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ANTI-GQ1B IGG ANTIBODY SYNDROME: MILLER FISHER SYNDROME AND BICKERSTAFF'S BRAINSTEM ENCEPHALITIS SUPERPOSITION

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Introduction: Miller Fisher syndrome (MFS) is clinically characterized by acute external ophthalmoplegia, ataxia, and areflexia; while impaired consciousness, external ophthalmoplegia and ataxia constitute Bickerstaff's brainstem encephalitis (BBE). Anti-ganglioside antibodies, anti-GQ1b, are associated with MFS, Guillain-Barré, acute isolated ophthalmoplegia and cranial polyneuropathy.

Objective: To present a pediatric patient with sequential development of MFS and BBE with positive serum anti-GQ1b IgG antibody.

Case description and methods: A 3 year-old previously healthy boy, presented with ataxia, ophtalmoparesia, areflexia, lower limb weakness and swallowing difficulties. With SMF diagnosis, he was treated with intravenous immunoglobulin showing marked improvement. A week later he presented with apathy, mutism, orolingual automatisms and impaired consciousness associated with pyramidalism. He was given high dose steroid treatment, after a BBE diagnosis was made, followed by complete recovery.

Results: Brain MRI showed midbrain and cranial nerve enhancement. Albuminocytologic dissociation in CSF was present. Nerve conduction study: motor and sensory polyradiculneuropathy, with facial involvement and blink reflex absence. Anti-GQ1b IgG antibody was positive.

Conclusions: Sequential development of MFS and BBE occurred in a pediatric patient with positive serum anti-GQ1b IgG antibodies. Immunotherapy helped recovery.

Serum IgG antibodies to GQ1b can cause different clinical phenotypes, with overlapping between them, MFS and BBE being considered part of a continuous clinical spectrum.

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THERAPEUTIC PLASMA EXCHANGE IN CHILDREN WITH IMMUNE-MEDIATED ENCEPHALOPATHY AND A PARTIAL RESPONSE TO IMMUNOMODULATORY TREATMENT

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Introduction: Early recognition of immune-mediated encephalopathies is fundamental given that the entities are rapidly progressive and potentially reversible with adequate treatment. Immunomodulatory treatment, however, may not be completely effective. The aim of this study was to assess the impact of therapeutic plasma exchange (TPE) in children with immune-mediated encephalopathies and a partial response to immunomodulatory treatment.

Methods: A retrospective study was conducted over the period 2011-2012. Immune-mediated encephalopathy was defined according to clinical and ancillary study criteria. TPE was performed in patients with persistent symptoms in spite of immunomodulatory treatment. The modified Rankin Scale (mRS) (0-6) was used at onset, immediate pre- and post-treatment, and at the last visit to assess clinical outcome. Data were analyzed with the Wilcoxon test.

Results: Seven of 18 children, with ages ranging from 4 to 14 years, underwent TPE. Three patients were positive for Anti-NMDA antibodies. Time between disease onset and TPE ranged from 18 to 180 days. The initial mRS score was 5 in all patients. A post-TPE decrease of the score of 1 to 2 points was found (p=0.011). Symptoms improved after the third TPE procedure and after day 9. Adverse events observed were hypocalcemia and catheter-related complications.

Conclusions: The present study shows the efficacy of TPE as a therapeutic option in children with immune-mediated encephalopathies with a partial response to immunomodulatory treatment, even in those with chronic disease.

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THE AETIOLOGY, OUTCOME AND MRI OF ACUTE CHILDHOOD ENCEPHALITIS IN A RETROSPECTIVE AUSTRALIAN COHORT; EMERGING ANTIBODY-MEDIATED ENCEPHALITIDES

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Introduction: Encephalitis is a common and potentially devastating illness in childhood. The aetiology includes infectious and non-infectious subgroups. Antibody-mediated encephalitis has emerged as an important and treatable subgroup of encephalitis. These autoantibodies bind to antigens on neuronal cell surface. They include the N-Methyl-D-Aspartate receptor (NMDAR), voltage gated potassium channel complex (VGKC)-complex and dopamine-2 receptor (D2R). This study particularly investigated the frequency of autoimmune encephalitis in children.

Methods: A retrospective study on 164 immunocompetent patients diagnosed with encephalitis at the Children's Hospital at Westmead from January 1998 to October 2010. The clinical features and MRI brain (n=149) data were reviewed. Autoantibodies were tested on stored ***acute*** samples of 78% of patients [acute disseminated encephalitis (ADEM) patients excluded].

Results: The most common identified aetiology in descending order of frequency was ADEM (21.3%), enterovirus (12.2%), mycoplasma ***pneumonia*** (6.7%), NMDAR encephalitis (6.1%), herpes simplex virus (5.5%) and VGKC-complex encephalitis (4.3%). The aetiology was unknown in 28% of patients. The outcome at median follow up of 6.5 years was abnormal in 56.1% of patients. The MRI brain was abnormal in 81.2% of patients. The most common MRI phenotypes were ADEM, multifocal grey matter, basal ganglia and limbic encephalitis.

Discussion: Immune-mediated encephalitis (ADEM, NMDAR, VGKC-complex and D2R) is the most common (34.1%) potentially treatable aetiology of childhood encephalitis in Australian children. The long-term outcome of childhood encephalitis remains a concern. The limbic, basal ganglia and multifocal grey matter encephalitis MRI phenotypes were more frequently seen in antibody-mediated encephalitis and mycoplasma encephalitis.

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HEMORRHAGIC ACUTE DIFFUSE ENCEPHALOMYELITIS ASSOCIATED WITH BORDETELLA PERTUSSIS

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Introduction: ADEM (Acute Diffuse Encephalomyelitis) is a demyelinating inflammatory white matter disease observed in any age group, especially in children under 10 years. Usually It hás a favorite prognosis, but sometime it can evolve into coma, convulsions and even death, mainly in cases of hemorrhagic ADEM.

Case report: Eight-months-old female, with one week lasting cough, presenting fever and focal seizures. Liquor had 2 leukocytes and 6,800 erythrocytes. She received ceftriaxone, phenobarbital, and phenytoin. Initial neurologic examination was normal. PCR was positive to Bordetella pertussis (BP). Brain CT (Computed Tomography) showed mixed lesions on the right fronto-parietal region, contrast enhancement, without mass effect. One week later, she presented left hemiparesis with no conscious compromise. Brain MRI (Magnetic Resonance Imaging) showed diffuse hypo and hyperintense lesions in grey and white matter, vasogenic edema and middle line deviation. She received pulse therapy with methyl prednisolone, but developed arterial hypertension, bradycardia, and conscious compromise, needing mechanical ventilation. A intracranial hypertension was installed, which was refractory to all therapeutic measures, such as manitol, hypersaline solution, dexamethasone, barbituric coma. Fifteen days later initial symptoms, cerebral death was diagnosed.

