MS/Neuroinflammatory Disease

P.080

Outcomes in Influenza and RANBP2 mutation associated Acute Necrotizing Encephalopathy of Childhood

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Background: Acute Necrotizing Encephalopathy (ANEC) is a rare neuroinflammatory disorder involving the deep grey matter following viral infection and has been associated with the RANBP2 gene. We aimed to evaluate clinical and imaging features in ANEC patients. Methods: This retrospective chart review of ANEC patients (2012-2020) seen at a tertiary pediatric center included analysis of outcomes including ANE-Severity Score, Expanded Disability Status Scale (EDSS) and the modified Rankin Scale (mRS), semi-quantitative imaging scores (degree of swelling or hemorrhage rated 0 (none)-5 (severe/massive)), and dichotomous outcomes including RANBP2 gene status, influenza status. Results: 20 patients were included (Avg. age at presentation 3.5 yrs IQR=3.56., F:M 2.33:1). 3/20 experienced recurrences. All patients with recurrences were positive for RANBP2 mutations. 10/20 patients were influenza positive. 7/20 were RANBP2 mutation positive. We observed higher likelihood of hemorrhage in influenza-positive compared to negative patients (W=78, p=0.048). Kaplan-Meier survival curve analysis revealed that patients without brainstem lesions were more likely to reach minimal/no disability (EDSS<=2) than patients with brainstem lesions (p=0.035). Conclusions: Hemorrhage is more likely to be seen in children with ANEC who are positive for influenza. RANBP2 status was predictive of relapse but not predictive of overall outcome.

P.081

Evolving treatment of pediatric-onset multiple sclerosis in Alberta

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Background: Pediatric-onset multiple sclerosis (MS) is associated with a high rate of disease activity. However, only a single DMT – fingolimod – has been approved by Health Canada for children, in 2018. In this study, we describe trends in the treatment of pediatric-onset MS in Alberta. Methods: We performed a retrospective review of Alberta administrative health databases, identifying cases of MS under 19 years of age from January 1, 2011 - December 31, 2020. Pharmacy dispenses of MS DMTs were identified by Anatomical Therapeutic Chemical classification code and grouped as injectables (glatiramer acetate, interferon-beta) or newer agents (all others). Results: 79 incident cases of pediatric MS were identified during the study period. 47/79 (59%) had at least one DMT dispense, with the first dispense occurring a median 263 days (IQR 134.5-988) from the index date at a median age of 17.2 years (IQR 16.0-18.6). Injectables accounted for all initial DMT dispenses < 19 years of age prior to 2019, while from 2019-2020 injectables accounted for only 3/15 (20%) initial dispenses, with rituximab (5/15, 33%) being the most common initial DMT in those years. Conclusions: The treatment of children with MS in Alberta has rapidly evolved, shifting shift towards earlier treatment using newer high-efficacy agents.

Neurocritical Care

P.082

Using machine-based learning to predict neonatal hypoxic-ischemic encephalopathy (HIE) severity based on patient-specific clinical factors

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Background: Severity of HIE is based on Sarnat classification; however, it is difficult to predict precise neurodevelopmental outcomes as this only provides a single snapshot in time. We aimed to use machine-based learning to better understand variables contributing towards HIE severity. Methods: Patients with HIE treated with hypothermia were studied between 2014 and 2020 at level 3 NICUs in Calgary, Alberta. Clinical information contained 23 features including specifics of clinical examination, blood work, MRI and EEG findings, and medications. Random forest models were trained to examine features most predictive of HIE severity. Results: Two hundred and six patients were eligible. By grouping patients based on the initial Sarnat score and post-cooling exam, features correctly predicted groups 43% and 73% of the time, respectively. Precision, accuracy, and recall was best for the mild group. Using MRI and day 1 seizures it was 54% and 67% predictive, respectively. Features contributing most included arterial pH, initial lactate, and overall EEG findings. There are ongoing analyses for further classification. Conclusions: Machine-based learning can improve predictive models for patient outcomes. There is benefit in using variables outside of the initial examination to improve classification. We aim to expand this model to include detailed neurodevelopmental outcomes to improve prognostication.

