 40 days and was treated at another institution with oral prednisone and acyclovir for three days. Ten days ago, he had started with ipsilateral palpebral ptosis, associated with binocular diplopia and divergent strabismus also on the left in the primary gaze position. He also complained that during this period he had worsened visual acuity and frequent falls, which he related to imbalance. Upon inspection, he presented maculopapular erythema on the back, abdomen and upper limbs, with ptosis on the left. On examination, he still had a stomping and dysbasic gait, paralysis of the II, III, and IV nerves bilaterally, as well as of the VII, IX, and X on the left, with reduced visual acuity bilaterally (worse on the left), in addition to a positive Romberg test — with a fall to the left. Furthermore, there was hyporeflexia in the biceps and triceps bilaterally, associated with areflexia in the Achilles and patellas, with symmetrical distal apalesthesia in the four limbs, and cutaneous-

plantar in bilateral flexion. Magnetic resonance imaging of the head and neck showed uptake by the contrast medium seen at the bottom of the internal auditory canal in the distal meatal and labyrinthine segments of the left facial nerve, without signs of intracranial expansive lesion and without detectable cervical abnormalities. Laboratorially, the patient had a positive treponemal test with a Venereal Disease Research Laboratory (VDRL) value of 1:128 and the serum test for Borrelia burgdorferi showed positivity for immunoglobulin G (with negative immunoglobulin M) on admission, and such tests were repeated and negative prior to discharge, probably being altered due to cross reaction, despite the history of travel for about two years to an endemic area (Canada). Rheumatological tests, tumor and granulomatous disease markers, and serology were negative. When studying the cerebrospinal fluid, we found pleocytosis with lymphomonocytic predominance and hyperproteinorrhachia, with negative VDRL and panel for meningitis. Therefore, the hypothesis of neurosyphilis causing involvement of multiple cranial nerves was suggested and treatment with intravenous crystalline penicillin G was started for 14 days. After the first day of antibiotic therapy, the skin lesions became more prominent (Jarisch-Herxheimer reaction), with spontaneous disappearance in the subsequent days. Even during hospitalization, the patient evolved with gradual improvement of the ptosis on the left, absence of tongue deviation to the right and partial improvement in gait. Intravenous antibiotic therapy was completed, rehabilitation was indicated and outpatient return was planned.

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550 – A 15-year-old boy with sudden short-memory impairment and ataxia

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A previously healthy 15 years-old male was taken to the hospital because of acute onset short-term memory impairment in the last two months. He would ask questions repeatedly and forgot about daily activities. He had no prodromal symptoms or any history of fever, nausea, or diarrhea. Physical examination was normal, except for an episodic memory deficit. Brain magnetic resonance imagin (MRI) was normal, cerebrospinal fluid (CSF) analysis revealed 9/mm^3 cell, Hem 255, protein 35, glucose 50. Serology for human immunodeficiency virus, syphilis, and hepatitis were negative. Complete blood count, thyroid, liver and renal function were normal. After two months, he evolved with cephalic and upper limbs tremor, speech disturbances and imbalance. Physical examination showed global ataxia and dysarthria. A novel MRI disclosed T2/FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) hyperintensities in cerebellar right vermis, with gadolinium enhancement suggesting glial tumor. At this point, CSF analysis showed positive oligoclonal bands. Electroencephalogram was unremarkable. Brain Positron Emission Tomography-Computed Tomography (PET/CT) revealed a nodular region of increased FDG uptake along the right cerebellum hemisphere and vermis. Brain biopsy showed a polyclonal immune cells infiltrate, chronic meningitis, and focal cerebellar atrophy. No neoplastic cells were found. The patient was first treated IV methylprednisolone and IVIG, with partial tremor and imbalance improvement, but persistent cognitive impairment. By this time, the antineuronal antibodies panel were positive for Anti-AMPA (Anti-alpha-amino-3-hydroxy-5-methyl-4-isoxazolepropionic acid receptor). As the patient developed partial improved he was started on Rituximab. Autoimmune encephalitis associated with anti-AMPA antibodies was first described in 2009 and the initial description involved adult patients with limbic encephalitis and seizures. We report an atypical presentation in an adolescent, with cerebellar pseudotumoral presentation. This report extends the clinical presentation of the disease among the young, and shows that the disease may initiate as cognitive impairment evolving with important cerebellar symptoms. Moreover, this case illustrates PET-CT may not differentiate between neoplasia and inflamamatory mechanism in AMPA encephalitis.

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551 – Critical review on the psycho-emotional effects on children due to the vacancy of classes due to the COVID-19 pandemic

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Introduction: With the advent of the coronavirus diseade 2019 (COVID-19) pandemic and social isolation policies many school institutions have had to suspend classes or adapt to the online system. However, the lack of this learning model and greater exposure to the Internet can cause health problems and psychic and emotional development.
Objectives: This study aims to critically analyze the implications in children due to the suspension of classes during the COVID-19 pandemic period.
Methods: Review of integrative literature, consulted the Databases PubMed, SciELO and Google Scholar. Chosen as descriptors (DeCS): "COVID-19", "Children" and "Classes" separated by Boolean connectors, and articles in English and Portuguese.
Results: The first years of teaching were the most affected by the absence of classes with a fall in learning from 6.02 to 5.64 in Brazil. In Latin America, school delay sits up to 15 months after the isolation period causing delays in learning and development. The problematic use of the Internet is defined as an excessive concern and use the internet to the point of causing clinical damage in various domains, such as the commitment of relationships and the education of the individual.
Conclusion: The total or partial interruption of classes causes negative effects in any school phase, but mainly in the phase of cognitive development, which is early childhood. Distance education did not work effectively for several children. Increased anxiety, irritation and financial instability causes students to leave schools.

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553 – Neurosyphilis - atypical presentation

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Neurosyphilis is a disease with high morbidity, whose incidence is alarming. We present a case of syphilis with an atypical neurological presentation, as tabes dorsalis was the most common form in the pre-antibiotic era and is currently uncommon in the antibiotic era. Neurosyphilis can be classified into ear-

ly and late forms. Early forms usually affect the cerebrospinal fluid, meninges, and vasculature, while late forms affect the brain and spinal cord parenchyma This case demonstrates the great variability of clinical manifestations of syphilis and the importance of maintaining a high degree of suspicion of the diagnosis. MLC, female, 64-year-old. She reports abdominal pain in the lower floor, followed by constipation, urinary retention, hypoesthesia in the right lower limb, being worse in the sella region. Physical examination on admission showed hypopalesthesia in lower limbs, saddle anesthesia, tactile sensory level at T4. In the investigation exams, she presented magnetic resonance imaging of the spine with an image suggesting extensive longitudinal transverse myelitis, with a more central involvement and discreet contrast enhancement. It features VDRL 1:16 and electrochemiluminescence reagent for syphilis, AntiHIV non-reactive. Clear and colorless cerebrospinal fluid with 24 erythrocytes, 94 mm^3 leukocytes with a predominance of mononuclear cells (98%) and proteinorrhachia (72.6 mg/dl) and glucose 46 mg/dl, with HGT 86 mg/dl. Once the diagnosis of neurosyphilis is made, the patient is treated with crystalline penicillin for 14 days, and at the end of the treatment there was an improvement in the condition of saddle anesthesia, urinary retention, no longer needing a bladder catheter for relief and improvement in constipation. Thus, since Neurosyphilis is a prevalent disease, its screening should be part of the routine investigation of neurological conditions with a possible infectious etiology.

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555 – Prevalence of high risk of obstructive sleep apnea in patients with stroke and its epidemiological profile

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Introduction: Obstructive sleep apnea (OSA) is a disease of upper airways diagnosed with polisomnography that is well associated with stroke throught diverse mechanisms. Either because of the cost of the diagnosis or because of the incipient topic, it lacks studies about its prevalence in Brazil and the epidemiological profile.

Methods: This study aims to elucidate the epidemiological profile of patients with stroke and OSA in Brazil. We performed a transversal study, part of a cohort of stroke patients. Patients were evaluated of risk of OSA with STOP-Bang and SOS scores; also,data of age,hipertension,diabetes,atril fibrillation (AF), obesity,body mass index (BMI), smoking,alcoholism,renal disfunction,dyslipid emia,asthma, sleep disorders and previous stroke were collected.

Results: We obtained 190 patients: 48.9% female, mean age 63.6 ± 6.43 years, mean BMI 25.69 ± 5.64 kg/m^2, mean SOS 11.65 ± 6.43, mean STOP-Bang 3.22 ± 1.45. Twenty-six were obese, 33 alcoholic, 37 smokers, 72 hypertensive, 71 diabetic, 15 with renal disfunction, 5 asthmatics, 8 with AF, 25 dyslipidemic, 28 with sleep disorders and 56 with previous stroke. 62.1% were classified as high risk with STOP-Bang (mean age 65.62±10.18, mean BMI 26.96±5.96) of which 71 were female, 23 obese, 23 alcoholic, 26 smokers, 102 hypertensive, 51 diabetic, 15 with renal disfunction, 1 asthma, 4 AF, 13 dyslipidemic, 19 with sleep disorders and 35 with previous stroke. 54.7% were high/moderate risk with SOS: (mean age 63.11±11.89, mean BMI 26.12±6.52) of which 55 were female, 19 obese, 13 alcoholic, 19 smokers, 82 hypertensive, 44 diabetic,12 with renal disfunction, 2 asthma, 5 AF, 12 dyslipidemic, 17 with sleep disorders and 32 with previous stroke.

Conclusion: An importante number of previous diseases were found in patients with high risk of OSA, which hypothetizes the role of obstructive sleep apnea in these diseases and the presence of them as comorbidities that lead to high rates of stroke and recurrence of stroke.

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558 – Stroke profile in patients with high risk of obstructive sleep apnea: a transversal study

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Introduction: Obstructive sleep apnea (OSA) is a disease of upper airways, diagnosed with polisomnography, that is well associated with stroke throught diverse mechanisms. Despite that, it still lacks studies about stroke profile in patients with high risk of OSA.

Objectives: This study aims to describe the diferences between stroke in patients with high and low risk of OSA.

Methods: We performed a transversal study, part of a cohort of stroke patients. Patients were evaluated of risk of OSA with STOP-Bang and SOS scores; also, data of wake-up stroke, stroke type and etiology, transient ischemic attack (TIA), thrombolysis, circulation evolved, ASPECTS (Alberta stroke program early computed tomography score), PC-ASPECTS (posterior circulation - acute stroke prognosis early computed tomography score), hemorragic transformation and its type (ECASS), ABCD2, National Institute of Health Stroke Scale (NIHSS) and death during internment were colectted.

Results: We obtained 190 patients. 62.1% were classified as high risk with STOP-Bang and (mean NIHSS 6.59 ± 5.13) and 8.5% died: 5.9% had a TIA (mean ABDC2 5.14 ± 1.21), 22% a wake-up stroke and 81.4% had a ischemic stroke (24 patients were cardioembolic, 34 atherotrombotic and 26 indeterminate), of which 12.7% were thrombolysed and 15.3% had hemorragic transformation. In ´patients with low risk, 2.8% died and we had mean NIHSS 7.64 ± 5.55, 18.3% had wake up stroke and 8.5% a TIA (mean ABCD2 4.17 ± 1.60), 74.6% were ischemic strokes (24% cardioembolic, 23 indeterminated); 12.7% patients were thrombolysed and 21% evolved with hemorragic transformation.

Conclusion: We verified a lower NIHSS mean and a higher rate of wake-up stroke, hemorragic transformation and TIA in patients with low risk of OSA. Althought this would suggest better outcomes in population with high risk, they had 5% more deaths: so, despite the aparent better initial state (that we can hypothesize about a type of "lucid interval" in this group), they still evolve worst.

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560 – Verheij syndrome: a rare cause of intellectual disability

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Case presentation: A 4-year-old boy was born to non-consanguineous parents at 38 weeks. Neonatal anthropometric measurements were normal. Since birth, he presented with global developmental delay, and muscular hypotonia. At present, he shows adequate psychosocial interaction. He has a healthy 3-year-old sister. On physical examination, there are dysmorphisms, such as prominent and pointed ears, long eyelashes, elongated face, flat oc-

cipital region, supernumerary teeth, and maxillary hypoplasia. His anthropometric measurements are normal for his age (p50). On neurological examination, he presents with apraxia of speech, axial and appendicular hypotonia, and reduced deep tendon reflexes. Brain magnetic resonance imaging showed a slight thinning of the corpus callosum and mild ectasia of the lateral ventricles. Transthoracic echocardiography and ultrasound of the kidneys and urinary tract were normal. On genetic investigation, no abnormalities were found in karyotype and CGH-Array. Whole exome sequencing showed a pathogenic variant in the *PUF60* gene (c.24+1G>C) in heterozygosis. Thus, the patient was diagnosed with Verheij syndrome. The patient is accompanied by physiotherapy, speech therapy, occupational therapy, and a psychopedagogy group.

Discussion: Verheij syndrome is a rare condition caused by variants in the *PUF60* gene, which encodes a component of the spliceosome. This syndrome is characterized by intellectual disability, delayed growth and neuropsychomotor development, facial dysmorphic features, and osteoarticular abnormalities. Also, there may be heart and kidney disorders. The spectrum of this disease's manifestations and severity still need to be further explored in future studies, as well as the treatment and prognosis of this condition. Multidisciplinary support is essential for managing the consequences of the disease. This case report reinforces the leveraged importance that genetics has had in the diagnosis of intellectual disabilities.

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561 – Constructing a low-cost lumbar puncture simulator for medical education

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Introduction: Simulation has progressively been recognized as a reliable teaching method for delicate medical procedures, including lumbar punctures, an important diagnostic and therapeutic tool. Despite its importance, studies show training to be insufficient, with medical students and residents feeling technically unprepared. Furthermore, multiple challenges lie ahead of implementing curricular training, including costs of simulators, available for prices ranging from US$ 1158,89 to US$ 2.897,21 a unit. Because the technique does not require complex events, selecting a type of simulator can be compatible with simpler but realistic devices that can be made at teaching institutions, not requiring third party suppliers.

Objectives: To construct a low-cost lumbar puncture simulator for medical education.

Methods: Based on literature and professional experience, critical points of construction and resources available on the market were assessed. A prototype was confectioned with processed vertebrae, sponges, a latex tube filled with water, attached to a syringe for pressure, and makeshift skin and soft tissue from a commercial suture kit, all mounted on a metal apparatus.

Results: The simulator concocted of the most cost-effective materials and only the most crucial components performed comparably to brand models according to pre-tests by selected students and experienced professionals. Currently, a more reproductible prototype is being constructed out of 3D-printed materials, which will be further evaluated and validated.

Conclusion: The rough prototype performs comparably to commercial models, and we believe the study will further corroborate modern tendencies to transform teaching into cost-effective techniques that enrich medical education in resource-limited settings.

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562 – A Creutzfeldt-Jakob disease mimics: case report

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Introduction: Creutzfeld-Jakob Disease (CJD) is an important differential diagnoses in patients with rapidly progressive dementia (RPD). Several inflammatory, immune-mediated, vascular, granulomatous and neurodegenerative conditions can mimic this disease.

Objectives: Report a curious case of CJD mimics that was a limbic encephalitis.

Methods: A 71-year-old man, with diabetes mellitus. His initial symptoms included visual hallucinations and temporo-spatial disorientation. Over the course of a month, he lost his critical judgment, and began with aggressive behavior and disinhibition. He progressed with gait apraxia and diffuse myoclonic jerks, as well as loss of both bladder and bowel sphincter incontinence and dysphagia. Blood tests results were normal. Cerebrospinal fluid presented increase in proteins (82) without any further alterations. Brain magnetic resonance imaging showed a bilateral increase in T2/FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) signal intensity from the hippocampus to the amygdala. Also presented alterations in white matter suggesting microangiopathy (Fazekas 2). Brain PET-CT (Positron Emission Tomography-Computed Tomography) scan showed hypermetabolism in bilateral basal ganglia and mesial temporal regions. EEG evidenced moderate to severe disorganization of background activity and myoclonic jerks with no electroencephalographic correlation. Detection of 14-3-3 protein in cerebrospinal fluid was negative.

Results: Patient received 1 g of methylprednisolone for five days and presented significant improvement of clinic symptoms, stopping myoclonic jerks and dysphagia with better tenacity, self-orientation and improved gait. However, he presented persistently short term memory alteration with signs of disinhibition (hyperphagia) that didn`t get better with a second immunotherapy treatment with cyclophosphamide.

Conclusion: immune-mediated conditions as encephalitis must be kept in mind when assessing differential diagnoses of RPD and treated as soon as possible

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563 – Poorer prognosis and higher chance of death in Guillain-Barré syndrome associated to COVID-19: a cohort study

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Introduction: Guillain-Barré Syndrome (GBS) is the most common neuromuscular complication in coronavirus disease 2019 (COVID-19) and dysimmune response has been considered as the most important mechanism. In the literature, there is a male preponderance and amongst the GBS-

variants, acute inflammatory demyelinating polyneuropathy (AIDP) is the most common type.

Methods: This is a retrospective cohort study of GBS hospitalization in the Brazilian health system from 1 February 2020 to 1 February 2023. Our objective was to compare the patients COVID-19 (COV19) against those of non-COVID-19 (nCOV19). The primary outcome was poorer prognosis and the secondary outcome was in-hospital mortality. SARS-COV-2 RT-PCR testing was performed on all patients.

Results: Sixty patients were included, 78.3% nCOV19 and 21.7% COV19 asymptomatic or with mild respiratory symptoms. They aged 4-79 years (IQ 27.3-61.5) with median age COV19 of 48 years (IQ 27.5-61) and nCOV19 of 43 years (IQ 25-56). The majority of COV19 were female (61.5%) and nCOV19 were male (57.4%). The most frequent subtype in both groups was AIDP, but we found 38.4% axonal forms in COV19 compared with 21.2% nCOV19 (OR 2.31; $P < 0.102$). When comparing Modified Erasmus GBS Outcome Score (EGOS ≥ 7), we found 46.1% in COV19 and 27.65% nCOV19 (OR 2.24; $P < 0.102$). In the Erasmus GBS Respiratory Insufficiency Score (EGRIS ≥ 3), we found 53.8% in COV19 and 36.2% nCOV19 (odds ratio [OR] 2.06; $P < 0.125$) with orotracheal Intubation in 30.7% COV19 and 27.6% nCOV19 (OR 1.16; $P < 0.048$). Finally, when comparing deaths, we found 15.3% in COV19 and 2.1% nCOV19 (OR 8.36; $P < 0.026$).

Conclusion: Despite COVID-19-associated GBS does not seem to differ in clinical presentation and treatment. However, in our cohort, COV19 had a relatively poorer outcome than GBS occurring otherwise, including higher odds of in-hospital mortality.

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564 – Characterization of de novo variants in exomes of individuals with autism spectrum disorder

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The main objective of this essay was to contribute to the characterization of the genetic architecture of autism spectrum disorder (ASD) based on an analysis of a Brazilian series, which is still little studied. To achieve this goal, we verified the proportion of cases of ASD caused by de novo variants in neurodevelopment genes (genes from the SFARI bank and those associated with neurodevelopment described in the DECIPHER bank). Sixty-three trios were evaluated, composed of parents and probands diagnosed with ASD treated at the Human Genome and Stem Cell Studies Center (CEGH-CEL, USP). Genealogy, clinical data, gender, age at the consultation, and parental age were collected. Whole-exome sequencing was performed through a collaboration with Mount Sinai, New York, United States (collaboration with the Autism Sequencing Consortium-ACS). Identification of De novo variants in candidate genes for ASD was performed using the LOVD program (LOVD v.3.0 – Leiden Open Variation Database). It was observed that most of the probands were boys (n = 55, 86%), and the minority had a family history of ASD (n = 4, 6%). It was also found that 40% (n = 25) of individuals had a delay in language development, and a small percentage had comorbidities such as ADHD and epilepsy (n=6, 10% and n=2, 3%, respectively). The mean parental age at the time of pregnancy was close to 30 years for both parents (29.7 and 32.5 for the mother and father, respectively). Nine de novo pathogenic or potentially pathogenic variants were identified in candidate genes: for TEA: six in SFARI genes (four pathogenic variants in NF1, TLK2, DNAH17, BRSK2 genes, and two probably pathogenic variants in ARHGAP5 and HUWE1) and three in genes of neurodevelopment of DECIPHER (Deciphering Developmental Disorders Study, 2015) (two pathogenic variants in the ERLIN2, ST3GAL3 genes and one probably pathogenic in COL11A1). When performing the gene enrichment analysis of genes with pathogenic or potentially pathogenic variants, we observed the enrichment of genes for intracellular protein transport. The clinical picture of individuals with pathogenic or potentially pathogenic variants was expected, as previously described in the literature. This study suggests that de novo variants are also an essential mechanism for the etiology of ASD in Brazil, explaining the genetic architecture of 9.5% of cases.

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565 – "Wake-up" onset of pharyngocervicobrachial variant of Guillain-Barré syndrome: a case report

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Guillain-Barré syndrome (GBS) is the most common cause of acute flaccid paralysis in the world. There are variants. Of the cranial forms, the following stand out: the classic form, facial diplegia with distal paresthesias, pharyngo-cervico-brachial form, polyneuritis cranialis, Miller-Fisher syndrome and Bickerstaff encephalitis. This study aimed to report the case of a 73-year-old male patient, diabetic, former smoker and alcoholic, who presented at the Neurology outpatient clinic of a tertiary hospital in Pernambuco, after emergency care and 19 days of symptoms, reported as sudden cervical weakness, dysarthrophonia, dysphagia and weakness in the right hemiface, three weeks after vaccination (influenza and triple viral) and flu syndrome. He had dyspnea since the onset of the condition, with no progression or fluctuating complaints. The neurological examination showed multiple cranial nerve syndrome (right peripheral pattern facial palsy, reduced elevation of the soft palate and cervical extension paresis) associated with global hypo/arreflexia. Complementary exams showed, in addition to leukocytosis and signs of bronchopathy on chest tomography, cerebrospinal fluid with 00 cells and 48 proteins and electroneuromyography with predominantly sensitive axonal polyneuropathy, decrease in bilateral facial motor amplitude, needle with myopathic pattern. Brain magnetic resonance imaging without alterations. With the possibility of cranial polyradiculoneuritis and a history of dyspnea raised, he was admitted to the intensive care unit to monitor his breathing pattern and dysautonomia. He received antibiotic therapy for seven days due to pneumonia and pulse therapy with IVIG (2 g/kg for five days). He maintained progressive improvement of symptoms. He was discharged with a multidisciplinary outpatient follow-up scheduled. It is concluded, therefore, that the recognition of GBS and variant forms is necessary.

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568 – Neurosyphilis causing bilateral optic perineuritis

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Introduction: Optic perineuritis is a rare disorder with multiple possible etiologies, including systemic autoimmune or infectious syndromes and, more rarely, neurosyphilis. The aim of this report is to describe an uncommon manifestation of this reemerging infectious disease. Case report based on a retrospective analysis of the medical records of the patient.

Case report: A previously healthy 35-year-old female presented to the hospital with a seven-day history of progressive right eye vision loss and pain on extraocular movement, with development of a pink maculopapular rash on the trunk. On the sixth day of her disease, similar symptoms appeared in the contralateral eye. Physical examination showed reduced fotomotor reflex and finger counting at 50 cm in both eyes. Magnetic resonance imaging revealed bilateral and circumferential thickening with enhancement of the optic nerve sheath, suggesting bilateral optic nerve perineuritis. Serum Venereal Disease Research Laboratory (VDRL) was 1/64 and cerebrospinal fluid (CSF) showed 440 cells (86% lymphocytes), proteins 97 mg/dL, glucose 47 mg/dL, lactate 21 mg/dL and VDRL 1/4. She was treated with IV crystalline penicillin for 21 days and had partial improvement of bilateral vision within two months. At follow-up, her visual acuity was 20/40 in the right eye and 20/100 in the left; her CSF normalized, with nonreactive VDRL.

Conclusion: Neurosyphilis is a treatable cause of optic perineuritis, and its recognition is important to establish the right antibiotic treatment.

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570 – Case report: an atypical case of post-poliomyelitis syndrome and its radiological correlations

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A.V.S., 57-year-old, had delayed motor development milestones, walking at the age of three with slow progression of symptoms which later stabilized. At the age 31 he noticed a new worsening, with proximal lower limbs weakness and the need for unilateral support while walking. He reports a history of Poliomyelitis. Neurological examination showed thoracic kyphosis and lumbar hyperlordosis, flaccid areflex and asymmetric tetraparesis predominantly in lower limbs (worse on the left side) and proximally in upper limbs. He also had muscle atrophy on those sites with fasciculations and a pawing gait. The hypothesis of Spinal Muscular Atrophy (SMA) was raised. The electroneuromyography identified chronic severe denervation in L3-S2 and moderate in C5-C7 myotomes. Thighs and legs magnetic resonance imaging identified liposuction in the anterior, posterior and lateral compartments bilaterally with myoedema. A gene panel testing showed no variants that would justify the case. Considering the asymmetry as well as its evolution and the possible infection by Poliomyelitis, we considered the hypothesis of Post-Poliomyelitis Syndrome (PPS) to be more likely, admitting SMA with lower limbs predominance as a differential diagnosis. Poliomyelitis was a prevalent disease during pre-vaccination era and is currently restricted to surviving patients who may present a phenomenon of progression after years of stability, triggered by overtraining with dysfunction of motor neurons, recognized as PPS, with impairment of functionality. There are few reports of muscle imaging in PPS that could be crucial for investigation of atypical cases and a potential marker of progression and natural history of the syndrome.

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576 – Effects of transcranial magnetic stimulation on the working memory of people with multiple sclerosis

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Introduction: Multiple sclerosis (MS) causes various cognitive symptoms. Transcranial magnetic stimulation (TMS) is a resource used for stimulation.

Objectives: To evaluate the effects of TMS on the working memory of people with MS.

Methods: A double-blind crossover study was carried out with 29 people diagnosed with MS (18 relapsing-remitting, 6 primary progressive and 5 secondary progressive), aged between 29 and 68 years (mean = 47.2, standard deviation [SD] = 10.9 years), 9 men (31%) and 20 women (69%), with EDSS from 0 to 6.5 (mean = 4.3 and SD = 1.85) and diagnosis time between 1 and 24 years (mean = 9.5 and SD = 6.57 years). The active group received 10 TMS interventions (primary motor cortex (Cz): 10Hz, 50 pulses per time, 30 trains, 20 seconds apart, totaling 1,500 pulses at 90% of the resting motor threshold and pre -left dorsolateral front (F3): 10 Hz, 50 pulses per train, 40 trains, 20 second interval, totaling 2,000 pulses at 110% of resting threshold) for 10 consecutive working days. The sham group received the inactive TMS and participated in the physical activities. After 30 days, there was an inversion of the active and sham groups for a new sequence of 10 days. For evaluation, an interview was conducted for data collection and

the subtest Digits of the Wechsler Intelligence Scale (WAIS-III) was applied at the beginning and end of the stimulations. The chi-square test was used for statistical analysis.

Results: 55.2% of people who received active TMS improved, against only 27.6% of people who received sham TMS. This difference was significant with P = 0.033.

Conclusion: TMS appears to be an important resource for treating the working memory of people with MS. This result may encourage further research.

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577 – Patient-reported outcome measures with multiple sclerosis on the effects of transcranial magnetic stimulation on communication difficulties

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Introduction: Multiple sclerosis (MS) can trigger difficulties in communication. Transcranial magnetic stimulation (TMS) is a treatment option.

Objectives: To analyze the self-reported communicative ability of people with MS undergoing TMS.

Methods: The protocol consisted of 10 interventions by TMS (primary motor cortex, left dorsolateral pre-frontal cortex). The domain 1 of the self-administered WHODAS 2.0 Disability Rating Scale was used before and after (30 days) TMS to identify patient's opinion of communication. Scores ranged from 0 = minimum difficulty to 100 = maximum.

Results: The study comprised 30 people with MS: 22 women, 8 men, age 33–68 years (mean = 50.0, standard deviation [SD] = 10.6), EDSS 0-6.5 (mean = 4.6, SD = 1.9), time diagnostic 1–24 years (mean = 10.0, SD = 7.2). All patients mentioned preserved ability for general understanding of the interlocutors' speech. Prior to TMS, n = 18/60% had moderate difficulty concentrating (including speech content/subject), moderate/severe to n = 24/80% in recent memory, n = 10/33%) in problem solving, and n = 10/33% in learning, fundamental mental and reasoning activities for communication, activity reported with a moderate degree of difficult in n = 18/60% to start and maintain conversations due to inability to recall words (language disorder) or impairment in speech intelligibility (dysarthria). Self-reports pointed to a decrease in difficulties after TMS.

Conclusion: We observed that WHODAS 2.0 seems to be a favorable both to identify the patient-reported outcome measures about the communicative difficulties, and to analyze the effects of TMS, which, in turn, showed potential to improve verbal communication and requires longitudinal investigation.

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578 – Treatment with transcranial magnetic stimulation combined with physiotherapy on manual dexterity and gait of people with multiple sclerosis

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Introduction: Transcranial magnetic stimulation (TMS) has good therapeutic effect and clinical application value when associated with therapeutic interventions.

Objectives: To evaluate the effect of TMS combined with physiotherapy on the manual dexterity and gait of people with multiple sclerosis (MS).

Methods: Participated 20 people with MS, 14 women and 6 men, aged 33 to 68 years (standard deviation 50.0) and EDSS between 0 and 6,5. The protocol consisted of 10 TMS sessions and 6 physiotherapy sessions. Participants were randomized and divided into two 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 1,500 pulses at 90% of resting motor threshold and left dorsolateral prefrontal cortex (F3): 10Hz, 50 pulses per train, 40 trains, 20 seconds interval, totaling 2,000 pulses at 110% of resting threshold), and Group II) sham, that received the application without TMS stimulation (10 patients). All underwent physiotherapy. The Box and Block Test and Timed up and Go Test (TUG) was applied before the combined interventions and after 60 days.

Results: It was found that 90% of participants showed improvement in manual dexterity, and 10% a plateau on Box and Block Test. At the TUG, it was found 70% of improvement and 30% of decrease in performance.

Conclusion: This study suggests that TMS can be incorporated into the physiotherapy in the rehabilitation of manual dexterity and gait in people with MS.

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581 – A new presentation or a new disease? An acute leukoencephalopathy resembling Canavan's

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Case presentation: We describe the case of a 75-year-old female, with no past relevant comorbidities. The onset was of mental confusion and imbal-

ance, within two weeks progressing to dysphonia, dysphagia and spastic quadriparesis. Brain MRI showed hyperintense T2/FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) lesions in white matter, with cortical sparing, restricted diffusion and gadolinium enhancement. Demyelinating diseases and central nervous system lymphoma hypothesis were made. Two months later a brain biopsy was performed. Diffuse white matter vacuolar impairment was found and no inflammatory infiltrate, resembling Canavan's disease. The patient had progressive clinical worsening, with prolonged hospitalization and poor general condition at discharge, not enduring further investigation. She deceased after 8 months of symptoms onset.

Discussion: The diagnosis of acute leukoencephalopathies is challenging and neuroimaging may be helpful. Diffusion, contrast enhancement and corticosubcortical relation in magnetic resonance imaging (MRI) can present valuable clues. In this case, for example, acute disseminated encephalomyelitis was less likely due to the MRI evidencing dissemination in time. To better address malignancy, brain biopsy was mandatory. Not only cancer was excluded, but the demyelination was revealed to possibly be neurodegenerative, which was neither clinically nor radiographically evident. The vacuolar pattern found is described in Canavan disease, a rare leukoencephalopathy with onset at 1–4 months age, and mean survival of months to few years. In literature there is only another report, cataloged in our Pathology department, of a 43-year-old male with similar clinical, imaging and histological findings. This case may illustrate a not yet cataloged disease, maybe an unknown presentation of Canavan disease spectrum, adding one more differential diagnosis for acute leukoencephalopathies.

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582 – Incidence of neurologic syndromes after diagnostic of arboviroses in reference center

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Introduction: Arboviruses, such as Dengue (DENV), Zika (ZIKV), and Chikungunya (CHIKV), are infectious diseases that have become a public health challenge in Brazil. These viruses are transmitted by mosquitoes and can cause a range of symptoms, from mild flu-like illness to severe and sometimes fatal neurological syndromes.

Objectives: To analyze the prevalence of neurologic syndromes related to the arboviroses in the state of Paraíba.

Methods: This is a cross-sectional, documentary, descriptive, quantitative and observational study. Analysis of sociodemographic data, neurological symptoms and imaging findings from the medical records of patients with a history of ZIKV, DENV or CHIKV infection was performed.

Results: This study analyzed 580 medical records, with 52 records showing arbovirus before neurological symptoms. Among which, 42 medical records were selected, 13 with confirmed serology. The majority of patients were female (78.6%). The most common arbovirus was chikungunya, followed by dengue. Regarding neurological symptoms, 24 patients had multiple sclerosis, 4 had myelitis, 3 had optic neuritis (ON), 3 had neuromyelitis optica (NMO), 1 had acute disseminated encephalomyelitis (ADEM), and 7 are under investigation. Of the patients with ON, 3 reported previous infection with dengue and 2 with chikungunya (CKG). Of the patients with myelitis, 3 reported previous CKG, 1 dengue and 1 zika. The most prevalent symptoms were paresthesias (43%) and fatigue (24%). Imaging exams, 8 patients without alterations; the most frequent was a sign of hypertension in the brain (29%).