Conclusion: Although usually occurring as an auto-immune response after infectious diseases or post-vaccination procedures,

ADEM remain with no clear etiology. Fortunately it is a rare situation. The hemorrhagic form of ADEM is even rarer, but can be lethal. In our case, BP was identified in a case that evolved into hemorrhagic ADEM. The possible cause-effect relation between this infection and neurologic outcome is controversial, but plausible.

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MESENTERIC ISCHEMIA FOLLOWING IMMUNOGLOBIN INFUSION FOR TREATMENT OF ANTI-NMDA RECEPTOR ENCEPHALITIS

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Introduction: Anti-NMDA receptor (NMDAr) encephalitis is an autoimmune disease that presents with psychiatric symptoms, epileptic seizures and movement disorders. Early diagnosis is essential for efficacy of immunotherapy; however, anti-NMDAr encephalitis is frequently mistaken for other autoimmune or infectious diseases.

Case description: A five-year-old boy presented with abnormal movements, epileptic seizures and progressive encephalopathy. The magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) were normal. Antiepileptic drugs were started and after seven months of the beginning of the symptoms he was seen in our service. He was unresponsive and with severe choreoathethosis. The MRI showed FLAIR signal hyperintensity bilaterally in insular cortex and CSF was normal. Electroencephalogram showed disorganized activity with electrographic seizures in right parasagital area. Anti-NMDAr antibodies were positive in CSF and serum. Investigation for associated tumors was negative. A course of methylprednisolone was attempted with no improvement. The child was treated with intravenous immunoglobulin, however in the fourth day he developed mesenteric thrombosis and underwent a large intestinal resection. He remained in intensive care unit for months and presented with several complications. After two months of immunotherapy, when he was clinically stable, the choreoathethosis improved and he was significantly more active and communicative.

Conclusion: Despite severe neurological features, anti-NMDAr encephalitis is a potentially reversible disease. Delay in diagnosis is frequent due to misleading presentation and may cause inferior response to immunotherapy. Our case illustrates some of the problems in the diagnosis and follow-up of patients with anti-NMDAr encephalitis, including rarely described adverse effects of immunotherapy.

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CASE REPORT: SUBACUTE SCLEROSING PANENCEPHALITIS

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SUMMARY: Subacute sclerosing panencephalitis (PES) is a progressive inflammatory disease of the central nervous system related persistent infection and abnormal measles virus (Paramyxovirus), presenting high mortality and poor prognosis. It affects primarily children and young adults, in 50 % of cases, infection history Paramyxovirus below 2 years of age; being age a risk factor for ESP, due to immaturity of the immune system. The incidence is 1-4:100.000 cases1, 7. The disease progresses slowly with progressively divided in 4 stages.

OBJECTIVE: Present a case diagnosed with Subacute Sclerosing Subacute and compare the data collected with the current scientific literature.

CASE REPORT: Patient, 16 years old female with history of social isolation and difficulty in school since October 2011; evolved with changing behavior, frequent falls, myoclonus, difficulty speaking and perform their daily activities. Reported Measles box with 1 year of age without complications. Neurological examination presents poor contact lack of understanding; Walking with broad-based, with momentary stoppages due to periodic myoclonus associated with facial expression. The EEG showed characteristic for this disease stroke, as well as antibodies and hypergammaglobulinemia in the CSF.

CONCLUSION: The diagnosis is made by the previous history of Measles virus infection, presence of complexes Radermacker and anti-measles antibodies in the blood and cerebrospinal fluid; histopathological findings in brain biopsy or autopsies. It is a disease with fatal course, and no treatment.

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OPTIC NEURITIS IN JUVENILE IDIOPATHIC ARTHRITIS PATIENT

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Optic neuritis (ON) was rarely reported in juvenile idiopathic arthritis (JIA) patients, particularly in those under anti-tumor necrosis factor alpha blockage. However, to our knowledge, the prevalence of ON in JIA population has not been studied. Therefore, 5,793 patients were followed up at our University Hospital and 630 (11%) had JIA. One patient (0.15%) had ON and was reported herein. A 6 year old male was diagnosed with extended oligoarticular JIA, and received naproxen and methotrexate subsequently replaced by leflunomide. At 11 years old, he was diagnosed with aseptic meningitis followed by a partial motor seizure with secondary generalization. Brain magnetic resonance imaging (MRI) and electroencephalogram showed diffuse disorganization of the brain electric activity and leflunomide was suspended. Seven days later, the patient presented acute ocular pain, loss of acuity for color, blurred vision, photophobia, redness and short progressive visual loss in the right eye. Fundoscopic exam detected unilateral papilledema without retinal exudates. Orbital MRI suggested right ON. The anti-aquaporin 4 (anti-AQP4) antibody was negative. Pulse therapy with methylprednisolone was administered for five days and subsequently with prednisone, he had clinical and laboratory improvement. In conclusion, a low prevalence of ON was observed in our JIA population. The absence of anti-AQP4 antibody and the normal brain MRI not exclude the possibility of demyelinating disease associated with chronic arthritis. Therefore, rigorous follow up is required.

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NEUROMYELITIS OPTICA IN CHILDREN – A SEVERE DISEASE.

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Introduction: Neuromyelitis optica (NMO) is an inflammatory demyelinating disease with predilection for the spinal cord and optic nerves. In relation to Multiple Sclerosis, has lower prevalence, however, the number and severity of relapses is higher, with a worst response to acute phase treatment.

Methods: Report of two cases ***Patient1:*** JKS, female, presented her first outbreak at seven years old with tetraparesis. MRI: extensive transverse myelitis. Nowadays she is 14 years old and presented five other outbreaks. MRI showed inflammation of optical nerves, what may represent a subclinical outbreak. Asymptomatic using azathioprine and oral prednisone - EDSS 1, 0.

Patient 2: MAS, female, fist symptoms at 11 years old with a series of self-limited episodes of crural paresis. At 14, presented tetraplegia. She did not search medical attention for this severe episode which resolved spontaneously. At 15, presented blurred vision, that progressed to bilateral amaurosis. After three days of pulsotherapy with steroids she partially recovered vision.

MRI: longitudinally extensive high intensity signal admitted at our hospital with irregular usage of oral prednisone, changed to monthly pulse therapy for non-adherence. Nevertheless, presented with another three outbreaks. Present medication is azathioprine plus monthly pulsotherapy, without clinical relapses, but with great sequelae (EDSS 5.5).Both patients were positive for anti-aquaporin 4 antibody.

Discussion: We present two distinct cases and consider differences in treatment and outcome according to the time of diagnosis and treatment, considering early recognition and adherence to treatment as important variables for reducing future outbreaks and sequelae.