Neuromuscular Disease and EMG

P.084

5q spinal muscular atrophy Canadian Paediatric Surveillance Program – 2020-2021 results

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Background: Spinal muscular atrophy (SMA) is the leading genetic cause of infant death and the second most common

autosomal recessive disorder; most cases are due to homozygous deletion of *SMN1* gene. Methods: This study uses the Canadian Paediatric Surveillance Program to determine the minimum annual incidence of 5q-SMA from birth to 18 years of age in Canada. The protocol can be accessed at www.cpsp.cps.ca/surveillance. Results: Eighteen cases were reported in 2020-2021. Ten (55%) cases were reported from Ontario and the remaining cases were reported from Atlantic Canada and Western Canada. Their median age was 11 months (IQR 4–21); 61% were male. The most common presenting symptoms were hypotonia and delayed motor milestones in 12 (86%) and 10 (71%) cases respectively. On average, the diagnosis was delayed after onset of symptoms by three months for SMA Type 1, by eight months for Type 2, and by 18 months for Type 3. Twelve (86%) cases received nusinersen as their first disease-modifying treatment. Conclusions: Early recognition and newborn screening are essential to reduce diagnostic delay and enable timely treatment of SMA. Other data sources including the Canadian Neuromuscular Disease Registry and molecular genetic laboratories will be used to estimate the annual incidence of pediatric SMA in Canada.

Other Child Neurology

P.085

The relationship between sleep and behavior in attention deficit/hyperactivity disorder

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Background: Attention-Deficit/Hyperactivity Disorder (ADHD) is a neurodevelopmental disorder that is associated with long-term reduced quality of life and impaired functioning. ADHD is commonly associated with sleep disturbances that can contribute to many difficulties in a child's life. This study aims to elucidate this complex relationship by utilizing a subset of the Adolescent Brain Cognitive Development (ABCD) database. Methods: The population included a group of children with ADHD age 10-13 years (n=212) and a matched typically developing (TD) group (n=212). Sleep data was obtained through Fitbit actigraphy measures, and the Parent Sleep Disturbance Scale (SDS). Behavioural and emotional subscores were obtained from the Child Behaviour Checklist (CBCL). Results: There were no significant correlations between the actigraphy and SDS sleep data. SDS sleep data were significantly different between ADHD and control groups, while actigraphy data was not. Sleep latency (measured by actigraphy) and 3 out of 6 of the SDS subscores were significantly related to behavioural scores. Conclusions: The results of this study indicate that sleep may not be an important mediator of behaviour and emotional responses in children with ADHD. Future studies should explore both influences on sleep parameters as well as behaviour and other measures important to families.

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Risk factors for term born periventricular white matter injury in children with cerebral palsy: a case control study

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Background: The aim of this study was to identify possible risk factors associated with term-born newborns with cerebral palsy and PWMI on imaging. Methods: This is a case-controlled study with cases from the Canadian Cerebral Palsy Registry and controls from Alberta Pregnancy Outcomes and Nutrition Study. PWMI was diagnosed based on MRI reports and 160 cases were compared to 1950 controls. Risk factor were selected *a priori*; including pregnancy complications, toxin exposure, perinatal infection, sex, small for gestational age, and perinatal adversity. Multivariate regression binomial model was used to calculate odds ratios(OR) and 95% confidence intervals(CI). Results: Multivariable analyses suggested PWMI was associated with pregnancy complications(OR=3.35; 95% CI=2.23-4.94), antenatal toxin exposure(OR=2.45; 95% CI=1.67-3.55), perinatal infection(OR=3.61; 95% CI=1.96-6.29) and perinatal adversity(OR=2.03; 95% CI=1.42-2.94). Term born males were not more likely to have PWMI compared to females(OR=1.37; 95% CI=0.98-1.93). Multiple regression analyses suggested independent associations between PWMI and pregnancy complications(OR=3.63; 95% CI 2.40-5.40), antenatal toxin exposure(OR=2.62; 95% CI 1.77-3.84), perinatal infection(OR=3.42; 95% CI 1.83-6.05) and perinatal adversity(OR=2.49; 95% CI= 1.71-3.69). Conclusions: Risk factors such as pregnancy complications, toxin exposure, perinatal infection and perinatal adversity are associated with PWMI in term-borns, suggesting a 'two-hit' model that could involve an interaction among both antenatal and perinatal variables.

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The landscape of paediatric tuberous sclerosis complex (TSC) neurological care in canada: results from a national survey

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Background: Tuberous Sclerosis Complex (TSC) is a genetic disease that affects multiple body systems with the neurological manifestations causing the greatest disease burden. The objective