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Clinical trials in children with Down Syndrome : now and future

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Background: Down syndrome (DS) is the most common genetic cause of intellectual disability.Although progress in managing co-morbidities has improved life expectancy, no therapeutic options have showed to significantly improve intellectual deficiencies.The current focus of the pharmacological treatment of DS is on the improvement of the cognitive impairment that is probably due to neurodevelopmental alterations,neurotransmitter alterations and neurodegeneration,and is also targeted to the overexpression of selected genes on HSA21. **Methods:** We review the clinical trials of the last 5 years focusing on the cognitive improvement of children with DS. **Results:** We report the results of therapeutic trials concerning selective negative allosteric regulators of the GABAAa5 receptor, NMDA antagonists, Kinase inhibitors of DYRK1A, folinic acid and thyroid hormone supplementation, activators of serotonergic and cholinergic system. **Conclusions:** The incomplete understanding of individual phenotypic variability, natural history, lack of biomarkers ,no adapted neuropsychological tests, placebo effect, epigenetic effect have limited our capacity to succeed, even when promising drugs are tested.We need new tools and models will allow a better understanding of the pathophysiology. We also need to create more sensitive and realistic outcome mesures to quantify disease and therapeutic efficacy. The association of different therapeutic agents (epidrugs included) with cognitive stimulation could be a future strategy.

P.079

4H leukodystrophy: a case series of siblings with an unusually mild phenotype

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Background: 4H leukodystrophy is a genetic disorder typically characterized by hypomyelination, hypodontia and hypogonatotropic hypogonadism. Previously reported patients had considerable cognitive and motor deficits. We present a pair of siblings with a less severe phenotype. **Methods:** Patient data was obtained from medical records from the Children's Hospital of Eastern Ontario. **Results:** The first patient was diagnosed with 4H leukodystrophy at the age of 21 years after genetic testing revealed a POLR3B mutation with a homozygous V523E variant. She has hypomyelination on MRI and a history of optic neuritis, as well as intermittent sensory and motor symptoms in the context of a diagnosis of multiple sclerosis. She has no clinical manifestations of 4H leukodystrophy. The patient is now 26 years old and has only mild neurological deficits. Her younger brother was diagnosed with 4H leukodystrophy at the age of 18 years and found to have the same genetic mutation as his sister. He has a history of seizures and mild learning disabilities. He is now 23 years old with no typical symptoms of 4H leukodystrophy. **Conclusions:** 4H leukodystrophy is usually associated with a severe, disabling phenotype and a poor prognosis. Our patients illustrate that a much milder phenotype exists.

P.082

Cannabinoids in the treatment of behavioural symptoms of autism: a rapid review to guide practice

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Background: Legalization of recreational cannabis in Canada has increased the presence of cannabis in the public mind. There are online parent advocacy groups which are already calling for the use of cannabinoids in pediatric developmental and behavioural conditions such as Autism Spectrum Disorder (ASD). We set out to perform a rapid review of existing literature regarding use of cannabinoid products in the treatment of the behavioural domains of ASD. **Methods:** Key search terms were identified in collaboration with a medical librarian and combined into standardized search filters. A total of 55 articles were identified, of which only two included primary data regarding the use of cannabinoids to control behavioural symptoms of ASD in pediatric populations. **Results:** Both studies found significant reductions in the behavioural measures examined - which included inappropriate speech, irritability, stereotyped behaviours and self injurious behaviours - after treatment with Cannabinoids. **Conclusions:** The minimal existing evidence indicates the use of cannabinoid products may be useful in improving behavioural difficulties in children with ASD. However, there is a complete lack of well powered, rigorous studies. Further studies with larger cohorts are needed before any recommendations can be confidently made for or against the use of cannabinoids in this population.

Clinical Neurophysiology (CSCN)
Epilepsy and EEG

P.080

Effects of REM sleep in anti-NMDA receptor encephalitis with extreme delta brush pattern

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Background: Seizures are rare in REM sleep. To our knowledge, the effects of different sleep stages in anti-NMDA encephalitis have not been studied. **Methods:** Case report. **Results:** 32 year-old healthy female presented with acute fluctuating level of consciousness with episodic impulsivity, disorientation, and emotional outbursts following 3 days of headache. Her temperature was 37.8^{o}C and she was started on ceftriaxone, vancomycin, and acyclovir. CSF only showed a total nuclear cell count of 182 and pleocytosis. MRI revealed increased T2 hyperintense right lateral temporal and parietal cortical thickening. EEG revealed right frontotemporal seizures with left sided rhythmic jerking, and an extreme delta brush pattern. Interestingly, seizures and extreme delta brush disappeared in REM sleep. After HSV PCR was negative, she was immunosuppressed with corticosteroids, IVIG, rituximab, and cyclophosphamide, given the extreme delta brush pattern. Repeat CSF analysis eventually yielded highly positive anti-NMDA antibodies. Ten weeks later, she became seizure free. **Conclusions:** For the first time, we report REM sleep

in a case of anti-NMDA encephalitis during which there was absence of extreme delta brush and ictal EEG patterns. This observation may provide insight into the ongoing debate over whether extreme delta brush is an ictal EEG pattern.

