study population comprised 164 patients (65 males) 6.25-year-old on average at absence onset. 22% had treatment-resistant seizures. The first ASM was Ethosuximide in 63.4%, Valproic acid in 23.2%, and Lamotrigine in 6.7%. Statistical differences between response groups included developing a second seizure type specifically GTC, the second and third ASM, and absence of EEG normalization. At last follow-up, 43.3% of children were seizure-free off ASMs. 32.9% of children had learning disabilities, 28% ADHD, and 12.8 % anxiety. **Conclusions:** 22% of children with CAE had treatment-resistant seizures. Photoparoxysmal response was not predictive of treatment resistance. Neuropsychiatric problems were common with learning disabilities increased with refractory absences.

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Midline Spikes and Intractable Seizures in Pediatric Epilepsy

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Background: Epileptic discharges localized to the midline vertex are rare. However, they have been associated with intractable seizures and severe long-term consequences in the developing brain. Our study aimed to understand the etiology of pediatric midline seizures and define post-surgical seizure outcomes. **Methods:** We reviewed charts, electroencephalography (EEG), and neuroimaging studies of ten pediatric patients with epileptic discharges localized to the midline vertex in the Comprehensive Epilepsy Program. The seizures were classified according to the International League Against Epilepsy criteria, patient age, sex, neuroimaging results, seizure etiology and outcomes were obtained. **Results:** Age of seizure onset was within the first 10 years of life in 90% of patients, with focal seizures being the most prevalent. Focal cortical dysplasia (FCD) was the most common etiology present in 50% of patients. These children had normal neuroimaging studies and intractable epilepsy. However, seizure freedom was achieved following surgical resection of the epileptogenic zone. **Conclusions:** We demonstrated that patients with midline epileptic discharges are associated with intractable focal seizures and early seizure onset. Despite normal neuroimaging reports, FCD was the most common pathology. Thus our study suggests early localization and resection of the epileptogenic zone may be beneficial for achieving seizure freedom in children with this electroclinical syndrome.

P.104

Children with Trisomy 21 and Lennox-Gastaut Syndrome with predominant myoclonic seizures

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Background: Lennox-Gastaut syndrome (LGS) is a severe form of pediatric epilepsy that is classically defined by a triad of drug-resistant seizures, characteristic EEG patterns, and intellectual disability. Long-term prognosis is generally poor with progressive intellectual deterioration and persistent seizures. At present, there are few reported cases of LGS and Trisomy 21 (T21) in the literature. To further delineate the spectrum of epilepsy in T21, we reviewed children with T21 and LGS at one center over 28 years. **Methods:** This is a retrospective case series. At our institution, all EEG results are entered into a database, which was queried for patients with T21 from 1992-2019. Pertinent electro-clinical data was obtained from medical records. **Results:** 63 patients with T21 and epilepsy, 6 (10%) had LGS and were included in the study. Four of the six patients were male and 5/6, had neuro-imaging, which was normal. Follow-up ranged from 3-20 years. Notably, 5/6 had predominant myoclonic seizures throughout the course of their epilepsy, associated with generalized spike-wave discharges. **Conclusions:** Myoclonic seizures appear to be a predominant seizure type in patients with T21, suggestive that T21 patients may have a unique pattern of LGS.

P.106

Intravenous lacosamide use in pre-school children

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Background: Data on intravenous lacosamide use in young pediatric patients is scarce, especially of pre-school age. **Methods:** We retrospectively reviewed the medical records of all patients less than 6 years old who received intravenous lacosamide at our tertiary pediatric hospital. Data on dose, timing and order of administration was collected. Clinical and electrographic response was independently assessed with EEG interpretation blinded to time of administration. For adverse effects surveillance, heart rate was noted before and 1 hour after dose. **Results:** Eleven patients (8 boys), received lacosamide between 2013 and 2018. Mean age was 2 years (11 days – 5,3 years). Medical indications were: refractory status epilepticus (n=6), repetitive seizures (n=4), and inability to take oral lacosamide (n=1). On average, lacosamide was the fifth (1^{st}-8^{th}) IV antiepileptic drug administered 78 hours (SD 11 hours) after presentation. The most frequent dose was 5 mg/kg. Clinical response was confirmed in 7 patients, while electrographic response was proven in 3 patients. Seizure relapse at 24 hours was noted in 6 patients. No bradycardia occurred post-lacosamide. **Conclusions:** Although very safe, therapeutic response to lacosamide in young pediatric patients was inconclusive, mostly due to delay in administration, suboptimal dose, and high number of other IV antiepileptic drugs previously given.