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RECURRENT ACUTE DISSEMINATED ENCEPHALOMYELITIS (ADEM): A CASE REPORT

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ADEM is usually monophasic demyelinating disease of the central nervous system associated a clinical presentation of focal neurologic signs few days to several weeks after clinical onset of the viral infection or vaccine. Recurrent ADEM occur in 10 to 29% of children which is characterized by a second attack, involving the same anatomic areas, more than three months after the initial event. We describe a case of a boy with 7 years old, who was diagnosed epilepsy and hemiparesis. Patient had normal development, when at 5 years, he presented generalized tonic-clonic seizures and right hemiparesis. Carbamazepine was introduced for six months and an overall improvement was seen. During one year, he was asymptomatic and anticonvulsivant was suspended. Six months later, the epilepsy return and CBZ was restarted. One year later, he was admitted with abdominal pain, vomiting and right proportional hemiparesis. Neurological exam constated right low strength. The diagnostic procedures was performed by brain and neuroaxial MRI, which registered hyperintensity on T2 in the periventricular white matter , subcortical region , the right half of the midbrain and the right middle cerebellar peduncle. Presence of lesions in contact with the corpus callosum . Multiple foci of hyperintensity in the cervical and thoracic spinal cord , especially at the C1-C2 level. There isn't contrast enhancement. Neurophysiologic study shows slow activity in the left frontal- central -temporal regions, compatible with focal epilepsy syndromes. Emphasize the importance to discuss ADEM and epilepsy like initial symptom.

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STRIVING FOR THE BEST TREATMENT FOR PEDIATRIC ACUTE DEMYELINATING SYNDROMES: RESULTS FROM COHORT ANALYSIS

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INTRODUCTION: The 2013 International Pediatric MS Study Group report brings awareness to the difficulties in managing the diagnostic work-up and treatment of Pediatric Acute Demyelinating Syndromes (ADS), and highlights their differences from adult onset forms and the need for pediatric guidelines. Taking into account that pediatric ADS course with increased inflammatory response, frequent relapses, long term disabilities and quality of life losses, a specialized outpatient clinic to follow-up children with ADS was created in our hospital. Cohort analysis of these patients and work-up/therapeutic strategies are described in this study.

METHODS: Prospective cohort analysis from a Pediatric ADS clinic population over the past year.

RESULTS: Fourteen patients (7 boys), mean age of 7.3yo (range: 1.8-13yo), at their first episode of ADS were seen in clinic, and protocols were created to be used through their differential diagnosis, relapses and remission phases. Four presented with multiple sclerosis (MS), 5 with Acute Disseminated Encephalomyelitis (ADEM), 2 with Neuromyelitis Optica (NMO), and 3 with Clinically Isolated Syndromes. Six patients presented ADS symptoms prior to temporal-spatial dissemination diagnosis. Serum from all patients were tested for inflammatory biomarkers: IgG index calculation, oligoclonal Bands, lymphocyte counts, cytokines (IFN-gamma, TNF-alpha, IL-17, IL-4 and IL-10). Currently, three patients with MS are treated with subcutaneous Interferon beta-1A, and a fourth with Glatiramer acetate due to prior treatment failure. NMO is treated with Azathioprine or Glatiramer acetate. Immunotherapy/cyclophosphamide treatment is used for ADEM with good response.

CONCLUSION: This preliminary cohort analysis describes our current work-up and therapeutic protocols for ADS at our hospital.

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EXTENSIVE UNIHEMISPHERIC WHITE MATTER LESION AS ATYPICAL PRESENTATION OF X-LINKED ADRENOLEUKODYSTROPHY

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Introduction: X-linked adrenoleukodystrophy (X-ALD) is a recessive neurodegenerative disease with impaired beta-oxidation of very long-chain fatty acids (VLCFA). The typical presentation is symmetrical parieto-occipital white matter lesions in magnetic resonance imaging (MRI), but a few rare cases of atypical clinical presentation and neuroimaging were described.

Case description: A ten-year-old boy, the only son of non-consanguineous marriage, presented with aggressive behavior and right hemiparesis. The MRI showed white matter lesion in left hemisphere. He was initially diagnosed as acute demyelinating encephalomielopathy (ADEM) and treated with steroids with little results. The boy later presented with worsening of clinical features, epileptic seizures and loss of speech. The mother reported darkening of the skin. At nine months after the beginning of the symptoms he was seen in our service. Another MRI was performed, showing white matter lesion in bilateral frontal lobes with extension along cortifugal pathways, left parietal lobe and midbrain with periperal enhancement. The cerebrospinal fluid was normal and electroencephalogram showed right epileptic activity. He underwent brain biopsy with unspecific inflammatory changes and blood screening with high levels of VLCFA confirmed the diagnosis.

Discussion: Atypical forms of X-ALD were described in less than 5% of cases, including only frontal lesions and asymmetric involvement. In these situations differential diagnosis with neoplastic processes can be challenging and delay in diagnosis is frequent. In our case, the skin changes suggestive of adrenal insufficiency and the absence of brain edema alerted to the diagnosis of X-ALD.

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ASSOCIATION BETWEEN ACUTE DISSEMINATED ENCEPHALOMYELITIS AND GUILLAIN-BARRÉ SYNDROME IN A CHILD

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Introduction: Acute disseminated encephalomyelitis (ADEM) and Guillain-Barré Syndrome (GBS) are commonly recognized as separated entities. Both are hypothesized to be related to a viral-induced autoimmune response. There are only few reports of concomitant ADEM and GBS.

Case report: A 2-years-old boy began with vomiting, irritability, and three days later with eyelid and scrotal edema. There was no history of recent fever or infection. Forty days before, he received anti-polio and tetravalent vaccines. Scrotal echography showed edematous infiltration. No other laboratorial alterations occurred. After 72 hours, irritability worse and somnolence was noted. He developed flaccid paraplegia, distal weakness of inferior limbs, absence of reflexes, sphincter dysfunction, and paralyses of the right sixth nerve. CNS MRI showed multiple cerebral and cerebellar lesions on the white matter, hyperintense on T2 and flair sequences. No medullar lesion occurred. CSF showed 30 leukocytes with mononuclear predominance, glucose 68, total proteins 95, negative for viruses. He received pulse therapy with methyl-prednisolone with partial improvement. One week later, MRI showed improvement of cerebral lesions and enhancement of spinal roots. He received immunoglobulin. Control CSF had protein-cytology dissociation. Electromyography showed intense sensitive-motor polyneuropathy. Visual evoked potential was normal, and auditive was abnormal on right side. Hands strength and axial tonus improved. Three weeks after, immunoglobulin was repeated, with improvement of sphincter function, but still with paraplegia. He was discharged with motor physiotherapy and oral prednisolone.

Conclusion: The occurrence of these 2 entities, simultaneously or sequentially during a short period, is not well understood and is probably underdiagnosed.

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ANTI-NMDA RECEPTOR ENCEPHALITIS. FIRST CASE WITH CONFIRMED CIRCULATING ANTIBODIES IN ECUADOR

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INTRODUCTION: Anti-N-methyl-D-aspartate receptor (anti-NMDAR) encephalitis is a newly described form of encephalitis in children

associated with prominent psychiatric and neurological symptoms at onset. There are few cases described in Latin America with confirmed circulating anti-NMDAR antibodies. According to literature review this is the first case described in Ecuador.

METHODS: Review of medical history and complementary studies.