Neuromuscular Disease and EMG

P.081

Critical illness neuropathy and clinical correlates in severely burned patients

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Background: Reported incidence of critical illness neuropathy (CIN) in burn patients is between 7-41%. **Methods:** Retrospective review including patients admitted to quaternary ICU for burn injuries between 2010-16. Variables include demographics, burn and neurologic characteristics, EMG reports, and measurements of illness severity. Patients with and without neuropathies were compared. **Results:** Of 147 patients admitted to ICU, thirteen had EMG studies and eight met CIN criteria. Five had electrophysiological CIN evidence, three had clinical diagnosis. Six EMGs focused on upper limb injuries only, insufficient to diagnose CIN. One patient was diagnosed with critical illness myopathy and nine had superimposed focal mononeuropathies or plexopathy. CIN patients had a mean of larger burns (TBSA 63% vs 21%), more operations (8 vs 2) and escharotomies performed (63% vs 12%), longer ICU admissions (23 vs 9 days), longer ventilation (28 vs 8 days), higher revised Baux score (101 vs 76) and initial APACHE II scores (21 vs 15) than those without. **Conclusions:** CIN was identified in 5.4% of burn patients admitted to ICU, lower than previously reported in literature, and associated with higher illness severity. CIN may be under recognized if not screened for. Unit examinations should include screening neurological measures and indicated EMGs to evaluate for CIN.

Neuroradiology (CSNR)

Neuroimaging

P.083

Radiology reporting of low-grade glioma growth underestimates tumor expansion

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Background: Surveillance by serial magnetic resonance imaging (MRI) is important in the management of diffuse low-grade gliomas (LGGs). Radiological interpretations of LGG scans, however, are typically qualitative and difficult to use clinically. **Methods:** We retrospectively compared radiological interpretations of LGG growth/stability to volume change measured by manual segmentation. Tumour diameter was measured to evaluate methods for assessing glioma progression, including RECIST criteria, Macdonald/RANO criteria, and mean tumour diameter/ellipsoid method. **Results:** Tumours evaluated as stable by radiologists grew a median 5.1 mL (11.1%) relative to the comparison scan. Those evaluated as having grown increased by 13.3 mL (23.7%). Diameter-based measurements corresponded well but tended to overestimate segmented volumes, and overestimation error increased with tumour size. Agreement with segmented volume improved from a mean difference of 17.6 to 4.5 to 3.9 mm for diameter and from 104.0 to 25.3 to 15.9 mL for volume with measurements in one, two, and three dimensions. **Conclusions:** Given evidence that LGG volume and growth are prognostic factors, lesions should be accurately measured. Current radiological reporting workflows fail to appreciate and communicate the true expansion of LGGs. Volumetric analysis remains the gold standard for growth assessment, but diametric measurements in three dimensions may be an acceptable alternative.

Neurovascular, Stroke and Neurointerventional

P.085

Perioperative endovascular procedure utilization in transsphenoidal surgery patients at two tertiary-care academic centres

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Background: Utilization of endovascular procedures in the perioperative management of patients undergoing transsphenoidal surgery is uncommon but plays a critical role in preventing and treating potentially life-threatening vascular complications. **Methods:** We performed a retrospective review of all patients over a 10-year period who underwent transsphenoidal surgery at two tertiary-care institutions and identified all pre-operative and post-operative endovascular procedures performed. **Results:** 18 perioperative endovascular procedures were performed including 9 pre- and 9 post-operative. Pre-operative procedures included balloon-test occlusion (n=4), aneurysm coiling (n=4), and parent artery occlusion (n=1). One aneurysm coiling was complicated by coil migration requiring coil retrieval with a snare device and one balloon-test occlusion was complicated by pituitary apoplexy. Pituitary apoplexy following balloon-test occlusion has not been reported and the potential pathophysiology is reviewed. Post-operative procedures included embolization for epistaxis (n=2) and embolization with or without parent artery sacrifice for carotid and anterior cerebral artery vascular injury (n=7). Arterial vascular injury was managed with coil embolization and/or with detachable balloons. Review of anatomical features predisposing to vascular injury are discussed. **Conclusions:** Patients undergoing transsphenoidal surgery should be managed with a multidisciplinary team ensuring that endovascular treatment options are made available during the perioperative period.