MS/Neuroinflammatory Disease

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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.

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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

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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