CASE DESCRIPTION: A nine-year-old-boy was admitted because of a head injury secondary to a transient loss of consciousness. Subsequently he developed progressive disorders of sleep/wake cycle, psychomotor excitability, choreoathetoid movements, orofacial dyskinesias, unintelligible speech, dysautonomic alterations and focal seizures. CT scan was unremarkable. DWI-MRI showed hyperintense bithalamic lesions. An EEG showed diffuse slowing with focal occipital spikes. Herpesvirus PCR and CSF cultures were negative. Paraneoplastic screening studies were also negative**.** A partial response to Methylprednisolone, Levetiracetam and Diazepam was seen. Nevertheless, concomitant Immunoglobulin treatment significantly improved symptoms with independent return to his daily activities without extrapyramidal movements and seizure-free. During steroids weaning a relapse featuring psychomotor hyperactivity, orofacial dyskinesias, alternating sensory and visual hallucinations had been reversed with. IV methylprednisolone. Currently he is on multidisciplinary follow up with almost total recovery of his daily activities. Circulating anti-NMDA receptor antibodies were confirmed in CSF samples.

CONCLUSIONS: Early recognition of the clinical manifestations of this condition will allow better recovery of acute symptoms, as well as a lower rate of sequelae. The identification of more cases in Latin America will also allow to define whether there are own characteristics of this disease in our population.

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A PEDIATRIC COHORT OF RECURRENT CENTRAL NERVOUS SYSTEM DEMYELINATION; EXPERIENCE OF A TERTIARY CENTRE FROM NORTH INDIA

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Introduction: Inflammatory demyelinating disease spectrum of the central nervous system comprises of Acute Disseminated Encephalomyelitis (ADEM), Multiple Sclerosis (MS), Clinically Isolated Syndrome (CIS) and Neuromyelitis Optica (NMO).Early diagnosis is imperative for optimal functional and neurological outcome.

Methods: Overall 35 cases presented between 2009 and 2013, with 11(31.4%) having recurrent episodes. These were classified according to the International MS study group and revised McDonald criteria (2010).

Results: Amongst the 11 patients with recurrent demyelination, majority were multiple sclerosis (8/11, 72.7%, 3 oligoclonal bands positive), followed by NMO (2/11, 18.2%) and multiphasic ADEM (1/11, 9%). The predominant clinical manifestations in MS were hemispheric syndrome (36.4%) followed by visual symptoms (22.7%) and encephalopathy(18.2%). One patient with NMO presented with recurrent episodes of encephalopathy with subclinical myelopathy without any visual involvement. In multiple sclerosis, lesions were predominantly juxtacortical and periventricular. One patient had diffuse bilateral involvement and another had tumefactive lesions. The acute attacks in MS and NMO were treated with steroids and azathioprine has been continued in all as the only disease modifying agent. Only one episode of relapse in one patient has been noted after starting azathioprine.

Conclusion: Atypical features like encephalopathy, tumefactive and diffuse bilateral lesions on neuroimaging have been infrequently reported in pediatric MS. Azathioprine, currently underutilized, could be a suitable alternative to interferon and glatiramer acetate, particularly in resource limited settings.

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VARIABLE MANIFESTATIONS OF ANTI-NMDA RECEPTOR ENCEPHALITIS IN CHILDREN: CASE SERIES FROM TERTIARY CARE CENTRE IN INDIA

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Introduction: Five cases of Anti-NMDAR encephalitis described with variable manifestations, and good response to immunotherapy.

Case 1: 4 years-old girl presented with fever, acute encephalopathy (A), seizures (B), and later dystonias and choreo-athetoid movements (C), perioral dyskinesias (D), urinary retention and episodes of alternating hypertension and hypotension, cardiac arrhythmias, recurrent cardiac asystole-all attributed to severe autonomic dysfunction. After failure of IV Methyl prednisolone (IVMP) and IVIg, she responded well to Rituximab.

Case 2: 5 years-old boy (Nigerian ethnicity); had symptoms A, B, C, and D as case 1. After failure of IVMP, he responded well to Rituximab given 2 months after onset.

Case 3: 5 months-old boy presented with symptoms A, B, C, and D as case 1, 3 weeks after febrile illness. He had suboptimal response to IVMP and plasmapheresis, but responded well to Rituximab.

Case 4: 8 years-old boy had A, B, C, and D as case 1, but also had severe catatonia and mutism. He had poor response to IVMP and IVIg (3rd week of illness), but excellent response to Rituximab.

Case 5: 8 year-old boy had acute-onset visual hallucinations and seizures. He improved spontaneously within 48 hours. Anti-NMDAR Ab positive in CSF but negative in serum.

Investigations: MRI-Brain was normal. CSF showed mildly elevated proteins; positive Anti NMDAR Ab in CSF and serum in all (except case 5-only CSF positive).

Follow up and conclusion: All had good recovery except case1 (partial recovery), and no relapse. High index of suspicion and early Rituximab therapy improves outcome.

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CLINICAL PROFILE AND OUTCOME OF CHILDREN WITH OPSOCLONUS MYOCLONUS ATAXIA SYNDROME (OMA)

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Introduction: Opsoclonus Myoclonus Ataxia syndrome (OMA) in children is an under recognised disorder and literature on its clinical profile and outcome are sparse.

Aim: To study the clinical profile and outcome of OMA in children.

Materials and methods: Retrospective chart review of children with OMA diagnosed from March 2006 to October 2013.

Results and discussion: Clinical data of 11 children with OMA were analyzed. Mean age at onset of symptom was 26 months (±7.3). Nine children (81.8%) had post infectious / idiopathic cause and 2 (18.2%) children had paraneoplastic etiology. Clinical symptoms at presentation were recurrent falls, refusal to stand, tremulousness, myoclonus, eye movement abnormalities and irritability. Mean delay in diagnosis was 6 months (varying from 1 to 20 months) and it was more for post infectious /idiopathic etiology when compared to paraneoplastic etiology. Younger age group, extreme irritability, early eye movement abnormalities and male sex predilection were found in paraneoplastic group. In paraneoplastic group, one had well differentiated neuroblastoma in suprarenal region and the other had poorly differentiated neuroblastoma in retroperitoneal area. Post infectious group sustained good remission with immunotherapy for prolonged periods of time. Paraneoplastic group did poorly with recurrent relapses and increased severity of symptoms in our series, although diagnosed early.

Conclusion: Paraneoplastic OMA had increased severity of symptoms and early eye movement abnormalities. Relapses were more common and severe in paraneoplastic group.

