

functional studies; and determined the effect of diazepam and insulin on wild type and mutant GABA receptors. **Results:** The patient is a 10-year-old girl with EE, treatment-resistant seizures, intellectual disability and autism. Her GABRA1 (R214C) mutation dramatically decreased whole-cell GABA-evoked currents by reducing GABA surface receptors, decreasing single channel open time, and altering channel kinetic properties. The combination of diazepam and insulin partially repaired these effects by enhancing channel activity and increasing the number of surface receptors, respectively. **Conclusions:** Diazepam and insulin partially mitigated a *de novo* GABRA1 (R214C) mutation's effects on GABA receptor number and function. Given the risks of insulin use, pharmacological agents with similar mechanisms of action but fewer side effects, such as IGF-1, should be studied and considered for clinical application.

P.017

Results of a Pilot feasibility study to develop reduce wait times strategy in the evaluation of children with new onset epilepsy

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Background: The goal was to understand factors leading to prolonged wait times for neurological assessment of children with new onset seizures. A second objective was to develop an innovative approach to patient flow through and achieve a reduction in waiting times utilizing limited resources.

Methods:

1. Audit of the referrals, flow through, wait times
2. Identification of bottlenecks
3. Development of triaging strategy:
 - i. Suspected Febrile seizures and non-epileptic events;
 - ii. Suspected benign and absence epilepsies;
 - iii. Suspected other Focal epilepsies, generalized epilepsies, epilepsy under 2 years
4. Initiation of early telephone contact and support
5. Development of a ketogenic diet

Results: Using a triaging strategy and focusing on timely access to investigations, wait times for clinic evaluations were shortened despite larger numbers of referrals (mean wait time reductions from 179 to 91 days). Limiting factors such increase in referral numbers, attrition in support staff, interfered with sustainability of reduced wait times achieved in the initial phase of the program. **Conclusions:** This pilot study highlights the effectiveness of an innovative triaging strategy and improvements in patient flow through in achieving the goals of reduction in wait times for clinical evaluation and timely investigations to improve care for children with new onset seizures. Insights into limitations of such strategies and factors determining sustainability are discussed.

P.018

Forced normalization after vagal nerve stimulation in a case of intractable Lennox-Gastaut syndrome

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Background: Forced normalization is the development of psychiatric symptoms in a patient experiencing remission of intractable seizures. The mechanism of this phenomenon is unknown. We present a complex case of Lennox Gastaut syndrome that experiences forced normalization after vagus nerve stimulation (VNS). **Methods:** This case details a 31-year-old male with seizures since early childhood. The patient has intractable epilepsy and failed AEDs, VNS, and a partial callosotomy. **Results:** The patient was in remission from 2-12 years old, when seizures returned at a frequency of 2-5 per day. He has multiple types of seizures including drop attacks, absences, and tonic-clonic seizures. Patient experienced status epilepticus multiple times. Twelve AEDs were failed before VNS was started in 2010, which helped curb the severity of seizures and the potential for clusters. Forced normalization developed over the course of treatment with VNS. The patient behavior was characterized by aggression, paranoia, and hallucinations. VNS was turned off late in 2010 and then re-started in January of 2011. Patient proceeded to cycle between several days of seizures without psychiatric symptoms and several days of psychosis without seizures. **Conclusions:** Vagus nerve stimulation gave way to forced normalization, characterized here as aggressive behaviour and psychosis. Forced normalization is seen commonly after epilepsy surgery, but rarely following VNS.

P.020

Novel GRIN2A variant in family members with variable phenotypic expression of epilepsy

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Background: Epilepsy aphasia spectrum of disorders is characterized by developmental and language regression with EEG abnormalities that include electrical status epilepticus of sleep (ESES). Landau-Kleffner syndrome (LKS) and epileptic encephalopathy with continuous spike-wave during sleep (CSWS) are the most severe presentations. GRIN2A mutations have been recognized as causative. **Methods:** we present two sisters with different epilepsy phenotypes. A variant of unknown clinical significance (VUS) in GRIN2A gene was found in one of the sisters and her similarly affected father. **Results:** The first sister presented with focal onset seizures at the age of 3 years accompanied by language and cognitive regression and EEG features consistent of ESES, meeting criteria for LKS. Multiple anticonvulsants were tried until she responded well to steroids regaining developmental milestones. Her 5-year-old sister recently presented with focal onset seizures. Her language development is appropriate. Her EEG showed independent multifocal spikes but no ESES during sleep. Her seizures were controlled on monotherapy anticonvulsants. **Conclusions:** We observed a variable EEG-clinical phenotype and different severity among these family members as

expected with GRIN2A-related disorders. This report contributes to evidence of the GRIN2A variant pathogenicity.