Conclusion: This study highlights the importance of early detection and management of arbovirus infections to prevent the development of neurological complications. Further research is necessary to better understand the pathophysiology of these diseases and develop effective treatments.

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583 – Atypical manifestation of Anti-Gaba-Br autoimmune encephalitis

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Introduction: Anti-GABA-Br is an autoantibody associated with autoimmune encephalitis and small cell lung carcinoma. Clinical seizures, including status epilepticus, represent the most common feature of anti-GABA-Br related disorders. This case reports a previously healthy 40-year-old man with rapidly progressive dementia and Anti-GABA-Br positive cerebrospinal fluid (CSF). This is a case report based on retrospective analysis of a single patient's medical record.

Case report: From May 2019 to October 2020, the patient presented with dulling, decreased speech frequency and gait disturbance. He became unable to perform all basic and instrumental activities of daily living. Physical examination was marked by: pancerebellar dysfunction and impaired working memory. Brain magnetic resonance imaging (MRI) showed global brain volumetric reduction with bilateral T2/FLAIR (T2-weighted-Fluid-Attenuated Inversion Recovery) hypersignal in cortical regions of insula and mesial temporal lobes, without contrast enhancement. Analysis of CSF: 8.3 cells/mm^3 (96% lymphocytes) and protein 64 mg/ml. There was no evidence of: clinical/electroencephalographic seizures or pathological patterns on electroencephalograms. He received methylprednisolone for five days followed by cyclophosphamide infusions (one per month for seven months). Azathioprine started after tapering prednisone. On March 2022, the patient underwent a new MRI with hypersignal improvement although atrophy had persisted. CSF after treatment had no inflammatory changes and was anti-GABA-Br positive. The patient presented a partial recovery and was still dependent on instrumental activities.

Conclusion: Anti-GABA-Br encephalitis has a predilection for males, affects variable ages and is best characterized by generalized seizures with evolution to refractory status epilepticus although behavioral changes may occur. There are few reports of seizure free cases.

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584 – Super-refractory status epilepticus after intoxication by quetiapine and carbamate

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Introduction: Quetiapine, an atypical antipsychotic, works through the agonism of multiple brain receptors, including dopaminergic, serotonergic, and adrenergic receptors. Compared to typical antipsychotics, quetiapine has a more favorable profile of adverse reactions. Myoclonus is infrequent. However, there have been reports of quetiapine overdose associated with generalized myoclonus. Electroencephalogram (EEG) modifications linked to the use of quetiapine are unusual. Aldicarb, commonly known as 'chumbinho' in Brazil, is a carbamate used as a rodenticide. Its toxicity is caused by the inhibition of acetylcholinesterase. A few reported cases of myoclonus related to its use may be due to the anticholinesterase effect causing hyperactivity. This is a case report based on retrospective analysis of the patient's records.

Case report: A 23-year-old woman was admitted after a suicide attempt by ingesting carbamate and quetiapine 36 hours after the attempt. Her initial symptoms were vomiting and sialorrhea, followed by generalized tonic-clonic seizures and coma. At admission, she was already sedated with midazolam, with her last tonic-clonic generalized seizure two hours earlier. Her pupils were myotic, with persistent tachycardia and diffuse muscular hyperexcitability – clonic movements were evoked with minimal stimuli. Brain imaging and cerebrospinal fluid analysis showed no alterations. A loading dose of 20 mg/kg of phenytoin was administered. Upon admission, an EEG showed status myoclonus. Midazolam was then titrated to 0.8 mg/kg/h, and clobazam (30 mg/day) and levetiracetam (1.500 mg/day) were added, with resolution of the status epilepticus after three days.

Conclusion: Status epilepticus should be considered a possible presentation of quetiapine and carbamate intoxication.

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585 – Neuropathic pain as consequence of rifle injury: a case report and literature review

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Rifle injuries are relatively common in emergency cases. It may result in several acute damages or it could lead to chronic consequences to the patient. This paper reports a case of a rifle injury on the leg that resulted in neuropathic pain and venous insufficiency. To treat this patient, a multidimensional approach was prefered, using comprehensive therapy, drug treatment and neuromodulation. The case reported in this paper was gathered by appreciation of the patient's records and interviews and was discussed in the light of the most recent literature. Neuropathic pain is caused by lesion or disease of the somatosensory nervous system, according to The International Association for Study of Pain that affects over 7 to 8 percent of general populations and corresponds to 20 to 25 percent of chronic pain, most frequently in women and man older than 50-years, diagnosed exclusively via clinical criteria. It might be classified by the clinical manifestations: spontaneous pain, evoked pain, after sensations, hyperpathia and referred pain. The details of pathophysiology of neuropathic pain are yet to be clarified once most of the data in the matter comes from animal testing and should be interpreted carefully, majorly in the long-term assessments. It is clear, though, that several mechanisms are involved in the pathogenic process, depending on anatomical location and etiology, and that different classifications of neuropathic pain could be clustered to form several subgroups based on the symptoms combination, each reflecting different mechanisms. Understanding these underlying mechanisms is crucial to a proper treatment of the patient. In this paper, we report a patient who suffered from a fire gun injury in the left feet resulting in a neuropathic pain. A 40-year-old male comes to the office referring to allodynia, hyperalgesia, burning pain and paroxysmal pain, characterizing persistent neuropathic pain, intensity 8 in the one-to-ten scale, and edema during the day in the left inferior member. The patient reports being shot, one year and half before, with a 5.56 caliber rifle at the left feet, riching the fifth metatarsal No remarkable finding was present in the medical history prior to the incident. At physical examination, it was perceived that the patient developed venous insufficiency, for which was prescribed a ⅞ compression socks (30-40 mmhg). The neuropathic pain was firstly treated with pregabalin 50 mg/day and Duloxetine 30 mg/day, showing relevant response. Then, electroneuromyography revealed intermedial dorsal cutaneous nerve injury, being managed with a local block, prepared with 8 ml of 2% Lidocaine and 2 ml of 4 mg Dexamethasone, and a 30 Hz neuromodulation, terminating the pain. Ballistic knowledge is essential to a proper management of gunshots injury. In this case report, the patient suffered a 5.56 caliber rifle injury, a high-velocity gun, resulting in an intermediate dorsal cutaneous nerve and the fifth metatarsal lesion. In these injuries, the projectile transfers energy to the affected tissue, expanding and crushing it. This brief expansion creates a subatmospheric pressure resulting in bacterial and foreign material suction into the wound. It is worth to note that gunshot-related injuries in the feet, due to scarce soft tissue, numerous bones and articulations, are more propense to vascular and neural lesions. Although pathophysiological mechanisms of neuropathic pain are not fully understood, it is described post-traumatic peripheral ectopic nerve activity and central sensitization that could help to understand the pain origin. Moreover, coexisting psychological and emotional triggers might be associated with neuropathic pain association. As the electroneuromyography shows, the intermediate dorsal cutaneous nerve, the smallest branch of the fibular nerve, was injured at the fifth metatarsal level rising neuropathic pain. In addition, as mentioned above, gunshot injuries might lead to bacterial invasion, triggering the inflammatory response. In this setting, it is important to point out that evidence suggests inflammatory mediators and proinflammatory cytokines as an inductor factor for pain hypersensitivity. Another possible gunshot related etiology that must be considered is lead toxicity in the composition of gun projectiles, which is reportedly associated with neuropathies. For its heterogeneity of etiologies, mechanisms and presentations, it is essential to understand the underlying causes and its consequences to a proper treatment resulting in partial or full pain relief. In this case, the patient was treated with Duloxetine, a serotonin-norepinephrine reuptake inhibitor and

first line to treat neuropathic pain, and pregabalin, an anticonvulsant used to inhibit neuronal excitatory transmission. The synergistic action of these drugs is superior compared to monotherapy. Auxiliary, a local block was performed using Lidocaine analgesic, sodium channel blocker, and Dexamethasone, an antiinflammatory corticosteroid, testifying in favor of mechanical and inflammatory pathogenic mechanism originated by the gunshot injury. Finally, it is worth noting that gunshot wounds might give rise to venous insufficiency as a result of vascular injury and must be carefully considered in order to provide the proper treatment, revascularization for exemple. Another possible cause is deep venous thrombosis and leg injury accounts as a risk factor. In this case, it was opted for compression therapy, a conservative treatment, for increased venous return. Concluding that, neuropathic pain is a heterogeneous condition with a limited action mechanism understanding. It is required from the physicians to comprehend the multiple dimensions and main etiology of this disease in order to provide a proper treatment. As gunshots may contribute to the rise of several damage mechanisms, this case report highlights the importance of acknowledging ballistic properties and possibilities wound-related.

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586 – Rare diagnosis of dystonia and ataxia: aceruloplasminemia

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Introduction: Aceruloplasminemia is a rare autosomal recessive disorder, characterized by transmembrane cerulopasmin deficiency. Impaired iron metabolism occurs, leading to its systemic accumulation, including brain. This is a case report based on retrospective analysis of a single patient's medical record.

Case report: This case reports a 57-year-old female, previously functional, with metabolic syndrome, admitted due to dystonia and ataxia with compound heterozygosis for ACP gene. Over the course of two years and six months, she manifested: depression, anedhonia, some instrumental activities limitations, plus walking and speech disturbances. Her physical examination presented disorientation in time/space, dysarthrophonia, saccadic intrusions, dysdiadocokinesia, enlarged base gait, oral dyskinesia and cervical dystonia. Brian magnetic resonance image (MRI) demonstrated GRE/SWI signal attenuation diffusely in cortex and bilaterally in lentiform and caudate nucleus. Laboratory tests: ferritin 2.540 ng/ml; serum iron 27 ug/dL; serum ceruloplasmine below 8 mg/dL; negative infectious and rheumatological serologies; normal levels of cobalamin, hemoglobin and thyroid hormones. Genetic testing confirmed compound heterozygosis for ACP gene (variant C1149 G>A |p.trp 383*), compatible with the aceruloplasminemia hypothesis.

Conclusion: Attention should be drawn to this patient's MRI showing SWI hyposignal on basal ganglia and cortex, because it's a highly suggestive finding. Individual case reports indicate the effectiveness of Deferiprone as a treatment in individuals with aceruloplasminemia, however there is no universally accepted treatment regimen.

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587 – History of neurology in Belo Horizonte: a systematic review

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The modern neurology in Belo Horizonte was inaugurated in the late 1950s, influenced by the Rio de Janeiro´s Medical School and the international scenario. This article aims to analyze the emergence of Neurology in the capital of Minas Gerais and the influences it received. To this end, it was conducted a systematic search in electronic databases, such as Google Scholar and Scielo, using the keywords "History", "Neurology" and "Minas Gerais", articulated with boolean descriptors, selecting a total of three articles focused on historical approaches. In this regard, in the XIX century, modern neurology has been established on the basis of three major neurological schools in Germany, England and France, in which neurology was initiated as a medical speciality in 1882, at the Medical Faculty of the University of Paris. In Brazil, until 1911, Neurology was taught by professors of Medical Clinic and, later, by neuropsychiatrists, inaugurating the discipline of Neurology, distinct from Psychiatry, in 1912, in Rio de Janeiro´s Medical School, ruled by the "Father of Brazilian Neurology", the professor Antônio Austregésilo Rodrigues Lima. In Belo Horizonte, the Medical School of the Universidade Federal de Minas Gerais (UFMG) was created in 1911. From this regard, until the 1950s, neurology, in Minas Gerais´ capital was exercised by neuropsychiatrists and neurosurgeons, when it was introduced modern Neurology by the doctors José Geraldo Albernaz, who, in 1962, assumed the position of full professor on neurological clinic and starts the Medical Residency in Neurosurgery at the Hospital das Clínicas of the UFMG, and Gilberto Belisário, who was oriented by Professor Albernaz and was elected, in 1997, Professor Emeritus of the Faculty of Medicine of UFMG. Therefore, neurology has gone a long way to becoming recognizedly a medical specialty in medical schools. Currently, it is the area of study of diseases of the Nervous System, whose specialist in diagnosing and treating them is the neurologist doctor.

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588 – Gradenigo's syndrome: an old, but still alive, cause of sixth nerve palsy

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Introduction: Gradenigo Syndrome is characterized by suppurative otitis media, pain in the distribution of the trigeminal nerve, and abducens nerve palsy. Sixth nerve palsy is secondary to nerve entrapment through the Dorello canal, petroclinoid ligament and petrous apex of the temporal bone. It is a report of a single case based on a retrospective analysis of the patient's medical records.

Case report: A 12-year-old girl was admitted due to new-onset binocular horizontal diplopia, worse with far vision, and right-side headache. She also

complained of otalgia and fever for one week before admission. She has an unremarkable past medical history and up-to-date vaccination. On her neurological examination, she was alert and oriented, there was right eye abduction palsy, reduced right corneal-palpebral reflex and pain in the distribution of the right ophthalmic branch of the trigeminal nerve; meningeal irritation signs were absent. Otoscopy revealed a tender right tympanic membrane. Brain and ear computed tomography revealed abnormal right temporal petrous apex aeration and opacification of the petromastoid air cells. Brain and ear magnetic resonance imaging show abnormal hyperintensity compatible with right petrous apicitis. Gradenigo syndrome secondary to otitis diagnosis was made. An empirically-guided intravenous antibiotic was started and a myringotomy tube was placed by the ENT team. The culture of the middle ear revealed a *Streptococcus pneumoniae* infection. The patient was discharged after 14 days with an improvement in neurological symptoms.

Conclusion: Gradenigo syndrome is a cause of sixth nerve palsy. The syndrome is classically described after suppurative otitis media, but it has already been described after complications of trauma and skull-base metastases. Nowadays, it is more uncommon due to early and widespread treatment with antibiotics to avoid complications of otitis media.

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590 – Case report: motor neurone symptoms as initial clinical manifestation in a patient with Creutzfeldt-Jakob disease

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Female, 54-year-old, began with weakness in her right hand and loss of dexterity. Four months after initial symptoms, patient developed sudden onset vertigo. She progressed with difficulties in coordination, dystonic posture and involuntary movements in her right arm. Family also reported insomnia and cognitive impairment. Ten days later, she had no recognition of family members, and developed myoclonia. After two weeks, progression of ataxia rendered her unable to walk and she also had urinary incontinence. Neurological examination revealed perseveration of speech, oculomotor apraxia, severe gait and limb ataxia, intention tremors and generalized hiperrreflexia. She had marked tenar, first dorsal and palmar interosseus amyotrophy. Video-EEG showed markedly disorganized activity and generalized periodic complexes. Neuroimaging had no signs of spinal cord suffering. Electroneuromyography showed preganglionic chronic neurogenic involvement in cervical and lumbosacral myotomes, and acute denervation bilateral of C5–C7. Cerebrospinal fluid examination showed discrete hyperproteinorraquia and positivity for protein research 14-3-3. Genetic testing showed M129M variant in the PRNP gene. Brain tissue neuropathology revealed histologic spongiform encephalopathy associated with reactional gliosis and neuronal loss. Creutzfeldt-Jakob disease is a rare neurodegenerative disease caused by prion propagation. It has been mainly associated with central nervous system involvement, but lower neuron signs have been described. We report a case of motor neurone symptoms as initial clinical manifestation of Creutzfeldt-Jakob disease.

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593 – Correlation between neurological deficit at admission and functionality at discharge of patients from a clinical hospital stroke program

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Introduction: Stoke is one of the main causes of disability in Brazil, with an average hospital stay (HS) of 11 days and prolonged HS can generate functional impairment to patients.

Objectives: To correlate neurological deficit at admission with functionality and days of HS.

Methods: Patients were selected from the clinical program of post-stroke care, from March to December 2022 with HS time greater than or equal to 4 days. They were divided into 2 groups: Group 1, between 4 and 11 days of HS and group 2, greater than 11 days of HS. The Functional Independence Measure (FIM) and the National Institute of Health Stroke Scale (NIHSS) were applied at admission and discharge. Data analysis was performed with Microsoft Excel© 2016 with descriptive statistics, Pearson's Correlation Test was also performed, considering $P < 0.05$ for the variables studied.

Results: 86 subjects were included. 41 were excluded due to lack of informations in medical records, and 45 patients were analyzed. Group 1: Included 32 patients, 24 with ischemic stroke (IS), 2 hemorrhagic stroke (ICH) and 6 transient ischemic attack (TIA), 56.2% were female and 69 ± 15.2 years and 5.9 ± 15.9 days HS with 5.2 ± 4.8 points in the FIM/days of hospitalization and NIHSS admission of 2.9 ± 4.2. Group 2: 13 patients, 11 with IS, 2 ICH, 61.5% male, with 67±15.2 years, NIHSS admission of 7.3 ± 4.2, there was a gain of 1.14 ± 4.7 points in the FIM/days of hospitalization and 21.3 ± 15.7 days HS. There was a negative correlation between Days of Hospitalization and FIM/days of hospitalization ($r = -0.3$), Days of Hospitalization and FIM at Discharge ($r = -0.57$) and NIHSS admission and FIM of discharge ($r = -0.6$).

Conclusion: We observed a correlation between neurological deficit at admission, functional outcome at discharge and time of HS related to systematized assistance to a clinical stroke program.

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594 – Multifocal motor neuropathy atypical presentation: case report

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Case report: Woman, 34 years old, with paresis that started four years ago during pregnancy, predominantly distal in the lower limbs and progression to the upper limbs. No involvement of cranial nerves and sensitivity. Initially consid-

ered a diagnosis of Myasthenia Gravis and clinical gain was observed with the use of pyridostigmine. After a few months, the patient's symptoms recurred with asymmetric flaccid tetraparesis, predominantly in the lower limbs, associated with global areflexia and oral immunosuppressive therapy was initiated, with improvement. Electroneuromyography with normal sensory nerve conduction studies, but reductions in amplitudes in proximal compound muscle action potentials, with markedly reduced persistence in F-wave studies of the four limbs. Diffuse neurogenic changes were observed on exertion and activities such as positive sharp waves, fibrillation, fasciculations and myokymia at rest. Repetitive nerve stimulation at 3Hz without changes. A hypothesis of multifocal motor neuropathy (MMN) was then made. Treatment with intravenous immunoglobulin was performed, with significant recovery of symptoms.

Discussion: MMN is a rare disease (prevalence of 0.6 per 100,000 individuals), with a predominance in men and a mean age of onset around 40 years. The differential diagnosis includes motor neuron disease and other demyelinating neuropathies. It is immune-mediated by antianglioside antibodies (anti-GM1), but they are not identified in all patients and may be present in other neuropathies. It is defined by muscle weakness predominantly distal, asymmetrical, predominantly in the upper limbs, slowly progressive, associated with reduced deep reflexes in the affected regions. The main electrophysiological characteristic is the presence of motor nerve conduction blocks (CB) outside the usual sites of compression.

Conclusion: The finding that CB presents in patients with MMN suggests that nerve conduction should be extensively studied in every patient with a lower-motor-neuron syndrome to identify patients who might respond favorably to immunomodulating treatment.

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598 – Prolactinoma compromising visual acuity and causing adrenal insufficiency in a young male patient: case report and literature review

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This article reports on a case study of a 27-year-old male patient with prolactinoma, a pituitary adenoma that secretes prolactin, leading to a loss of visual acuity. The data were obtained from the patient's clinical history, as well as complementary exams and procedures. The article emphasizes the importance of correct and early diagnosis, particularly in young patients, as the patient in this case had a relevant family history of brain tumors and experienced a year of symptoms before diagnosis. The patient was diagnosed with prolactinoma after a canon magnetic resonance imaging revealed a macroadenoma pressing on the optic chiasm, causing visual complaints, weight gain, milk production, and erectile dysfunction. The patient's serum prolactin level was higher than 200 ng/mL, and he began treatment with Cabergoline after discovering adrenal insufficiency. While the patient reported an improvement in symptoms after two months of medication, his vision did not return. The report highlights the importance of proper screening and thorough evaluation, including atypical patients who may not fit the typical epidemiology of the disease. It took a year of research in the ophthalmological area to realize that the patient's symptoms were neurological, and this delay in diagnosis may have allowed complete blindness in the left eye and progression with mild symptoms in the right eye. However, proper treatment led to significant improvement in symptoms. In conclusion, the article emphasizes the importance of early and assertive diagnosis, particularly in young patients, whose complaints are often neglected. The case study illustrates the consequences of delayed diagnosis and the need for a complete anamnesis and correct screening.

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602 – Sleep disorders and degree of disability in patients with multiple sclerosis

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Introduction: Multiple sclerosis (MS) is a chronic disease that manifests through neuronal inflammation and demyelination. Patients exhibit symptoms that can reduce their physical and mental well-being, including worsened sleep quality.

Objectives: To verify if there is a relationship between the level of Expanded Disability Status Scale (EDSS) and sleep quality in patients with MS.

Methods: This is an observational, field and documentary study with a cross-sectional, descriptive, and quantitative approach. Sociodemographic parameters were evaluated, as well as the relationship between chronic use of Interferon Beta 1a and sleep disorders assessed by the Pittsburgh Sleep Quality Index (PSQI) and the Epworth Sleepiness Scale (ESS).

Results: A hundred and twenty nine people were observed, subjected to inclusion and exclusion criteria, and 90 patients were selected. After analysis, a predominance of women (78.78%) was noted. The most commonly used drug was Interferon Beta 1a, which showed excessive daytime sleepiness in 5% of patients when compared to the control group. When sleep pattern was investigated through PSQI, the score was 9.1, with a prevalence of poor sleep reaching 67.78%. No correlation was observed between EDSS and poor sleep quality, with EDSS values ranging from 0 to 6, totaling an average of 2.4. In addition, EDSS was observed in 21% of patients, but a negative relationship between EDSS and ESS was found.

Conclusion: The prevalence of poor sleep and excessive daytime sleepiness in the studied population was evidenced. The research was limited by the target audience with recurrent-intermittent MS, which may explain the lack of correlation between variables. This opens up space for possible investigations of other progressive forms of MS and their relationship with sleep disorders.

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603 – Acute cerebellar ataxia due to Dengue fever: case presentation and four-year follow-up

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Case presentation: A 70-year-old female was admitted due to imbalance associated to high fever, headache, photophobia, myalgia, and emesis for

5 days. She denied other symptoms or predisposing agents. At examination, there was a global asymmetric cerebellar syndrome, without other findings. Laboratory work-up showed lymphocytosis, leucopenia, thrombocytopenia, and increased liver enzymes. Serologic exams were negative for syphilis, HIV, hepatitis B and C viruses, 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. During hospital stay, there was complete recovery of 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. First evaluation was 54 days after onset; she had a mild subjective imbalance complaint, with normal neurological exam. She has been followed for four years since onset, remaining asymptomatic, without any cerebellar signs.

Discussion: Neurotropism in Dengue fever, as direct viral or immunomediated damage, has been well recognized. Its spectrum comprises many presentations; however, association with cerebellar syndrome is extremely rare, with only seven other cases published. Acute cerebellar ataxia is characterized by normal neuroimaging and complete recovery up to three years. In Dengue cases, recovery happened from one week to two months. Currently, there are no other reports with long-term follow-up.

Conclusion: Dengue virus is a rare, but probably underdiagnosed, cause of acute cerebellar ataxia. Recovery is fast, and, in our case, persistent, without recurrence or evolution to any degenerative disease.

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605 – Cerebral cryptococcoma in immunocompetent patients: a series of four cases

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Introduction: Cryptococcoma is a rare CNS infection caused by Cryptococcus neoformans, with a granulomatous lesion. It's even rarer in immunocompetent patients. Diagnosis requires histopathological analysis and treatment with amphotericin B, often with surgical resection. The aim is to report four cases of cerebral cryptococcoma In Immunocompetent Patients. This is a case report study. The information of this work was obtained through review of the medical record.

Case report: We report on four cases of cerebral cryptococcoma in immunocompetent patients. All cases presented with images of expansive lesions and compatible histopathological analysis. The first case was a 62-year-old man with dysphasia and decreased strength on the right side, and underwent total surgical resection, followed by 23 days of amphotericin B treatment. The second case was a 62-year-old woman who presented with orofacial dyskinesia and decreased strength on the right side, and underwent stereotactic surgery, followed by antifungal treatment. After 10 days of hospitalization, the patient developed respiratory insufficiency, progressed to cardiorespiratory arrest and brain death. The third case was a 54-year-old woman who presented with a skin infection caused by Cryptococcus. During treatment with fluconazole, the patient experienced altered sensory perception and left hemiparesis. Surgical resection of the lesion was performed, followed by 30 days of antifungal treatment. The fourth case was a 54-year-old man who presented with pulsatile headache and right hemiparesthesia, and underwent stereotactic neurosurgery, followed by 12 days of amphotericin B treatment.

Conclusion: Cryptococcus neoformans infection in the CNS, manifested in the form of cryptococcoma, although rare, should be considered as a differential diagnosis for brain expansive lesions in immunocompetent individuals.

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607 – Multiple sclerosis associated with adalimumab treatment in psoriatic arthritis: a case report

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Introduction: Tumor necrosis factor (TNF) antagonists have emerged as one of the most effective treatment options for patients with autoimmune diseases, including psoriatic arthritis. However, these agents carry a risk of severe adverse effects, including demyelinating disorders. The aim to report a case of multiple sclerosis associated with the use of adalimumab. This is a case report study. The information of this work was obtained through review of the medical record.

Case report: A 40-year-old female with psoriatic arthritis on adalimumab (ADM) treatment. She developed paresthesia in her lower limbs, which was initially attributed to anxiety. Six months later, she presented with decreased visual acuity in her left eye, which was diagnosed as optic neuritis. She received pulse therapy with methylprednisolone and underwent investigation for multiple sclerosis (MS). Magnetic resonance imaging (MRI) of the brain revealed multiple lesions with high signal intensity in T2/FLAIR on periventricular regions bilaterally. Furthermore, MRI of the thoracic spine showed high signal intensity on posterolateral cords at levels T5 to T9. These findings fulfilled the revised McDonald Criteria for MS and suggested a link to ADM treatment. On neurological examination, mild left lower limb paresis, bilateral plantar-cutaneous reflex, hyperreflexia in the lower limbs, and mild hypopalesthesia in her right leg. ADM treatment was discontinued, and although she reported a slight worsening of her skin condition, there were no further signs of psoriatic arthritis.

Conclusion: Neurological adverse events associated with TNF antagonists are rare but must be considered. If neurological disorders develop during therapy, prompt discontinuation of the drug is recommended.

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608 – Melatonin: a safe and effective treatment for rem sleep disorder in an elderly patient - a case report

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Introduction: REM sleep disorder is a parasomnia characterized by the loss of atonia during REM sleep, resulting in dream enactment behavior. It is com-

monly associated with dementia with Lewy bodies (DLB) and is prevalent in up to 90% of individuals with this condition. Treatment for REM sleep disorder typically involves a combination of antipsychotics and melatonin, which can regulate sleep and reduce symptoms. The aim is to report a case of using melatonin for REM sleep disorder in an elderly patient. This is a case report study. The information of this work was obtained through review of the medical record.

Case report: A 87-years-old female with DLB. After 36 months of dementia onset, the patient experienced dysregulation of the sleep-wake cycle, with nocturnal awakenings, visual hallucinations, and vocalizations and complex motor behaviors during REM sleep. The patient was initially prescribed olanzapine 5 mg once a day, which led to three episodes of dysregulation per week. Clonazepam 0.5 mg was added to the treatment, in combination with environmental measures and maintenance treatment for DLB, resulting in two episodes per week. After three months, clonazepam was replaced by melatonin 3 mg, which led to a reduction in episodes to five per month. The dosage of melatonin was later increased to 6 mg, which reduced the frequency of dysregulation to three episodes per month. Olanzapine was eventually replaced by quetiapine 25 mg once daily, leading to only one episode per month.

Conclusion: Melatonin was found to be effective in reducing agitation and other symptoms without causing any side effects in the patient. It is important for healthcare professionals to consider the use of melatonin in the treatment of REM sleep disorder, particularly in elderly patients.

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611 – History of neurology in São Paulo at the first half of the 20th century: a systematic review

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Neurology in São Paulo was initiated at the beginning of the XX century, with the creation of the Paulista School of Medicine. This article aims to analyze the emergence of Neurology in the state of São Paulo and the influences it received. To this end, we conducted a systematic search in an electronic database, Scielo. In this context, in the 20th century, neurology studied in São Paulo was greatly influenced by the science practiced in France, since in 1882 the specialty was initiated at the University of Paris. Discussions about the diseases in this area were discussed at the Santa Casa de Misericórdia in São Paulo. Another important place for the development of neurology in São Paulo was the creation of the neuropathology and cerebrospinal fluid laboratory, in 1924, at the Hospital de Juqueri. In the 1920s, the teaching of clinical neurology was created at the Faculty of Medicine by Enjolras Vampré, known as the father of neurology in São Paulo. In the 1930s, there were major changes in the areas of brain pathology, diagnosis of diseases that affect the central nervous system, and therapy. All of this was used at Hospital de Juqueri and Faculdade Paulista de Medicina. Among the prominent methods used were pneumoencephalography, cerebral arteriography, and cerebral leucotomy. Some literary works were fundamental for the dissemination of knowledge of Neurology in the society of São Paulo, namely "Memories of the Hospital of Juqueri" from 1924 and "Archives of Assistance to Psychopaths" from 1936. Neurology underwent a major evolution at the end of the first half of the twentieth century, however, it became expensive and inaccessible to a large part of the population, especially at a time before the creation of the Unified Health System, making the poorest marginalized concerning the treatment of neurological diseases that affected the society of São Paulo at the time.

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613 – Primary coenzyme Q10 (COQ10) deficiency: clinical presentation of a new variant in COQ7 gene

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Cases reports: Two females from different families, 34 and 16 years old, who started at puberty with distal weakness in lower limbs and difficulty in walking. The youngest had a history of parental consanguinity. The oldest also developed symptoms of cerebellar ataxia. Both patients had joint hypermobility. After physical exercise, both showed increased serum levels of creatine phosphokinase, but only one of them showed increased lactate. Both electroneuromyography showed motor neuropathy, predominantly in lower limbs, with axonal and demyelinating pathophysiology, with probable superimposed pre-ganglionic involvement. Both genetic tests showed homozygous pathogenic variation in *COQ7* gene, described as Chr16:19.067.667, which leads to methionine substitution and impaired protein traduction.

Discussion: Primary COQ10 deficiency is a heterogeneous group of mitochondrial disorders caused by defects in proteins involved in COQ10 biosynthesis. It's inheritance usually is autosomal recessive. Mutations in 10 genes directly involved in coenzyme Q10 synthesis and collectively named "*COQ* genes" have been identified. Clinical spectrum may overlap encephalomyopathies, ataxia, neuromyopathy, spastic paraplegia and even impairment of another organs. Only four COQ7 deficiency patients have been reported so far. In addition, cases reported here are related with a new variation in *COQ7* gene. All reported COQ7 deficiency patients have asian ancestry, which is not the case of patients related here. Some improvement can occur by COQ10 supplementation, which was initiated in both cases.

Conclusion: The diagnosis of primary COQ10 deficiency is limited by factors like rarity, heterogeneous phenotypes and unavailability of genetic testes, which favors under or misdiagnosis. Discovery of new cases and mutations can increase our knowledge about this condition, make possible the diagnosis, consequently look for dysfunction of other organs and try a specific treatment.

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614 – Association between risk of obstructive sleep apnea and stroke recurrence: partial results of a cohort

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Introduction: Obstructive sleep apnea (OSA) is a disease of upper airways,diagnosed with polisomnography,that is well associated with stroke through diverse mechanisms.Despite that,it still lacks studies about its role in prognosis of stroke,especially in stroke recurrence.

Objectives and methods: This study aims to elucidate the possible association between stroke recurrence and risk of OSA.We performed a cohort. Patients were evaluated of risk of OSA with STOP-Bang and SOS scores and

remotely evaluated with Rankin, Barthel, questioned about stroke recurrence and another hospitalization during 3.6 and 12 months. We performed univariate analysis (T test,Mann-Whitney or chi-square) and a multivariate analysis with logistic regression (considering $P < 0.05$).

Results: We obtained 190 patients: 7 patients already finished the study and 78 started. 48.9% female, mean age 63.6 ± 6.43 years, mean body mass index 25.69 ± 5.64 kg/m^2, mean SOS 11.65 ± 6.43, mean STOP-Bang 3.22 ± 1.45. Rankin in 3 months mean 2.89 ± 1.81; 6 months mean 2.67 ± 1.89; 12 months mean 5 ± 1.95. Barthel 3 months 62.51 ± 33.44; 6 months 72.40 ± 34.78; 12 months mean 35.40 ± 46.59. 3.7% were hospitalized due to cardiovascular causes; 2.1% due to infection causes and 4.7% due to other causes. 2.6% evolved to death and 1.6% recurred with stroke. Considering STOP-Bang, we had in univariate analysis Rankin 6 months ($P = 0.06$), Barthel 12 months ($P = 0.07$) and hospitalization of other causes ($P = 0.1$). Considering SOS, we obtained stroke recurrence, Rankin 12 months ($P = 0.1$) and death ($P = 0.03$). In multivariate analysis, to STOP-Bang, we had Barthel 12 months ($P = 0.03$); to SOS, it was not possible to perform due to reduced number of stroke recurrence and death.

