P.121

Leading the Way to the Future: Implementing Novel Therapeutics for Rare Pediatric Neurological Disorders

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Background: Children and Adolescents with rare neurogenetic disorders often have no known cure or disease modifying treatments. Recent advancements in treatments are offering much needed hope to these patients and families. However, these treatments are extremely costly, have complex administration requirements and have many unknown long-term risks and outcomes. **Methods:** In this presentation, we will discuss our experiences with the implementation process, including developing intricate care pathways, collaborating with multiple disciplines and services, supporting and advocating for our patients and families, and interacting with government agencies and pharmaceutical companies. Case studies will highlight the positive impact these treatments are making on the lives of children and adolescents with rare neurological disorders. **Results:** Spinal muscular atrophy and Neuronal Ceroid Lipofuscinosis Type 2 are both rare and devastating neurodegenerative conditions with significant morbidity and mortality. Health Canada and government funding agencies recently approved Nusinersen, Onasemnogene abeparvovec for the treatment of SMA and Cerliponase alfa for the treatment of CLN2, leading us to swiftly integrate these treatments into our standard of care. **Conclusions:** While implementing these novel therapies into clinical practice can be both challenging and rewarding, neuroscience nurses are positioned at the forefront to be leaders in this process at both organizational, national, and international levels.

P.122

The Pediatric Neuroirritability Management Protocol at the Stollery Children's Hospital – Inspired by an Irritable Infant with GM3 Synthase Deficiency

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Background: We describe an infant with a diagnosis of GM3 synthase deficiency, presenting with severe neuroirritability from birth. He required multiple admissions due to extreme agitation and caregiver burnout. Multiple pharmacological agents were tried, and the effect of each medication was modest and short-lasting at best. The literature on the management of neuroirritability in children with progressive genetic and metabolic conditions is sparse, and a neuroirritability management protocol has yet to be developed at our institution. **Methods:** We searched for relevant primary research and articles on PubMed. We reviewed the evidence of each pharmacological agent and added non-pharmacological strategies. We developed management guidelines for neuroirritability at our hospital. This protocol was reviewed by several pediatric neurologists and pediatric palliative care specialists at the Stollery and SickKids Hospitals. **Results:** We present the Pediatric Neuroirritability Management Protocol for the Stollery Children's Hospital. **Conclusions:** Further study is required to assess whether this protocol can be adapted to treat irritability in the context of other neurological conditions such as hypoxic-ischemic encephalopathy and non-accidental injury. In addition, we will expand our guidelines to include other symptoms such as spasticity, dystonia, and autonomic dysfunction.

P.123

Severe DNM1 Encephalopathy with Dysmyelination due to Recurrent Splice Site Pathogenic Variant

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Background: Patients with DNM1-encephalopathy almost exclusively have missense variants, mostly in the GTPase domain of DNM1. Delayed myelination has been reported in at least three patients with DNM1-encephalopathy, all with missense mutations in the DNM1 central domain. Only one *DNM1* splice-site variant has previously been reported, and the authors questioned whether the variant accounted for all aspects of the patient's phenotype. **Methods:** Case-Report. **Results:** Our patient had hypotonia and brief multifocal tonic seizures from age-1-month. He still has profound global developmental delay, daily seizures and microcephaly. MRI-Brain at age-21-months showed T2 hyperintensity in the bilateral periventricular and subcortical white matter; spectroscopy showed a questionable lactate peak and an elevated choline peak relative to N-acetylaspartate. Clinical gene-panel identified a heterozygous de novo pathogenic variant in intron 9 of DNM1 (c.1197-8G > A; IVS9-8G>A). In-silico tools categorized this variant as deleterious secondary to a splicing defect. RT-PCR analysis on peripheral blood was unsuccessful as DNM1 expression is extremely low outside of the brain. **Conclusions:** Our patient carried the same DNM1 variant previously reported, indicating this is a recurrent pathogenic splice-site variant. The spectroscopic abnormalities suggest a possible element of demyelination in *DNM1* variants of the central domain, though the mechanism remains unclear.