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THE CLINICAL NATURAL COURSE AFTER A FIRST EPISODE OF CENTRAL NERVE SYSTEMS DEMYELINATION IN CHILDREN

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The central nervous system inflammatory demyelinating disorders of childhood include both monophasic self-limited and chronic lifelong conditions, which can be indistinguishable at the time of initial presentation. In 2007, an International committee proposed diagnostic criteria for CNS demyelination in children. In this study, we review the recent consensus definitions for acquired inflammatory demyelinating diseases and analyze the clinical course and predictor of increasing risk of the second demyelinating event in Korean children. A retrospective review was performed about the medical records of the

pediatric patients who were diagnosed with acquired inflammatory demyelinating diseases at Samsung Medical Center, Seoul, Korea. We identified 40 patients (23 female, 17 male), including CIS 80%, ADEM 17.5% and NMO 2.5%. Average age of onset was 8.7 years. Nineteen patients (19/40, 47.5%) experienced the second event. Average interval between first and second event was 21 (0.8-69) months. Finally, 27.5% patients diagnosed with MS, during a mean follow up of 6.2 (2.4-12.9) years. The age, sex, initial clinical presentation, presence of abnormality in the initial brain MRI and CSF study findings did not influence upon the recurrence in CIS group. With the diagnosis of initial inflammatory demyelinating disorder, a thorough investigation of the other CNS areas to find out concomitant lesions and the serial follow up over at least 2 years are recommended for the early diagnosis and immediate treatment.

P392

SCREENING AUTOIMMUNE SYNAPTIC ANTIBODIES IN PEDIATRIC PATIENTS SUSPECTING AUTOIMMUNE ENCEPHALITIS

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Background: This study was aimed to identify and describe pediatric cases of autoimmune encephalitis confirmed by autoimmune synaptic antibodies tests.

Methods: From July 2011 to May 2013, acute encephalitis patients who were tested for NMDAR, LGI1, CASPR2, AMPA1/AMPA2, and GABA-BR were included. All patients who met all of the following criteria were enrolled; immunocompetent, acute onset encephalopathy, no proven etiology by initial screening.

Results: Among 20 enrolled cases, six patients (30%) were positive for NMDAR antibody and one patient (5%) was positive for CASPR2. The mean age of NMDAR Ab+ patients was 9.7 years (range, 2-16) and four of them (67%) were female. Two patients experienced prodromal illness such as fever, headache, or respiratory symptoms. All patients showed early onset of seizure, and some of them showed dyskinesia(5/6, 83%), psychosis(3/6, 50%), language disturbance(4/6, 67%) or autonomic instability(3/6, 50%) in the hospital course. Only one had a tumor, unilateral ovarian teratoma. All patients received steroid and intravenous immunoglobulin and 2 patients received adjunctive rituximab. Mean follow-up period was 10 months (range, 4-18 months), and none of the patients had significant sequalae. The patient with positive CASPR2 antibody was 8 years old girl, with similar clinical course and treatment outcome with NMDAR+ patients.

Conclusion: Anti-NMDA receptor encephalitis was the most common cause of autoimmune encephalitis. Multistage illness and favorable response to immunotherapy in all patients were in line with previous studies. We also identified the first pediatric case of anti-CASPR2 encephalitis, expanding the spectrum of pediatric autoimmune encephalitis

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POSTERIOR REVERSIBLE LEUKOENCEPHALOPATHY SYNDROME. REPORT OF TWO CASES.

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Introduction: Posterior reversible leuko-encephalopathy syndrome (PRLS), is a clinical-radiological condition characterized by headache, altered mental functioning, seizures and visual alterations, associated with cerebral oedema predominantly in the bilateral occipital white matter. Different reports have associated it with numerous processes and possibly with chemotherapy and immunosuppressive therapy.

Casereport: We report two cases with PRLS.

Case 1: Male sixteen years old, associated to chemotherapy treatment (methotrexate, cytarabine) who presented headache, spasticity, altered sensorium and visual disturbances. The MRI revealed lesions in the bilateral occipital white matter, hyperintense in T2 and FLAIR, and hypointense in T1.

Case 2: Male two years old, associated to immunosuppressive therapy (cyclosporine) who presented seizures, altered sensorium, visual disorders. The MRI showed lesions in the bilateral occipital-frontal white matter; and in the cortex, situated in bilateral fronto-temporo-parietal regions; hyperintense in T2 and FLAIR. The time between the start of treatment and the apparition of the neurological symptoms, was 2 days for the case 1 and one week for the case 2. A prompt diagnosis, an appropriate therapy (antiepileptic drugs, physical therapy and change of immunosuppressive therapy) were the keys for the complete clinical recovery.

Conclusions: The pathogenesis of SPRL is still not fully understood. Multiple clinical conditions can act as triggers. The onset of neurological symptoms in a patient with chemotherapy or immunosuppressive therapy should alert neurotoxicity and think of a PRLS.

P394

NON-PARANEOPLASTIC LIMBIC ENCEPHALITIS- CLINICAL COURSE IN 10-YEARS-OLD BOY

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Limbic Encephalitis is an inflammatory disease of limbic system, in which a triad of symptoms occurs: recent memory impairment, affective disorders and temporal lobe seizures. It may manifest as paraneoplastic syndrome (PLE), may be caused by autoimmune process or viral infection.

Most of documented cases of limbic encephalitis were reported in adults and most of available information concerns PLE. Data of the occurrence of this disease in children is less available, its course is diverse and prognosis difficult.

We are reporting a case of a 10-years old boy with progressive encephalopathy, classified as limbic encephalitis. He presented increasingly affective disorders, recent memory impairment and complex focal seizures. Those symptoms were followed by dysphagia and speech disturbances. Empiric therapy with Acyclovir and Ceftriaxone did not led to improvement. Screening to exclude neoplasm was carried out. After immunosupressive therapy (intravenous immunoglobulins and steroid therapy) noticeable improvement was observed. The patient was discharged in a good general condition, seizure-free, with circadian rhythm disorders, bradycardia and memory disorders. He required pedagogical care and further antiepileptic treatment.

We would like to show diagnostic difficulties of this disease as a result of limited knowledge about limbic encephalitis in children and also non-homogenous process. The described patient was initially being diagnosed for a month by a psychiatrists because no pathology in neurologic examination at that time was found and MRI findings have not shown any changes characteristic for LE.

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SPINAL CORD INVOLVEMENT IN CHILDREN AND ADOLESCENTS WITH MULTIPLE SCLEROSIS

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Introduction: The recognition of multiple sclerosis (MS) in children and adolescents increases recently, however, initial brain MRI may not be sufficient to establish the proper diagnosis in many children. According to McDonald's criteria, spinal cord lesions may be important for the diagnosis. The purpose of our study was to determine the prevalence of spinal cord lesions revealed by MRI in children and adolescents with clinically definite MS.

Material: and methods: Initial spinal cord MRI in 57 children and adolescents (37 girls and 20 boys, aged from 7 to 18 years, mean 14.1 years) with clinically definite MS were reviewed. All patients had typical MS lesions revealed on brain MRI and none met the diagnostic criteria for neuromyelitis optica (NMO). Total lesion count, lesion location, and gadolinium enhancement, as well as clinical symptoms were analyzed.

Results: Spinal cord lesions were identified in 35 (61.4%) patients. Contrast enhancement was present in 37.1% of them. In 36 patients, the clinical symptoms of spinal cord involvement were revealed by neurological examinations. There was no correlation between clinical symptoms of spinal cord involvement and MRI.

Conclusions: The prevalence of spinal cord lesions in children and adolescents with MS is high. Therefore, spinal cord MRI with contrast should be performed in every child suspected as having MS, irrespective of clinical presentation.