P.021

Childhood small vessel primary angiitis of the CNS: a potentially treatable cause of super-refractory status epilepticus

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Background: Childhood primary angiitis of the central nervous system (cPACNS) is a rare inflammatory disease of brain vessels. The small vessel subtype is diagnosed on brain biopsy and often presents with cognitive and behavioural changes, headaches and seizures. However, there are few reported cases of super-refractory status epilepticus. **Methods:** We present a case of small vessel cPACNS complicated by super-refractory status epilepticus and review the literature. **Results:** Our patient is a previously healthy 11-year-old boy who presented with new-onset seizures and encephalopathy in the context of fever. He developed super-refractory status epilepticus, requiring burst suppression for four weeks with various IV infusions. During this time, he was on the ketogenic diet and tried eight anti-seizure medications. Extensive investigations included brain biopsy confirming small vessel cPACNS. He was treated with IV methylprednisolone, oral steroids, IVIG, and cyclophosphamide. After prolonged rehabilitation, he recovered almost completely and has a normal neurological examination with no epileptiform activity on EEG. **Conclusions:** Small vessel cPACNS should be considered in the differential diagnosis of super-refractory status epilepticus. Despite being in SE for four weeks, symptomatic management of seizures and immunosuppression to treat the underlying pathology resulted in favourable neurological outcomes. This is one of the long-term cases of SE in small vessel cPACNS in the literature.

FUNCTIONAL NEUROSURGERY AND PAIN

P.022

Hemifacial Spasm due to dolichoectatic vertebrobasilar artery compression

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Background: Hemifacial Spasm (HFS) is rarely caused by a dolichoectatic vertebrobasilar artery (eVB) compression of the Facial Nerve. This can pose a surgical challenge when performing microvascular decompression as vessel mobilization is often difficult due to atherosclerosis, tethering from brainstem perforators, and large size. These patients are often not considered for surgery. **Methods:** A retrospective chart review of patients who were surgically treated by the senior author between 2003 and 2017 with an admitting diagnosis of HFS was performed. Patients with preoperative neuroimaging demonstrating eVB compression of their facial nerve/root were included. **Results:** During the 15-year review, 315 patients underwent microvascular decompression for HFS and 21

(6.7%) had dolichoectatic vertebrobasilar compressions. At final followup (>3 months) 19 patients (90.4%) experienced reduction in symptoms with 15 (71.4%) having complete resolution. One patient required re-operation and benefitted from subsequent symptom relief. The majority of culprit compression was found proximally on the pontine surface. Mobilization of the culprit vessel was achieved successfully in the majority of cases with Teflon pledgets. There were no perioperative strokes or death. Complications are presented. **Conclusions:** Microvascular Decompression for Hemifacial Spasm caused by dolichoectatic vertebrobasilar artery compression can be performed with a high rate of safety and success in the setting of a high case volume centre.

P.023

Clinical and patient satisfaction outcomes after partial sensory rhizotomy for refractory trigeminal neuralgia among MS patients

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Background: MS related trigeminal neuralgia (MS-TN) is associated with high recurrence and retreatment rates. Optimal treatment and role for partial sensory rhizotomy (PSR) for MS-TN remains to be determined. **Methods:** We analyzed time to treatment failure (TTF) after PSR (n=14) versus other prior procedures (n=53) among 12 consecutively treated MS-TN patients. Kaplan-Meier curves and Log-Rank tests were utilized to compare BNI pain scores and TTF after PSR vs prior procedures using the same patient cohort as their own control group. Subsequent analysis compared TTF after PSR to other procedures (n=93) among a second cohort of 18 MS-TN patients not undergoing PSR. **Results:** TTF was significantly longer after PSR compared to prior procedures among the PSR cohort (p<0.0001) with median TTF of 79 vs 10 months respectively. Similarly, there was a longer TTF after PSR compared to prior procedures among the MS-TN cohort with median TTF 79 vs 16 months respectively (p<0.001). PSR resulted in a higher proportion of excellent pain scores when compared to prior procedures in the MS-TN cohort (77% vs 29%, p<0.001). **Conclusions:** TTF was significantly longer following partial sensory rhizotomy compared to other prior procedures in MS-TN patients. A higher proportion of patients achieved excellent BNI pain scores after PSR.