P.124

Baseline Assessment of Attention and Executive Function Deficits in Children with Neurodevelopmental Disorders: Data from a Speciality Attention Clinic

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Background: Attention and executive function (EF) deficits in children negatively impact academics, social interactions, and overall quality of life. Children with other brain-based disorders are at high risk for attention and EF concerns, but the effects of these impairments are not well studied in the literature. The Complex Attention and Executive Function Clinic at the Alberta

Children's Hospital collected baseline data on patients referred for concerns of attention deficits co-occurring with diagnosed neurologic illness/injury, or neurodevelopmental disorder (NDD). **Methods:** The Behaviour Rating Inventory of Executive Function (BRIEF-2), Behaviour Assessment System for Children (BASC-3), Parenting Stress Index (PSI-4) and medical and past treatment information were collected on initial clinic visit for patients aged 5-15 years. **Results:** BRIEF-2 Global Executive Composite demonstrated 88.9% of children had clinically elevated scores. Clinically significant scores were observed in 55.5% for BASC-3 Adaptive Skills index and 40% of parents in PSI-4 Total Stress scores. **Conclusions:** Children with neurologic illness/NDDs are at high risk of clinical impairments in attention and EF. In children referred for attention and behavioural regulation, there is clinically significant increased reporting of executive function impairment out of proportion to other behavioural difficulties. The clinic aims to improve overall functioning through treatment of unmanaged attention and EF deficits.

Clinical Neurophysiology (CSCN)

Epilepsy and EEG

P.125

Discriminating sharp-wave ripples and interictal epileptiform discharges in patients with mesial temporal epilepsy using intracranial EEG recordings

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Background: Interictal epileptiform discharges (IEDs) are known as epilepsy biomarkers for seizure detection, and It is essential for clinicians to detect them from from physiological events with similar temporal frequency characteristics. **Methods:** We analyzed the SEEG recordings obtained from patients with medically-resistant epilepsy (MRE) implanted with DE at the Western University Hospital Epilepsy Unit. The data were cleaned, denoised, montaged and segmented based on the clinical annotations, such as sleep intervals and observed Ictals. For event detection, the signal waveform and its power were extracted symmetrically in non-overlapping intervals of 500 ms. Each waveform's power across all detected spikes was computed and clustered based on their energy distributions. **Results:** The recordings included thirteen sessions of 24 hours of extracellular recordings from two patients, with 312 hours extracted from four hippocampus electrodes anterior and posterior hippocampus. Our results indicate IEDs carrying the most different characteristics in the bands [25-75] Hz; SWR, on the other hand, are distributed between [80-170] Hz. **Conclusions:** Our algorithm detected and successfully distinguished IED from SWRs based on their carrying energy during non-sleep periods. Also, the most powerful spectral features that they were distinguished from occur in [15-30] Hz and [75-90] Hz.

P.126

Incorporation of Objective Structured Clinical Examination into EEG/Epilepsy Fellowship Training

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Background: For over 40 years, the objective structured clinical examination (OSCE) has been a part of medical education, eventually finding its way into most aspects of clinical training and evaluation. However, the EEG/epilepsy fellowship training has not classically involved OSCE evaluations. **Methods:** We designed and implemented a formative OSCE for pediatric and adult EEG/epilepsy fellows in Montreal, Quebec, Canada. The examination was offered in French and English. Stations included: technical issues, short cases, a long case, and communication. We solicited post-examination feedback from all participants via anonymous electronic survey after they had completed the Canadian Society for Clinical Neurophysiology (CSCN) EEG examination. We asked questions surrounding utility of the examination, areas for improvement, and whether the participant had been successful in passing the CSCN examination. **Results:** Six fellows took the initial formative OSCE. All six reported passing the subsequent CSCN examination. All participants reported the OSCE as useful in examination preparation. The communication station was consistently ranked as the least useful station, an unsurprising finding given that the CSCN examination does not involve a communication component. **Conclusions:** OSCE is an effective tool in assessment of the level of competence of EEG/epilepsy fellows, and as preparation for the CSCN EEG examination.

P.127

Ultra-high field 7-Tesla magnetic resonance imaging and electroencephalography findings in epilepsy

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Background: Assessment of patients for temporal lobe epilepsy (TLE) surgery requires multimodality input, including EEG to ensure optimal surgical planning. Often EEG demonstrates abnormal foci not detected on clinical MRI. 7T MRI provides improved resolution and we investigated its utility to detect potential abnormalities associated with EEG. **Methods:** Images were acquired on 7T MRI scanner (N=13) in oatients with TLE. Evaluation of 7T imaging for focal abnormalities was performed. Correlation of 7T MRI findings with EEG of focal slowing or interictal epileptic spikes (IEDs) and seizures was performed. **Results:** Assessment of 7T MRI demonstrated concordance with TLE in 8/13 cases. Three cases exhibited abnormal 7T MRI abnormalities not detected by 1.5 T MRI. Eleven out of 13 cases had EEG findings without anatomic correlates on MRI, with IEDs localizing to contralateral temporal, frontal, and parieto-occipital lobes. 7T images did not reveal focal anatomical abnormalities to account for the EEG findings in these patients. **Conclusions:** To our knowledge, this is the first study to