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THE CLINICAL AND PREDICTIVE FACTORS FOR RELAPSE AFTER AN INITIAL EVENT OF ACUTE DISSEMINATED ENCEPHALOMYELITIS IN CHILDREN.

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Introduction: Acute disseminatedencephalomyelitis (ADEM) is an immune-mediated inflammatory and demyelinating condition of the central nervous system (CNS) that occurs mainly in the pediatric population. The diagnostic criteria for the disorder include multifocal clinical and neuroimaging findings associated with encephalopathy. The disorder usually has a monophasic course, but multiphasic and recurrent cases have been reported in the literature, following the International Pediatric Multiple Sclerosis Study Group Consensus definitions published in 2007. In this study, we aimed to recognize clinical, neuroradiological, and cerebrospinal fluid findings present at the initial event of ADEM, which may represent possible predictive factors of relapses that may predispose these children to develop recurrent or multiphasic ADEM forms.

Methods: A retrospective observational study was performed which was based on the definitions proposedby the International Pediatric Multiple Sclerosis Study Group in 2007. Thirteen subjects were included in the study.

Results: Seven children had a mild encephalopathy at the first event. Eleven patients had supratentorial and infratentorial lesions, and in six of the patients, the lesions were greater than 1 cm. In our study, seven of the thirteen patients with ADEM went on to relapse. Six of these patients developed multiphasic ADEM, and one patient developed the recurrent form of ADEM.

Discussion: In the study sample, no clear predictive factors forADEM relapse were found among the clinical, neuroradiological and cerebrospinal fluid findings. Therefore, more studies are needed to characterize the possible underlying risk factors that may predispose patients with ADEM to develop relapses of the condition.

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ACUTE NECROTIZING ENCEPHALOPATHY OF CHILDHOOD IN A NON-ASIAN 4-YEAR-OLD CHILD AFTER VARICELLA IMMUNIZATION

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Acute Necrotizing Encephalopathy of Childhood (ANE) is a rare form of acute parainfectious encephalopathy occurring most commonly in Eastern countries. ANE has a variable clinical spectrum with a homogenous biochemical, neuroimaging and neuropathological profile among the cases described in the literature. This entity has a variable prognosis with a mortality rate of up to 30% of cases. We describe the case of a four-year-old boy who was diagnosed with ANE several days after receiving a varicella immunization and following the development of a nonspecific viral febrile illness associated with acute gastroenteritis, seizure episodes and rapid deterioration in the level of consciousness. Laboratory analysis was remarkable for an acute marked elevation of serum aminotransferases without associated hyperammonemia and CSF analysis did not revealed pleocytosis. There was evidence of bithalamic-restricted diffusion on brain MRI as well as additional bilateral lesions on cerebellar hemispheres and vermis, which are part of the radiologic diagnostic criteria for this condition. Clinical, laboratory and radiological findings in this case pointed to the diagnosis of ANE and served to differentiate it from other similar syndromes that must be considered in the differential diagnosis. In addition to the marked improvement from initial neurological deficits and surviving despite a reported 30% mortality, the significance of this case relies on the fact that it represents an atypical disease in Western countries and the report of an association with varicella immunization.

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AUTOIMMUNE ANTI-N-METHYL-D-ASPARTATE RECEPTOR(ANTINMDAR) ENCEPHALITIS: 2 CASE REPORTS FROM SAUDI ARABIA

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Background: Anti-NMDAR encephalitis is an autoimmune disorder characterized by memory deficit, decreased level of consciousness, autonomic dysfunction and movement disorders. It occurs commonly in children and young adolescent .We report 2 cases with anti-NMDAR encephalitis presented to the pediatric emergency department at King Khalid University Hospital (KKUH).To our knowledge this is the first report from Saudi Arabia.

Case (1): A 4-year-old girl presented with sudden onset of seizure and behavioral changes followed by deterioration in her mental status and abnormal movements. Her Clinical Examination revealed normal vital signs, poor social interaction, Oro-facial dyskinesia, chorioathetosis and dystonia. Brain MRI was unremarkable. Electroencephalography (EEG) showed delta slowing. Cerebrospinal fluid (CSF) analysis was negative for infection. Anti-NMDAR antibodies was positive in both serum and CSF. Screening for underlying malignancy was negative. Marked improvement was noted after treatment with corticosteroid, intravenous immunoglobulin (IVIg) and rituximab.

Case (2): A 6- year-old girl presented with five days history of behavioural changes and insomnia associated with fluctuation in the level of consciousness and involuntary movements .Her clinical examination revealed minimally interactive, autonomic manifestations, oro-facial dyskinesia and choreoathetosis. MRI brain was unremarkable, EEG showed diffuse delta slowing .CSF analysis was negative for infection. Anti-NMDAR antibodies were positive in both serum and CSF. Screening for underlying malignancy was negative. She was given Ivy, corticosteroid and rituximab and showed marked improvement.

Conclusion: Anti-NMDAR encephalitis is a new, under-recognized disorder. Early recognition and high index of suspicion is very important in initiation of different modality of treatments which lead to favorable outcome.

P399

NEUROPSYCHOLOGICAL OUTCOMES OF ANTI-NMDA RECEPTOR ENCEPHALITIS IN CHILDREN: A CASE SERIES

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Introduction: Anti- N-methyl-D-aspartate (NMDA) receptor encephalitis is an autoimmune encephalopathy that is being increasingly diagnosed in children and adolescents with seizures, altered mental status and movement disorders. We report the clinical features and neuropsychological outcomes of three children who were diagnosed with anti-NMDA receptor encephalitis.

Case Description: We describe three children, 2 male and 1 female, with anti-NMDA receptor encephalitis. All three were adolescents, who presented with acute behavioural change, seizures and movement disorders. All cases were seropositive for NMDA receptor antibodies and were not associated with tumours. At presentation, the patients had poor executive function, inappropriate behaviour with disinhibition and impulsivity, as well as memory impairment. Based on serial neuropsychological assessments, these impairments were noted to improve with immunosuppressive therapy. The range of follow-up after diagnosis was 4 months to 4 years. The female patient was followed up for 4 years and although she showed initial clinical improvement, had a relapse after completing therapy which was characterised by acute onset of cognitive decline and behavioural change.

Conclusion: We report the neuropsychological and clinical outcomes of children with anti-NMDA receptor encephalitis. The cognitive deficits associated with this condition reduce in severity over time with immunosuppressive therapy, but executive impairment persists. Children with autoimmune encephalopathies such as anti-NMDA receptor encephalitis should undergo serial neuropsychological assessments to monitor response to therapy and relapse.

P400

ANTI-N-METHYL-D-ASPARTATE RECEPTOR ENCEPHALITIS IN TAIWAN

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Introduction: Since the antibodies against the N-methyl-D-aspartate receptor discovery in 2007, anti-N-methyl-D-aspartate receptor encephalitis has entered the mainstream of neurology and other disciplines worldwide. We aimed to reportthe clinical features of patients with this disorder in Taiwan.