Conclusion: Preliminary results already showed association between OSA and recurrence stroke, which can lead us to a differential approach. Considering the most successful research about this theme obtained 1.2% of recurrence, the study is on the way to elucidate this question.

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615 – Atypical presentation of progressive multifocal leukoencephalopathy in a patient with ataxic syndrome and retrovirus with infra-tentorial involvement: a case report

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Introduction: Progressive Multifocal Leukoencephalopathy (PML) is a rare demyelinating disease of the central nervous system caused by reactivation of the JC virus. It usually occurs in immunocompromised patients. The majority of PML cases present with supratentorial involvement. In contrast, infratentorial involvement is exceedingly rare in PML. The aim to report an atypical presentation of PML in a patient with ataxic syndrome and retrovirus with infra-tentorial involvement. This is a case report study. The information of this work was obtained through review of the medical record.

Case report: A 38-year-old, female, HIV-infected since 2015 and started antiretroviral therapy only in 2018. In June 2019, she developed paresthesia in lower limbs, imbalance and difficulty walking, requiring support. Two months later she evolved with dysphagia, dysarthria, and limitation in activities of daily living. In September 2019, she presented for evaluation in a wheelchair. On neurological examination, she had bi-directional horizontal nystagmus, cervical dystonic tremor, global ataxia, altered deep sensation and spastic gait. Her metabolic profile was normal. Brain magnetic resonance imaging (MRI) showed atrophy of the cerebellar leaflets and vermis associated with extensive T2/FLAIR inversion recovery hyperintensities involving the cerebellum and bilateral middle cerebellar peduncle. Cerebrospinal fluid analysis revealed hyperproteinorraquia and posterior examination revealed positive JC virus DNA.

Conclusion: In the diagnosis approach of acquired ataxias, it is essential to establish a propaedeutic directed to the patient's clinical context. Investigating opportunistic infectious diseases is essential in care and treatment especially for immunosuppressed patients.

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616 – Impact of obstructive sleep apnea in imediate outcomes of stroke patients: a transversal study

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Introduction: Obstructive sleep apnea (OSA) is a disease of upper airways, diagnosed with polisomnography, that is well associated with stroke through diverse mechanisms. Despite this, it still is not clear about the impact of this disease in the prognosis of the patients, especially in immediate outcomes.

Objectives: This study aims to elucidate the impact of high risk of obstructive sleep apnea in immediate outcomes of stroke patients.

Methods: We performed a transversal study, part of a cohort of stroke patients. Patients were evaluated of risk of OSA with STOP-BANG and SOS scores. We compared hospitalization time, CTI time, hemorragic transformation, type of hemorragic transformation, infection, delirium, new stroke and death during hospitalization between high/moderate risk and low risk of OSA using T test, Mann-Whitney or chi-square.

Results: We obtained 190 patients. Considering STOP-Bang, we obtained significance with delirium ($P < 0.001$), infection ($P < 0.001$) and death during hospitalization ($P = 0.04$). Considering SOS score, we obtained CTI time ($P < 0.001$) and death during internment ($P < 0.001$).

Conclusion: The important role of OSA in death during internment was clear with the results of this study. Other outcomes that can lead to death or increase the risk of death, such as the infection itself and CTI time, were essential. Thus, it is imperative that we need to be aware of these questions, in order to reduce CTI time in adequate condition, so we can have better outcomes in this type of patient.

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617 – Laboratory markers of hemorragic transformation and mortality in patients with stroke: a transversal study

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Introduction: Hemorrhagic transformation is an important cause of death after stroke and/or thrombolysis. Its relevance is clear, but lack studies about the predictors of this event,in order to prevent it.

Objectives: This study aims to describe laboratory predictors of hemorrhagic transformation and mortality in stroke patients.

Methods: It is a transversal study, part of a cohort of stroke patients, and collected data about hemorrhagic transformation and its type (ECASS), death, thrombolysis, admission INR, platelets, creatinine clearance, partially activated thromboplastin time, admission and highest creatinine and urea. We performed an univariate analysis with T test or Mann-Whitney and a multivariate analysis with logistic regression (considering $P \leq 0.05$).

Results: We obtained 190 patients, of which 17.4% evolved to hemorrhagic transformation and 6.3% evolved to death. Both outcomes showed positive association with each other and weak correlation ($P < 0.001$; $r = 0.02$); death and hemorrhagic type showed no correlation. INR ($P = 0.01$), highest ($P = 0.05$) and last ($P = 0.04$) creatinine were significant in univariate analysis; INR ($P = 0.007$) and last creatinine ($P = 0.004$) were independently associated with hemorrhagic transformation. Platelets ($P = 0.1$), highest urea ($P < 0.001$) and creatinine ($P = 0.008$), admission urea ($P = 0.01$) and creatinine ($P = 0.05$), last creatinine and urea and clearance ($P < 0.001$) were significant in comparison to death; last urea and clearance ($P = 0.03$) were significant in multivariate analysis.

Conclusion: INR are essential parameters to be aware of, which lead us to a unique monitoring of patients with altered INR. Renal conditions showed importance,so it is also a laboratory marker and a modifiable factor to be careful and have an adequate management. The correlation between both outcomes encourages the best assistance to those who evolved with hemorrhagic transformation, independent of its type — it is mandatory, necessary and can lead to outcomes other than death.

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618 – Impact of previous comorbidities in stroke patients with high risk of obstructive sleep apnea: a transversal study

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Introduction: Obstructive sleep apnea (OSA) is a disease of upper airways well associated in literature with stroke through diverse mechanisms. Despite that, it still is not clear the impact of the regular diseases in the context of high risk of OSA and stroke.

Objectives: This study aims to elucidate the impact of previous comorbidities in imediate outcomes (hospitalization time, CTI time, hemorragic transformation,death, infection and delirium during hospitalization) of patients with high/moderate risk of OSA and stroke. **Methods:** We performed a transversal study, part of a cohort of stroke patients. Patients were evaluated of risk of OSA with STOP-BANG and SOS scores. Data of previous comorbidities (obesity, alcoholism, smoking, hypertension, diabetes, renal disfunction, dyslipidemia, asthma, atrial fibrillation – AF, heart failure, previous Rankin and sleep disorder) were analysed with T test or Mann-Whitney and they were submitted to multivariate analysis with logistic regression (considering $P < 0.05$).

Results: We obtained 118 patients (hospitalization time 15.41 ± 15.16; CTI time 1.87 ± 5.02). Considering hospitalization time, we had previous Rankin ($P = 0.1$), AF ($P = 0.1$) and dyslipidemia ($P = 0.07$); to CTI time, we had dyslipidemia ($P = 0.04$); to hemorragic transformation, we had asthma ($P = 0.03$) and smoking ($P = 0.05$); to death, we had alcoholism ($P = 0.1$) and renal insufficiency ($P = 0.07$). To infection, we had previous Rankin ($P = 0.002$), diabetes ($P = 0.009$) and renal insufficiency ($P < 0.001$); to delirium, we had previous Rankin ($P = 0.007$), obesity and sleep disorder ($P = 0.07$), diabetes ($P = 0.1$) and renal insufficiency ($P < 0.001$). In multivariate analysis, renal disfunction to infection were significant ($P = 0.02$; odds ratio = 2.34 confidence interval 1.70-3.23).

Conclusion: Renal disfunction appeared to be important in most of outcomes, and the only significant in multivariate analysis.Conditions that affect metabolism showed importance, that lead us to better control renal and metabolic disorders.

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619 – Schistosomal myeloradiculopathy: a case report of a disease that still exists

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Introduction: Schistosomiasis affected 164 individuals during 2022, of which 56% were from Northeast region, being a disease of public concern. One of atypical presentations is the myeloradiculopathy, that can evolve — if without adequate treatment and eradication — with incapacity.

Case report: 44-years-old patient, with no previous comorbidities, artisanal fisherman, positive epidemiology for schistosomiasis. Admitted in emergency with backache (lumbar and sacral regions), evolving with paraparesy of lower members associated to progressive (two months) fecal and urinary hesitation. Physical examination showed an alert patient, oriented in time and space, no alteration in cranial nerves and coordenation exams. Flacid paraparesy in lower limbs (muscular force III/V), fasciculations and hypoesthesia in L3/L4 myotomes. Positive Lasegue, cremasteric reflex absent. Metabolic, inflammatory and sorologies negatives. Oriented to proceed with pulsotherapy and methylprednisolone (1 g) during five day and, after, praziquantel 50 mg/kg twice a day, during three days, and after, prednisolone 60 mg/day. Two months later, patient returned with positive sorology to schistosomiasis (1:128) and evolved with partial improvement of paraparesy (muscular force IV/V) and fecal/urinary hesitation, although maintained hypoesthesia (L3/L4) and lumbar pain. Considering the persistence of symptoms, a new cicle of praziquantel and corticoid was done, just as motor physiotherapy and eradication actions.

Conclusion: Antihelmintic treatment associated with corticoid use is still an adequate strategy to this disease, but it is a problem of public health, demanding major measures to its erradication.

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621 – Renal predictors of mortality in stroke patients: partial results of a transversal study

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Introduction: Renal factors are known as important, and a controllable factor, to define prognosis,mortality and hospitalization time in most of diseases. It still lacks studies that clarify in a specific way the role of this factors.

Objectives: This study aims to elucidate the impact of renal factor in outcomes of stroke patients (hospitalization time, CTI time, delirium, infection and death during internment).

Methods: We performed a transversal study, part of a cohort. Data of previous renal insufficiency,admission,highest and last creatinine and urea and the highest creatinine clearance during hospitalization. We performed univariate analysis, with T test, Mann-Whitney or chi-square tests;after, multivariate analysis with logistic regression (considering $P < 0.05$).

Results: We obtained 190 patients. Admission creatinine was significant to death ($P = 0.05$), delirium ($P = 0.07$) and infection ($P = 0.06$); highest creatinine to hospitalization time, delirium, infection ($P < 0.001$) and death ($P = 0.008$);and last creatinine to death ($P < 0.001$), delirium ($P = 0.003$), infection ($P = 0.01$) and CTI time ($P = 0.1$). Previous renal insufficiency showed significance to death ($P = 0.03$), delirium and infection ($P < 0.001$). Highest urea showed significance to all outcomes ($P < 0.001$), including CTI time ($P = 0.004$); admission urea to death ($P = 0.01$); and last urea to death ($P < 0.001$), hospitalization time ($P = 0.1$) and delirium ($P = 0.06$). Clearance only did not show significance to CTI time.Multivariate analysis to each outcome obtained: last creatinine to death ($P = 0.03$); highest creatinine to hospitalization time ($P = 0.02$); admission ($P = 0.01$), highest ($P = 0.01$) creatinine, admission urea ($P = 0.03$) and clearance ($P = 0.01$) to delirium; renal insufficiency ($P = 0.009$), admission ($P = 0.007$), highest ($P = 0.002$) and last ($P = 0.02$) creatinine, highest urea ($P = 0.005$) and clearance ($P = 0.003$). None was significant to CTI time.

Conclusion: All parameters of renal function showed to be important measures of prognosis in stroke patients; thus, it is a factor we need to be aware of during hospitalization time.

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622 – Congenital myasthenic syndrome due rapsyn mutation presenting with predominant ocular symptoms and good therapeutic response with salbutamol

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Introduction: Rapsyn mutations are found in Congenital Myasthenic Syndromes (CMS), usually described with early onset, hypotonia and respiratory insufficiency. Late onset phenotypes had already been described, with mild limb weakness and semiptosis. Episodic respiratory symptons and lack of ophthalmoparesis are considered hallmarks of rapsyn – CMS. The aim is to describe an unusual phenotype of a commom mutation of Rapsyn CMS and therapeutic response with salbutamol.

Case report: A 29-year-old female patient was referred complaining of diplopia and fatigable assymmetrical semiptosis during the last six years. She pratices physical exercise, with no fatigue or weakness in limbs. Her past medical history was unremarkable except for drop neck episodes triggered by viral infectons during childhood, with first episode taking place around two years of age. Her parents were consaguinous, howeve no similar clinical picture was reported in family members. In clinical examination besides assymetrical semiptosis and ophthalmoparesis, was noted winged scapula, with normal muscle trofism and strength in four limbs. Laboratory investigation showed negative acetylcholine receptor antibodies (anti-AChR), and negative muscle-specific kinase antibodies (anti-MuSK). Single fiber electromyography revealed abnormal jitter. Genetic panel found a pathogenic homozygous mutation in the RAPSN gene (chr11:47.448.079G>T; p.Asn88Lys). She was treated with pyridostigmine and showed poor response. We opted to start salbutamol with marked clinical improvement.

Conclusion: Rapsyn-CMS may present as late episodic diplopia, with semiptosis and ophthalmoparesis, and no limb or respiratory muscle weakness. That diagnosis possibility should be also considered in patients with late onset ocular symptons, incomplete response to pyridostigmine and negative assays for auto antibodies.

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625 – Renal predictors of mortality in stroke patients with high risk of obstructive sleep apnea: partial results of a transversal study

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Introduction: Renal factors are known as important, and a modifiable,to define prognosis in most of diseases. Lately,the relation between stroke and obstructive sleep apnea is extensively studied,but the role of renal component in outcomes in the context are not yet elucidated.

Objectives: This study aims to elucidate the impact between patients with different risks of OSA of renal factor in outcomes (hospitalization and CTI time, hemorragic transformation, delirium, infection and death during internment).

Design: It is a transversal study, part of a cohort.

Methods: Patients were evaluated of risk of OSA with STOP-BANG and SOS scores. Data of previous renal insufficiency, admission, highest and last creatinine and urea and the highest creatinine clearance during hospitalization was collected. We performed univariate analysis, with T test/Mann-Whitney/chi-square tests; after, multivariate analysis with logistic regression (considering $P < 0.05$).

Results: We obtained 118 patients. Clearance was significant to delirium,infection ($P < 0.001$), death ($P = 0.004$) and hemorragic transformation ($P = 0.01$); renal insufficiency to delirium,infection ($P < 0.001$) and death ($P = 0.09$). To hemorragic transformation, we obtained significance with admission ($P = 0.07$) and highest ($P = 0.007$) creatinine. To death, all the variables were significant. To delirium, only admission urea did not showed significance ($P = 0.18$). To infection, we obtained all creatinine parameters, clearance, highest and last urea; to hospitalization time, we obtained highest creatinine ($P = 0.008$) and urea ($P < 0.001$); to CTI time, highest urea ($P = 0.02$). In multivariate analysis, last creatinine was associated to death ($P = 0.04$); highest urea to hospitalization time ($P = 0.02$); highest creatinine to delirium ($P = 0.04$); to infection, admission ($P = 0.01$) and highest ($P = 0.008$) creatinine, clearance ($P = 0.01$) and renal insufficiency ($P = 0.01$).

Conclusion: The role of renal parameters in this type of stroke patient is an important measure to better control outcomes.

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628 – Charcot-Marie-Tooth type 1F with bi-allelic NEFL mutation

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Introduction: Charcot-Marie-Tooth disease (CMT) is the most common hereditary neuropathy, with a diverse phenotypic and genotypic spectrum. The main clinical features are onset during infancy, slowly progressing symptoms and foot deformities, especially if there is a positive family history, although the lack of family awareness can be present. Over 70 distinct genes have been associated, however, their genetic diagnosis can be challenging, especially if we consider the fact that the same gene can transmit disease either dominantly or recessively. The aim to describe a case of CMT1F as a rare case of recessive demyelinating hereditary neuropathy.

Clinical case: A 25-year-old woman, born of consanguineous parents, had history of distal weakness and burning sensation in lower limbs, with onset in infancy. Her childhood was marked for abnormal gait and falls, and evolved with foot deformities, requiring surgical corrections. The symptoms progressed slowly and reached upper limbs in few years. On physical evaluation was noted: muscle weakness of upper and lower limbs, predominantly distal, associated with atrophy, foot drop and absent reflexes. Electroneuromyography demonstrated signs of chronic demyelinating polyneuropathy. An initial sequencing analysis of the *PMP22* gene was indicated, with normal results. A panel for neuropathies was performed, showing a homozygous frameshift mutation in *NEFL* (p.Lys362Glufs*2; c.1084_1085delAA), classified as probably pathogenic variant.

Conclusion: CMT due to bi-allelic *NEFL* mutations is a rare condition that should be considered in hereditary demyelinating neuropathy, especially when recessive inheritance is suspected. Our study illustrates this condition and brings attention to the importance of the disponibility of high throughput genetic tests.

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630 – Cerebral tuberculoma as a differential diagnosis for extra-axial lesions in immunocompetent patient: case report

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Introduction: Cerebral tuberculomas are a rare and serious form of tuberculosis, representing 0.2% of intracranial expansive processes. In the absence of conclusive diagnostic tests, doctors often make treatment decisions based on clinical judgment, cerebrospinal fluid (CSF) profile, and radiological findings. Magnetic resonance imaging (MRI) commonly shows discrete lesions with ring enhancement, with perilesional edema, being a relevant differential diagnosis in ring lesions, as their clinical and imaging can mimic brain tumors. The aim to report a case of cerebral tuberculoma in an immunocompetent patient. This is a case report study. The information of this work was obtained through review of the medical record.

Case report: G.L.F, female, 26 years old, immunocompetent, with a history of generalized tonic-clonic seizure. MRI showed a right temporal extra-axial lesion, with heterogeneous contrast enhancement and adjacent dural thickening, associated with vasogenic edema. CSF was collected for investigation of inflammatory disease, without any significant findings. Excisional biopsy for anatomopathological analysis revealed an epithelioid granulomatous process (complete tuberculoid granulomas) with central caseous necrosis. Based on clinical, laboratory, and imaging data correlation, the conclusion was reached that it was a cerebral tuberculoma. Treatment followed the standard tuberculosis regimen for one year.

Conclusion: *Mycobacterium tuberculosis* infection in the central nervous system, manifested as tuberculoma, although extremely rare, should be considered as a differential diagnosis for expansive brain lesions, especially in regions with a higher prevalence of the disease.

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631 – Botulism in the southeast region of Brazil: an epidemiological analysis from 2007 to 2021

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Introduction: Botulism is a rare, serious neurological disease caused by a toxin produced by Clostridium botulinum. Although rare, it has a high lethality rate, reaching up to 15%. The Southeast region of Brazil is the most populous and may have more cases. Epidemiological analysis is essential to understand its occurrence, identify risk groups, and adopt preventive measures.

Objectives: To perform an epidemiological analysis of botulism in the Southeast of Brazil between 2007 and 2021, in order to evaluate its incidence, geographic and temporal distribution.

Methods: Data on botulism cases in the Southeast region of Brazil were obtained from the Brazilian Ministry of Health's Notifiable Diseases Information System (SINAN). The study included confirmed cases between 2007 and 2021 and analyzed the number of cases per year, gender, age range, and mode of transmission.

Results: 41 cases of botulism were reported, with an average of 2.73 cases per year. The year 2009 presented the highest number of cases (n = 6), followed by 2007 and 2008 (n = 5 in each year). The majority of cases occurred in males (n = 29, 70.7%) and the most affected age group was 30 to 59 years old (n = 21, 51.2%). The most frequent mode of transmission was foodborne (n = 39, 95.1%). No significant correlations were found between the number of cases and gender, age range, or mode of transmission.

Conclusion: Botulism in Southeast Brazil has the highest incidence in the country, with a variable temporal and geographic distribution. Foodborne transmission is the most common and predominantly affects young adult males. Health professionals must be vigilant for early diagnosis and treatment, and prevention measures must be intensified, particularly for proper food handling and storage.

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632 – Quality of life assessment in individuals with recurrent remitting multiple sclerosis at a referral center

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Introduction: Multiple sclerosis (MS) is a demyelinating and autoimmune disease that affects the central nervous system (CNS). It is the most common CNS disabling disease in young adults, predominantly in the 20-40 age group. Patients with MS have a compromised quality of life. The Short Form Health Survey 36 (SF-36) is the most widely used questionnaire to assess quality of life in patients with MS.

Objectives: To analyze the quality of life of patients diagnosed with relapsing-remitting multiple sclerosis (RRMS), in a Reference Center for Multiple Sclerosis in Paraíba.

Methods: This is a cross-sectional field work of quantitative approach, of descriptive and exploratory nature. The SF-36 questionnaire was applied in April 2021, at the Reference Center for Multiple Sclerosis. At the study site, 390 patients with Multiple Sclerosis were registered. For the research to present a confidence level of 90% and an expected error of 10%, a sample of 58 patients was determined. The statistical data were made by IBM SPSS statistics. The study was approved by the Research Ethics Committee of Joao Pessoa University Center.

Results: The data found in the sample are compatible with what is described in literature, reaffirming the predominance of MS for the female gender of 77%. As for the analysis of the patients' quality of life, the domains with the most impairment were limitation by emotional aspects, followed by vitality and limitation by physical aspects. The use of Betainterferone 1a was associated with a better quality of life in six of the eight domains studied.

Conclusion: Patients with RRMS have a decreased perception of Quality of Life. Other factors that affect the quality of life of these patients are gender and the use of betainterferone 1a.

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633 – Chronic paraneoplastic polyradiculoneuropathy during colorectal cancer: a case report

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A 70-year-old woman, four months before admission in our Hospital started with hypoesthesia in the hands, progressing to the lower limbs, followed by sensory ataxia and decreased strength, with frequent falls, losing 8 kg in that interim. The electroneuromyography demonstrated a pattern compatible with chronic demyelinating polyradiculoneuropathy (CIDP). Pulse therapy (PT) with Methylprednisolone was administered, with remission of symptoms. She underwent neoplastic investigation, being diagnosed with moderately differentiated and invasive sigmoid adenocarcinoma (SA), with indication for colectomy (CTC). No other solid tumors (ST) more commonly related to CIDP were found. Onconeural antibodies were not evidenced by blood analysis. 5 days after treatment, she has returned to her sensory-motor clinical condition, now attacking the bulbar muscles, developing dysphagia and dyspnea, requiring orotracheal intubation (OTI) and mechanical ventilation. Submitted to drug therapy with Intravenous Human Immunoglobulin and new PT for five days, with progressive improvement in strength, being extubated 11 days after OTI. As disease-modifying therapy, Rituximab was administered, one month before CTC. The association between SA and the development of CIDP was infrequently reported in the literature, lacking data that correlate the pathophysiology of this paraneoplastic (PNP) syndrome. Immunomodulatory therapy has an impact on the functional recovery of the patient and on preventing the progression of the symptoms, but the definitive treatment is CTC. Despite the low prevalence, the search for ST of the gastrointestinal tract should be included in PNP screening in the face of a recent diagnosis of CIDP, aiming at early identification and treatment in the initial stages, with an impact on oncological and neurological prognosis.

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634 – Partial involvement of the upper division of the oculomotor nerve of microvascular etiology: a case report

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A 70-year-old woman with uncontrolled hypertension woke up with a vertical binocular diplopia worsened when looking upward, without pain during ocular movements. After two days, she presented with left eyelid ptosis. She denied any history of trauma. At admission in hospital, was ruled out macular and/or retinal pathologies. Upon neurologic examination, she had restricted upward gaze in the left eye and eyelid ptosis, without pupillary involvement or other cranial nerve deficits, consistent with involvement of the upper division (UD) of the left third cranial nerve (CN III). The patient underwent extensive investigation for cranial nerve syndrome, including magnetic resonance imaging (MRI), cerebrospinal fluid analysis, and laboratory tests. All of these exams had normal results, and microvascular (MV) etiology was then considered. Ischemic involvement of the CN III often presents with limitation of extrinsic eye movement of all muscles innervated by this nerve. When the limitation is restricted to the UD of the CN III, the most common etiologies are related to extrinsic mechanical compression of the orbit, mainly due to neoplastic and infiltrative processes, in addition to skull base trauma. MV etiology due to involvement of the vasa-nervorum should be considered a diagnosis of exclusion in patients with high cardiovascular risk. In these cases, structural causes should be initially investigated with Orbital MRI and the image should be carefully evaluated for mechanical compression. After ruling out such etiology, MV disease responsible for the irrigation of the CN III should be considered, actively searching for risk factors such as hypertension and diabetes mellitus, which, if present, should be promptly controlled with lifestyle changes and appropriate medication treatment.

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635 – Assessment of the stess score as a prognostic tool in a Latin American hospital: a retrospective cohort study

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Introduction: Status epilepticus (SE) is a neurological emergency, with variable mortality according to several factors, such as etiology, age and type of seizure. It is currently unclear whether the aggressive therapeutic approach promotes better outcomes for all patients. In this context, the clinical score "Status Epilepticus Severity Score" (STESS), which considers four factors: age, history of seizures, type of seizure and impaired level of consciousness, is a tool with the potential to identify patients with a better prognosis and prevent aggressive treatments that can cause complications and worsen the outcome.

Objectives: Evaluate the use of the STESS score to predict the risk of in-hospital death in patients with status epilepticus (SE) at Santa Casa de Misericórdia de São Paulo.

Methods: Forty-six patients with SE evaluated at Santa Casa de Misericórdia de São Paulo, from March to December 2022, were included. Variables associated with mortality in previous studies were evaluated, such as age, etiology and type of crisis and, mainly the STESS score.

Results: Forty six patients, intra-hospital mortality was 28.3%, higher than that shown in previous studies. The STESS score showed a statistically significant relationship ($P < 0.05$) with the cutoffs ≤ 1, ≤ 2 and ≤ 3, but with a better sensitivity and specificity ratio with the cutoff ≤ 2, according to the receiver operating characteristic curve, justifying the use of this as the best predictor of in-hospital mortality, with a negative predictive value of 95% for death.

Conclusion: STESS is a useful tool to predict intra-hospital mortality, regardless of other risk factors, with potential to guide treatment and avoid aggressive therapy and its inherent risks in patients with a good prognosis.

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636 – NfL levels significantly decrease in response to treatment with patisiran or vutrisiran in hATTR amyloidosis with polyneuropathy

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Introduction: Neurofilament light chain (NfL) is a potential biomarker of disease progression and treatment response in patients with hereditary transthyretin-mediated (hATTR) amyloidosis with polyneuropathy (PN).

Objectives: To analyze NfL levels in patients from the APOLLO and HELIOS-A studies to further assess its potential utility in hATTR amyloidosis.

Methods: NfL plasma levels were measured at different time points in healthy controls and in a subset of patients from APOLLO or HELIOS-A.

Results: NfL levels at baseline were slightly higher in APOLLO (69.4 pg/mL) than in HELIOS-A (58.2 pg/mL). In the APOLLO placebo arm, NfL increased from baseline at 4 months (+19.0 pg/mL, $P < 0.001$) and 18 months (+36.3 pg/mL, $P < 0.001$). In the APOLLO patisiran arm, NfL decreased from baseline at 4 months (-20 pg/mL, $P < 0.001$) and 18 months (-23.2 pg/mL, $P < 0.001$). Similarly in HELIOS-A, in both patisiran and vutrisiran groups, NfL decreased from baseline at four months (-9.7 pg/mL and -11.0 pg/mL, respectively; $P < 0.05$), and these decreases were maintained at 18 months (-16.4 and -19.9 pg/mL, respectively; $P < 0.01$).

Conclusion: NfL may serve as a biomarker of treatment response as early as 4 months following initiation of patisiran or vutrisiran. The observed decreases in NfL from baseline are maintained through 18 months of treatment, in contrast to increased levels in untreated patients, making it potentially useful for monitoring disease progression and treatment response over time in hATTR amyloidosis with PN.

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637 – Cerebral venous thrombosis (CVT) and its interface with arboviroses

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Introduction: Cerebral venous thrombosis (CVT) is a rare condition. The presence of headache, papilledema, alteration of the VI cranial nerve, focal deficits and changes in consciousness are highlighted. Attention is needed for the interface between arboviruses, and their impact on the Central Nervous System to better understand the disease. This aim is to report a case of CTV, addressing how the arboviroses affect the Central Nervous System. The information contained in this work was obtained through review of the medical record.

Case report: 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. Admission exams revealed platelets of 20.000 mm^3, positive polymerase chain reaction for dengue and positive immunoglobulin M and negative immunoglobulin G serology. However, during hospitalization, headache intensity worsened, associated with nausea, vomiting, visual blurring and unprecedented tonic-clonic seizures in four limbs. Due to clinical worsening, underwent cranial resonance and angioresonance which revealed venous thrombosis in the transverse 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. On this occasion the platelets were 175.000 mm^3. Hematological and rheumatological examinations were normal. Anticoagulation was started with low-molecular-weight heparin and later warfarin 5 mg/day. The patient was discharged without complaints, with oral anticoagulation and follow-up at the CVT outpatient clinic.

Conclusion: It is essential to be aware of the alarm signals (red flags) as a form of suspected secondary causes of headache as the TVC, especially in women with persistent headache in the context of arboviroses.

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638 – An uncommon neurological manifestation of acute myeloid leukemia: longitudinally extensive transverse myelitis

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Case presentation: A 68-year-old male patient presented with progressive difficulty walking over the course of two months, associated with sphincter dysfunction and weight loss of 25 kg. On examination, the patient presented fluctuation in the level of consciousness, bilateral papilla edema and asymmetric tetraparesis with crural predominance. During investigation, the magnetic resonance imaging showed areas of hypersignal in T2/FLAIR with foci of contrast uptake in the region of cerebellar peduncles and hemispheres, bulb and spinal cord of C1-C8, associated with diffuse meningeal enhancement. Cerebral spine fluid (CSF) showed 20 cells with 56% of monocytes; 61 red blood cells; 419 protein and 5 glucose. After extensive negative investigation, CSF immunophenotyping showed 92% of anomalous monocytes, associated with bone marrow study without evidence of atypical cells, the diagnosis of primary acute monocytic leukemia (AML) of the central nervous system (CNS) was made.

Discussion: CNS infiltration in lymphoproliferative diseases may be present at the time of diagnosis or develop over the course of the disease, but there are few reports in the literature of CNS involvement preceding peripheral blood and bone marrow involvement. The clinical presentation can be very varied, and may even manifest as longitudinally extensive transverse myelopathy. CSF examination usually shows pleocytosis and hyperproteinorrhachia associated with hypoglycorrhachia, being a differential diagnosis of neuroinfectious diseases, and the diagnosis is usually made with the result of immunophenotyping that shows anomalous myeloid lineage.

Conclusion: Primary CNS AML is a rare condition, however it should be considered as a differential diagnosis of myelopathy. This disease is diagnosed through the evaluation of CSF immunophenotyping. Therefore, one should consider performing this test in cases of myelopathy as described above.

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640 – Pure motor-variant CIDP associated with immune checkpoint inhibitor therapy

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Introduction: Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an autoimmune disease with many possible etiologies. There are only a few reports of CIDP secondary to the use of immune checkpoint inhibitor therapy. The aim is to describe a case of CIDP secondary to the treatment of metastatic melanoma with immune checkpoint inhibitors (ipilimumab and nivolumab).

Case report: A 52-year-old male patient, with arterial hypertension and hypothyroidism, presented with paresthesia and pain in the hands and forearms in November/22, that progressively spread and affected the feet in a one-month period. He then presented proximal and distal tetraparesis in January/23, leading him to depend on a wheelchair. At the time, he was being treated with immunotherapy for metastases in the pectoral muscles due to a melanoma. Symptoms' onset and progression coincided with the therapy infusions. On examination, there were a grade 4 strength in flexion, extension, abduction and adduction of the thighs and grade 5 in other movements, with global areflexia and tactile hypoesthesia in the feet. Cerebrospinal fluid examination (CSF) showed 11 cells, predominantly with lymphocytes, and 283 mg/dL of proteins. Electromyographic studies revealed focal demyelinating neuropathy of the medians at the wrist level, with moderate to severe intensity on the right and moderate on the left, suggestive of pure motor-variant CIDP. Methylprednisolone 1g/day was given for five days with significant improvement of the condition.

Results: The 2021 EFNS/PSN criteria provide diagnostic guidelines for CIDP based on clinical, electromyographic and CSF studies.

Conclusion: CIDP secondary to the use of immune checkpoint inhibitors has distinct characteristics such as lymphocytic pleocytosis with slightly increased CSF cellularity and severe neuropathic pain as an initial symptom.

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641 – Neurosarcoidosis as a differential diagnosis of tolosa-hunt syndrome: a case report

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Introduction: Neurosarcoidosis (NS) is a rare and often underdiagnosed manifestation of sarcoidosis, which is an inflammatory disorder that can affect multiple organs, including the nervous system. The main clinical feature is cranial neuropathy, especially involvement of the optic nerve and facial nerve. These alterations make a differential diagnosis with Tolosa-Hunt syndrome. The aim is to report a case of neurosarcoidosis. This is a case report study. The information of this work was obtained through review of the medical record.

