neurodevelopmental evaluation at TEA in infants born between 29-36 weeks GA. Methods: Prospective cohort study of preterm infants born 29-36 weeks GA with 1 hour EEG recording at TEA. EEG discontinuity index (proportion <25mcV amplitude) and spectral power densities were calculated as well as the mean and maximum values of interburst intervals. At TEA, neurodevelopment was evaluated using the *General Movement Optimality Score (GMOS)*. Linear regression analyses were used to evaluate the association between EEG features and neurodevelopmental assessment. Results: Eighty-two children (median GA 33.6 weeks) were included (47 males). Median GMOS was 29.0 (IQR 24.3-35.0). A greater EEG discontinuity index was associated with reduced GMOS (B -6.85; 95% CI -12.13,-1.57; p=0.012). Conclusions: At TEA, a greater EEG discontinuity index was associated with a more abnormal neurodevelopmental assessment. Ongoing longitudinal neurodevelopmental assessments are needed to better evaluate the prognostic potential of TEA EEG.

P.077

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 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.

P.078

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

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Background: 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: 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: 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: 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.

Metabolic Disease

P.079

MT-TA: A mitochondrial genome cause of developmental and epileptic encephalopathy

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Background: *MT-TA* (OMIM 590000), one of 22 mitochondrial transfer-RNA (mt-tRNA) genes, encodes the mt-tRNA for alanine. Pathogenic variants in mt-tRNA genes affect the translation of respiratory chain complexes I, III, and IV; which leads to mitochondrial dysfunction and a clinically variable phenotype. *MT-TA* pathogenic variants have been described in only seven patients, all of whom had isolated myopathy Methods: Case report. Results: Our patient initially presented with drug-resistant West syndrome, later evolving towards a Lennox Gastaut phenotype. Although she had hypotonia, serum creatine kinase and electromyography were normal. Brain-MRI showed bilateral symmetric hypointense T1, hyperintense T2-fluid-attenuated-inversion-recovery and restricted-diffusion signal changes in the dentate nuclei. Mitochondrial genome testing identified a previously published pathogenic variant in *MT-TA* (m.5591G>A) with 14% blood heteroplasmy and 16% urine heteroplasmy. The variant was absent in serum sampled from the patient's mother Conclusions: Our case extends the phenotypic spectrum of *MT-TA* variants to include developmental and epileptic encephalopathy, in the apparent absence of muscle disease. We hypothesize that our patient may have the greatest degree of heteroplasmy in brain tissue; however, animal models and induced pluripotent stem cell (iPSC) models are needed to identify the precise mechanism by which *MT-TA* dysfunction results in variable phenotypes with variable degrees of heteroplasmy.