Methods: Information was obtained by the authors or referring physicians and those published in English literature about anti-N-methyl-D-aspartate receptor encephalitis in Taiwan from 2007 to 2013. The clinical features were analyzed.

Results: 12 patients (10 female) aged between 7 years and 28 years with anti-N-methyl-D-aspartate receptor encephalitis were identified. 6 patients (50%) were less than 18 yearsold and 1 of them was male. 3 of them had an underlying tumor. 91.6% presented with mood, behavior, or personality change. 91.6% developed seizures, 100% stereotyped movements, 83.3% autonomic instability, and 66.7% hypoventilation. Responses to immunotherapy were slow and variable. Overall, 63.6% patients had substantial recovery after immunotherapy or tumor removal. Neurological relapses occurred in 9.1%.

Conclusions: Anti-N-methyl-D-aspartate receptor encephalitis is increasingly recognized in Taiwan. It is characterized with characteristic clinical features, predominantly affects females with/without an ovarian tumor and apotentially treatable disorders. Early recognition and early intervention ofthe disease can improve the outcome. It is important for neurologist to befamiliar with the clinical presentations of the disease in children and young adults.

P401

HASHIMOTO ENCEPHALOPATHY PRESENTING AS SEIZURE AND ACUTE PSYCHIATRIC FINDINGS

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Hashimoto encephalopathy (or Steroid Responsive Encephalopathy with Autoimmune Thyroiditis) Steroid-responsive encephalopathy with autoimmune thyroiditis is a relatively uncommon entity in the pediatric population. Seizures of unknown etiology are a common presenting sign in the pediatric intensive care unit, and steroid-responsive encephalopathy with autoimmune thyroiditis should be considered as a possible cause of recurrent seizures. The clinical presenting with variable symptoms ranging from behavioral and cognitive changes, myoclonus, seizures, pyramidal tract dysfunction, involuntary movements, and cerebellar signs to psychosis and coma, with relapsing and progressive course. We report an 14 year old boy with generalized seizures and behavioural changes. She was found to have sub-clinical hypothyroidism and elevated anti-thyroid peroxidase antibodies.

P402

LONGITUDINALLY EXTENSIVE TRANSVERSE MYELITIS IN A-17-MONTH OLD GİRL PATİENT WİTH RECURRENT HYPOTONİA ATTACKS TREATED WITH INTRAVENOUS IMMUNGLOBULIN

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Introduction: Longitudinally extensive transverse myelitis (LETM) is defined as involving 3 or more consecutive vertebral levels. In clinical practice, the first-line treatment of noninfectious immune-mediated transverse myelitis is intravenous pulse methylprednisolone. We report a seventeen–month girl patient, who had recurrent hypotonia attacks and panmyelitis in her spinal magnetic resonance images (MRI) treated with intravenous immunoglobulin and she had clinical improvement without any change in MRI findings.

Case: A Seventeen-month-old girl presented at the pediatric neurology clinic with generalized hypotonia and weakness. Her spinal magnetic resonance imaging showed diffuse demyelination in whole spinal cord and additionally T2 hyperintensity was present in the medulla oblongata. Serum Aquaporin 4 antibody was negative. Although the radiological lesions did not recovered, clinical improvement became obvious one week after admission with increased control of head and body. After 2 weeks the patient was able to sit, stand and to take a few steps. Fifteen days later from discharge she presented again with weakness. She had three flaccid weakness attacks on follow-up and treated with monthly IVIg.

Discussion: Although the accepted first line treatment for transverse myelitis is steroids, other treatment modalities include IVIg and plasmapheresis should be kept on mind especially in the infectious etiologies. We observed clinical improvement after first IVIg administration within a week in our case. And as well, recurrent weakness attacks were relieved with IVIg. As far as we know this is the youngest patient, who had whole spinal cord and medulla oblongata involvement and recurrent hypotonia attacks.

P403

HEMICONVULSION-HEMIPLEGIA-EPILEPSY (HHE) SYNDROME PRESENTING WITH NO APPARENT HEMICONVULSION: A CASE REPORT.

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Introduction: Hemiconvulsion-Hemiplegia (HH) has been described in patients following prolonged focal, often febrile convulsions and has heterogeneous aetiologies, ranging from structural brain disorders across to immune-mediated causes. Here we describe a child with no apparent convulsion prior to the onset of a dense hemiplegia with imaging changes characteristic of HHE, whose condition improved dramatically following immunotherapy.

Case history: A 7 year old boy with a recent febrile prodrome, presented with a dense right hemiplegia and progressive encephalopathy after a lucid 24 hour interval following a short spontaneously recovering event. He has a well controlled seizure disorder (diagnosed age 2) and his treatment had recently been stopped after 3 years of seizure freedom. His repeat neuroimaging 24 hours later following right sided focal seizures revealed left hemispheric oedema and left parieto-occipital restricted diffusivity. His electroencephalogram showed only attenuated left hemispheric cerebral rhythms. HH syndrome was considered despite the lack of hemiconvulsion due to the striking radiological features. Intravenous corticosteroids and immunoglobulins was commenced as an immune aetiology has been proposed. He made a full motor recovery after 2 weeks and remains seizure free at 3 months follow-up, although gliotic changes in the temporal and occipital region are revealed on follow-up imaging. Metabolic, immune and genetic (CACNA1A) investigations undertaken have not revealed an aetiology to date.

Conclusion: HHE should be considered even in the absence of hemiconvulsion and the inflammatory response either primary or secondary may respond to early treatment.

P404

ELECTROPHYSIOLOGICAL FEATURES OF CHILDHOOD AUTOIMMUNE ENCEPHALITIS

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Introduction: Early electroencephalogram (EEG) revealing extreme delta-brush,[1] and other focal and unilateral electro-clinical features have been reported in N-Methyl-D-Aspartate receptor (NMDAR) antibody encephalitis. We evaluated the electrophysiological features of children with a range of autoimmune encephalitides, to identify similar and other features.

Methods: 34 consecutive patients (identified from cohort of 40) with presumed autoimmune encephalitis as previously defined with acute and/or follow-up EEG recordings available for blinded review by a paediatric neurophysiologist, were studied.

Results: 6 NMDAR antibody positive patients with focal or multifocal seizures had EEG demonstrating either diffuse slowing with spike wave discharges (n=1), unilateral hemispheric slowing (n=3) or focal slowing (n=2). 4/6 voltage-gated potassium channel (VGKC) antibody positive patients had focal temporal slowing with 3 having additional diffuse changes, of which 2 had ictal activity. In the remaining antibody positive group (n=4; glycine receptor, thyroid, basal ganglia, ganglioside); and negative group (n=18), EEG features ranged from normal (n=4), focal slowing (n=6), diffuse slowing (n=11) and PLEDS (n=1), with ictal activity observed in 7. Overall, the antibody positive group were more likely to have persistingly abnormal EEG (7/12 vs 7/22); and these changes were also abnormal for longer (mean 12 vs 6 months).