Case report: Female, 39-year-old. In 2005, she started pain and paralysis of the lateral movement of the left eye, which was resolved with mucocele removal surgery. Presented three similar cases in 2010, 2015, and 2020, resolved with the use of anti-inflammatory drugs. In 2021, she presented left facial paralysis, left eyelid ptosis, visual blurring and headache. During hospitalization, the possibility of Tolosa-Hunt syndrome was investigated. Imaging exams and laboratory tests were performed without significant alterations. In 2022, she presented a similar clinical manifestation. Magnetic resonance imaging with angiography were performed, which showed thickening and enhancement of the pachymeninges in the occipital regions and cerebellar hemispheres, suggesting the possibility of meningeal granulomatous involvement. A biopsy with histopathological and immunohistochemical analysis was performed, confirming the diagnosis of neurosarcoidosis.

Conclusion: This case report highlights the importance of considering neurosarcoidosis as a differential diagnosis in cases of multiple cranial neuropathy, such as in Tolosa-Hunt syndrome. Early recognition and diagnosis can lead to timely treatment and improved outcomes for patients.

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642 – Clinical aspects of multiple sclerosis in childhood

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Introduction: Multiple sclerosis (MS) is an autoimmune, demyelinating, and inflammatory disease affecting the central nervous system. This condition has an unknown etiology and variable prevalence, being more common in young adult females. The prevalence of MS in childhood is low, ranging from 2% to 4% of all cases.

Objectives: This study aims to describe the clinical development and symptoms of MS in children, contrasting them with the development of MS in adults.

Dessign: This is an integrative literature review.

Methods: Ten articles were selected from the PubMed and SciELO in English and Portuguese between 2010 and 2020. The keywords used were Multiple Sclerosis and Pediatric.

Results: The prevalence of MS in children presenting with clinical symptoms is estimated to be between 2% and 5% of all MS cases. Despite variations in prevalence, the relapsing-remitting subtype and female predominance are similar to MS in adults. The female-to-male ratio ranges from 1.13:1 to 3.88:1. The relapsing-remitting subtype accounted for 85.7% to 100% of cases. While there is no typical presentation of clinical symptoms, optic neuritis, motor alterations, sensory changes, and cerebellum and brain stem dysfunctions are frequently reported. Unlike what is observed in adults, MS in children typically presents in a more diverse range of ways and progresses more rapidly and severely, with multifocal presentations. Also, children experience slower illness progression due to higher neuroplasticity but reach the impairment milestone before adults.

Conclusion: The clinical presentation of multiple sclerosis in childhood is diverse, presenting different characteristics of adults. There is no consensus on the most prevalent symptoms. However, there are similarities in gender and subtype between children and adults with the disease.

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643 – Syringomyelia in children: a case report

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Introduction: Syringomyelia or hydromyelia is a rare neurological disorder of cerebrospinal fluid circulation caused by a pathologically dilated central spinal canal, manifested as dissociated sensory loss, chronic pain and motor deficits. The pathogenesis is not completely understood, usually being a diagnostic challenge. The authors describe a case of syringomyelia in a ten-year-boy associated with depression, as a challenging condition, emphasizing diagnostic difficulties and the multidisciplinary approach.

Clinical case: The patient was admitted to the emergency room with pain, gait abnormality, arthralgia and dysesthesia in limbs with subacute onset. Neurological examination revealed tetraparesis with hypoesthesia and hypopallesthesia in limbs, associated with emotional lability. Feelings of worthlessness and abandonment in psychiatric evaluation resulted in diagnosis of depression. For persistent pain assessment, a magnetic resonance imaging of the spinal cord showed dilatation of the central canal, compatible with hydromyelia. To establish if the sensitive symptoms were secondary to damage of spinothalamic pathway, a laser evoked potential was performed, demonstrating integrity of Aδ-fibers, excluding, therefore, impairment of the nociceptive pathways only in the segments analyzed. In view of the findings, the patient was discharged from the hospital, still with some gait impairment, taking selective serotonin reuptake inhibitors for depression and carbamazepine for chronic pain with progressive improvement.

Conclusion: This report explores the complexity of diagnosis of hydromyelia in children, which are difficult to examine accurately, particularly on sensitive examination, whose objective alteration in this case made us consider this rare disease. We believe that rigorous clinical follow-up, with multidisciplinary intervention, is extremely important to achieve the best response to treatment.

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645 – A 26-year-old woman presenting with a history of epileptic crisis, ataxia and cognitive impairment

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A 26-year-old woman was referred to a neurology outpatient clinic due to a 9-month history of generalized tonic-clonic seizures, gradually more frequent since onset. She additionally reports developing insidiously over four years ago, an asymmetrical tremor in the upper limbs (worse on the right hand), difficulty walking, speech disorders and decreased visual acuity on the right eye. She had a past medical history of major depressive disorder, but normal neuropsychomotor development in childhood, and did not drink alcohol or smoke tobacco. There is no family history of neurological conditions (she has three brothers and two healthy children). She reports consanguinity (maternal grandparents). Upon neurological examination, the patient was alert, attention was impaired and was not oriented to place or time. Speech was scanned. Her visual acuity was decreased in the right eye (20/100), right gaze-evoked nystagmus and slow saccades. Fundi in both eyes were normal and examination of the other cranial nerves was unremarkable. Based on Medical Research Council grading, the patient had a power of 5/5 in all muscle groups of the lower and upper limbs, deep tendon reflexes in upper limbs were brisk, normal in lower limbs, and plantar responses were flexor bilaterally. Sensory exam was also unremarkable in all four limbs. Appendicular ataxia was present in all members, with rest and intention tremor in upper limbs. During gait she had a noticeable widened base, and steps were unsteady and irregular. Meningismus was absent. A mini-mental exam was done: 21/30 (eight years of study), with impairment mainly in attention, language and planning. Routine blood tests including full blood count, fasting glucose, B12 level, renal profile, electrolytes, liver function tests, C-

reactive protein, serum protein electrophoresis, thyroid function test and erythrocyte sedimentation rate were normal. Serologic tests for syphilis (venereal disease research laboratory), viral hepatitis B and C, and HIV serology were negative. Cerebrospinal fluid analysis showed 01 white blood cell/L, protein 32, glucose 68 mg/dL and absence of oligoclonal bands. Magnetic resonance imaging (MRI) sequences showed significant cerebellar atrophy. Electroencephalogram was normal. A genetic panel was done which shows a mutation on *TPP1* gene, compatible with neuronal ceroid lipofuscinosis-2 (CLN-2, OMIM #204500). Neuronal ceroid lipofuscinosis (CLN) is a progressive neurodegenerative lysosomal storage disease caused by the accumulation of lipofuscin in the cerebellum and cerebral cortex, which results in neuronal death. There is an estimated incidence of < 0.5 per 100,000 live births in Europe; in Brazil its prevalence is unknown. With the identification of molecular defects, the CLNs are classified according to the underlying gene defect, regardless of the age at onset. CLN2 is caused by a deficiency of the tripeptidyl peptidase 1 (TPP1) enzyme secondary to mutations in the *CLN2* gene, being the most prevalent type observed and the only treatable one. The clinical course includes refractory epilepsy to antiepileptic medications, progressive mental regression and deterioration, ataxia, myoclonus, and visual loss. On MRI, most patients have diffuse cerebellar atrophy, corroborating the clinical finding of central nervous system progressive degeneration.

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652 – Benefits of yoga on quality of life of people with remitting-recurring multiple sclerosis

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Introduction: Multiple sclerosis (MS) is an autoimmune and demyelinating disease of the central nervous system. MS can be accompanied by physical, psychological, and/or cognitive symptoms, which can limit the quality of life of patients. Therefore, in addition to pharmacological treatment, it is required the participation of integrative and complementary therapies such as Yoga.
Objectives: To analyze the benefits of Yoga on the quality of life of people with relapsing-remitting MS (RRMS).
Methods: This is an observational, exploratory, descriptive, cross-sectional, and qualitative field research. The final sample of the study was four people. Quality of life was evaluated using the SF-36 questionnaire, before and after seven Yoga classes.
Results: Improvement in the intervention of emotional state in daily and work activities was observed in 75% of cases. 25% had an improvement in self-perceived health, and 100% improved the amount of time they dedicate to work. Additionally, after the classes, there was an improvement in the quantification of pain and its interference in work. Finally, regarding the comparative perspective between their health situation compared to the population, 50% thought their health would mostly worsen, followed by 75% who then thought it was definitely false that this would happen. Most participants also began to affirm that their health was excellent.
Conclusion: Improvement was observed in the physical and emotional health of the participants. However, more research with a larger sample is needed to validate these benefits.

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653 – Body fat percentage and Alzheimer's disease biomarkers in people with mild cognitive impairment

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Introduction: Patients with mild cognitive impairment (MCI) have an unnatural cognitive loss of aging and have an increased chance of developing Alzheimer's Disease (AD). Another factor that also increases this risk is the chronic inflammation caused by obesity, described by a body fat percentage (BF%) above healthy values.
Objectives: This study aimed to investigate whether BF% in older adults with MCI correlates with AD biomarkers, such as total TAU protein and beta-amyloid.
Methods: Twenty-one older adults with MCI were evaluated, 11 men and 10 women with a mean age of 66.3 (standard deviation, SD: ±5.88) years. The participants were submitted to the collection of 10 ml of cerebrospinal fluid (CSF) via lumbar puncture and evaluated regarding the dosages of CSF total TAU protein and beta-amyloid protein using immunoenzymatic ELISA kits: INNOTEST® ß-AMYLOID (1-42), INNOTESTR h TAU Ag (Innogenetics, Gent, Belgium). They were also evaluated for BF% on a bioimpedance scale (Tanita® BC-108). Kendall's tau correlation coefficient was applied for statistical analysis using the IBM SPSS Statistics 22 Software.
Results: Both men and women had an average BF% that classified them with higher BF (men: 27% (SD: ±4.23) and women: 36.7% (SD: ±4.84). We found a weak positive correlation ($\tau = 0.360$*) between BF% and Total TAU protein concentration and between BF% and beta-amyloid protein concentration ($\tau = 0.341$*).
Conclusion: Our results provide an insight into the possible influence of BF% on concentrations of biomarkers related to neurodegeneration in elderly people at increased risk of developing AD. The higher the BF%, the higher the protein concentration that reflects neuroinflammation. Studies with more individuals with MCI and with different degrees of obesity are suggested to contribute to the investigation of the relationship between AD biomarkers and BF%.

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654 – Use of lamotrigine in impulse control and social cognition in patients with temporal lobe epilepsy

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Temporal lobe epilepsy (TLE) is a type of focal epilepsy that can begin in one or more regions of the temporal lobe and spread to adjacent brain tissue via neural connections and can be divided into two types according to the Classification of Epileptic Syndromes (ILAE 2017). The most common is mesial temporal lobe epilepsy, which affects temporal regions such as the hippocampus, entorhinal cortex, amygdala, and parahippocampal

gyrus. The second type is lateral or neocortical, where seizures occur in the temporal neocortex (superior, medial and inferior temporal, temporooccipital and temporoparietal gyri and associative senses for auditory, visual and verbal functions). Approximately 60% of patients with mesial TLE associated with hippocampal atrophy are unable to control their seizures even after optimal treatment with various antiepileptic drugs. For these patients, epilepsy surgery can be an effective alternative treatment. After a series of preoperative studies, including medical history and careful neurological examination, complex neurophysiological studies (surface, surface and invasive electroencephalographic video electroencephalogram), neuroimaging studies and neuropsychological evaluations for selected cases. Notably, according to Wiebe and Engel, 2012, surgical treatment of TLE is superior to long-term medical therapy in these selected cases. Because the pathophysiological course of mesial TLE may favor preservation of epileptogenesis even after removal of the primary regions, effective cure in these patients is not always guaranteed. Furthermore, due to the location of mesiotemporal lesions, patients with TLE suffer from stigma, associated with seizure and psychiatric disorders, which affects the quality of life and functioning of these patients. Therefore, this study aims to investigate the efficacy of using antiseizure medications, especially lamotrigine on impulse control, which is also impaired in some mood disorders. Bear Fedio Inventory (BFI) was used to study the effect of lamotrigine and other antizeiures medications on impulse control in patients with TLE. Patients with TLE confirmed by clinical semiology and magnetic resonance imaging findings treated with lamotrigine or other antiseizure medications were included. Only patients older than 18 years and younger than 60 years were investigated. Patients with psychotic symptoms were excluded from this analysis. The BFI was used and applied together with the International Personality Disorder Examination (IPDE). All participants received the questionnaires and were allowed to omit any demographic data that they felt might lead to disclosure of their identity. Ethical approval was obtained from the Ethics Committee of the Botucatu Medical School. The inventory consists of 100 items that must be marked as true or false. Each group of five statements examines one of the following areas: writing tendencies, hypermorality, religious beliefs, anger and impatience, tendency to organize or order, decreased libido, fear and anxiety, guilt, seriousness, sadness, emotion, suspicious and detail-oriented, cosmic interest, belief in personal predestination, persistence and reproducibility, hatred and revenge, addiction, euphoria, and somatization. A high score is 2 or more true items in each domain, or 20 or more items marked true in total. The IPDE, on the other hand, describes personality traits according to ICD-10 and identifies them based on a set of 5 responses with at least two being true to assume that the respondent has that trait, such as impulsivity or borderline. 36 respondents answered the questionnaires and the responses were stored and categorized into two groups, those who take lamotrigine medication and those who do not. With this separation in mind, the answers that defined the personality trait according to the inventories were selected and grouped, the answers were yes or no, and the accumulation of the answers and the score of the accumulation were applied, and the positive and negative cases for the trait were grouped so that the chi-square test could be applied. Nine of the 36 respondents were taking lamotrigine and 27 were taking other medications. For the IPED with the score of impulsivity, there were 7 positives and 2 negatives, the 27 who did not use lamotrigine, 21 with a positive score and 6 negatives. For the BFI, the Hate and Vengeance and Euphoria traits were selected for comparison and to test the hypothesis of decreased impulsivity traits. There was no change in the respondents who use lamotrigine, of the 9, only 2 had a positive score and 7 a negative score, for the non-users tested in this criterion 16 positive and 11 negative. There was not difference for hatred and revenge trail between the groups (P = 0.0543). For the euphoria trait, the values for lamotrigine users were 8 positive and 1 negative, and for non-users were 21 positive and 6 negative (P = 0.466). This preliminary investigation did not show difference for impulse control between patients taking lamotrigine or not. A larger sample size is currently underway to support this observation.

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655 – A case study on dysarthria present in coexisting autoimmune diseases

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Introduction: Autoimmune diseases are characterized by immune dysfunction, autoantibody production and chronic inflammation. The aim is to describe dysarthria in a case that combines autoimmune diseases.
Case report: This study reports the case of a 43-year-old woman with complaints of speech difficulties and a diagnosis of dysarthria due to Multiple Sclerosis, Sjögren's Syndrome, Raynaud's Phenomenon, Systemic Lupus Erythematosus and Hashimoto's Thyroiditis. Assessments were performed using the GRBASI scale, Praat software and Voice Related Quality of Life Protocol for six years consecutive. During the period, the patient sought speech therapy support annually and reported symptoms of low and hoarse voice, slurred speech and tiredness when speaking. Negative impact on quality of life was self-reported in the presence of speech difficulties, with mean scores in the VRQOL domains: total = 75, physical = 71, and socioemotional = 82. The most frequent signs were a breathy and asthenic hoarse voice with marked instability, and reduced intensity (56 decibels), slowed speech, and imprecise articulation, characterizing mixed dysarthria. The signs and symptoms did not always occur simultaneously with the expressions of the diseases and had intermittent, heterogeneous manifestations of varying severity.
Conclusion: This study shows that dysarthria affects verbal communication, family, social and professional interaction and compromises quality of life. Considering the complexity of these chronic diseases, periodic specialized speech therapy monitoring is recommended for analysis of speech subsystems, guidelines and personalized approaches.

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657 – Longitudinal extensive myelopathy (LEM): a case with investigation pitfalls

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Introduction: Systemic lupus erythematosus (SLE) can affect the central nervous system, with different presentations. The aim is to describe a case of LEM and its pitfalls investigation. Literature review in the PubMed and LILACS electronic library databases.
Case: A 45-year-old man presented with a history of low back pain, radiat-

ing to the lower limbs, evolving with weakness in legs and hands, episodes of falling and tingling in lower limbs. After a week, he was restricted to bed, with urinary and fecal retention. His neurological examination shown weakness in hands 4/5 and in lower limbs 0/5. Bilateral lower limbs reflexes were absent and there was hypoesthesia under the level of T12. Initial investigation showed VDRL 1/8 and positive HIV serology. Spinal cord magnetic resonance imaging revealed LEM, with central predominance. Due to his probable immunocompromised status, ganciclovir and crystalline penicillin were started empirically. Cerebrospinal fluid study revealed lymphomonocytic pleocytosis (120 cells), protein (96 mg/dl), lactate (34 mg/dL), glucose (48 mg/dL), negative VDRL and PCR for CMV, VZV, HSV and EBV. A confirmatory HIV test was negative and the complementary investigation showed FAN 1/160, undetectable complement and positive anticardiolipin, CVT and bilateral PTE, in addition to subnephrotic proteinuria. At this point, we made a diagnosis of SLE and Antiphospholipid syndrome myelopathy. Immunosuppression was started with a reduction of the myelitis and further negative VDRL.

Discussion: Lupus myelopathy is characterized by inflammation and damage to the spinal cord, which can lead to a variety of symptoms. Treatment for lupus myelopathy typically involves immunosuppressants, and biologic agents.

Conclusion: Lupus myelopathy is a serious complication that requires prompt diagnosis and treatment.

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658 – Neurophobia in Brazil: a literature review

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Introduction: In 1994, Josefowicks used the term neurophobia to designate the fear or aversion towards neurology and neurosciences among the medical students, resulting in: indifference about neurology, less confidence in the care of neurological patients and less interest in the specialty as a medical residency worldwide, with impact in medical education and healthcare.

Objectives: Identify the scientific literature produced in Brazil focusing on the study of neurophobia in medical graduation: objectives, population, risk factors and strategies to combat the problem.

Methods: This is a literature review that in a first moment researched and collected data from: Scielo, PubMed, BVS and Google Scholar, followed by evaluation, analysis and interpretation of the data collected.

Results: Ten articles were selected: 4 cross-sectional; 3 exploratory studies; 1 literature review and 2 expert opinions, published between 2014 and 2021. Half of them sought to diagnose the neurophobia in medical students and in all of these it was positive and prevalent, among public and private institutions located in north, northeast and southeast of the country. The following risk factors stand out: poor articulation between neurosciences and neurology, limited time of practice and need for knowledge of neuroanatomy. Team based learning was the strategy used by two studies, but with limited scenario. For "neurophilia" the opinions of specialist corroborate with other studies: integration between neurosciences and clinical, with an increase in time for practice and a curriculum aimed at generalists.

Conclusion: The presence of neurophobia in medical students in Brazil, the risk factors and the proposed strategies converge with those described in the world literature. However, our scientific production is scarce, focuses on detecting the student's perception and lacks studies with higher levels of evidence, highlighting the importance of expanding scientifically based knowledge about the subject in our country.

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661 – Pediatric onset multiple sclerosis: defying disability – a case report

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Introduction: Multiple sclerosis (MS) is the main demyelinating disturbance of the central nervous system in childhood and, compared to adulthood, the disease presents in a more inflammatory way and with greater probability of younger disability if not well managed in adequate time.

Objectives: Case report of a patient with pediatric onset multiple sclerosis (POMS) and high lesions burden in need of high efficacy medications in early time.

Methods: The present case report was based on data collected through literature review and medical records.

Results: We found the case of a 12-year-old girl presenting with monoparesis of right leg and being first diagnosed with acute disseminated encephalomyelitis due to lesions with a demyelinating character in a skull magnetic resonance (MRI) in January 2022 after COVID-19 vaccination. After six months, she presented with diplopia and ataxia, besides worsening of radiological pattern of the previous lesions and evidence of spinal involvement, being submitted to therapy with methylprednisolone. Further, the diagnosis of POMS was stablished due to oligoclonal bands in cerebrospinal fluid and negativity to serum anti aquaporin 4 and serum antibody against myelin oligodendrocyte glycoprotein; treatment with natalizumab was started. After four months, she presented with worsening of previous neurological deficits and progression of disease was seen in a new skull and neuroaxis MRI, being submitted to a new cycle of methylprednisolone and five sessions of plasmapheresis. After these therapies, she went home with an expanded disability status scale of 4 and rituximab was the chosen drug to continue treatment according to literature and the neurological state.

Conclusion: Early intervention in pediatric onset multiple sclerosis is necessary to avoid the maximum of future disability and high inflammatory level frequently demands high efficacy therapies to control the disease.

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662 – Ataxia unit of the Federal University of São Paulo: 16 years of experience and a description of frequency and etiology

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Introduction: Cerebellar ataxias are a heterogeneous group of diseases comprising genetic and non-genetic etiologies. Sporadic ataxias are a group

of neurological disorders with no family history for ataxia or its related symptoms and signs. Hereditary cerebellar ataxias and hereditary spastic paraplegias (HSP) are rare genetic neurodegenerative disorders with considerable clinical overlap.

Objectives: To report a descriptive analysis of the frequency of different forms of cerebellar ataxias evaluated over 16 years in the Ataxia Unit, Federal University of São Paulo, Brazil.

Methods: We reviewed charts of patients who were being followed at the Ataxia Unit at Federal University of São Paulo, from January 2007 to December 2022. We used descriptive statistics to present our results as frequencies and percentages of the overall analysis. Patients were classified into the following 8 groups: sporadic ataxias, autosomal dominant cerebellar ataxias, autosomal recessive cerebellar ataxias (ARCA), mitochondrial ataxias, congenital ataxias, X-linked ataxias, HSP, and others.

Results: Overall, 1225 Brazilian patients with ataxias or spastic paraplegias. There were 683 patients with a definite diagnosis, which 89 were sporadic ataxias, 322 autosomal dominant cerebellar ataxias, 169 ARCA, 4 congenital ataxias, 6 X-linked ataxias, 16 autosomal dominant HSP, and 35 autosomal recessive HSP.

Conclusion: This study describes the frequency of cerebellar ataxias

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663 – Myoclonic nonconvulsive status epilepticus as a differential diagnosis in intensive care patients

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Case report: Analyzed the case of three patients with myoclonus in an intensive care setting, all with heart failure with reduced ejection fraction (HFrEF), admitted due to decompensation of the cardiac disease resulting from infectious conditions and the need for moderate to high doses of dobutamine. One of the patients had a concomitant ischemic stroke in border territory. During hospitalization, they presented negative myoclonus in the trunk and lower limbs, an electroencephalogram was performed where in two of the three cases, a generalized epileptiform disorder and electroclinical seizures were diagnosed. The resolution of nonconvulsive status epilepticus (NCSE) of the myoclonic type in the three was obtained after the introduction and titration of valproic acid, infectious control and, in two of the cases, the association of clobazam was necessary.

Discussion: The NCSE common to the three patients was the myoclonic one. The etiologies, in the context of critically ill patients, can be multifactorial, such as: ischemic stroke, hypoxia/anoxia, metabolic infectious disorders and induced by drugs, such as dobutamine. The electroclinical classification helps in the management and prognosis of these patients. The initial treatment performed in the three cases was a combination of intravenous benzodiazepine to abort the NCSE and antiseizure medications: that did not induce coma, with good response, in addition to correction of the subjacente cause.

Conclusion: The presence of myoclonus in intensive care patients should be quickly investigated. It is important to carry out the differential diagnosis with NCSE and identify comorbidities. The recognition and adequate treatment of this often multifactorial condition is fundamental for the prognosis.

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664 – Quality of life evaluation in individuals with multiple sclerosis in a reference center in Paraíba

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Introduction: Multiple sclerosis (MS) is the most prevalent chronic inflammatory disease of the central nervous system (CNS). It affects over 2 million people worldwide and is currently incurable. Similarly to autoimmune diseases, multiple sclerosis can significantly affect the patient's lifetime, especially in terms of compromising their quality of life (QoL).

Objectives: To analyze the QoL in patients with MS at a Reference Center for Multiple Sclerosis in Paraíba (CREM/PB).

Dessign: A transversal, exploratory, and descriptive study.

Methods: The short-version structured interview of the World Health Organization Quality of Life Questionnaire (WHOQOL-BREF) and the Kurtzke Expanded Disability Status Scale (EDSS) were applied to measure the impact on quality of life and the dysfunctions in functional systems with disease progression, respectively. The sample had 46 patients. Data collection took place between February and March 2020. Statistical data were analyzed using IBM SPSS statistics version 26.0.

Results: The Spearman correlation showed that there is a negative and reasonable correlation between the EDSS and the physical domain of the QoL assessment ($P = -0.392$; $P < 0.05$) and a negative and fair correlation between the EDSS and the related field of the social aspects of the QoL assessment ($P = -0.303$; $P < 0.05$). A positive correlation exists between the environment domain and the educational level component ($P = 0.541$; $P < 0.05$).

Conclusion: Individuals with MS experience a decrease in the perception of QoL in the physical domains and social relationships, with a higher perception among men to the detriment of women. There is a correlation between the level of disability and quality of life. Therefore, the necessity for a multidisciplinary approach is reinforced.

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665 – Etiological evaluation of hypertrophic pachymeningitis in a tertiary general neurology department

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Introduction: Hypertrophic pachymeningitis (HP) is the thickening of the cranial or spinal dura. This process causes cranial nerve palsy, vascular events (stroke and venous thrombosis), and intracranial hypertension. The diagnosis of HP is disclosed by magnetic resonance imaging, showing dural thickening and con-

trast enhancement. Several etiologies are possible for HP, but infection and autoimmune diseases are the most frequent. The gold standard for diagnosis is the dural biopsy, but cerebrospinal fluid (CSF), blood and other tissues analysis are essential guides to the correct diagnosis. A quick diagnosis is crucial to early treatment and this avoid permanent deficits. However, the diagnostic approach can be a complicated process, which could delay the beginning of the treatment.
Objectives: This study aims to analyze the population of patients with magnetic resonance imaging showing HP.
Methods: Its epidemiological, clinical and radiological aspects was evaluated through a retrospective and descriptive study to elucidate the etiological diagnosis.
Results: A total of 45 patients was included with some different etiologies: granulomatosis with polyangiitis (n = 7); idiopathic (n = 14); probable neurosarcoidosis (n = 9); seronegative rheumatoid arthritis (n = 1); neurotuberculosis (n = 1); plaque meningioma (n = 2); IgG4 related disease (n = 4); Inflammatory myofibloblastic tumor (n = 1); neurosyphilis (n = 1); diffuse B-cell lymphoma (n = 1); Erdheim-Chester disease (n = 1); iatrogenic secondary to radiotherapy (n = 1) and cryopyronopathy (n = 1).
Conclusion: The diversity of etiologies found in this study demonstrates the complexity of HP and how important it is to follow a diagnostic algorithm in order to institute appropriate and early therapy for a better clinical outcome for patients.

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666 – The impact of acute hepatic porphyria on mental health: results from the porphyria worldwide patient experience research (power) study

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Introduction: Acute hepatic porphyria (AHP) is a group of rare genetic diseases of heme biosynthesis characterized by life-threatening attacks that could be associated with psychiatric symptoms.
Objectives: To investigate the burden of AHP on mental health.
Methods: Adult patients who experienced >1 AHP attack within the past two years or receiving prophylactic treatment were recruited from the United States, Italy, Spain, Australia, Mexico, and Brazil. Patients taking givosiran were excluded. Participants took an online survey that used standardized questionnaires and validated screening instruments to evaluate the impact of AHP symptoms on social life, personal goals, and mental health.
Results: Ninety-two patients with AHP completed the survey, mean age 41.1 years and 90% female. Patients reported substantial impact on social life – 76.1% reported that most of their symptoms were hidden and that people in their social circle did not know they had AHP; 72.8% reported feeling guilty/upset that their symptoms and disabilities affect others. Patients also reported impacts in their personal life/goals, >80% having had to modify or give up goals important to them, more than half decline in their mental and physical health, and over a third lost their sense of purpose. PHQ-8 scores indicating moderate to severe depression in more than half of patients with AHP (58.7%) regardless of attack rate or prophylactic treatment status. GAD-7 scores indicating moderate to severe anxiety in 48.9% of patients and were highest in patients experiencing recurrent attacks (56.8%).
Conclusion: Patients with AHP, regardless of attack rate, treatment received, or duration of active disease, experience a high mental health burden on their personal and social life and high frequency of moderate to severe anxiety or depression, highlighting the importance of mental health monitoring as part of disease management.

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668 – "Descartes' error": evidences for the role of brain regions and their connections in introspective thoughts and self-identity

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Introduction: René Descartes was a French philosopher, considered the father of rationalism and one of the most important thinkers in history. Based on his methodical doubts' technique, the author concluded that he exists as a thinking thing, that if "I think, therefore I am". He establishes that he had no more doubts that he really existed as thought and as a body, however, he could not think that thought and body were two equal substances, since they had certain different properties, therefore, the author came to believe that mind and body were distinct substances. Since the 17th century, advances in neuroscience have helped to clarify which brain regions are involved in introspective and self-referential thinking.
Methods: We reviewed through searches in PubMed, MEDLINE, SciELO and Scopus the brain regions involved in the self-referential and introspective thinking.
Results: Recent studies show that self-referential and introspective thinking are located mainly in regions related especially to the Default Mode Network, mainly located in the frontal regions. Especially the "autobiographical self" shows greater activity in memory-related regions (hippocampus and posterior cingulate cortices), medial prefrontal cortex, and insula cortices, but the establishment of the "self" as the individual's relationship to the world around it, uses regions related to exteroception, such as secondary and tertiary sensitive areas. For self-referenced thinking, there is greater engagement of the medial frontal cortex regions, more specifically in Brodmann's area 10, and this structure is also related to the DMN.
Conclusion: The advancement of neuroscience has allowed demonstrating that mental processes and thoughts happen as consequences of certain brain connections and processes. Thus, we ask poetic license to the great philosopher to say "the brain connections exist, therefore we think".

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669 – A467T variant of the polg gene: description of two clinical cases

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Introduction: Variations in the POLG gene are the most common causes of mitochondrial disease of autosomal inheritance, and may be present in about 2% of the population.
Case report: Case 1. CMAM, male, 48-year-old, complaining of bilateral eyelid ptosis with onset in adolescence. Since the age of six, he has been diagnosed

with epilepsy. After five years of follow-up, he developed sensory ataxia. After 10 years he began to present dysarthria, dysphagia, tremor and pyramidal syndrome. Case 2. ASB, female, 42 years old, at 20 years old presented generalized clonic tonic crisis during the second and third trimesters of pregnancy; at 35 years of age she complained of tingling in plants and legs; at 37 years she noticed bilateral eyelid ptosis and at 39 years she noticed the presence of slurred speech and fatigue on small efforts. He has 3 siblings with similar symptoms and great difficulty walking. No history of consanguinity. Propedeutics: Normal serum lactate and CPK dosage; muscle biopsy showed variation in the caliber of muscle fibers, with the presence of "ragged red fibers" in Gomori's Trichrome stain. Cranial magnetic resonance imaging: mild cerebellar atrophy in patient 1 and normal in patient 2. Electroneuromyography reveled absence of sensory action potentials in all nerves studied in both cases. New generation sequencing myopathy panel revealed pathogenic variant in homozygosis in the POLG c.1399G>A gene (p.Ala467Thr).

Results: The patients received the diagnosis of mitochondrial disease, presenting complex clinical phenotype.

Conclusion: DNA polymerase gamma is the enzyme responsible for replicating and maintaining mitochondrial DNA, encoded by nuclear DNA. The c.1399G>A variant in exon7 causes a replacement of an alanine with threonine (A467T), and is one of the causes of ataxia, such as spinocerebellar ataxia with epilepsy; autosomal recessive mitochondrial ataxia, sensory neuropathy, dysarthria and ophthalmoparesis and myoclonic epilepsy, myopathy and sensory ataxia. However, most of the time, they present a continuum between the phenotypes described.

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670 – Giant cell glioblastoma: a case report of a long-term survival

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Glioblastoma (GBM) is the most common primary brain tumor in adults, classified as grade 4 by World Health Organization, with a mean overall survival of 15–18 months. The histopathological subtype described as giant cell GBM corresponds to < 1% of this tumor, more frequent in young patients, affecting the temporal and parietal lobes in a more circumscribed and subcortical way. We present a case report of a woman diagnosed with GBM giant cell (GC). Data was collected through electronic medical record. A 19-years-old woman, healthy, during investigation of headache with visual and speech alteration, a cranial tomography was performed in January 2009, demonstrating a supratentorial lesion. Cranial magnetic resonance imaging showed a solid-cystic, infiltrative lesion with perilesional edema, located in the left parietal region and heterogeneous peripheral enhancement. Subtotal resection of the lesion was performed in February 2010. The anatomopathological study was compatible with GC, and adjuvant treatment was performed with radiotherapy (60 Gy, in 30 fractions) and concomitant chemotherapy, temozolamide (TMZ), followed by adjuvant TMZ, monthly for seven cycles. The patient developed osteomyelitis and it was necessary to suspend the adjuvant chemotherapy in November 2010. Clinical and radiological follow-up showed stability of the lesion and staying functionally independent, with progression-free survival >13 years. This is an atypical case of a patient with glioblastoma with prolonged survival. Some studies suggest that the dominant presence of giant cells, extent of surgical resection in addition to some molecular may be predictors of long-term survival. Besides the rarity of this malignancy and heterogeneity response with the treatment, it's important to recognize this subtype of GBM and to treat aggressively for the possible of achieve better outcomes, until better therapeutic data are available.

