to initiation of quinidine, this patient experienced 22 electrographic seizures over 24 hours. At target dose, this patient experienced greater than 70 seizures over 24 hours. *Conclusions:* Quinidine has previously been reported to be effective in patients with MMPSI with the same and different mutations. We report the second case of a patient with MMPSI and KCNT1 mutation R428Q with poor clinical response to quinidine.

P.037

Role of epilepsy monitoring unit in the investigation of patients with epilepsy and developmental delay

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Background: A significant part of the developmental delay (DD) population has epilepsy (26-70%) and live in an institution. These patients tend to have atypical presentation of epileptic seizures with higher risk of misdiagnosis. Distinguishing their ictal events from paroxysmal behaviors can be challenging.There often is a lack of description of the spells or inadequate history from the caregivers or the patients. These patients often have drug resistant epilepsy requiring polypharmacy with increased risk of morbidity and mortality. The aim of this study was to determine usefulness of Epilepsy Monitoring Unit (EMU) in diagnosis and management of these patients. *Methods:* This is a retrospective observational study of the patients with epilepsy and DD living in institutions that were admitted to the EMU. *Results:* Four patients met the inclusion criteria for this study. The mean age was 45(29-71), 3/4 (N=3) were male and 3/4 had focal epilepsy. All patients had mood disorders and 2 were taking antipsychotic medication. The mean admission-time was 6,25 days (2-15) and there was a correlation with the events and seizures in 2/4 of the patients and the rest had a combination of behavioural-changes and seizures. *Conclusions:* EMU admission can provide an accurate diagnosis of spells in patients with DD and epilepsy, and improve their quality of life.

P.038

Clinical experience with perampanel for refractory pediatric epilepsy in one Canadian center

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Background: Perampanel (PER) is a new anti-seizure medication that inhibits the α-amino-3-hydroxy-5-methyl-4-isoxazole-propionic acid (AMPA) class of glutamate receptors. It is available in Canada for children since 2014. It is important for physicians to be aware of the efficacy and tolerability of drugs in the post-marketing phase. *Methods:* We did a retrospective review of our experience with PER at BC Children's Hospital. Patients on PER were identified. Clinical data, including demographics, efficacy, tolerability, adverse effects (AE) and retention rates were obtained by review of clinical records. *Results:* Of 24 patients pediatric patients prescribed PER, 21 (87%) had focal and three had symptomatic generalized epilepsy. Ten (42%) had greater than 50% reduction in seizures. In fifteen patients, (63%) PER was discontinued due to AE or poor response. Twelve (50%) had behavioral AE and eight (33%) had non-behavioral AE. PER was effective, at lower doses than required for adults. One third experienced serious AE. One patient experienced oculogyric crisis, not previously reported with PER. AE were not associated with high doses and were reversible. Possible risk factors for behavioral AE include behavioral problems with other medications and pre-existing behavioral co-morbidities. *Conclusions:* It is important for clinicians to be aware of and counsel patients about serious AE, particularly behavioral, when prescribing PER.

P.039

mTOR inhibitors as a new therapeutic strategy in treatment resistant epilepsy in hemimegalencephaly: a case report

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Background: Hemimegalencephaly (HME) is a hamartomatous malformation of one cerebral hemisphere, resulting in refractory epilepsy, intellectual disability, and autistic features. Hemispherectomy is the definitive treatment, but there is risk of high morbidity and mortality, especially when done in early infancy. Various preclinical studies have shown that dysregulation of the mTOR pathway has an integral role in the development of various epilepsy syndromes, including tuberous sclerosis complex (TSC), focal cortical dysplasia and HME. Recently, mTOR inhibitors were proven to be effective in treating seizures in TSC. *Methods:* We present a case of a 6 day old female with refractory epilepsy despite the trial of 9 anti-seizure medications and the ketogenic diet. As the patient was awaiting epilepsy surgery, an mTOR inhibitor, rapamycin was initiated. *Results:* After 1 week of the initiation, she had over a 50% reduction in seizures. At two weeks, the parents felt that for the first time, she was making developmental gains. She also appeared brighter and more interactive. Due to her response to treatment, her hemispherectomy was deferred to when she is older, so there will be a decreased risk of complications from the surgery. *Conclusions:* This case exemplifies how mTOR inhibitors should be considered as a treatment option for patients with HME and refractory epilepsy.

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