Conclusion: Previously reported EEG features of NMDAR antibody encephalitis were identified in our patients. Other than the unilateral hemispheric slowing, the other features identified in patients with autoimmune encephalitis were shared across the antibody positive and negative group.

P405
ANTI-MOG ANTIBODIES IN PAEDIATRIC DEMYELINATING DISEASE

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Objectives: Anti-myelin oligodendrocyte glycoprotein (anti-MOG) antibodies have been identified in paediatric acute demyelinating encephalomyelitis (ADEM). Their relevance remains uncertain. We aimed to define the features of cases with anti-MOG antibody in our paediatric population.

Materials and Methods: Case series of paediatric patients with monophasic or recurrent demyelinating episodes who demonstrated anti-MOG antibodies in serum.

Results: We describe five children with serum anti-MOG antibodies; three females, two male. All cases were serum aquaporin-4 antibody negative. Mean follow-up was 9.8 months (1-30). Mean age at onset was 6 years (5-7). Every case presented with ADEM, monophasic in four cases. In one, unilateral optic neuritis (ON) developed 8 weeks later and ON subsequently recurred after three months. Neuroimaging was typical for ADEM in all cases, although initial scans were normal in three cases. Two cases had spinal cord lesions. All had cerebrospinal fluid (CSF) pleocytosis, with marked lymphocytosis in two cases, and one had markedly elevated protein. Each case received intravenous steroids at diagnosis, followed by reducing doses of oral steroid.

Conclusions: In this case series, anti-MOG antibody positivity was associated with monophasic ADEM in all but one case, in which there was also recurrent optic neuritis. Each child made a full recovery, suggesting that presence of the antibody does not necessarily confer a poor short-term prognosis. Although anti-MOG antibody is a useful marker for autoimmune aetiology, it is yet to be seen if this predicts recurrence or risk of multiple sclerosis in the longer term.

P406
KINSBOURNE SYNDROME. SHOULD WE MODIFY OUR THERAPEUTIC BEHAVIOR?

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Introduction: Opsoclonus-myoclonus syndrome or Kinsbourne syndrome is a rare immunomediated disorder characterized by opsolconus, myoclonus, ataxia, irritability and sleep disorders. It is associated with neuroblastoma in up to 45% of cases. Neurological sequelae occur in more than 80% of cases. Treatment is based on immunosuppressive and immunomodulatory therapies, and surgery resection in neuroblastoma cases. For many years corticosteroids have been the gold standard. Afterwards, intravenous immunoglobulin was associated. Due to high prevalence of corticosteroid dependency, adverse events and poor outcome, multiple drug protocols have been proposed.

Method: We reviewed clinical records of patients admitted with Kinsbourne syndrome between 2001 and 2013 at Pereira Rossell Children`s Hospital, Montevideo, Uruguay. We analyze clinical features and treatments. Neurological outcome was evaluated in the five children whose follow-up was greater than 2 years.

Results: 8 patients were diagnosed in this period, with a mean age of 21 months. Table shows data obtained. All patients were treated with corticosteroids with initial improvement. Six children had cortical dependency, the other two had less than one year of follow-up. In three children with partial response to intravenous immunoglobulin and corticosteroids, or with cortical dependency, Rituximab was performed with clinical improvement. The five children with more than 2 years of follow-up had neurological sequelae including speech delay, cognitive deficits, motor delay, persistent ataxia and behavioral disturbance.

Discussion: In our experience Kinsbourne syndrome has a poor clinical outcome so we support multiple drugs treatment, including corticosteroids, intravenous immunoglobulin and rituximab.

P407
KLEINE-LEVIN SYNDROME WITH RAPID CYCLING – CASE REPORT AND REVIEW OF THE LITERATURE

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OBJECTIVE: Describe a patient with Kleine-Levin Syndrome (KLS) with rapid cycling of hypersomnia episodes and autonomic changes.

BACKGROUND: KLS is a rare disorder (1.5 cases/million), typically in adolescent males and preceded by a triggering event, such as infections or head trauma It exists in the borderland between psychiatry and neurology, often misdiagnosed as depression, bipolar disorder, psychosis or seizures.

DESIGN/METHODS: A 14 yo boy with a previous history of absence epilepsy, off medications for 1 year, presented with hypersomnia after an upper airway infection. He slept over 15h/day, when awake exhibiting derealization and speaking in a regressed fashion. Family reported dishinhibition, bradyphrenia and alternating hyper/hypophagia. Symptoms improved without treatment but he relapsed every 2-3 weeks, with cycles lasting 10-14 days. Investigation included two lumbar punctures, with non-inflammatory results and negative bacterial/viral studies. Long-term EEG and imaging were unrevealing. Toxicology, thyroid function, B12, Folate, Thiamine, serological, paraneoplastic and vasculitic/autoimmune panel were negative. Psychiatric evaluation did not elicit diagnostic criteria for mood, somatic or other mental health conditions.

RESULTS: Repeat EEG showed intermittent delta slowing. Cardiac monitoring displayed alternating bradycardia/tachycardia. HLA typing was positive for DQB1*0201, associated with KLS and suggesting autoimmune etiology. He was started on modafinil with improved hypersomnia; preventive treatment with valproic acid and lithium was unsuccessful for the rapid cycling.

CONCLUSIONS: KLS has significant morbidity if unrecognized. We review and discuss updated diagnostic criteria, pathophysiology and management. This case exemplifies an unusual rapid cycling pattern of hypersomnia episodes associated with classic KLS

P408
EVALUATION OF NEUROINFLAMMATION IN PEDIATRIC MULTIPLE SCLEROSIS PATIENTS

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Introduction: Both gray and white matter pathology, including neuroinflammation, have been incriminated in the pathogenesis of multiple sclerosis (MS), though not-much is known about brain inflammatory changes in pediatric MS population. Increased expression of translocator protein (TSPO) imaged by positron emission tomography (PET) can detect neuroinflammation, mediated by activated microglia, in normal-appearing white matter and cortex in adults with MS. Therefore, we applied PET imaging using the TSPO radiotracer (11) C-[R]-PK11195 (PK PET) in children with MS.

Methods: PK PET was performed in 8 children (age: 10-18 years) with MS associated with typical cerebral MRI findings. The PET images were evaluated by calculating the regional binding potential, based on a simplified reference region model, and the values were compared to a pediatric PET database (Kumar et al., J Neuroinflammation, 2012) to identify brain regions with neuroinflammation.

Results: All but one child showed regions with increased binding on PET, affecting individually variable regions including thalamus (n=4), whose values were also increased bilaterally on the group level;

increases were also seen in centrum semiovale (n=2), lentiform nucleus (n=2), frontal (n=3) and parietal cortex (n=2), brainstem (n=2), and cingulate (n=1); some of which were not detected on MRI. Focal areas with increased binding were more restricted than MRI abnormalities, likely representing PK uptake in active lesions only.

Conclusions: PET imaging of activated microglia can detect areas of neuroinflammation in both gray and white matter in children with MS, and may identify regions with active disease thus complementing clinical MRI.