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671 – Impact of acute hepatic porphyria attack frequency on patient-reported outcomes: results from the Porphyria Worldwide Patient Experience Research (POWER) study

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Introduction: Acute hepatic porphyria (AHP), a group of rare genetic diseases, is characterized by neurovisceral attacks.

Objective: This study evaluated its impact in patients with sporadic or recurrent attacks.

Methods: Adult patients having >1 attack within the past two years or receiving prophylactic treatment were recruited from the United States, Italy, Spain, Australia, Mexico, and Brazil for an online survey. Patients taking givosiran were excluded. Differences between patients with sporadic attacks (annualized attack rate [AAR], <6 within past two years) and recurrent attacks (AAR, ≥6) were evaluated. Attacks included those leading to hospitalization, emergency room, outpatient doctor, or self-management. Outcomes were assessed with Generalized Anxiety Disorder-7 (GAD-7), Patient Health Questionnaire (PHQ-8) and burden of chronic symptoms.

Results: Of the 92 patients who completed the survey, 60% reported sporadic attacks and 40% recurrent attacks. Most patients were female (sporadic 92.7%; recurrent 86.5%), and the most frequent diagnosis was acute intermittent porphyria (sporadic 83.6%; recurrent 59.4%). Most patients in the sporadic (52.7%) and recurrent (67.6%) groups reported a PHQ-8 score indicating moderate to severe depression; 43.6% in the sporadic and 56.8% of recurrent groups reported a GAD-7 score indicating moderate to severe anxiety. Pain was one of the most burdensome chronic symptoms in the sporadic (50.9%) and recurrent (59.5%) groups. Most patients reported muscle weakness as a top three symptom that limited their daily activities (sporadic 83.3%; recurrent 90.6%).

Conclusion: While disease burden appeared greater for AHP patients experiencing recurrent attacks, both sporadic and recurrent group patients experienced a substantial impact on physical, mental, and emotional quality of life.

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674 – COVID-19 related rombencephalitis: 2 cases report

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O coronavirus diasease 2019 (COVID-19) may be associated with severe neurological complications, among them, encephalitis. Neurological manifestations are assigned to several mechanisms, such as direct viral infection and immune-

mediated processes. This case series aims to describe patients who presented rhombacephalitis after or during severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection. Case report 1: Female, 26-year-old, had mild COVID-19 infection, followed by nausea and vomiting, which evolved to headache and vertigo, later associated with dysarthria, dysphagia, diplopia, blurred visual, hypoacusis, postural instability and right hemiparesis. Brain resonance image showed hypersignal in the midbrain, pons and medula oblonga. Normal cerebrospinal fluid. Therapy with methylprednisolone was performed, as the same time with ampicilin therapeutic test, followed by plasmapheresis,all without response. Progressed with difficulty in trunk support, use of nasogastric tube and Cognitive impairment. After neurological decline and septic shock, he evolved to death. Case report 2: Female, with suspected psychiatric diagnosis, 27-year-old, interned due to left peripheral facial paralysis associated with right hemiparesis, diplopia, ataxia, dysarthria, strabismus and fever. Positive for COVID-19. Brain resonance with lesions in the cerebellum, midbrain, bridge and white matter of the cerebral hemispheres, some with contrast uptake. Normal cerebrospinal fluid. Performed empirical treatment with ampicillin and acyclovir, in addition to therapy with methylprednisolone, partial improvement. In eleven months, it evolved with worsening of symptoms, associated with seizures, convulsive status and coma. Repeated corticosteroids, followed by immunoglobulin and plasmapheresis. She was discharged with severe neurological sequelae and is still in rehabilitation. This is an extremely relevant and current topic, considering the world scenario of COVID-19 and the lack of evidence for the treatment of possible neurological conditions related to SARS-CoV-2 infection.

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675 – Neuro Behçet's disease: report of a rare disease with neurologic presentation

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Introduction: Behçet's disease is a multisystem disease consisting of oral lesions, genital ulcers and uveitis with unpredictable evolution. Vasculitic and parenchymal involvement of the central nervous system is uncommon. The aim is to describe a case of systemic rheumatologic disease with unusual central involvement in clinical practice.
Case report: Woman, 57-year-old, with a previous history of rheumatoid arthritis for 23 years, presented oral and genital ulcers in addition to paresthesia in the right hemiface and subacute change in visual acuity and bilateral ocular pain in 2017. She was diagnosed with Behçet's Disease with neurological involvement, being submitted to pulse therapy with Cyclophosphamide and maintenance treatment with Azathioprine. She recurred with the same neurological symptoms in 2020, readmitted in the context of severe thrombocytopenia, being diagnosed with idiopathic thrombocytopenic purpura. Pulse therapy with Methylprednisolone was performed, with improvement of symptoms, but thrombocytopenia was maintained. He opted for the maintenance of Immunoglobulin then, with subsequent initiation of Rituximab. Bilateral ocular sequelae persisted with low acuity and hypoesthesia in the path of the trigeminal nerve on the right. Maintains good control of the disease using Rituximab at weaning and maintenance dose of Prednisone.
Conclusion: The prognosis of Behçet's disease is poor when the central nervous system is affected and studies about treatment still have limited evidence. Early identification of the disease is essential to enable rapid and adequate treatment.

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677 – Poems syndrome: a case report

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Introduction: Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes syndrome (POEMS) is a rare monoclonal disease with multisystemic presentation and therefore challenging diagnosis. The aim is to report a case of a rare hematological disease with unusual characteristic neurological manifestation.
Case report: Male, 59-year-old, with a report of anesthesia and paresthesia in the feet that started in 2018. On examination, he presented tactile and painful hypoesthesia in a path innervated by L5 to S1, in addition to hypoactive Achilles tendon reflexes. Electroneuromyography showing moderate-grade diffuse distal symmetric axonal sensorimotor polyneuropathy. In 2021, erythematous lesions appeared on the arms and hyperchromic macules on the left leg. Biochemical tests were performed with the presence of a monoclonal peak of gammaglobulins with a predominance of Kappa chains. In 2022, he presented worsening of tactile, painful and palesthetic sensations in the lower limbs, with no conscious proprioception. POEMS diagnostic hypothesis questioned. The myelogram revealed the presence of 12.4% of plasmocytes and the bone marrow biopsy showed cellularity of 40% with the presence of three lineages with slightly hyperplastic erythroid series. Hemogram presented polyglobulia. Imaging propaedeutics showed hepatomegaly. Confirmatory findings of monoclonal gammopathy and peripheral polyneuropathy, in addition to minor findings of skin changes, hepatomegaly, and polyglobulia, confirmed the diagnosis of POEMS.
Conclusion: Polyneuropathy is a neurological hallmark of the disease and, when associated with a detected monoclonal protein, should lead to evaluation for POEMS, among the differentials of paraproteinemias. Finally, early recognition is essential to enable adequate treatment, which helps to stop the progressive worsening of symptoms, especially the debilitating neurological ones.

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678 – Aneurysm in the vein of galenus in a newborn: case report

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Case presentation: M.L.S, 17-year-old, G1P0A0, 35 weeks, referred to the pediatric neurosurgery service for prenatal evaluation and postpartum surgical planning because the fetus has ventricular asymmetry on initial ultrasound examination. Previously the mother had no morphological ultrasound and no proper prenatal care, she was referred to the neurosurgery service but did not attend the appointment, which made it difficult to transfer to an appropriate hospital. After the initial examination, she underwent magnetic resonance imaging, a fetal morphology study and transcranial Doppler ultrasound that showed the presence of a bulky expandable mass compatible with a galenary vein aneurysm. The patient presented pulmonary hypertension that made surgical intervention impossible and died on the 12th day of life due to refractory heart failure.
Case discussion: Galen's vein has a great importance because it drains the

central portion of the brain. Prenatal developmental aneurysmal malformation of the gallenary vein begins in the first trimester of gestation, with variable severity and clinical manifestations that can manifest intrauterine with signs of heart failure, hydrocephalus or oligohydramnios. The patient evolved with significant respiratory distress and acute pulmonary edema abs the severity of the condition made it impossible to perform the surgery. Vein of Galen aneurysm constitutes less than 1% of the cases of congenital vascular malformation, its early diagnosis is of great importance, especially since the condition has a high lethality rate. The recognition of this condition based on ultrasound in the prenatal period is a fundamental factor for the diagnosis of the aneurysm and its subsequent referral to a reference center. In this way it is possible to define the best treatment model.

Conclusion: It is assumed that if it were diagnosed and referred properly during the intrauterine period, it would be possible to evolve successfully and diminish or nullify the pathophysiological consequences of the newborn aneurysm of Galen's vein.

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679 – Evaluation of the NINDS-CSN 5-minutes protocol as a cognitive screening test to detect Parkinson's disease dementia: a study of a Brazilian sample

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Introduction: Many cognitive screening tests have been investigated for the diagnosis of Parkinson's disease dementia (PDD), due to its prevalence and its interference in the evolution of the disease, quality of life and in the treatment response of the patients with Parkinson's disease (PD). Therefore, more effective and faster cognitive screening tests are needed.

Objectives: To evaluate the usefulness of the NINDS-CSN 5-minutes protocol assessment (NC5MPA) in PD patients as screening test for the detection of PDD, as well as to test if the association with the cube drawing test (CDT) can increase the test accuracy.

Methods: A total of 98 patients with PD were evaluated using the NC5MPA, combined with the CDT, Mini Mental State Examination and the Montreal Cognitive Assessment (MoCA). These patients were also evaluated for mild cognitive impairment (MCI) and dementia by the Clinical Dementia Rating Scale (CDR).

Results: There was a good correlation (with P value < 0.00) between the test scores and PDD, but the results of the 3 tests for MCI was > 0.05. The NC5MPA test has had sensitivity of 78.5%, specificity of 85.7%, accuracy of 82.6%, positive predictive value of 80.4% and negative predictive value of 84.2%, in addition to demonstrate an average performance time of 3.2 minutes (3.08–3.31). The association with the CDT has led to a little significant increase in sensitivity and has showed a decrease in specificity and accuracy, besides increase the test performance time. In assessing the interference of education level, the results were influenced by the small sample size of the 5 to 8 years of education group.

Conclusion: The NC5MPA test has proven up to be a good screening test for PDD, being even faster and easier to perform, but more tests with larger populations are necessary to assess the accuracy of this test for MCI and to assess if there is interference of education level in the test accuracy.

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680 – Poems syndrome: report of a rare disease with often neurologic presentation

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Introduction: Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes (POEMS) syndrome is a rare monoclonal lambda plasma cell disorder, with multisystem presentation leading to a challenging diagnosis. The aim is to report a case of a rare hematologic disease, with neurologic manifestation widely seen in daily practice.

Case report: Male, 35-year-old, with a history of distal weakness of the lower limbs, symmetrical, subacute onset and progressive worsening, associated with ascending numbness, lower back pain and fatigue. Neurological examination showed motor deficit in extremities (plantar flexion and extension), hypoactive reflexes of lower limbs, in addition to distal sensory deficit (tactile and painful). Electroneuromyography confirmed axonal sensory-motor neuropathy in both lower limbs. Biochemical testing revealed a monoclonal gammaglobulin peak with a predominance of lambda chains and an elevated serum vascular endothelial growth factor (VEGF) concentration. Magnetic resonance imaging detected sclerotic lesions in vertebral bodies of thoracic and lumbar spine. Furthermore, patient also had mild pigmentary changes in the skin of the extremities of lower limbs and polycythemia. Accordingly with the evaluation was possible to make POEMS diagnosis and patient is currently in preparation for an autologous bone marrow transplant.

Conclusion: Polineuropathy is a neurological hallmark of the disease and should lead to evaluation for POEMS, when associated with a monoclonal protein detected. Elevated serum VEGF can be a biomarker that supports the diagnosis. In summary, early recognition is essential, because enables adequate treatment, which contributes to stop progressive worsening of symptoms, especially debilitating neurologic ones.

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681 – Anti-N-Methyl D-Aspartate (NMDA) receptor encephalitis after herpes virus meningoencephalitis: a case report

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Anti-N-methyl D-aspartate (NMDA) receptor encephalitis is an autoimmune disease of the central nervous system characterized by psychiatric and neurological symptoms. This pathology may be associated with paraneoplastic syndrome and viral infections, especially the Herpes simplex virus. However, a considerable number of cases may remain without an identifiable triggering factor. We report the case of a 17-year-old girl who presented with headache, fever and meningeal signs. Cerebrospinal fluid (CSF) analysis showed a herpes virus infection. After treatment using Acyclovir for 21 days, the patient showed improvement in neurological symptoms. However, new symptoms

were evidenced: behavioral alterations, aggressiveness, delirium, hallucinations, thoughts of self-extermination, catatonia and cognitive impairment. Psychiatric evaluation suggested the use of an atypical, but no beneficial effect was observed. Considering the refractoriness of the condition, a new CSF analysis showed positivity for the NMDA antibody. Thus, the patient underwent immunomodulatory therapy with Methylprednisolone for five days, obtaining little behavioral improvement. Due to subsequent dysautonomia and seizures, the patient underwent plasmapheresis for five days, obtaining significant improvement in symptoms. The present case emphasizes the importance of the differential diagnosis of NMDA receptor encephalitis after infections of the central nervous system that present psychiatric symptoms.

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682 – Meningeal sporotrichosis in an immunocompetent host: case report

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Endemic in Brazil, sporotrichosis usually presents in its cutaneous form. Meningeal involvement is rare and generally affects immunocompromised hosts. Here we describe a case of meningeal sporotrichosis in an immunocompetent patient without concurrent skin lesions. A 78-year-old woman, previously healthy, developed cognitive deficits with fluctuating mental confusion over the course of three months, until she went to the emergency department with fatigue and stupor. Laboratory workup showed severe hyponatremia due to inappropriate ADH secretion. She was treated with fluid restriction and discharged. Two months later, she was referred to a neurologist because of persistent mental confusion in spite of controlled sodium. She had no headache, fever, seizures, or focal deficits. Cerebrospinal fluid (CSF) analysis revealed chronic meningitis: 232 cells (95% lymphocytes), elevated protein (3.021 mg/dl) and low glucose; cultures and polymerase chain reaction (PCR) for *Mycobacterium* were negative. Brain magnetic resonance imaging (MRI) revealed basal meningeal enhancement. She was hospitalized and empirically treated for meningeal tuberculosis, with no clinical nor CSF improvement after one month. In a careful history review, we found out that one year earlier she had been exposed to cats who died from sporotrichosis. One month after that, she had had a single skin lesion which spontaneously disappeared, followed by polyarthritis, which also resolved. At the time of our evaluation, she had no cutaneous lesion nor arthritis; CTs of the chest and abdomen were normal. PCR for *Sporothrix* in the CSF came out positive. She received liposomal amphotericin for 1 month, followed by a 6-months use of itraconazole. She fully recovered her cognitive capacities; follow-up MRI showed no alterations; her late CSF was nearly normal (5 cells, protein 55 mg/dl, no glucose consumption). Our report draws the attention to atypical neurological presentations of sporotrichosis, a treatable condition that may unrecognized.

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683 – PRES in the context of an infectious insult

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Introduction: Posterior reversible leukoencephalopathy syndrome is an acute/subacute clinical-radiological syndrome characterized by headache, changes in consciousness, seizures, focal neurological deficits, visual deficits and that may be preceded by uncontrolled blood pressure in the day before. It does not commonly occur concomitantly with prolonged infectious conditions. The most typical imaging finding is white matter edema in the posterior cerebral hemispheres. Treatment is based on adequate control of blood pressure and epileptic seizures, as well as metabolic and electrolyte control. The prognosis in most cases is excellent with significant improvement and complete reversal over days to weeks.

Case report: Female, 37-year-old, after one month of underwent an endoscopic retrograde cholangiopancreatography was hospitalized due to a perirenal abscess, and treatment with antibiotics was initiated. During this treatment, she developed a sudden onset of bilateral amaurosis, continuous horizontal nystagmus, mental confusion, right upper limb weakness and hyperreflexia, in addition to an epileptic seizure and fencing response posture requiring intravenous phenytoin. Brain tomography demonstrated hypodensity in the occipital hemispheres and magnetic resonance imaging identified signal alteration in the white matter diffuse without diffusion restriction compatible with posterior reversible leukoencephalopathy syndrome. She evolved with adequate control of blood pressure and epileptic seizures, progressive improvement in visual acuity and muscle strength, until he returned to normality 10 days after the onset of symptoms. The antiepileptic drug was suspended, and she was discharged with antihypertensive medication.

Conclusion: Posterior reversible leukoencephalopathy syndrome is an important differential diagnosis in conditions of sudden deficits and seizures, especially in this infectious context, because it is a reversible clinical condition with a good prognosis.

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684 – Complete recanalization of large vessel occlusion in ischemic stroke with intravenous thrombolysis: case report

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Introduction: Intravenous recombinant tissue plasminogen activator (rtPA) treatment for acute ischemic stroke works by achieving recanalization of intracranial occlusion resulting in restoration of flow and prevention of infarct expansion. Data on recanalization after intravenous thrombolysis are performed by angiographic methods and transcranial Doppler monitoring studies. We aimed to report one case of complete recanalization of large vessel occlusion in ischemic stroke with intravenous thrombolysis with rtPA.

Case report: Pacient, 27 years old, male, brown, with coronary artery disease and ischemic cardiomyopathy, with clinical presentation of weakness in the

left side and speech alteration 75 minutes ago, with National Institutes of Health Stroke Scale (NIHSS) of 9 points. At admission, magnética ressonante imaging of the head showed restriction in the diffusion-wighted imaging sequence in basal ganglia and internal capsule topography, with magnetic resoance angiography (MRA) showing a stop in the M1 segment of the right middle cerebral artery (MCA). The patient underwent IV thrombolysis with rtPA 160 minutes after the ictus, uneventfully (NIHSS 0 points at the end of treatment). Control MRA (four hours after thrombolysis) showed complete recanalization of the M1 segment of the right MCA.

Discussion: Despite the known limitation of intravenous rtPA in recanalization of large vessel occlusion and the increasing use of mechanical thrombectomy, it is necessary to value the role of intravenous thrombolysis in this setting, considering the difficulties in accessing endovascular treatment still present in our country. More studies are needed on recanalization rates ntravenous thrombolysis in large vessel occlusion cases.

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685 – Comparison of efficacy outcomes with Vutrisiran vs. Patisiran in hATTR amyloidosis with polyneuropathy: post-hoc analysis of the HELIOS-A study

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Introduction: Vutrisiran and patisiran are approved RNAi therapeutics that reduce TTR protein production to treat hereditary transthyretin-mediated (hATTR) amyloidosis with polyneuropathy (PN).

Objectives: Assess the relative efficacy of RNAi therapeutics for hATTR amyloidosis with PN.

Methods: In the Phase 3 HELIOS-A study (NCT03759379), patients with hATTR amyloidosis with PN were randomized to vutrisiran (25 mg SC, Q3M) or patisiran (0.3 mg/kg IV, Q3W). Comparison of TTR reduction between vutrisiran and patisiran was included as a secondary endpoint. Here, post-hoc analyses comparing vutrisiran and patisiran arms on clinical outcomes are reported: neuropathy (mNIS+7), quality of life (Norfolk-QOL-DN), gait speed (10-MWT), nutritional status (mBMI), and disability (R-ODS).

Results: TTR reduction with vutrisiran was non-inferior to that observed with patisiran (median difference [95% confidence interval], 5.28% [1.17, 9.25]). Least-squares mean (±SE) changes from baseline to Month 18 for vutrisiran and patisiran, respectively, showed similar effects: mNIS+7 (0.06 ± 1.48 versus 1.53 ± 2.59; P = 0.6248), Norfolk-QOL-DN (−2.5 ± 1.8 versus −0.8 ± 3.0; P = 0.6472), 10-MWT (-0.019 ± 0.025 versus −0.053 ± 0.043 m/s; P = 0.4936), mBMI (21.8 ± 9.2 versus 7.6 ± 15.8; P = 0.4378), and R-ODS (−1.2 ± 0.5 versus −1.3 ± 0.9; P = 0.9266).

Conclusion: Vutrisiran and patisiran showed numerically and statistically similar efficacy for treating the polyneuropathy manifestations of hATTR amyloidosis and have similar pharmacodynamic effects in terms of TTR lowering.

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686 – Response of neuropathic pain to intravenous immunoglobulin in diabetic amyotrophy: a case report

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Introduction: A 58-year-old male patient, diagnosed with diabetes mellitus (DM) five years ago, started with neuropathic pain in the lumbar region radiating to the right thigh, and later to the left thigh, progressing to difficulty walking and significant weight loss over six months. On examination, patient with grade IV proximal paraparesis, slightly asymmetrical, associated with muscle atrophy and abolished reflexes. Regarding the upper limbs, the examination was normal. During the investigation, electroneuromyography (ENMG) showed sensory and motor involvement in the lower limbs, with a mixed pattern, associated with cerebrospinal fluid with hyperproteinorrachia. The rest of the exams did not show alterations, raising the hypothesis of diabetic amyotrophy (DA).

Discussion: DA affects less than 1% of all diabetic patients. Its typical clinical presentation is the presence of acute pain in the proximal region of the lower limbs, initially unilateral, evolving with paresis and muscle atrophy, as well as involvement of the contralateral limb in more advanced cases. The diagnosis is based mainly on clinical suspicion. ENMG demonstrates a pattern more related to axonal degeneration than demyelination, generally sparing upper limbs. Currently, there is no evidence to support or contraindicate any immunotherapy in the treatment of DA. In this case, IVIg was performed with excellent response in pain control.

Conclusion: Despite being a less common complication related to DM, DA becomes important in a scenario of increased incidence of metabolic diseases in the Brazilian population. It is important to know its presentation to aid in the diagnosis of a disease thais is rate but with an excellent response to IVIg infusion.

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691 – Thyroid dysfunction with the continued use of beta interferon in the treatment of multiple sclerosis

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Introduction: The treatment of multiple sclerosis (MS) with beta interferon has been shown to be effective in reducing the number of relapses and the inflammatory activity of the disease, as well as slowing its progression. Although there are controversies in the literature, recent studies have pointed to the possibility of thyroid dysfunction as a side effect of prolonged use of beta interferon.

Objectives: To analyze the presence of thyroid dysfunction due to the use of beta-interferon in patients with multiple sclerosis.

Methods: That is a documentary, observational, and analytical study, with a quantitative, retrospective, and cross-sectional approach. Sociodemographic variables, thyroid function laboratory tests, and medication usage time present in medical records were analyzed.
Results: From 118 medical records evaluated, after applying the exclusion and inclusion criteria, 35 were included in the study. 80% of the sample were female, mean age was 32.43. The mean values for TSH and free T4 before starting treatment with beta-interferon were 1.66 mIU/L and 0.96 ng/dL, respectively. After 6 months, the values for TSH and free T4 were 2.20 mIU/L and 0.91 ng/dL, respectively. It can be concluded that the continuous use of beta-interferon did not lead to significant changes in TSH and free T4 levels compared to baseline (P = 0.061 and P = 0.102, respectively). It is worth noting that there were two cases of subclinical hypothyroidism developed during the study.
Conclusion: MS patients using beta-interferon are mostly young women. No significant changes in their thyroid function were observed. Therefore, based on this information, the need for routine periodic laboratory tests to evaluate thyroid function in patients using beta-interferon is questioned. Further studies are needed with a larger sample size to definitively clarify this topic.

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693 – Cocaine abuse and risk of induced carotid artery dissection

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Introduction: Cocaine is among the most commonly used illicit recreational drugs worldwide, and its abuse significantly increases the risk of ischemic stroke. Preclinical studies have indicated that cocaine may stimulate arterial dissection, during which blood enters the arterial wall through an intimal tear and activates the coagulation cascade at the site of endothelial damage. This is a case report of a patient, who presented an ischemic stroke, with a history of cocaine abuse in the last 24 hours. The article was developed from data collection through literature review on the subject, as well as analysis of the patient's medical records.
Case report: A 28-year-old man with a history of previously healthy, with a report of cocaine abuse in the last 24 hours, found unconscious on a public road and upon waking up he had aphasia and right hemiplegia, scoring NIHSS 18. Patient was investigated with brain resonance which showed an extensive acute ischemic infarction compromising the frontal, temporal, insula and basal ganglia regions on the left. The etiological investigation discarded the hypothesis of a cardioembolic and/or autoimmunity phenomenon. The arteriography showed an occlusion of the internal carotid artery at its origin with an aspect of arterial dissection.
Conclusion: Cocaine-induced dissection of various arteries has been described in the literature, leading to cardiac and neurological complications. The combination of hypertension and intense vasoconstriction result in shear stress on the arterial wall, which may lead to atherosclerotic plaque rupture in those with vascular disease, and an increased risk of dissection even in those with normal arteries, as was the case with our patient. Therefore, public policies against drugs are also means of preventing strokes, especially in the younger population.

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695 – Evaluation of lung function and respiratory muscles in Duchenne muscular dystrophy

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Introduction: Duchenne muscular Dystrophy (DMD) is a genetic disease of recessive inheritance linked to the X chromosome, caused by a mutation in the dystrophin gene. This mutation will result in absence of the dystrophin protein, leading to the degeneration of muscle skeletal. The disease is the most common childhood-onset form of muscular dystrophy and affects males almost exclusively. DMD symptoms onset occurs in early childhood, usually between the ages of three and five years, with progressive muscle weakness and loss of gait in adolescence, progressive cardiomyopathy, and respiratory failure, leading to death. Spirometric parameters such as forced vital capacity (FVC) are used to monitor lung function. Muscle ultrasound has been increasingly used in neuromuscular diseases, being a possible tool for evaluating respiratory muscles individually and a non-invasive method of assessing diaphragm function.
Objectives: This study aims to evaluate lung function and respiratory muscles in patients with DMD at different stages of the disease.
Methods: This is a prospective observational study with 25 patients with DMD follow-up at the Hospital das Clínicas de São Paulo (HC/FMUSP), with Assessment of diaphragmatic thickness by ultrasonography in DMD patients and correlate with FVC.
Results: Diphragmatic thickness significantly decreased with age and with the reduction of the FVC in DMD patients. Some patients had pseudo hypertrophy of the diaphragm but without related reduction in lung function.
Conclusion: Ultrasound of respiratory and appendicular muscles can help in the assessment of respiratory function and possible indirect markers of worsening lung function.

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696 – Description of electroneuromyographic findings in leprosy: confirming a spectral neuropathy

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Introduction: Leprosy is the most common cause of peripheral neuropathy, leading to sequelae that perpetuate the stigma linked to the disease. Electroneuromyography (ENMG) is the gold standard to evaluate neural damage, as it can detect even subclinical alterations.
Objectives: To describe the electroneuromyographic findings in leprosy patients.

Methods: Retrospective analysis of the records from patients with leprosy in a Brazilian Reference Center, between 2014 and 2021. The sensibility conduction of the median, ulnar, radial, sural and peroneal nerves, and motor conduction of the median, ulnar, peroneal and tibial nerves were analyzed.
Results: Five hundred and thirteen patients with leprosy submitted to ENMG at the diagnosis were analyzed. The mean age was 45.8 years (±16.6) with 50.1% being male, and multibacillary represented 85.8% of the cases. Of these patients, 71.9% presented neural damage compatible with leprosy. In total, 2,627 damaged nerves were found, with a mean of 7.1 (±5.2) per patient. The most affected sensitive nerve was the superficial peroneal 15.8%, with a mean of 4.4 (±3.3) damaged nerves per patient. The most affected motor nerve was the ulnar 12.9%, with a mean of 2.7 (±2.5) damaged nerves per patient. Between the 369 patients with neural leprosy damage, 19.2% presented with mononeuropathy, while 80.8% presented two or more damaged nerves. Asymmetric sensorimotor axonal neuropathy with focal slowing of conduction velocity was the most common ENMG pattern 53.9%.
Conclusion: Leprosy is a spectral disease, therefore, the damage and neurophysiological characteristics can vary according to the clinical presentation. The ENMG pattern confirms that there is a greater involvement of sensitive nerves asymmetrically, mainly axonal, with localized non-uniform myelin damage.

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697 – Analysis of the distribution of neurologist among Brazilian regions

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Introduction: Brazil has 490,077 active physicians, but these professionals are concentrated in large urban centers, following a historical trend. However, there is a lack of data related to whether neurologists (1.81% of physicians) also follow such disproportion.
Objectives: To analyze the distribution of neurologists in Brazil in December 2022.
Methods: This is a descriptive analytical ecological study using data obtained from the IBGE 2022 Demographic Census and the National Register of Health Establishments (CNES) provided by DATASUS, regarding the number of neurologists in December 2022, using the variables: number by region, number by capital and works in SUS.
Results: In December 2022, there were 8841 neurologists in Brazil, representing a ratio of 0.043 neurologists/1000 inhabitants. The North (N) and Northeast (NE) regions had the lowest proportions, with 0.017 and 0.027 neurologists/1000 inhabitants, respectively. Of the total number of neurologists, 54% were located in the Southeast (SE) region, while only 3.38% were in the N region. Of these physicians, 63.60% worked in the SUS, with higher prevalence in the NE (76.02%) and N (73.58%) regions. In addition, 40.89% of neurologists were located in state capitals. This concentration ranged from 30.72% in the South (S) region — next to SE (36.09%), the only regions with less than half of their neurologists in capital cities — to 65.57% in the Midwest (SW) region.
Conclusion: The data presented point to an inequality in the distribution of neurologists in Brazil. The SE region concentrates the majority of professionals, while the N and NE regions have a relative deficit. Moreover, an even greater shortage was observed within these regions, along with the SW region. In the N and NE regions, most neurologists work in the SUS, ensuring access to health care to a historically neglected population.

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700 – Super refractory status epilepticus in a child: brain on "fires"

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Introduction: New onset of a refractory status epilepticus (NORSE) is a clinical presentation, without a clear cause, within the first 72h of admission of a non-epileptic patient. FIRES (Febrile Infection-Related Epilepsy Syndrome) is a NORSE subgroup with febrile prodrome 24 hours to 2 weeks before the onset of status. This case reports a previously healthy 7-year-old female with FIRES and super refractory status epilepticus (SE).
Methods: This is a case report based on retrospective analysis of a single patient's medical record.
Results: The patient presented with SE four days after low grade fever and airway infection. Midazolam (MDZ) 0.15 mg/kg, phenytoin (PFT) 30 mg/kg, phenobarbital (PB) 15 mg/kg were administered in sequence; followed by continuous infusion of MDZ 1 mg/kg/h and ketamine (KET) 30 mcg/kg/min. Empirical treatment started with ceftriaxone, acyclovir and ampicillin. Exams on admission: Brain magnetic resonance (MRI) with no abnormalities; normal serum leukocyte count and electrolyte levels; cerebrospinal fluid (CSF) with 61 cells (71% lymphocytes), protein 38, Lactate 15, glucose 61 (mg/dL). CSF on two different occasions was negative for infectious agents and autoimmune antibodies. Electroencephalograms (EEG) on the 3rd, 4th and 7th days after hospitalization revealed SE despite the followings drugs: Levetiracetam 60 mg/kg, Lacosamide 10 mg/kg, topiramate 10 mg/kg, MDZ 2 mg/kg/h, KET 2 mg/kg/h. Throughout hospitalization, she received a 3 days course of methylprednisolone 30 mg/kg/day, followed by five days of immunoglobulin 2 g/kg/day, ketogenic diet and thiopental 5 mg/kg/h. From the 13th day on, EEG maintained global suppression and no epileptiform activity. The patient died on the 18th day after a septic shock.
Conclusion: NORSE and FIRES are entities with unfavorable outcomes and high mortality rates. Autoimmune/inflammatory encephalitis represents 40% of NORSE causes.

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701 – Intraventricular baclofen for spasticity – a forgotten strategy?

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Introducion: Spasticity is persistent increased muscle resistance to joint movement. Treatments for this condition aim to prevent or minimize its evolution into severe movement restrictions. Among the therapeutic options are the use of oral medications or the surgical approach for continuous baclofen infusion, in which two forms of administration can be used: intraventricular or intrathecal. However, the intraventricular approach has been replaced by the intrathecal approach in most cases, making it necessary to

investigate in the literature the possible reasons for such a change. Perhaps it is a forgotten therapy?

Objectives: The present paper aims to address spasticity, as well as review the main indications and results of the use of intraventricular baclofen when compared to the intrathecal approach.

Methods: This is an integrative literature review. The database used was PubMed. Using the terms "Baclofen", "Intraventricular", "Intratecal" and "Spasticity", associated with the Boolean term "AND" to direct the search. Articles from the last five years that addressed the intraventricular and intrathecal techniques of baclofen administration were included.

Discussion: Spasticity has several forms of surgical treatment. The intraventricular approach is indicated in cases of cerebral palsy, spinal cord injury and stroke, but mainly for cases of dystonia and spasticity refractory to conventional drug treatment.