P.107

Response to the Ketogenic Diet in refractory epileptic spasms at BC Children's Hospital

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Background: Epileptic spasms (ES) are a devastating seizure type with poor neurodevelopmental outcome; 1/3 are resistant to

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treatment with first line therapies. Recently attention has been drawn to the ketogenic diet (KD) as a potentially effective therapy, though data regarding optimal time of initiation, and its sustained effectiveness, are lacking. **Methods:** Retrospective chart review of all patients with ES treated with KD at BC Children's Hospital between 2002 and 2020 (n=28) with comparison of spasm response based on age of initiation of KD in two groups: < 12 months (n=11) and ≥ 12 months (n=17). **Results:** Comparing the <12 months and ≥ 12 months groups showed: unknown etiology in 9% vs 25%; spasm freedom for 3 months on KD in 18% vs 41%; median time to spasm freedom was 2 vs 6 weeks; relapse after a period of spasm freedom occurred in 66% vs 70%. **Conclusions:** Although more effective in children ≥ 12 months of age in the first 3 months, spasm freedom in either group was not sustained with KD. KD is recommended as early therapy for refractory ES, but this study suggests clinicians be aware the KD has limited efficacy in long-term control of ES and must be used with other therapies.

Metabolic Disease

P.109

Diagnostic Yield of Targeted Exome Sequencing in West Syndrome

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Background: West syndrome (WS) is characterized by the onset of epileptic spasms usually within the first year of life. Global developmental delay with/without regression is common. Advances in high-throughput sequencing have supported the genetic heterogeneity of this condition. To better understand the genetic causes of this disorder, we investigated the results of targeted exome sequencing in 29 patients with WS. **Methods:** Whole exome sequencing (WES) was performed on an Ion Proton™ and variant reporting was restricted to sequences of 620 known epilepsy genes. Diagnostic yield and treatment impact are described for 29 patients with WS. **Results:** A definitely/likely diagnosis was made in 10 patients (34%), which included 10 different genes (*ALG13, PAFAH1B1, SLC35A2, DYNC1H1, ADSL, DEPDC5, ARX, CDKL5, SCN8A, STXBP1*) known to be associated with epilepsy or WS. Most variants were *de novo* dominant (X-linked/autosomal) except for *ARX* (X-linked recessive) and *ADSL* (autosomal recessive). 4 out of 10 (40%) had a genetic diagnosis with potential treatment implications. **Conclusions:** These results emphasize the genetic heterogeneity of WS. The high diagnostic yield, along with the significant genetic variability, and the potential for treatment impact, supports the early use of this testing in patients with unexplained WS.

MS/Neuroinflammatory Disease

P.111

Use of rituximab for pediatric central nervous system inflammatory disorders in Alberta

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Background: Rituximab is a B-cell-depleting monoclonal antibody whose off-label use is funded in Alberta by the Short-Term Exceptional Drug Therapy (STEDT) program. This study describes the use of rituximab for pediatric central nervous system (CNS) inflammatory disorders in Alberta. **Methods:** Rituximab applications for CNS inflammatory indications in patients < 18 years of age were identified from the STEDT database between January 1, 2012 – December 31, 2019. Patient information was linked to other provincial datasets, including the Discharge Abstract Database, Pharmaceutical Information Network, and provincial laboratory data. Analysis was descriptive. **Results:** 51 unique rituximab applications were identified, of which 50 were approved. New applications increased from one in 2012 to a high of 12 in 2018. The most common indication was autoimmune encephalitis (other than anti-NMDA receptor encephalitis; n=20, 39%). Most children were approved for a two-dose (n=33, 66%) or four-dose (n=16, 32%) induction regimen. Physician-reported outcomes were available for 24 patients, of whom 14 (58%) were felt to have fully met outcome targets. **Conclusions:** The use of rituximab for pediatric CNS inflammatory disorders has increased, particularly for the indication of autoimmune encephalitis. This study identified significant heterogeneity in dosing practices and laboratory monitoring, as well as regional disparities in use.

Neuromuscular Disease and EMG

P.112

5q Spinal Muscular Atrophy Canadian Paediatric Surveillance Program - 2020 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; the majority of 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 complete protocol can be accessed at

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