Conclusion: When comparing the use of intraventricular and intrathecal baclofen, the intrathecal approach, despite being a technique used for over 30 years and for similar conditions, is still an effective therapeutic measure with low complication rates, depending on the technique used.

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703 – Miller-Fisher syndrome in puerperium

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Introduction: Miller-Fisher syndrome (MFS) is recognized as a variant of Guillain-Barré syndrome, composed of the clinical triad of ataxia, areflexia and ophtalmoplegia. The association between MFS and pregnancy/postpartum has been described in few cases previously.

Case presentation: Woman, 28-year-old, nine days after normal partum present paresthesia and weakness in arms, a day later presenting with abnormal gate and diplopia. No recent infections or vaccination was described. The examination showed gait ataxia, right sixty nerve palsy, muscle strength in legs MRC grade 4 and hypotonia. The Patellar and Achilles tendon reflex was absent. No alterations in sensitive examination. Cerebrospinal fluid (CSF) showed 1 cell, protein 41 mg/dL, with infectious analysis negative. Laboratorial tests and magnetic resonance imaging were unmarkable. Electroneuromyography (ENMG) showed reduced amplitude of the sensory action potential in bilateral sural and ulnar nerves. The set of clinical and ENMG findings diagnosed MFS. She was admitted to the intensive care and realized five plasmapheresis sessions, showing significant improvement of symptoms. Four months later, she was asymptomatic of the neurological condition.

Discussion: In PubMed, we found a total of four cases in pregnancy were reported, and no case in puerperium. The effect of hormonal changes of pregnancy/puerperium presents on the immune system make these periods special situations in the understanding of the MFS. A particularity of our case is the normal CSF count of cells and protein levels, which may be normal in 30–50% of patients in first week of symptoms. Anti-GQ1b antibodies are found in up to 90% of patients with MFS, however it is not necessary to establish the diagnosis. When MFS is suspected, the recommendation is not wait for antibody tests to starting treatment.

Conclusion: The relationship of MSF and pregnancy/puerperium is few described, but the evolution of these cases seems to be like the other patients.

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704 – Bilateral infiltrative optic neuropathy in a patient with metastatic gastric cancer: a case report

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A 66-year-old woman, newly diagnosed with a gastric adenocarcinoma with signet-ring cells, presented with bilateral retro-orbital pain, with worsening eye movement. After seven days, she developed a loss of visual acuity in the right eye, with an altitudinal pattern and rapid evolution to amaurosis. A week later, she noticed the loss of contralateral visual acuity, with field constriction. She sought an ophthalmologist, who performed fundoscopy, finding occlusion of the central retinal vein on the right and bilateral papillary edema. Prednisone 60 mg was prescribed for seven days. Due to a lack of improvement, she searched our service for evaluation. Examination findings revealed only light perception in the left eye and no light perception in the right eye. A bilateral absolute afferent defect was also found. Fundoscopy revealed bilateral papillary edema. Magnetic resonance imaging revealed thickening of the optic nerves and chiasm, with diffusion restriction, in addition to FLAIR hyper signal next to the optic nerve sheath, which exhibits gadolinium enhancement with densification of the surrounding intraorbital fat. This may represent a metastatic spread of the underlying disease. Cerebrospinal fluid analysis showed 4 cells, with 33% of neoplastic cells. Glucose and protein values were within normal range and cultures for both bacteria and fungi were negative. Blood test findings were unremarkable. Computed tomography of the abdomen showed diffuse gastric thickening, in addition to a left adrenal lesion suggestive of secondary involvement. The patient was referred to an oncology service for treatment and follow-up of metastatic gastric neoplasia. Few cases of metastatic involvement of the optic nerve are reported in the literature. Therefore, the cautious approach of a patient with visual loss and cancer history would avoid misdiagnosis.

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705 – Epidemiological 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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Introduction: Multiple sclerosis (MS) is an inflammatory, immune-mediated disease of the central nervous system, characterized by multifocal areas of demyelination. There is a knowledge gap in understanding the disease, particularly the progressive forms in different regions of the country, such as the Northeast.

Objectives: 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.
Results: From 283 patients 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 and 18 (50%) were high educated. The median follow-up time was five years and the peak prevalence was between the 4th and 5th decades. 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 clinical-epidemiological profile of MS patients with a progressive phenotype is essential to guide the propaedeutic reasoning, guise the conduct and restructure the dynamics of health services.

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707 – Long thoracic nerve paralysis as a rare complication of anterior spinal surgery

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Introduction: Isolated long thoracic nerve paralysis causes weakness of the serratus anterior muscle and winging of the scapula. Several traumatic and non-traumatic causes of damage have been reported and should be considered for diagnosis.
Case: 53-year-old woman with neck pain and sensory symptoms, with irradiation to the upper limbs. She was then submitted to an anterior spinal release at levels C5-C6 e C6-C7. In this procedure the patient was positioned in the dorsal decubitus position with a rolled towel under her scapulae bilaterally. After two weeks she complained of weakness of the right upper limb during overhead activities, mild fatigue and pain in the right periscapular region. The right upper limb was intact for both sensory and motor innervation, but the right scapula was winged and she was not able to flex her arm forward over 60°. An electrodiagnostic study carried out and revealed an isolated acute axonal long thoracic nerve lesion.
Discussion: The long thoracic nerve innervates the serratus anterior muscle. When this muscle is paralyzed, the scapula's medial border, and especially its lower angle, stands out prominently and the patient cannot raise the arm fully or push. Due to its long and superficial course, the long thoracic nerve is vulnerable to damage at various levels. Several causes of its damage have been reported, including trauma, compression, and improper surgical technique. In this case, despite the operation was not that long it appears that external compression on the nerve during surgery was the most probable cause of the nerve palsy.
Conclusion: To avoid this injury during positioning of patients the course of the long thoracic nerve must be attended to carefully and the nerve should be protected from any external pressure.

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709 – Clinical features and treatment approach of sporadic inclusion body myositis: a single-center experience

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Introduction: Sporadic Inclusion Body Myositis (sIBM) is the most common idiopathic inflammatory myopathy after age 50, usually resistant to treatment. To report our clinical experience with sIBM, we performed a retrospective analysis of clinical data from 33 patients diagnosed by 2011 European Neuromuscular Center diagnostic criteria and followed in our outpatient clinic from 2017 to 2023.
Results: From the 33 patients included, 54.5% were male with median age of onset of 59.7 and median time of disease of 11.1 years. Related comorbidities were cardiovascular (60.6%), T-cell large granular lymphocytic leukemia (9%), HIV (6%), hepatitis C (3%), and monoclonal gammopathy (3%). About functional motor status, 48.4% of patients walked independently, 36.3% required unilateral or bilateral support to walk, and 15.1% were restricted to wheelchair. Dysphagia was reported in 57.6% of patients and 54.5% underwent physical therapy and/or speech therapy. Regarding pharmacological treatment, 64% of patients received methotrexate (MTX), associated with steroids in 2 patients (who had rapid clinical course) and with Intravenous immunoglobulins (IVIg) in other 4 patients; 2 patients received IVIg only. Among the 24 treated patients, the mean age was 61.2 years, 18 walked independently and 6 required support to walk, 14 had dysphagia. After a median period of 1.7 year of follow-up, 14/20 patients continued to use MTX considering a positive, occasionally transient, response with referred stabilization or improvement of muscular strength. All patients who received IVIG had improvement in dysphagia.
Conclusion: In our center, some patients with sIBM are treated with immunomodulator treatment, especially with MTX and IVIg, with stabilization and occasionally improvement. Further controlled studies are needed to define a subgroup that can have beneficial response.

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711 – Prevalence and impact on the quality of life of urinary incontinence in patients with ischemic stroke: a cross-sectional study

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Introduction: Acute stroke is the second most prevalent cause of death in the world and an important cause of disability in adults. Urinary incontinence (UI) is a complication present in the acute and chronic phases of stroke and is a strong predictor of mortality and need for institutional care.

Objectives and methods: Cross-sectional study that aimed to evaluate the presence of UI through the International Consultation Questionnaire on Incontinence - Short Form (ICIQ-UI SF) in patients with ischemic stroke followed in the neurovascular outpatient clinic of a tertiary hospital in São Paulo, Brazil. The ICIQ-SF assesses the impact of UI on quality of life and qualifies degrees of urinary loss. The final score ranges from 0 to 21, with higher values representing greater impact.

Casuistics and results: A total of 141 patients adequately answered the questionnaire, among which 75 were female and 66 were male. As for symptoms, 36.87% reported some degree of UI, 40.00% among women and 33.33% among men. The mean ICIQ-UI SF score across all patients was 9.90. For males the mean score was 10.36 and for females it was 9.56. Among symptomatic patients, only 9.21% reported urinary symptoms before the stroke.

Discussion: Almost 40% of the patients in the study acquired UI after stroke. This condition prevailed in women, but with less impact compared to men. Urinary incontinence can affect the quality of life of patients, causing psychological, social, physical, economic, personal and sexual relationships.

Conclusion: New studies are needed to evaluate prevention and treatment strategies for incontinence in these patients.

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713 – Lower limb weakness: a challenge diagnosis

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A 46-year-old male patient started a condition of lower back pain radiating to the lower limbs in 2016. He underwent lumbar arthrodesis surgery between L5-S1 due to a herniated disc and reports that after the surgery, his clinical condition improved, remaining asymptomatic over four years. In 2020, he started to experience hypoesthesia in the calf and dorsal region of his right foot, with progressive worsening throughout the year, with a tripping sensation associated with a numbness and decreased distal strength of the right lower limb, with subsequent difficulty in climbing stairs over the next year. He continued to follow up with orthopedics and began following up with neurosurgery, which did not identify a clear etiology for his clinical condition. In 2021, the patient was already complaining of worsening weakness in the right foot with foot drop and a decrease in trophism of the entire right lower limb. The patient denies sphincter alterations or sexual dysfunction throughout the course of the disease. In March 2022, the patient began using unilateral support due to falls and returned to the neurosurgery outpatient clinic with a referral to clinical neurology. He denies recent worsening of strength. In October 2022, during a consultation with neurology, treatment with prednisone 20 mg/day was started without improvement in the clinical condition, and during a follow-up visit in January 2023, the need for diagnostic elucidation led to the patient being hospitalized at the Hospital do Servidor Publico Estadual for etiological investigation. On neurological examination, the patient presented alterations in the right lower limb, with grade IV proximal strength and II distal strength of this limb, including plantar and dorsiflexion flexion, associated with global hypotrophy of the limb up to the gluteal region, with normoactive deep tendon reflexes and plantar reflex in flexion. With regard to sensitivity, the patient presented hypoesthesia in the lateral region of the right leg and the sole of the right foot, worse distally in the topography of L4 L5-S1, with preserved artresthesia and palesthesia. No other alterations were present on the somatic examination, except for a right-hand scraping gait with a component of proximal weakness. In complementary investigation, no signs of spinal cord compression lesions were found on magnetic resonance imaging, no inflammatory signs were found in cerebrospinal fluid results, and no systemic involvement was found in chest and abdominal computed tomography exams, except for asymptomatic hepatosplenomegaly. Electroneuromyography (ENMG) revealed motor impairment with preserved sensory parameters, leading to a preganglionic pattern of impairment, in addition to recent denervation findings with fibrillations and positive sharp waves, which were also visualized in the contralateral lower limb in a lesser degree of involvement. Despite the hypoesthesia on the physical exam, no sensory alterations were demonstrated on ENMG, which may be justified by the previous spinal surgery. Due to findings on the neurological and complementary exams, with impairment only in the right lower limb, substantial atrophy, and weakness with ENMG findings, the hypothesis of flail leg syndrome, a variant of amyotrophic lateral sclerosis (ALS) with lower limbs onset, was raised. Flail leg syndrome is a rare variant (6%) of atypical forms of amyotrophic lateral sclerosis, predominantly affecting males between 55–65 years of age. Its manifestation includes asymmetric and progressive distal onset weakness and wasting with restricted involvement of the lower limbs for at least 12 to 24 months. ENMG shows fibrillations and positive sharp waves, with little evidence of fasciculations. It has a significantly better prognosis in terms of median and 5-year survival rates compared to bulbar and limb onset ALS, due to its slower progression to other locations. FLS ir rare, but it should be considered as an alternative diagnosis when there is limited impairment of lower limbs after at least one year of clinical observation.

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714 – Primary CNS lymphoma mimicking demyelinating disease

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Case report: Male, 32-year-old, diagnosed with COVID-19 (coronavirus disease) in January 2022 and two weeks later present progressive vertigo, gait instability, binocular diplopia and right facial palsy. Three months later he was sent for investigation in our service. Magnetic resonance imaging (MRI) showed a FLAIR-hyperintense lesion in the pontomesencephalic transition with gadolinium enhancement. Cerebrospinal fluid (CSF) analysis shows increase in protein level (83 mg/dL). In this moment, considering the infectious antecedent and MRI characteristics, he has been diagnosed with acute disseminated encephalomyelitis (ADEM) and submitted to pulse therapy with methylprednisolone, showing complete improve of symptoms. In outpatient follow-up he presents with worsening of symptoms. Ten months after onset of symptoms he was hospitalized again, MRI showed new lesions on medulla and cerebellum. Submitted to pulse therapy followed by plasma exchange, with no improve. New MRI presents with infiltrative lesion without gadolinium enhancement. CSF analysis show 48 cells (98% lymphocytes), no evidence of neoplastic cells. A biopsy was performed on the lesion and showed atypical lymphoid infiltrate with B-Cells predominance, positive to CD20 and CD45, confirming diagnosis of Primary CNS Lymphoma (PCNSL). Initiated chemotherapy, but unfortunately patient die in consequence of neutropenia.

Discussion: PCNSL is an uncommon neoplasm and rarely affects brainstem. When it occurs the prognosis is poor, nearly half of patients died within 1 year after diagnosis. Glucocorticoids could promote tumor shrinkage. Neuroimaging characteristics could be confounding with infections, demyelinating lesions, and gliomas.

Conclusion: Our case illustrates a patient initially diagnosed with ADEM and submitted to corticotherapy with good response, however he worse in months later and the diagnosed reviewed to PCNSL. This shows the importance of considering atypical PCNSL in differential diagnosis of demyelinating diseases.

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715 – History and evolution of the neurological hammer: an integrative review

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Introduction: The neurological hammer has been a fundamental tool in neurological assessment for over a century. However, its history and evolution are still little known.
Objective: This study aims to investigate the history and evolution of the neurological hammer, based on an integrative review of the literature.
Methods: A Integrative review was conducted in the Scielo, PubMed, and Cambridge Core databases using the keywords "neurological hammer", "history of neurology", and "clinical neurology". Were analyzed seven articles, and selected one book with historical content to compose the review, published between 1998 and 2021. The analysis included the materials used in the construction of the hammer, its use, and the changes that occurred over time. The aim of this study was to investigate the history and evolution of the neurological hammer.
Results: The neurological hammer was invented by Ernst L.O. Oppenheim in 1888, with the aim of creating a softer tool to test reflexes in animals. The original instrument was made of wood and had a soft rubber end. Over the years, the hammer has evolved into a variety of shapes. New materials were introduced in its construction, such as metal, plastic, and harder rubber. In addition, the shape of the hammer's tip also changed, with the introduction of triangular tips, star-shaped tips, and pin-shaped tips.
Conclusion: The history and evolution of the neurological hammer is an important part of the history of medicine and the evolution of diagnostic tools. Knowledge of the evolution of the neurological hammer can help to better understand the origin and development of modern medical practice. In addition, the integrative review presented here can contribute to improving the construction and usage of the neurological hammer in neurological assessments, for the benefit of the patient's health.

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716 – Prevalence of sexual dysfunction in women with ischemic stroke: a cross-sectional study

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Introduction: Stroke is characterized by the acute onset of one or more neurologic deficits that persist for at least 24 hours, and is the result of a vascular disorder in a defined territory, being the leading cause of disability. Late complications, such as sexual dysfunction, have a direct impact on psychological and emotional aspects and directly affect the quality of life of these patients.
Objectives and methods: Cross-sectional study that aimed to determine the prevalence of sexual dysfunction in patients with ischemic stroke followed in the neurovascular outpatient clinic of a tertiary hospital in São Paulo, Brazil, using the Female Sexual Quotient (FSQ) questionnaire. The FSQ can range from 0-100 points. Higher values indicate better sexual performance/satisfaction and a score of 60 points or less was considered as having sexual dysfunction.
Casuistics and results: Sixty-five questionnaires were analyzed and classified according to the FSQ score. Mean age was 52 years and median FSQ score was 54 points. Of the interviewees, 30.7% had their sexual performance classified as poor-null, 13.85% of patients had poor-unfavorable sexual performance, 15.38% as unfavorable-regular. Regular-good and good-great sexual performance were found in 21.54% and 18.46%, respectively.
Discussion: In this study, only 40% of post-stroke women have regular-good and good-great sexual performance. The impact of the stroke on the sexual life of these patients is high, because most of the interviewees had sexual dysfunction according to FSQ.
Conclusion: The health professional must be aware of the non-motor sequelae caused by the stroke and be prepared to help these patients in coping with sexual dysfunctions.

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717 – Risk assessment for obstructive sleep apnea in stroke patients: a cross-sectional study

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Introduction: Stroke is the second most prevalent cause of death in the world and an important cause of disability in adults. Obstructive sleep apnea (OSA) is an independent risk factor for stroke and is associated with poor post-stroke functional outcome if left untreated.
Objectives and methods: In this cross-sectional study, we aimed to determine the prevalence of OSA in ambulatory stroke patients using the STOP-Bang (snoring, tiredness, observed apnea, blood pressure, body mass index, age, neck size, gender) questionnaire.
Casuistic and results: A total of 149 patients completed the clinical assessment. Mean age was 57.47 years, 75 (50.34%) were men and 74 (49.66%) were female. Among the patients, 56.37% were smokers or former smokers, and mean body mass index was 27.01 kg/m^2. The median STOP-Bang score was 3 (interquartile range: 2–4). 69.12% were considered moderate to severe risk for OSA (STOP-Bang score ≥3). For the identification of OSA, the questionnaire demonstrated sensitivity of 83.5%, accuracy of 75.2% and specificity of 45.5%. Among the 84 patients who answered the question about the presence of previous OSA symptoms, 59.53% had a negative answer.
Discussion: Approximately two out of three stroke patients were classified as moderate to high risk for OSA, reinforcing the need for screening for the disease in this population. In addition, almost 60% reported no symptoms prior to cerebrovascular disease. Finally, the high sensitivity in this study confirms the usefulness of the tool in screening patients for diagnosis and therapeutic intervention.
Conclusion: New studies are needed to evaluate OSA prevention and treatment strategies, as this measure will help in the global clinical recovery of the patient after the stroke.

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718 – Relationship between social support and spirituality and religiosity in epilepsy in adults

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Introduction: There is still no consensus in the literature on how religiosity/spirituality (R/S) would act as an instrument of social support in individuals with chronic diseases.
Objectives: To evaluate the perception of social support and R/S measures in adult patients with epilepsy (PCE).
Methods: Data from the Brief Multidimensional Measure of Religiosity/Spirituality (BMMRS-P) and the Social Support Satisfaction Scale (ESSS) of 44 PCE were evaluated. BMMRS-P and ESSS data were related to the clinical variables of epilepsy, with statistical tests, with $P < 0.05$.
Results: Gender was female in 28 (63.6%) cases, mean age was 44.3 ± 14.3 years, and mean disease duration was 28.1 ± 16.1 years. The reported religion was Catholic in 25 cases, Evangelical in 13 cases, other in 2 cases and no religion or agnostic in 4 cases. The total score on the BMMRS-P self-assessment was 4.2 ± 1.7, and the total score on the ESSS was 55.5 ± 4.0. Younger PCE have less organizational religiosity (Person correlation; -0.360; $P = 0.019$). There is a greater perception of forgiveness (0.367; $P = 0.024$) and religious and spiritual overcoming (0.437; $P = 0.006$) in PWE with younger age at the time of the first epileptic seizure. The duration of epilepsy was negatively correlated with daily spiritual experience (-0.310; $P = 0.040$), with religious support (-0.440; $P = 0.003$) and with organizational religiosity (-0.309; $P = 0.041$). Schooling correlated with organizational religiosity (0.401; $P = 0.009$). There was a significant correlation between the perception of social support in the friends dimension and religious support (0.325; $P = 0.031$) of the BMMRS-P. There was no significant difference in the perception of social support and R/S according to the epilepsy variables and the type of epilepsy syndrome.
Conclusion: Social support in epilepsy is important for PWE to deal with the unpredictability of epileptic seizures, with the stigma and social restrictions that these individuals are sometimes subject to. The use of R/S can positively contribute to coping with psychosocial aspects of epilepsy.

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719 – Epidemiological profile of Stroke hospitalizations in Brazil from 2017 to 2022 and the role of primary health care

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Introduction: Stroke is the most disabling neurological disorder and the second leading cause of death in Brazil. In turn, the acute and chronic clinical repercussions lead to the need for Primary Health Care (PHC) to act as a tool for access, continuity and quality of care, as well as to prevent risk factors.
Objectives: To identify the profile of patients hospitalized for stroke in Brazil and the role of PHC.
Methods: Quantitative, descriptive and ecological study based on TabNet data on stroke hospitalizations in Brazil during the years 2017 to 2022, using the variables processing year, region, sex and age group 1.
Results: There were 971,205 hospitalizations for stroke with emphasis on 2022, which recorded 181,190 cases, 13% higher than the previous year and 16% above the average of the previous 5 years. By regions, the Southeast is the leader (42.4%), followed by the Northeast and South. According to gender, there was a predominance of male patients 10% more than females, a pattern observed in all regions. Per age group, the elderly (72%) were the most affected, with prevalence between 70 and 79 years (26.2%), highlighting the relation of population aging with the pathophysiological process of the disease. In this sense, it denotes the importance of the role of PHC in acting in primary prevention and control of risk factors related to stroke, as well as in the secondary prevention of new events, to reduce the number of hospitalizations.
Conclusion: Stroke is a disease with high morbimortality. The epidemiological profile can be a potential tool for identifying and directing resources to PHC, especially considering regional sociodemographic aspects in order to offer prevention, quality care and resolve bad outcomes.

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720 – Ventilatory support and telemonitoring in patients with neuromuscular disease: a prospective study.

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Introduction: Neuromuscular disorders (NMD) are diseases of the peripheral nervous system with systemic manifestations. The ventilatory impairment could be one of the most serious complications and can lead to respiratory failure, either chronically or acutely.
Objectives: To describe the effects of non-invasive ventilation (NIV) associated with telemonitoring in patients with NMD.
Methods: A prospective study of patients with NMD and indication for NIV due to a reduced FVC on spirometry or respiratory symptoms was done. These patients were followed up with periodic exams, telemonitoring and adjustments of ventilatory parameters for six months.
Results: 37 patients were included, 56.7% male, average age 49.7 years (±12.6). 69.5% reported dyspnea before the indication of NIV and 10% needed medical attention. The average forced vital capacity (FVC) at inclusion was 67.7 (±14.9) and 72.4 (±15.7) after six months, with average diurnal oxygen arterial saturation of 95.22 (±5.4) at inclusion and 95.7 (±5.1) after 6 months. Throughout the study, 4 (10.8%) patients died and 2 (5.4%) underwent tracheostomy. Between inclusion and after 6 months, according to telemonitoring, there was an increase in the average daily use in hours from 5.5 (±2.2) to 5.6 (±2.0), with a drop in leakage (L/min) from 1.9 (±6. 6) to 1.0 (±7.0) and in the AHI from 3.15 (±4.9) to 2.2 (±4.3).

Conclusion: Diurnal oxygen arterial saturation was not a good parameter for indicating NIV, as well as FVC < 70, since most patients already had respiratory manifestation. Regular follow-up of patients associated with telemonitoring showed an improvement in ventilatory parameters, maintaining stability in adherence to therapy over the six months.

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722 – Ehlers-Danlos syndrome: an important differential diagnosis for congenital myopathies

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Kyphoscoliotic Ehlers-Danlos syndrome (kEDS) is a genetic collagen disorder, with early onset hypotonia, weakness, progressive kyphoscoliosis, joint hypermobility, and other features underlying connective tissue involvement. On the other hand, congenital myopathies (CMs) are genetic muscle disorders, with hypotonia and weakness usually from birth which can associate with skeletal deformities.

Case 1: RSS, 2-year-old, male. At birth, hip dysplasia and global hypotonia. On examination, proximal tetraparesis, global hyporeflexia and hypotonia, joint hypermobility, ogival palate, umbilical hernia, scoliosis and clubfoot. Genetic testing: Homozygous pathogenic variant in the *FKBP14* gene (ENST0000000222803-c.362_363insC, p.Glu122Argfs*7), associated with kEDS type II.

Case 2: RRO, 37-year-old, female. Global hypotonia at birth with congenital hip dislocation. On examination, mild proximal weakness, global hyporeflexia, joint hypermobility, scoliosis and ogival palate. Absence of marfanoid habitus. Genetic testing: Homozygous pathogenic variant in the *PLOD1* gene (ENST00000196061-c.2032G>A, p.Gly678Arg), associated with kEDS type I.

Conclusion: Uncommon clinical features in CMs should alert for an alternative diagnosis. Extreme joint laxity is more often a sign of a connective tissue disease, and can be associated with Marfanoid habitus, arterial dissection, bluish sclerae, umbilical hernia. Both patients presented nonspecific findings like neonatal skeletal deformities, hypermobility and mild weakness. A wide genetic test was definitive for the correct diagnosis. EDS must be included in the differential diagnosis of congenital myopathies, especially when associated with involvement of connective tissue. This is very relevant in the management care of the patients.

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723 – Cerebral cryptococcoma successfully treated by isavuconazole in an immunocompetent patient: a case report

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Introduction: Cryptococcosis has been recognized as an increasing cause of severe systemic mycosis in immunocompetent patients in recent years. Radial cryptococcomas are a more uncommon manifestation of cryptococcal meningitis, which are usually not included in the differential air masses. Thus, transmission of the disease occurs by inhalation of cryptococcus spores, initially infecting the lungs. Infectious propagules may also be restricted to the central nervous system, causing cryptococcal meningitis, meningoencephalitis, and respiratory cryptococcomas.

Case report: We report the case of a young immunocompetent woman who rapidly evolved with severe neurological deficits. She was finally diagnosed with cerebral cryptococcoma caused by cryptococcus neoformans and *Cryptococcus gattii*, and was treated with amphotericin B and isavuconazole. This is a case report on antifungal drug treatment in immunocompetent patients with cerebral cryptocoma. After several complications during hospitalization, including hydrocephalus and cerebellitis, she was discharged on isavuconazole. At follow-up, he complained only of anosmia. We reviewed the clinical and radiological findings of similar cases. It is the first time that this form of cryptococcal meningitis is favorably treated with isavuconazole and is caused by two species of Cryptococcus.

Conclusion: We emphasize that respiratory cryptococcomas should be suspected in immunocompetent patients who have brain masses or whose symptoms can progress rapidly, sometimes with a lethal outcome.

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726 – Prevalence of sexual dysfunction in men with ischemic stroke: a cross-sectional study

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Introduction: Stroke is the leading cause of disability worldwide. Late complications, such as sexual dysfunction, have a direct impact on psychological and emotional aspects and directly affect the quality of life of these patients.

Objectives and methods: We present a cross-sectional study that aimed to determine the prevalence of sexual dysfunction in patients with ischemic stroke followed in the neurovascular outpatient clinic of a tertiary hospital in São Paulo, Brazil, using the Male Sexual Quotient (MSQ) questionnaire. The MSQ can range from 0-100 points. Higher values indicate better sexual performance/satisfaction and a score of 60 points or less was considered as having sexual dysfunction.

Casuistics and results: Fifty eight questionnaires were analyzed and classified according to the MSQ score. Mean age was 60 years and median MSQ score was 64 points. Of the interviewees, 24.14% had their sexual performance classified as poor-null, 17.24% of patients had poor-unfavorable sexual performance, 6.90% as unfavorable-regular; regular-good and good-great sexual performance were equally found in 25.86% of the subjects.

Discussion and conclusion: In this study, almost half of post-stroke men have sexual dysfunction according to MSQ, indicating a high impact of such a condition in the quality of life of our patients due to its prevalence. Thus, the health professional dealing with post-stroke patients must be aware and prepared to help patients in coping with sexual dysfunctions.

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727 – Use of FilmArray® in the diagnosis of bacterial meningitis

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Introduction: The Meningitis/Encephalitis FilmArray is an automated multiplex polymerase chain reaction for identifying 14 central nervous system (CNS) care agents, including viruses, Cryptococcus, and bacteria. The following bacteria are tested: E. coli K1, H. influenzae, L. monocytogenes, N. meningitidis, S. agalactiae and pneumoniae. In this study we compared the performance of FilmArray® with conventional microbiological methods for bacterial meningitis.

Methods: We retrospectively evaluated data from 903 patients with CNS infection manifested by the method.

Results: 42 cases were positive for bacteria, *E. coli* K1=2, *H. influenzae*=7, *L. monocytogenes*=5, *N. meningitidis*=9, *S. pneumoniae*=20. Of these, only 14 (33.34%) were positive with conventional microbiological methods, including culture and/or bacterioscopy. Three patients were negative on FilmArray® and positive with other methods: 2 culture positive (*S. intermedius* and *Micrococcus*) and one Gram negative. All 28 cases positive only with FilmArray® adopted a cerebrospinal fluid infection pattern suggestive of bacterial meningitis: pleocytosis with neutrophilic predominance, increased protein and lactate, and hypoglycorrhachia.

Conclusion: The study confirms previous data indicating that FilmArray® increases the sensitivity of etiological diagnosis of bacterial meningitis.

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728 – Cardiovascular risk in adults with drug-resistant epilepsy submitted to the modified atkins diet

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Introduction: The modified Atkins diet (MAD) the carbohydrates are limited to 15–30 g/d and high fat intake is encouraged. It are used in children, being less studied in adults

Objectives: To assess the impact of MAD over a 6-month period in adults with drug-resistant epilepsy in terms of reduction in seizure frequency and cardiovascular risk.

Methods: Prospective and interventional study.Inclusion criteria:patients aged 18–60 yrs, with drug-resistant focal epilepsy, without surgical indication or in the postoperative of epilepsy surgery. Exclusion criteria were: use of corticosteroids, metabolic diseases, cholecystectomy or nephrolithiasis, inadequate nutrition, uncooperative parents or caregivers and inadequate socio-family dynamics. All started the diet at a 1:1 ratio and ASMs. Study periods: baseline (P0), six month (P6). Carotid and vertebral Doppler ultrasounds were performed at P0 and P6 in order to assess possible alterations related to of the carotid arteries the intima-media thickness.

Results: Twenty patients are being followed up, 12 of which started the MAD, and of these, 6 completed the six-month period of diet therapy and underwent pre and post-diet exams. The 6 patients showed no increase in mean total, low-density lipoprotein, triglycerides cholesterol levels.There was no change in the thickness of the myointimal complex of any patient, made by comparative analysis by ultrasonographic exam before and after six months of starting the diet. Four out of 6 patients had >50% reduction in seizures (two seizure free) and two had no change in frequency.

Conclusion: MAD for six months in adults with drug-resistant focal epilepsy had a significant effect on the reduction of epileptic seizures and did not reveal alterations in the myointimal wall of the carotid arteries.

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729 – Clinical utility of FilmArray® in the diagnosis of nervous system infections

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Introduction: The *FilmArray®* meningitis/encephalitis is a multiplex polymerase chain reaction for identifying 14 agents of central nervous system (CNS) infections, including bacteria, *Cryptococcus* and viruses. In this study, we retrospectively evaluated the clinical utility of using this method.

Methods: We retrospectively evaluated data from 101 patients with suspected CNS infection in which cerebrospinal fluid (CSF) was submitted to *FilmArray®* and its diagnostic usefulness was evaluated.

Results: Thirty-six CSF samples (35.6%) had a positive *FilmArray®* result. Twenty-six (72.2%) cases were positive for enterovirus, six (16.67%) were positive for HHV-6, two were positive for Parechovirus (5.56%), HSV-1, HSV-2, *Haemophilus* and *Listeria*=1 (2.78%). Of these 36 CSF samples, 34 (94.44%) had altered cytobiochemical CSF findings and 2 (5.56%) had no CSF abnormalities. None of these etiological agents were identified by other methods.

Conclusion: *FilmArray®* was able to identify neuroinfection etiology in cases in which other etiological identification methods did not reveal the causative agent. In two cases with positive *FilmArray®* the CSF general analysis was normal. Therefore, this method increases diagnostic sensitivity and allows an early identification of the etiological agent, contributing to the clinical management of CNS infections.

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730 – Neurodegeneration with brain iron accumulation associated with late-onset psychiatric symptoms

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Introduction: The neurodegeneration with brain iron accumulation (NBIA) is a heterogeneous group of rare genetic disorders that lead to accumulation of iron in the basal nuclei, which leads to numerous neurologic disorders. This condition has been associated with many genes, *PKAN2* being the most

common. The treatment remains mostly symptomatic and the approach based on a multidisciplinary team is recommended.

Objectives: The aim is to discuss a clinical case of a rare genetic disease that causes neurodegeneration with brain iron accumulation in the individual.

Methods: The case presented is from a 67-year-old male with iron accumulation bilaterally in the pale globes and has been having delirium for eight months, as well as visuospatial and executive dysfunction and behavioral alterations. The patient's magnetic resonance imaging (MRI) showed abnormalities and the "eye of the tiger" sign. Furthemore, the treatment of choice was the administration of atypical antipsychotic medication, which partially controlled the patient's condition.

Results: The most recurrent location of brain iron accumulation is in the basal ganglia and it is mainly linked with mutations in the pantothenate kinase 2 (*PKAN2*), which leads to psychiatric manifestations of insidious progression and can be inferred when the "eye of the tiger" sign is found in a MRI.

Conclusion: The patient was diagnosed with late onset psychotic disorder, mild cognitive deficit and parkisonian syndrome. Genetic tests were not performed to diagnose the etiology. Therefore, the case is considered a NBIA of unknown cause with atypical characteristics and probable mutation linked to *PANK2*.

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731 – Multisegmental ultrasonography of peripheral nerves in leprosy neuropathy

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Introduction: Leprosy is the most common treatable peripheral neuropathy worldwide. The detection of peripheral nerve impairment is essential for its diagnosis and treatment.

Objectives: To identify neural thickening through multisegmental ultrasonography (US).

Methods: US measurements of cross-sectional areas (CSAs) of ulnar, median and tibial nerves at two points (in the osteofibrous tunnel and proximal to the tunnel), and also of the common fibular nerve at the fibular head. CSA index (ΔCSA) was calculated as the absolute difference between CSAs from the contralateral side. This is a cross-sectional study composed of 53 leprosy patients (LP) and 53 healthy volunteers (HV).

Results: US evaluation detected a total of 138 enlarged nerves, implying neural impairment in 71.7% (38/53) of LP (3.6 enlarged nerves/LP). We detected only mononeuropathy on patient in 23.7% (9/38), and multiple mononeuropathy in 76.3% (29/53). The ulnar and tibial were the most frequently affected nerves. The mean values of CSA of all nerves were significantly higher in LP. We also observed higher ΔCSA measurements for all nerves in LP, with significantly higher values for the ulnar and tibial nerves, both in the tunnel and proximal to the tunnel points. All clinical forms of leprosy evaluated showed neural enlargement through US.

Conclusion: The multisegmental US is a useful method for diagnosing leprosy neuropathy, revealing that asymmetry and irregular thickening are characteristics of the disease. Furthermore, that neural involvement is common in different clinical forms of leprosy, reinforcing the importance of including US evaluation of peripheral nerves in the investigation of all leprosy patients.

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734 – Erdheim Chester: an atypical presentation

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Introduction: We present a case of a patient with atypical manifestations of Erdheim-Chester Disease. We aim to describe the neurological manifestations of Erdheim-Chester disease and the main radiologic findings in magnetic resonance imaging. We carried out a literature review in the PubMed and SciELO electronic library databases.

Case: 71-year-old men presented with memory loss, inattention and apathy, developed in the last month. Within the symptoms onset, he had ictal episodes of transient loss of conscience, oromastigatory automatisms and right gaze deviation, with daily recurrence. Physical examination revealed disorientation, right hemiparesis and ideomotor dyspraxia. Magnetic resonance imaging revealed subdural laminar collections on the left hemisphere, with focus of restricted diffusion, thickening of the pachymeninges and also a nodular lesion in the left posterior corona radiata, with granulomatous aspect. Cerebrospinal fluid analysis showed lynphomonocytic pleocytosis (26 cels) and high protein (229 mg/dL). A meningeal biopsy was performed and the anatomopathological examination showed lymphoplasmocytic and histiocytic infiltrate with xanthomized histiocytes, positive for CD68, negative for CD1a and Protein S-100. At this point, we made a diagnosis of Erdheim-Chester disease.

Discussion: Erdheim-Chester disease is a non-Langerhans histiocytosis with diverse clinical manifestations. Neurological symptoms include headache, motor deficit, ataxia, encephalopathy and seizures. Radiologic features are heterogeneous, including tumoral, vascular and pseudodegenerative presentations, as well as pachymeningitis and subdural collections.

Conclusion: Erdheim Chester disease has multiple neurological presentations and must be considered as a differential diagnosis in the setting of pachymeningitis and granulomatous solid lesions.

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735 – Carotid artery stenting in a stroke patient with carotid web: a possibly safe treatment

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Introduction: Carotid web (CW) is a shelf-like luminal protrusion of the intimal layer of the carotid artery, mostly located in the posterior wall of the carotid bulb. It is a rare pathologic finding and an underrecognized cause of stroke. It can be detected on computed tomography angiography (CTA) or digital subtraction angiography (DSA) and treat-

ment options include medically management and carotid revascularization with carotid endarterectomy (CEA) or carotid artery stenting (CAS). CEA is the therapy generally chosen by physicians. The aim is to report a patient with ischemic stroke and CW who was successfully treated with CAS.

Case description: A 46-year-old female, current smoker with no comorbidities, was referred to a tertiary hospital in stroke protocol. Two months prior, she started with headache and visual blurring, evolving with reduced strength, hypoesthesia on the right side of the body and speech alteration. She was lucid and the National Institutes of Health Stroke Scale on admission was 3. She presented mild dyslalia and comprehension impairment, in addition to hypoesthesia in the right hemiface and right hand. Reflexes and muscle strength were normal on all four limbs. Her skull computed tomography demonstrated left temporoparietal hypodensity. A CTA and DSA were performed, revealing a shelf-like filling defect along the posterior wall of the bulb of the left internal carotid artery, characteristic of CW. She was treated with dual antiplatelet treatment and statin before she underwent a CAS with placement of a stent at the origin and proximal segment of the left internal carotid artery, seven days after the ictus. There were no complications and the patient was discharged two days after the procedure with the same drugs.

Conclusion: Stent placement can be a safe treatment option for carotid web.

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736 – Bilateral radiological presentation in Rasmussen´s encephalitis with good surgical outcome

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Case presentation: A 4-year-old and 5-month-old child with a history of sudden onset of clonic seizures that started in the left hallux and progressed to the ipsilateral hemibody and face, lasting up to five minutes. After that, she presented walking and learning difficulty and was unable to perform everyday activities (drawing, writing), which evolved in five months. Electroencephalogram (EEG): diffuse slowing predominating in the right hemisphere; very frequent, almost continuous bilateral interictal epileptiform activity in frontocentroparietal regions mainly on the right. Brain magnetic resonance imaging showed right hemispheric atrophy, with predominantly frontoparietal cortical thinning and reduced brainstem volume on the right, in addition to a nonspecific signal alteration in the left thalamic-mesencephalic area, with possible inflammatory or demyelinating origin. She received CBZ and LEV without seizure control. Cerebrospinal fluid analysis was unremarkable including autoantibodies. At age 5-year-old and 2-month-old she developed progressive left hemiparesis. She received two cycles of pulse therapy (methylprednisolone 30 mg/kg for three days), without significant improvement, with continuous clonic seizures in left hallux. Video-EEG revealed right sided epileptiform activity with midline involvement. At age 6-year-old and 8-month-old the child underwent right hemispherotomy and has been seizure free since then, presenting left hemiparesis and taking clobazam, LEV, ESM and CBZ. Histopathology revealed signs of Rasmussen's encephalitis (RE), with frequent microglial nodules and perivascular lymphocytic infiltrate, and some lymphocytes surrounding neurons.

Discussion: Bilateral brain disease in RE is debated, but it is probably very rare. Only two out of 200 published cases had evidence of bilateral disease.

Conclusion: Although rare, the possibility of bilateral involvement in RE should be considered to stress early surgical treatment and to improve outcome.

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737 – Clear and colorless cerebrospinal fluid among suspected central nervous system infections

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Introduction: The physical examination of the cerebrospinal fluid (CSF) is part of its routine evaluation and is easily performed as soon as CSF is obtained. However, its predictive value of CSF abnormalities is poorly known.

Objective: In this study we evaluated the predictive value for CSF abnormalities leukocytes count among clear and colorless CSF in cases of suspected central nervous system (CNS) infection.

Methods: We retrospectively evaluated data from CSF samples collected from suspected CNS infection that were classified as clear and colorless. This visual categorization was performed by the collector physician and by biomedical staff experienced in CSF analysis. The CSF cell count were considered normal when CSF white blood cells were up to 3 cells/mm^3, protein was below 45 mg/dL, glucose above 50 mg/dL, and lactate bellow 19.6 mg/dL. The proportion of normal CSF and normal CSF parameters were recorded.

Results: 34,434 CSF samples from suspected CNS infection were included. Of those, 22,960 were classified as clear and colorless (66.7%). Among clear and colorless CSF samples 13,137 (57.2%) had normal CSF cytology and biochemical findings. The probability of CSF abnormalities in the cytological and/or biochemical examination in clear and colorless CSF samples obtained from patients with suspected CNS infection was 42.8%.

Discussion: In this study, we showed a high frequency of cytological and/or biochemical abnormalities in clear and colorless CSF obtained from patients with suspected CNS infection.

Conclusions: Viral infections and other inflammatory CNS diseases are among the conditions that may present with clear and colorless CSF. Therefore, a clear and colorless CSF in suspected CNS infections has a little predictive value for CSF abnormalities.

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739 – Case report: baló concentric sclerosis in a patient presenting with dyslexia, dysgraphia, and agraphesthesia

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Introduction: Baló Concentric Sclerosis (BCS) is a rare inflammatory demyelinating disease of the central nervous system. Commonly reported symptoms and its clas-

sic neuroimaging findings can lead to a mistaken diagnosis of expanding lesions. The aim is to report on a case of a rare etiology of altered parietal lobe cortical functions, and the importance of topographic reasoning. Review of the patient's file for clinical history, neurological exam, and complementary tests was performed.

Case report: A 36-year-old male patient presented to the emergency room (ER) with complaints of sudden difficulties in reading and writing. He also recalled a perception of numbness on the right side of his body throughout the 15 days before ER evaluation. On neurological exam, the patient presented with dyslexia, dysgraphia, right agraphesthesia, and right hemihypesthesia to all sensory modalities. Magnetic resonance imaging showed a lesion on the left inferior parietal lobe with alternating isointense and hypointense concentric ring-like bands on T1-imaging; hyperintense alternating lamellae on T2-sequences; high signal intensity at the lesion edge on DWI. Its location matched our topographic hypothesis, based on the neurological exam, and its appearance was typical for BCS. We observed other demyelination lesions fulfilling the dissemination in time and space criteria. Cerebrospinal fluid was collected, without infectious signs. Treatment was started with methylprednisolone 1 g/day for seven days and five plasmapheresis sessions.

Conclusion: BCS usually presents with subacute elevated intracranial pressure symptoms and classic focal demyelinating deficits, like hemiparesis and ataxia. Unlikely, cortical disturbances can be the main presentation, making topographic reasonig fundamental. Occasionally, BCS has an acute onset, mimicking a stroke event.

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740 – High-resolution ultrasonography: a diagnostic tool for early diagnosis of neural impairment in leprosy

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Introduction: Leprosy household contacts (HC) have high chances to develop leprosy. Anti-PGL-I IgM seropositivity also increases the risk of illness.

Objectives: Identify neural impairment in leprosy HC by analyzing differences in high-resolution ultrasonographic (US) measurements of peripheral nerves between HC and healthy volunteers (HV).

Methods: Seventy nine seropositive household contacts (SPHC), 30 seronegative household contacts (SNHC) and 53 HV underwent dermato-neurological examination and laboratorial analysis, followed by high-resolution US evaluation of cross-sectional areas (CSAs) of the median, ulnar, common fibular and tibial nerves.

Results: Neural thickening was clinically observed in only 2% of SPHC and none of the SNHC. None of the HC presented skin lesion, sensory symptoms or muscular weakness. The US evaluation detected neural thickening in 26.5% of the SPHC and only in 3.3% among the SNHC. Among SPHC with thickening detected by US, the mean number of nerves affected was 1.8 per contact, while 53.8% presented mononeuropathy and 46.2% presented multiple mononeuropathy. The nerves most affected were the common fibular and the tibial nerves. The CSA of the common fibular, tibial at the tunnel and tibial proximal to the tunnel were significantly higher in SPHC compared to the other groups, being asymmetrical in the fibular and tibial nerves. SPHC presented a 10.5-fold higher chance of neural impairment.

Conclusion: There is a prevalence of neural thickening in SPHC and US in very important to the early diagnosis. The positive anti-PGL-I serology can identify individuals with greater chances of developing leprosy neuropathy, who should be referred for US examination.

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741 – Kappa index and oligoclonal bands detection in cerebrospinal fluid samples of patients with suspected multiple sclerosis

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Introduction: The evaluation of the intrathecal production of immunoglobulin G (IgG) can be useful in the evaluation of multiple sclerosis (MS) suspected cases and the most important test to verify it 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.

Objectives: 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 MS.

Methods: Patients with suspected MS underwent CSF analysis. Intrathecal synthesis of IgG was evaluated with BOCs, IgG index, Reiber nomogram, and kappa index.

Results: One hundred and seventy-one serum and CSF samples were included, 76 of them had positive CSF BOC. Among the BOC+ cases, 48 (63.2%) were also positive with the Reiber nomogram, 53 (69.7%) were also positive with the IgG index, and 74 (97.4%) were also positive with the kappa index. Among the patients without BOCs, 5 (5.2%) cases had a positive Reiber, eight had a positive IgG index (8.4%), and eight (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.

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742 – Prevalence of depressive symptoms in stroke patients: a cross-sectional study

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Introduction: Sequelae are frequent in patients with a history of ischemic

stroke and result in decreased quality of life, increase in morbidity, mortality and healthcare costs. One often overlooked complication of ischemic stroke is its association with depression and depressive symptoms.

Objectives and methods: This cross-sectional study aims to assess the prevalence of depression and depressive symptoms in patients with previous ischemic stroke followed in the neurovascular outpatient clinic of a tertiary hospital in São Paulo, Brazil, through the application of the Patient Health Questionnaire-9 (PHQ-9) filled in by the patients.

Casuistics and results: A total of 173 participated in the study, but only 154 answered all PHQ-9 questions. Mean age was 52 years; 56.82% were women; 44 patients (28.6%) had a score ≥ 10, resulting in sensitivity and specificity for post-stroke depression of 86 and 79% respectively; 118 (68.2%) did not present depression or depressive symptoms before the stroke, 33 (19.1%) had previous symptoms and 22 (12.7%) did not answer this specific question.

Discussion and conclusion: The results indicate that depression is an important comorbidity in patients with ischemic stroke. In this cohort, most patients did not have depressive symptoms before the stroke. Further studies are important to better assess this association as well as to determine strategies to prevent and to manage this condition.

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743 – Young patient with pineoblastoma and four-year progression-free survival, still under follow up: a case report

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Pineal tumors comprise 1% of all primary central nervous system neoplasms. They are classified as Pineocytoma, Pineal parenchymal tumor of intermediate differentiation, and Pineoblastoma, as well as Papillary tumor of the pineal region. Pineoblastomas are most commonly seen in children and teenagers and exhibit aggressive clinical behavior, with frequent metastases throughout the craniospinal axis. Most cases commonly presents with findings of elevated intracranial pressure (headache, nausea/vomiting, and decreased level of consciousness) as a result of obstructive hydrocephalus from compression of the cerebral aqueduct by the tumor mass. Parinaud syndrome (upgaze paralysis, convergence nystagmus, and near-light dissociation) is also seen, as a result of compression of the dorsal midbrain structures. Here we represent a case of a female patient, who started with progressive headache at the age of eighteen, in 2017. After further investigation, she was diagnosed with obstructive hydrocephalus, which was found to be caused by a tumor in the pineal region. Admission to the São Paulo Hospital happened on february 2019 (two years after starting the symptons), when she was submitted to biopsy. Histopathological analysis shower grade IV Pineoblastoma. She underwent radiotherapy from march to april 2019 and two cycles of chemotherapy from march to september 2019, with Ifosfamide, etoposide, carboplatin and vincristine. Despite the fact that the median overall survival of the PB is around 20 months after maximal resection and adjuvant chemotherapy/radiotherapy, our patient has been under follow up since 2019, performing cranial magnetic resonance imaging every six months and maintaining clinically and radiologic signs of stable disease.

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745 – Comparative analysis between cure without sequelae and cure with sequelae in the therapeutic approach of acute flaccid paralysis in Brazil

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Introduction: Acute Flaccid Paralysis (AFP) is a serious condition that causes sudden muscle weakness and loss of deep reflexes. It is important to note that there are various causes of AFP, and one of them is the Guillain-Barré Syndrome (GBS), an autoimmune disorder that affects the nervous system, causing muscle weakness and sometimes paralysis.

Objectives: The objective of this study is to compare the cases of cure without sequelae with the cases of cure with sequelae of AFP, based on data obtained between 2007 and 2021, using the records of Notifiable Diseases Information System (SINAN).

Methods: A descriptive ecological study was conducted. Data on cure without sequelae and cure with sequelae of AFP were collected from all Brazilian regions between 2007 and 2021. The data were statistically analyzed and compared between the regions.

Results: The data obtained indicated a total of 4,784 cases of AFP during the analyzed period, with 3,476 cases (72.6%) resulting in cure without sequelae, while 1,308 cases (27.3%) resulted in cure with sequelae. The Northeast region presented the highest number of cases of cure without sequelae (1,355), while the Southeast region presented the highest number of cases of cure with sequelae (408). The statistical analysis showed a higher proportion of cases of cure without sequelae in all regions.

Conclusion: The results of this study demonstrate that cure without sequelae is more frequent than cure with sequelae in the therapeutic approach of AFP in Brazil. The Northeast region presented the highest number of cases of cure without sequelae, while the Southeast region presented the highest number of cases of cure with sequelae. These results highlight the importance of immunization and appropriate treatment of AFP to prevent sequelae and improve the quality of life of patients.

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746 – Clinical manifestations and self-limited course of a low-flow carotid-cavernous fistula

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An 80-year-old female patient with a history of long-standing systemic arterial hypertension presented with a gradually progressive frontal headache, diplopia and decreased right eye movement for one month. On admission

to the hospital, her blood pressure was 190/120 mmHg, despite the regular use of four antihypertensive medications. She had right eye proptosis, hyperemia and chemosis. Neurological examination revealed a fixed mydriatic pupil, light perception visual acuity on the right eye, plegia of all extrinsic ocular muscles and absence of ocular murmur. A brain computed tomography imaging revealed right orbital congestion, enlargement of the right extrinsic ocular muscles and right periorbital fat, as well as of the convexity of the lateral wall of the right cavernous sinus. Contrast-enhanced imaging showed arterial flow in the right ophthalmic vein. Her laboratory test results, including serology for HIV, syphilis, viral hepatitis, rheumatology laboratory screening and tumor biomarkers were unremarkable. Her cerebrospinal fluid was clear and colorless, opening pressure of 13 mmHg, 3.3 cell, 4 erythrocytes, protein 67 mg/dL, glucose 74 mg/dL and lactate 21.6 mg/dL. Following a week of adequate blood pressure control, her symptoms improved. Digital subtraction angiography was performed, revealing an increase in the right intraorbital arterial flow and the size of the ophthalmic vein, consistent with thrombosis of the cavernous sinus but with no evidence of an arteriovenous shunt. Improvement in symptoms was due to the spontaneous closure of the fistula caused by local thrombosis. A left renal artery stenosis was found as the cause of refractory systemic arterial hypertension, with no need for surgical intervention. This case exemplifies the clinical symptoms and the possibility self-resolving nature of a low-flow carotid-cavernous fistula.

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747 – Stroke-like symptoms in Lyme Disease, a case report

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Introduction: First described in Brazil in 1992, Lyme disease (LD) is a multisystem bacterial infection caused by Borrelia burgdorferi and transmited by infected ticks of the genus Ixodes or Amblyomma. LD prevails in temperate climate countries, with some few cases reported in Brazil. In early disseminated LD, main neurologic findings are meningitis, cranial neuropathy, radiculoneuropathy, cerebellar ataxia and encephalomyelitis. Neurologic features of late LD include confluent mononeuropathy multiplex, peripheral neuropathy and encephalopathy. Diagnosis is essencially clinical, with possibility of serologic testing. The treatment includes antibiotics and must be individualized.

Objectives: We aim to present a LD case with stroke-like presentation.

Results: A 43-year-old man presented with central facial palsy, vertical nystagmus, headache, vertigo and nausea. Magnetic resonance imaging (MRI) showed hyperintense oval lesion in T2/FLAIR in the left middle cerebellar peduncle, with diffusion restriction. At first, he was diagnosed with of ischaemic stroke. After two weeks of symptoms onset, MRI showed an enhancement in the previous cerebellar lesion with normal angiogram. Cerebrospinal fluid analysis was also normal. The patient received a 5-day pulse therapy, with partial clinical response and yet, a new MRI showed an increased lesion with the same previous features and CSF protein of 71. Viral CSF screening, cryptococcal and cysticercosis antigens, oligoclonal bands were negative. LD serology came out positive, allowing the treatment with ceftriaxone for 28 days. Due to partial response, alternative treatment with benzathine penicillin, doxycycline and tinidazole was instituted, achieving satisfactory results.

Conclusion: LD may mimic other neurologic syndromes, thus neurologists frequently ought to consider it as a differential diagnosis.

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749 – Prevalence of fatigue in patients with ischemic stroke: a cross-sectional study

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Introduction: Stroke is the leading cause of disability worldwide, not only due to motor impairment, but to many sequelae in other domains. Fatigue is one of the most impacting symptoms in the quality of life in post stroke patients, with estimated prevalence around 30%.

Objectives and methods: We present a cross-sectional study that aimed to determine the prevalence of fatigue in patients with ischemic stroke followed in the neurovascular outpatient clinic of a tertiary hospital in São Paulo, Brazil, using the Fatigue Assessment Scale (FAS) questionnaire. It can range from 0-50 points, proportional to symptom intensity; scores equal or over 24 points indicate fatigue in such individuals.

Casuistics and results: One hundred and fifty seven questionnaires were analyzed and classified according to the FAS. Mean age was 58 years, 74 (47.13%) individuals were men and median FAS score was 21 points (interquartile range: 18–29). Of the interviewees, 64 individuals (40.76%) were classified as having fatigue by the established cut-off; in this subgroup, mean age was 55 years and 28 (43.75%) were men.

Discussion and conclusion: In this study, around 40% of post-stroke patients have fatigue, indicating a high prevalence in our population. Fatigue has a high impact on the quality of life of patients, also worsening in functionality and mortality, due to lesser participation in rehabilitation and physical activities. Thus, the health professional dealing with post-stroke patients must be aware and prepared to help patients in coping with fatigue.

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753 – Elderly man presenting with focal seizures, encephalopathy and tumefactive lesion on imaging scans

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An 84-year-old man with cardiomyopathy and a pacemaker was admitted to our hospital due to a focal impaired awareness seizure. Computed

tomography (CT) scan on admittance evidenced right parieto-occipital hypodensity with mild mass effect. A non-contrast CT scan realized in an outer institution seven months earlier showed that such hypodensity, considered then as a stroke, was present, but slowly progressing. He was submitted to therapy with phenytoin upon entry and his electroencephalogram showed increased slow-wave activity. Cerebrospinal fluid showed hyperproteinrachia, normal cell count and slightly reduced glycorrhachia. With such findings, the main hypothesis of neoplasm and neuroinfection emerged. Magnetic resonance imaging (MRI) could not be realized at first due to his pacemaker. Empiric treatment with acyclovir was initiated but, in spite of that, he maintained somnolence and left hemiparesis. The cardiology team was activated, and his pacemaker was set to do an MRI. His scan evidenced a T2/FLAIR hyperintense mass lesion on the right parieto-occipital area. SWI sequences showed microbleeds along the cerebral cortex and chronic lobar hematoma simulating cortical superficial siderosis. Hence, Cerebral Amyloid Angiopathy-Related Inflammation (CAA-ri) was diagnosed and pulse therapy with methylprednisolone was indicated. By closure of this report, he was started on antibiotics for urinary tract infection, and would be initiated on corticosteroids after 48 hours. CAA-ri is a rare yet reversible etiology of seizures, encephalopathy and focal neurological signs in patients with amyloid angiopathy, an entity that occurs mainly in the elderly as a deposit of amyloid protein on vessel walls. MRI is essential as it shows characteristic cortical-subcortical hemorrhagic lesions. Suggestive findings allows treatment with corticosteroids, optimizing neurological recovery and minimizing future deficits.

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754 – Prevalence of connective tissue abnormalities in patients with spontaneous cervical artery dissection in a multiethnic population

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Objectives: To investigate the prevalence of characteristics phenotypic associated to connective tissue abnormalities in patients with craniocervical artery dissections (CAD).

Methods: We assessed phenotypic detectable signs of connective tissue aberration in a series of patients with CAD and patients with ischemic stroke unrelated to CeAD (non-CAD) by a standard protocol including 68 items.

Results: The study group included 20 patients with CAD (mean age, 44.5 6 7.8 years; 66.7% men) and 12 patients with non-CAD. Connective tissue abnormalities were detected more frequently in the group of patients with CAD than in the group of those with non-CAD IS (63% in CAD IS and 58% in non-CAD IS; P = 0.001). The mean of detectable sign in the CAD group was 2.86 compared with 1.9 in the group with non-CAD IS (P = 0.005).

Conclusion: Connective tissue abnormalities are frequent in patients with CAD IS. This reinforces the hypothesis that systemic aberrations of the connective tissue might be implicated in the pathogenesis of the disease.

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756 – Neuro-Behçet as a cause of acute onset cerebellar ataxia: a case report

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Case presentation: A 37-year-old woman, current smoker and previously addicted to cocaine, was brought to the emergency department with complaints of "imbalance and altered speech" that started eight days prior to admission, acutely. The patient, who was previously asymptomatic, also reported fever, headache and myalgia during the same period. Emotional lability was reported by the family members. At the initial evaluation, the patient was somnolent but reactive. Scanning speech, left-sided central facial palsy, sings of bilateral pyramidal involvement, axial and appendicular cerebellar ataxia were also observed. After a thorough examination, cicatricial lesions of previous genital ulcers were observed and the patient reported the presence of recurrent orogenital ulcers in the previous three months. Also, sporadic episodes of conjunctival hyperemia and hematuria were reported. CSF analysis demonstrated a cellularity of 63 leukocytes/mm3 (100% lymphocytes), increased total protein (105 mg/dL) and a low glucose level (cerebrospinal fluid/plasma ratio of 53%). The magnetic resonance imaging showed FLAIR hyperintensity at the midbrain, left cerebral peduncle, pons (base and tegmentum), posterior limbs of both internal capsules and left posterior thalamus. A pontine lesion with peripheral contrast enhancement, leptomeningeal enhancement at the level of the prepontine cistern and enhancement of the VII and VIII complexes were also observed. High-dose steroid therapy with methylprednisolone 1 g/day was initiated (total of five days of therapy) and the patient presented significant improvement of the symptoms, returned to walk independently and no longer presented emotional lability. At the time of discharge, a mild cerebellar ataxia was still observed. The pathergy test was positive and the diagnosis of Neuro-Behçet was confirmed. The steroid treatment was transitioned to oral prednisolone and, later, immunosuppressive therapy with infliximab was started.

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758 – Ultrasound detection of carotid web in a recurrent stroke patient: a case report

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Introduction: Carotid web is an intimal variant of fibromuscular dysplasia, leading to a shelf-like linear defect in the internal carotid bulb. It is a rare, but important embolic etiology for stroke in young patients without conventional cardiovascular risk factors, with a high recurrence rate. Carotid ultrasound is a low-cost, widely available tool in stroke investigation and can be useful as a diagnostic tool for carotid web, as described in this case.

Methods: Case report of a patient with recurrent stroke secondary to carotid web, detected by ultrasound, obtained by medical records.

Case report: A 57-year-old female patient, with a history of obesity treated with bariatric surgery 7 years ago, presented three episodes of stroke, all in the right anterior circulation. In all of them she presented left side weakness and paresthesia. She remained only with left side paresthesia after those events. The recurrent events happened despite using aspirin and double antiplatelet treatment. A carotid ultrasound was performed and detected a shelf-like, thin band of intima in the posterior wall of the right internal carotid artery. Endarterectomy treatment was performed, and she has been stable ever since with aspirin.

Conclusion: Ultrasound is an increasingly available, non-invasive method performed bedside. Although it is a useful diagnostic tool to detect carotid web, it is still a challenging diagnosis, probably due to the lack of awareness among examiners. Given the high rates of stroke recurrence related to carotid web, a careful ultrasound examination and meticulous imaging analysis probably allows earlier diagnosis and appropriate treatment.

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759 – Early non-invasive ventilation indication in amyotrophic lateral sclerosis and other neuromuscular disorders: a prospective study

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Introduction: Neuromuscular diseases (NMD) affect the peripheral nervous system and may lead to progressive respiratory failure. Ventilatory assistance reduces dyspnea and improves quality of life.

Objectives: The aim of this study is to compare patients with amyotrophic lateral sclerosis (ALS) or other NMD and the effects of early indication of non-invasive ventilation (NIV) (FVC > 70).

Methods: A study was carried out in patients with NMD and indication for NIV, accompanied by periodic examinations, weekly telemonitoring and adjustments of ventilatory parameters for six months.

Results: The study found that ALS patients had greater adherence to NIV, with mean daily use increasing from 4.7 to 6.0 hours and a decrease in leakage (L/min) from 2.5 to 1.5 and in the apnea-hypopnea index (AHI) from 5.85 to 4.85. Patients with other NMDs had a decrease in mean daily use from 6.1 to 5.3 hours, with a decrease in leakage from 1.2 to 0.5 and AHI from 3.1 to 1.9. Patients with early indication for NIV had a lower rate of deaths and noncompliance compared to those with late indication. Patients with early indication had 6 hours (±2.0) of average daily use and maintained it after 6 months (±1.3) with a decrease in leakage from 1.9 to 0.2 and AHI from 3.1 to 2, two. In the late indication group, there were 4 deaths and 2 dropouts, with an increase in daily use from 4.7 to 5.5 hours, a decrease in leakage from 1.9 to 1.5 and AHI from 3.2 to 2.2.

Conclusion: This study suggests that early NIV indication can improve ventilatory parameters and outcomes in patients with NMDs, especially ALS, and should be considered for patients with FVC > 70%.

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760 – Mutation in the REEP1 gene related to SPG31 (Autosomal Dominant Hereditary Spastic Paraplegia type 31)

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Introduction: The *REEP1* gene is associated with a spectrum of overlapping autosomal dominant conditions including hereditary spastic paraplegia 31 (*SPG31*), distal hereditary motor neuropathy 5B. It's a neurologic condition limited to progressive lower extremity spastic weakness and associated atrophy, distally predominant, hypertonic urinary bladder, and mild lower extremit vibratory sensation loss. *SPG31* has an estimated prevalence of one to 12 per 100.000 individuals.

Objectives: To report a rare case of mutation in the *REEP1* gene related to *SPG31*.

Case presentation: A 38-year-old woman, born to non-consanguineous parents, presented with difficulty walking and frequent falls since the age of 11, with slow progressive evolution, accompanied by cramps in the lower limbs. The patient hasd an extensive family history (father, dizygotic twin sister, paternal grandmother, and three paternal uncles). Physical examination of the lower limbs revealed spasticity, proximal muscle strength grade 4+ and distal muscle strength grade 4-, grade 4 hyperreflexia in the patellar and Achilles tendons, with an inextinguishable plantar clonus and bilateral Babinski sign, and slight hypotrophy of the gastrocnemius muscles were observed, as well as spastic gait. The patient hasd urge urinary incontinence with partial remission using Oxibutynin. Baclofen 20 mg/day is being used to control spasticity. Genetic testing was performed and revealed the c.128_138dup mutation (p.Phe48Tyrfs*25) in heterozygous in the REEP1 gene related to SPG31 (Autosomal Dominant Hereditary Spastic Paraplegia type 31).

Conclusion: As the diagnosis of hereditary spastic paraplegia 31 associated to a REEP1 gene mutation is rare, neurologists may have limited knowledge of this condition, precluding its adequate diagnosis. The urge urinary incontinence is a distinctive feature, and should raise awareness for this condition.

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763 – Mastocytosis, pachymeningitis and systemic neoplasia

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Mastocytosis is a condition characterized by abnormal growth and accumulation of dysfunctional mast cells in one or more organs. Diagnosis is challenging due to its rarity, non-specific symptoms, and imaging findings, requiring a combination of physical examination, histopathological findings, genomic testing, and laboratory results. In this report, we present the case of a 40-year-old female patient with a history of systemic mastocytosis who presented to the emergency department with a 10-day history of headache, dizziness, and visual disturbances. Physical examination revealed bilateral sixth cranial nerve palsy, papillary edema, and a hardened right breast. The cerebrospinal fluid study showed an opening pressure

of 76 cmH_2O and a closing pressure of 34 cmH_2O. Magnetic resonance imaging of the skull and neuraxis showed pachymeningitis, a recent ischemic lesion in the left cerebellar hemisphere, bilateral optic neuropathy, multiple bone lesions, and pathological fractures throughout the thoracic and lumbar spine.Ppositron emission tomography-computed tomography showed hypermetabolism in the right breast and liver, with a breast and liver biopsy confirming neoplasia. Mastocytosis can cause acute and chronic nervous system disorders, including headache, anxiety, memory loss, and vertigo/dizziness, with mostly unspecific magnetic resonance imaging imaging. The etiology of the intracranial hypertension finding was extensively discussed by the assisting team. A deep literature review was conducted on mastocytosis with pachymeningitis, which is extremely rare. No sign of meningeal carcinomatosis was found, leading us to believe that mast cell infiltration may have been possible. A meningeal biopsy was suggested to confirm mast cell infiltration, but it was not performed due to the increased risk to the patient. The patient's complications were likely caused by mastocytosis, either through mast cell infiltration or an increased predisposition to neoplasms.

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764 – Hypertrophic pachymeningitis associated with systemic lupus erythematosus: a case report

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Introduction: Hypertrophic pachymeningitis is a chronic inflammatory process that results in focal or diffuse thickening of the dura mater. Nowadays, it's an uncommon cause of neurological symptoms and has been mainly associated with infections, inflammatory diseases and carcinomatosis. This is a report of a single case based on retrospective analysis of the patient's medical records.

Case report: A 48-year-old woman was admitted in the emergency department with a two-month history of severe headaches that worsened with neck flexion and four episodes of focal to bilateral tonic-clonic seizures. In addition to that, she also complained of intermittent gritty sensation and redness in both eyes and arthritis of the hand and wrist joints. Her past medical history included systemic lupus erythematosus, bilateral lower limb deep venous thrombosis and cerebral venous thrombosis (CVT). She was on anticoagulant therapy with rivaroxaban 20 mg daily at the time. On admission, patient presented with bilateral conjunctival hyperemia and slit-lamp examination showed mild anterior chamber flare reaction (grade of 2+). Neurologic examination revealed impaired attention and nuchal rigidity. Brain magnetic resonance imaging was performed and showed linear dural thickening and diffuse pachymeningeal post-contrast enhancement over cerebral convexities, falx cerebri and cerebellar tentorium, with hyperintense signal on T2-weighted sequence in the subcortical region of the right frontal lobes, which might have resulted from obstruction of venous reflux caused by previous CVT. Brain venogram demonstrated partial recanalization of superior sagittal sinus. Lumbar puncture revealed a clear cerebrospinal fluid (CSF) with 12 cells/mm^3 (82% lymphocytes, 13% monocytes, 5% plasmocytes), total protein of 46 mg/dL and glucose of 58 mg/dL. Search for infectious agents by microscopy, culture and molecular testing resulted negative. Systemic investigation was negative for infections or metabolic dysfunction, but it revealed elevated inflammatory markers and hypocomplementemia. Her panel of autoantibodies revealed an ANA titer of 1:640 with a nuclear homogeneous pattern and was negative for the others (anti-dsDNA, anti-Sm, anti-RO, anti-LA, anti-RNP, RF). The patient was then treated with intravenous methylprednisolone 1 g daily for five days and followed by maintenance treatment with oral prednisone 60 mg daily. She showed good improvement of symptoms, as well as a significant reduction of CSF leucocytes and protein content. Unfortunately, it was not possible to obtain a meningeal biopsy at the time. The patient was finally discharged from the hospital and, given the rare association between pachymeningitis and SLE, is expected to return for a follow-up consultation with neurology for additional investigation.

Conclusion: Although hypertrophic pachymeningitis has been related with some autoimmune disorders, it is an extremely rare form of nervous system manifestation in SLE. Despite the plausible relationship between the patient's SLE disease activity and hypertrophic pachymeningitis, further investigation is still required to rule out other etiologies. Nevertheless, we hope our report will raise awareness of hypertrophic pachymeningitis as a possible differential diagnosis in SLE patients with new-onset headache with nuchal rigidity.

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765 – Acquired hemichorea secondary to hyponatremia: a case report

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Introduction: Chorea is a movement disorder characterized by involuntary, random and irregular movements. In the emergency department, the most common causes of chorea are cerebrovascular disorders, but other conditions may also play an important role. The aim is to present a rare case of acquired hemichorea secondary to hyponatremia by reviewing the patient's records.

Case report: A 73-year-old female patient with a history of arterial hypertension, dyslipidemia, active smoking, and heart failure, taking enalapril, hydrochlorothiazide, furosemide and simvastatin, was admitted to the emergency department after falling from standing height and hitting her head, also presenting choreic movements in the right side of her body. The patient started amitriptyline for chronic low back pain in the last month and had been experiencing apathy and weight loss since; the choreic movements had started a week before admission and had progressively worsened. On admission, the patient presented with fluctuating level of consciousness and hemichorea. Complementary evaluation showed leukocytosis and hyponatremia (117 mg/dL); magnetic resonance imaging of the brain showed diffuse brain atrophy without any other findings compatible with hemichorea and the lumbar puncture did not reveal any additional findings; a pulmonary infection was detected by the chest computed tomography. The patient was treated for pulmonary infection and hyponatremia. Haloperidol was started, but the chorea was fully solved after hyponatremia correction over the next few days and did not return after haloperidol weaning.

Discussion: We presented the case of a 73-year-old woman with acquired hemichorea secondary to hyponatremia, which resolved completely after the metabolic disorder was corrected. Although this condition has been described, its incidence remains unclear, as few cases have been reported.

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766 – Effect and tolerance of acetazolamide in patients with idiopathic intracranial hypertension

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Introduction: Acetazolamide is commonly used to treat idiopathic intracranial hypertension (IIH). The Idiopathic Intracranial Hypertension Treatment Trial showed a modest improvement in visual field function in patients using acetazolamide with a low-sodium weight-reduction diet compared with diet alone. There is still insufficient data to establish its use.
Objectives: To describe acetazolamide effect in improving headache, visual acuity and papilloedema in patients with HII and assess patient's tolerance to it Design. Retrospective analysis of 37 patients with IIH taking acetazolamide in a headache clinic of a tertiary medical center in São Paulo, Brazil.
Results: The population analyzed was mostly female, with a mean age of 34.32 years, obese with a mean body mass index of 34.16, and were referred to the headache clinic from the emergency department. All 37 patients complained of headache and had increased cerebrospinal fluid opening pressure initially (mean 42.05). Improvement of headache was reported by 25 (67.56%) patients. Fundoscopy was described in the initial and last visit in 24 patients: 15 had papilloedema initially and 9 had a normal exam. At the last visit 9 patients improved (60%), 6 maintained papilloedema and 1 patient developed it. Visual acuity was described in the initial and last visit in 20 patients: 5 had normal visual acuity, 14 had low visual acuity and one patient was amaurotic. Of those with low visual acuity 2 (14.28%) got completely recovered, 5 (35.71%) recovered partially and 7 (50%) got worse. Two patients with the initial normal exam got worse during follow-up. Adverse effects were reported by 5 (13.51%) patients and led to discontinuation of acetazolamide — 3 of 5 reported paresthesias.
Conclusion: Acetazolamide reduced headache in 67.56%, improved visual acuity in 50% and papilloedema in 60%, and was well tolerated by 86.48% of patients.

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769 – Focal dystonia after an ischemic stroke: case report

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Case presentation: A 77-year-old female patient, previously hypertensive, was admitted to the emergency room due to involuntary movements in the right upper limb onset one day ago with no other associated complaints. During neurological examination she presented hemiparesis and hypoesthesia in the right dimidium of the body besides involuntary movements in the ipsilateral hand. Skull magnetic resonance imaging evidences hyperintensity on T2/FLAIR sequences in areas of periventricular and left parietal white matter in accordance to a previous ischemic event.
Discussion: Dystonia is a movement disorder caused by involuntary muscle contractions leading to repetitive or abnormal movements. It is related to hereditary, idiopathic or acquired causes, including the ones related to brain lesions of cerebrovascular etiologies, such as ischemia, hemorrhage and arteriovenous malformation and aneurysm. The diagnosis of dystonia is made clinically. The treatment consists of the application of botulinum toxin, oral medications such as clonazepam and baclofen.
Conclusion: A challenge due to the numerous etiologies involved, dystonias are movement disorders that negatively impact patients' self-esteem and independence. The present case is extremely relevant considering there was no reported prior complaint of focal neurological deficit, although an area consistent with cerebral ischemia was identified on cranial magnetic resonance imaging, correlated as the causative factor leading to the involuntary movement.

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770 – Pseudotumor demyelination lesion associated with bevacizumab in oncological treatment

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Introduction: Bevacizumab is a monoclonal antibody directed against the human isoform of vascular endothelial growth factor A (VEGF-A) and inhibits angiogenesis.
Case: Female patient, 54-year-old, with a history of intestinal adenocarcinoma associated with peritoneal carcinomatosis being treated with bevacizumab. The patient started to present a deviation of the labial rhyme to the right, which showed the presence of left facial paralysis with a central pattern on the neurological physical examination, without other alterations. Magnetic resonance imaging of the brain showed a nodular lesion centered in the subcortical white matter of the right frontoparietal transition, showing T1 hyposignal, T2/FLAIR hypersignal, with areas of pronounced SWAN hyposignal, without significant post-contrast enhancement and a thin halo of diffusion. It has no significant mass effect and measures 1.4 x 2.5 x 2.0 cm. Screening for HIV, toxoplasmosis, cryptococcus and mycobacterial infection was negative. Magnetic resonance imaging of the cervical spine and cerebrospinal fluid analysis were normal. The presence of a demyelinating lesion was observed in the exteriotaxic biopsy, which confirms the hypothesis of a pseudotumoral lesion and rules out the possibility of a metastatic lesion. Over four weeks, there was resolution of neurological signs with radiological improvement in subsequent examinations. There was no evidence of new inflammatory activity, clinically and on MRI over a 2-year period.
Conclusion: A definitive causal association between VEGF-A inhibition and central nervous system inflammatory demyelination could not be confirmed in this patient. However, the temporal relationship between institution of bevacizumab therapy and presentation with tumefactive demyelination is significant.

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771 – Case report: myofasciitis associated with the NFkB gene

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A male patient, 44-year-old, presents with severe abrupt myalgia in the upper and lower limbs, with evolution to muscle weakness after two weeks. After 40 days, he developed intermittent fever and night sweats. Personal history of anorectal abscess drainage. The neurological exam evidenced discreet muscular hypotrophy of the lateral and medial portion of the thighs, global hyperreflexia, proximal muscle weakness, and bilateral antalgic gait. A right vastus lateralis muscle biopsy showed muscle atrophy and congested vessels. Magnetic resonance imaging of the thighs visualizes diffuse inflammation of the fascia and muscles of the thigh. After beginning the use of corticosteroids, there was a significant improvement. A genetic test showing the c.1129G>A variant (p.Gly377Ser) in the *NFKB1* gene was also requested, followed by corticoid weaning and human immunoglobulin initiation.Myofasciitis is a painful inflammatory condition affecting the muscles and the tissues around them. One of them is the fascia, a fibrous connective tissue that surrounds and connects the body's muscles, tendons, and bones. The pathogenesis may be founded on the mutation of the *NFkB* gene, which regulates our body's inflammatory and immune processes, which may result in autoinflammatory diseases, immunodeficiencies, and, consequently, tissue damage. Symptoms are varied and can include muscle weakness, arthralgia, and skin rashes. Diagnosis is based on blood tests, imaging, muscle biopsy, and genetic testing. Treatment involves rehabilitation and immunosuppressive medications to control the immune system's response. Although NFkB-associated myofasciitis is rare, awareness and understanding of its symptoms are essential to ensure early diagnosis and appropriate treatment.

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772 – Migraine and painful headaches in Brazil's national health system: a descriptive epidemiological study (2010–2020)

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Introduction: Primary headaches are more common in females until the 5th decade of life and should be properly diagnosed to ensure appropriate management, as they can cause significant socioeconomic impairment and psychological damage.

Objectives: To describe migraine and other painful headaches (MPH) cases and costs in Brazilian National Health System (SUS) from 2010 to 2020.

Methods: This is a descriptive epidemiological study. Data was collected from the Brazilian National Health System's (DATASUS) database, describing MPH's morbidity (CID-10) between 2010 and 2020. As this is a public domain database, there was no need for ethics committee approval.

Results: The total number of hospitalizations for MPH in Brazil during the study period (2010–2020) was estimated at 89,372, with 65.6% being female patients. The number of hospitalizations increased each year since 2010, reaching its peak in 2019 (12,120 cases), and presenting a new drop in 2020 to 67% of its value, potentially indicating an impact of the coronavirus pandemic. The age group with the highest number of hospitalizations was between 30–49 years. In relation to the values of hospital services, a total of R$35,538,127.81 was spent in this time interval, with 63.9% of the funding allocated to the treatment of female patients. The region with the highest cost during the study period was the South (31.9%), while the lowest was the North (3.9%), which also presented the lowest number of hospitalizations among all regions of the country (5.1%).

Conclusion: Between 2010 and 2020, hospitalizations and costs of hospital services related to the treatment of migraine and other painful headaches were predominantly directed towards female patients. Middle-aged adults were the age group with the highest number of hospitalizations. The region with the lowest hospitalizations and hospital costs for this condition was the North.

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775 – Fluctuating spastic paraparesis with "owls sign" in spine MRI: a diagnostic challenge

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Case presentation: A 42-year-old female patient, with no previous comorbidities, complained of tremors and gait disturbance that had started two years before. She mentioned that her legs had fluctuating moving difficulties, associated with pain. At her first evaluation at clinical onset, the case was interpreted as episodic myoclonus, and clonazepam was prescribed with partial improvement. In the initial physical examination, it was observed pathologic glabellar reflex, bilateral hyperreflexia in the lower limbs, and bilateral foot and knee clonus, with no findings in tonus or muscle weakness. The patient was reassessed after one month, showing moderate spasticity in the lower limbs. A variable character of hypertonia was noticed in the following days. Ancillary tests, brain MRI and electroneuromyography was unremarkable. Spinal cord magnetic resonance imaging (MRI) showed hyperintensity in the corticospinal tract, with "owl-eyes" sign. Cerebrospinal fluid (CSF) analysis was unremarkable, except for the presence of oligoclonal bands. Other laboratory findings included serum anti-TPO 1200 U/mL and Anti-GAD65 in CSF 3081UI/mL. Therefore, the diagnosis of Stiff-Person syndrome was established. The patient was treated with human immunoglobulin with good clinical response.

Discussion: Stiff-Person syndrome (SPS) is a rare autoimmune disorder, characterized by insidious, fluctuating, progressive hypertonia. Some case reports

have shown signal changes in the spinal cord on MRI, especially associated with amphiphysin autoantibodies. The "owl-eyes" sign is a hypersignal observed in the anterior horns of the spinal cord in the axial MRI T2-weighted imaging, being more commonly described in medullary infarcts and compressive myelopathy. So, this sign is uncommon in SPS.

Conclusion: Although rare, Stiff-Person syndrome associated with GAD-65 can be included in the differential diagnosis of myelopathy with the "owl-eyes" sign, if the clinical history is suggestive.

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776 – Leprosy neuritis due to COVID-19: a case report

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Introduction: Leprosy is one of the oldest diseases of humanity and still one of the most neglected. The coronavirus disease 2019 (COVID-19) pandemic has caused several entities to be concerned about the impact of the disease caused by severe acute respiratory syndrome coronavirus 2 (Sars-CoV-2) in patients previously affected by other comorbidities.We report a case of leprosy neuritis due to COVID-19.

Case: A 39-year-old male patient was diagnosed with borderline-lepromatous leprosy in 2015. On physical examination, he presented nerve thickening and an asymmetric multiple mononeuropathy on electroneuromyography (ENMG). He was treated with Rifampicin-Ofloxacin-Minocycline regimen for two years, with no sequelae. In 2021, he started with pain and paresthesia in the plantar and dorsal regions of the feet. He evolved with difficulty in walking and physical examination showed diffuse paresthesia and evident allodynia on the soles and dorsum of the feet, bilaterally. The patient did not present proximal muscle weakness, nor impairment of deep osteotendinous reflexes. Cerebrospinal fluid analysis was normal. About fifteen days prior to this, he was detected with COVID-19, manifested in a mild way, requiring only symptomatic medications. The ENMG showed signs of an asymmetric multiple mononeuropathy, predominantly axonal and sensorial, of severe intensity, with multiple associated focal myelin impairments, such as temporal dispersion. There were a severe impairment of the left ulnar, left fibular and tibial nerves. He was then diagnosed with type 1 leprosy reaction and underwent monthly pulse therapy with methylprednisolone, with an important clinical response.

Discussion: Since the beginning of COVID-19, the possibility of an interaction between SARS-CoV-2 and *Mycobacterium leprae* has been raised, especially due to the immunological component of both diseases. Some authors have warned of the possibility of the influence of COVID-19, especially during its inflammatory phase, in triggering leprosy reactions, since infections are risk factors for their occurrence. Post-infectious reactional episodes always represent a challenge in clinical practice, as they are the main cause of disability and maintenance of leprosy-related stigma.

Conclusion: The case reports a significant neurological impairment in a patient previously diagnosed with leprosy after SARS-CoV-2 infection. The importance of verifying possible post-pandemic impacts on historically neglected pathologies is highlighted.

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777 – Sex differences in Parkinson's disease: is there an impact on the national health system? a ten-year analysis

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Introduction: Parkinson's disease (PD) is the second most prevalent neurodegenerative disease worldwide. National data on its incidence and impact on public health are scarce. Moreover, there are marked differences in sex-related factors, with males exhibiting a higher risk of both developing the disease and experiencing mortality.

Objectives: To compare hospitalizations, deaths, and associated costs related to PD between sexes from 2010 to 2020.

Methods: This is a descriptive epidemiological study. Data was collected from the Brazilian National Health System's (DATASUS) database, describing PD cases (CID-10) between 2010 and 2020. As this is a public domain database, there was no need for ethics committee approval.

Results: The total number of hospitalizations for PD in Brazil during the studied period (2010–2020) was estimated at 9,993, with 57.4% being male patients. The year with the highest number of hospitalizations was 2016 (1,154 cases), which contrasts with 2020, where only 58.6% of this amount was reported, potentially indicating an impact of the coronavirus pandemic. Regarding mortality, the data show a total of 38,898 deaths associated with PD during the studied period, with 55% being male patients. Although 2010 had the lowest number of reported deaths, the value has increased over the years. In relation to the values of hospital services for PD provided by the National Health Service (SUS) in this time interval, a total of R$27,386,399.29 was spent, with 62.6% of the funding allocated to the treatment of male patients.

Conclusion: Between 2010 and 2020, hospitalization, mortality, and public hospital service expenditure associated with Parkinson's disease were predominantly higher for male patients compared to their female counterparts. These results are consistent with the current literature.

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778 – Bilateral pallido-nigral lesions in a patient with subacute chorea after diabetic ketoacidosis: case-report

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Case presentation: A 54-year-old female patient, with prior history of arterial hypertension and type 2 diabetes, presented a one-year history of bilateral weakness in lower limbs, cognitive impairment and irregular hyperkinectic movements in upper limbs and face. The clinical onset was subacute after

hospitalization due to severe pneumonia and diabetic ketoacidosis, which required intensive care unit care and ventilatory assistance for more than 15 days. On physical examination, she presented cognitive impairment with dysexecutive predominance, spastic paraparesis and facial and upper limbs chorea, mainly on the left side. Laboratory findings included a vitamin B12 level of 184 pg/mL. Brain magnetic resonance imaging revealed marked hypointensities in the globus pallidus and mesencephalic substantia nigra bilaterally, besides global encephalic atrophy and nonspecific white matter hyperintensities. Patient improved of chorea with administration of risperidone.
Discussion: Toxic and metabolic lesions can involve the basal ganglia symmetrically, due to the high rate of metabolism and oxygenation required by these structures. The most common etiology of bilateral pallidal lesions are poisoning by carbon monoxide and cyanide intoxication. In this case, diabetic ketoacidosis was observed as a possible precipitating factor for the pallidonigral lesions, which is an uncommon association for this type of injury.
Conclusion: Patients with acute and subacute movement disorders should be extensively investigated for injuries in basal ganglia associated to hypoxic or metabolic etiologies.

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781 – Brain metastases from lung adenocarcinoma with dramatic response to anti-egfr therapy – case report

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A 60-year-old man developed, within a month, right thoracic pain, shortness of breath and weight loss. He had a history of working in a metallurgical company and was exposed to chemical paints. He was admitted into the emergency department and a chest computed tomography revealed a large solid mass at the left superior lung lobe, measuring 80 mm x 40 mm. A biopsy was performed and confirmed the diagnosis of Lung Adenocarcinoma. Cancer staging was performed and revealed liver, bone and brain metastases. The brain magnetic resonance imaging disclosed several metastatic lesions at the right postcentral gyrus, occipital lobes and both cerebellar hemispheres. He underwent whole brain radiation therapy and received five cycles of palliative chemotherapy (carboplatin in combination with paclitaxel, and gemcitabine in combination with cisplatin). A second biopsy was performed at the liver metastasis, to search for possible targeted therapies. The results came positive for Epidermal Growth Factor Receptor (EGFR) exon 19 deletion, and an anti-EGFR was prescribed. The patient was started on Osimertinib, 80 mg once a day. The one year follow up showed great response and good drug tolerance, with an important improvement of muscle strength, asthenia and sensory deficits. The patient became fully ambulatory with a Karnofsky Performance Status Scale score of 90 and on the Neurologic Assessment in Neuro-Oncology Scale a score of 3 out of 23. Imaging studies revealed a smaller number of brain metastatic lesions, with decreased contrast enhancement. Therefore, the patient has had a dramatic response to the targeted therapy, both clinical and imaging. Targeted therapy is a growing field in neuro-oncology, leading us to an era of personalized medicine and tailored treatments, with great improvement in overall survival and quality of life. Unfortunately, in Brazil targeted therapies are still not widely available, due to high costs.

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782 – Acute psychotic episode after surgical complication of endoscopic septoplasty with iatrogenic sella turcica injury and pituitary apoplexy

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Introduction: The nasal septum is an important structure, responsible for the centralization of the nose to the face. Septum deviation is a symptomatic condition, typically characterized by chronic nasal obstruction of the upper airways. Correction of deviated septum needs to occur through surgical therapy, notably through endoscopic septoplasty. The aim is to report and discuss the case of a man who had an acute psychotic episode after a surgical complication endoscopic septoplasty with iatrogenic saddle turmeric injury and pituitary apoplexy.
Case report: A 47-year-old man attempted suicide the night before presenting with psychosis following a septoplasty procedure to correct a deviated nasal septum six years ago. At the present date, in a psychiatric consultation, complaints of insomnia, restlessness and paranoid delusions. On psychiatric examination, he was lucid, with disorganized thinking and paranoid delusions. Properly medicated, the patient returns to medical care with an improvement in his general condition, but maintains the condition of psychic disorganization. However, the patient decided to suspend the prescribed medication and did not return for the appointment on the scheduled date. After a week, the patient developed psychotic symptoms, mystical delusions, insomnia and irritability that triggered a new suicide attempt. Referred to emergency and later to psychiatric hospitalization, where he remained for 22 days. The patient's diagnosis was mixed bipolar disorder.
Conclusion: The following case report depicts a rare case of hypopituitarism, resulting from an iatrogenic injury to the sella turcica and the pituitary gland, complications triggered by the performance of endoscopic septoplasty, which resulted in a severe psychotic condition.

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783 – Gut-microbiome-brain-axis: the crosstalk between the vagus nerve, alpha-synuclein and the brain in Parkinson's disease

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Introduction: The vagus nerve, the main component of the parasympathetic nervous system, is involved in the regulation of immune response, digestion,

heart rate, and control of mood. It can detect microbiota metabolites through its afferents, transferring this gut information to the central nervous system. Preclinical and clinical studies have shown the important role played by the gut microbiome and gut-related factors in disease development and progression, as well as treatment responses.

Objectives: To describe and discuss the close link between the microbiome, the gut and the brain in Parkinson's disease.

Methods: This is a critical review of the literature on the microbiome, gut, and brain in Parkinson's disease.

Results: The gut microbiota has been demonstrated to be a pivotal contributor to the promotion of health. Emerging data has indicated that, up to 20 years before the onset of motor symptoms, an alteration in the gut microbiome may be present in Parkinson's disease patients. This dysbiosis of the gut may lead to increased intestinal permeability and inflammation, as well as Lewis body formation, and can also cause neuroinflammation and decreased neurotransmitter production in the central nervous system.

Conclusion: More studies are needed to better understand the underlying biology and how this axis can be modulated for the patient's benefit.

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784 – Cryptococcal meningoencephalitis: case report

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Case presentation: A 44-year-old male, retired, schizophrenic, living in an urban area, with a recent history of hospitalization due to hypokalemia, was admitted to the emergency due to paresis in lower limbs for three days associated to dysarthria and somnolence. He was confused, presenting nuchal rigidity, divergent strabismus to the left and hypoesthesia in the right lower limb. Magnetic resonance imaging of the brain showed acute periventricular ischemic events, semi-oval centers, mesencephalon and pons; human immunodeficiency virus negative (2 samples); cerebrospinal fluid (CFS): pleocytosis 123 (L49%/N48%/M8%), glucose < 5, protein 103, fungal survey suggestive of Cryptococcus sp (China ink positive), cryptococcal antigen titer: >1/1024, cultures negative. Initiated Amphotericin B and Fluconazole evolved with deteriorating clinical condition to death.

Discussion: Cryptococcal meningoencephalitis is the main presentation in immunocompetent patients, usually caused by Cryptococcus gattii. The clinical presentation is assorted and may include fever, central nervous system signs or symptoms such as headache, behavioral changes, memory loss, and lethargy. Diagnosis is based on CSF analysis with China ink evaluation, its' routine studies, fungal culture and cryptococcal antigen testing. The treatment consists of Amphotericin B in association to Fluconazole by, at least, 14 days. The outcome depends on the degree of immunocompetence, associated diseases, CSF/serum cryptococcal antigen titer. Final considerations: The relevance of the case considers the patient's age and its rapid evolution, despite the protocoled therapeutic measures, emphasizing the importance of early identification and treatment of the neuroinfection.

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785 – Diabetic striatopathy: a case report

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Introduction: Striatopathy or non-ketonic hyperglycemia is a rare etiology, but well reported in the literature, of involuntary movements in patients with diabetes mellitus, being characterized by affecting, in general, elderly patients with hemicorrea-hemiballismus. The aim is to report a clinical case of the development of movement disorder caused by hyperglycemia.

Case report: Patient E.J.D.O, 71-year-old, male, hypertensive and dyslipidemic. Admitted to the emergency room with a history of involuntary movements in the upper left limb for about two months, with recent worsening. Upon admission, a hyperglycemic state (capillary blood glucose HI) was observed, until then, the patient had not been diagnosed with diabetes. The movements were present at rest, worsened on intention and were present during sleep, being characterized as clonic/myoclonic movements. In view of the clinical picture on admission, the hypothesis of subtle focal status epilepticus was suggested, and therefore Diazepam 2 mg EV was performed as a therapeutic test, with notable improvement of the picture. As a complementary propaedeutic, brain magnetic resonance imaging was performed. An electroencephalogram was also requested, which showed no epileptiform activity in status epilepticus. Cerebrospinal fluid collection was performed, showing possible reduction of diffusion. The patient was discharged after receiving valproic acid and clobazam, however, he returned later, reporting worsening of movements to which he associated the interruption of medications. The hypothesis of autoimmune encephalitis was raised, but no immunosuppressive therapy was instituted. During the second hospitalization, he had complications, including CRA, progressing to a state of hypoxic-ischemic encephalopathy.

Conclusion: In this report, we describe a patient with focal involuntary movements who made a differential diagnosis between epileptic seizures/focal status evil with movement disorders of metabolic etiology such as hyperglycemia, with the topographic radiological finding corroborating such differential diagnosis. In the literature, there is a consistent association between movement disorders and hyperglycemia, especially in elderly patients, which adds strength to the present case report. Korea.

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786 – Manganese neurotoxicity: a case report

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A 51-year-old female patient, teacher, presented initially with hematemesis associated with hemodynamic instability, with recurrence of the condition for two times and other several clinical complications. During hospitalizations,

she developed progressive weight loss and alteration of the mental state, in addition to reduced strength in the four limbs, cognitive decline of rapid evolution and loss of functionality and independency for basic activities of life daily. The cognitive exam revealed significant alterations with impaired sustained attention and verbal fluency, loss of executive and visuospatial functions. The somatic exam showed rigidity and bradykinesia in the four limbs, with reduction of blinking, in addition to important postural instability, with Cock-Walk Gait. Laboratory and abdominal imaging pointed signs of chronic liver disease and magnetic resonance imaging (MRI) of the brain showed a hypersignal on T1 sequence in bilateral globus pallidus (GP) with a symmetrical appearance. Such changes could be related to an acquired hepatocerebral degeneration associated with manganese (Mn) deposition in the GP. Treatment for parkinsonism and hepatic encephalopaty were established In outpatient return, an important improvement was seen, especially in the cognitive tests. The association between chronic liver disease and parkinsonism added cock-walk gait suggest the clinical of chronic acquired hepatocerebral degeneration, a rare case of deposit of Mn in the central nervous system (CNS), which causes neurotoxicity, neurological disorders and MRI findings. Chronic liver disease is associated with Mn clearance and binding mechanisms, as well as metal transport across the blood-brain barrier. Associated, there are several neurotoxicity mechanisms still under investigation, which cause the deposition of the metal in the GP. More investigations are needed to delineate the complex nature of the mechanisms of Mn-induced CNS toxicity.

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787 – Anterior cerebral artery: a borderland territory between neurology and psychiatry

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Introduction: The association of depressive and apathetic symptoms after lesions in the frontal region in the territory of the anterior cerebral artery is described, presumably due to interruption of complex subcortical frontal circuits.
Objectives: Illustrate and discuss a bilateral frontal syndrome resulting from stroke in the territory of the anterior cerebral artery, as well as, the possible confusion between functional and organic conditions presenting the same symptoms.
Results: 61-year-old man complaining of falls and left lower limb weakness. Moreover, had been interned due to suicide attempt recently. He is hypertensive and was addicted to alcohol and illicit drugs. Two years before, he had a left anterior cerebral artery ischemic that led to right lower limb paresis. At admission, he had poor interaction and presented hyperreflexia and Babinski sign at the right lower limb while normoreflexia at left. Computed tomography (CT) of the skull showed left frontal encephalomalacia. It was discarded metabolic and infectious disturbance as well as no signs of trauma from reported falls or high-risk lesions for new stroke. Psychiatric evaluations identified depression related to social, and economic factors. Pharmacological and behavioral measures were instituted. However, the condition worsened with the onset of mutism, akinesia, and abulia, which was attributed to the worsening of depression. Antidepressant regimen was adjusted again and new brain CT was performed. It disclosed a new stroke of the right anterior cerebral artery. Initially, the symptoms were attributed to a functional condition and no organic justification was found. Nevertheless, the worsening of the basal status evolved to a catatonic manifestation, in parallel to a new frontal stroke. Previous contralateral frontal stroke might have turned the patient vulnerable to this condition. Nonetheless, the course of time eased by pharmacological strategy used (lithium, amitriptyline, and risperidone), was able to reverse the condition, except for left leg monoplegia evidenced after the restoration of spontaneous motricity.
Conclusion: The case illustrates the clinical-anatomical correlations of anterior cerebral artery stroke, but also highlights the interface between neurology and psychiatry.

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790 – Real World Safety and exploratory efficacy of gene therapy for patients with 5q-Spinal Muscular Atrophy in a Brazilian cohort

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Introduction: Spinal muscular atrophy (SMA) is a motor neuron disease associated with progressive muscle weakness, ventilatory failure, and reduced survival. Onasemnogene abeparvoves is the first gene replacement therapy (GT) approved to treat this condition.
Methods: Observational retrospective study to assess adverse events and efficacy of gene therapy in types 1 and 2 SMA patients through motor milestones and functional motor scales.
Results: Forty-one patients with SMA (24 females, 58.5%) were included, including 33 (80.1%) SMA type 1, seven (17.1%) type 2, and one pre-symptomatic. The mean age at GT dosing was 18 (±6.4) months. Thirty-six patients (87.8%) were under previous treatment with nusinersen, and 10 (24.4%) continued nusinersen after GT dosing. Mean CHOP-INTEND increased 13 points after 6 months and 22 points after 12 months of GT ($P < 0.001$). CHOP-INTEND increase at six months did not differ between groups according to nusinersen maintenance after GT ($P = 0.949$). Among SMA type 1 patients, 14 (46.6%) reached the ability to sit alone, and three (10%) stood with support. Liver transaminases elevation at least two times higher than the upper limit of normal value occurred in 29 (70.7%) patients. Thrombocytopenia occurred in 13 (31.7%) patients, and one case presented thrombotic microangiopathy. Older age (> two years) was associated with more prolonged use of corticosteroids ($P = 0.021$).
Conclusion: GT is effective in SMA patients, and combined nusinersen after GT does not present additional gain in motor function. Close monitoring of adverse events is necessary, and older age is associated with prolonged corticosteroid use.

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