

non-dominant MTLE, the smaller dominant HV at 2-3y period correlated with decline of verbal memory ( $p < 0.05$ ). **Conclusions:** Post-operative progression of non-epileptic hippocampal atrophy was found with significantly more pronounced in patients with older age at surgery and larger pre-operative non-epileptic hippocampus. After the epileptogenic hippocampus is resected, the remaining hippocampus alone might exhaust to maintain the memory, especially in elders.

## P.013

### Convulsive status epilepticus due to intracranial hypotension

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**Background:** Intracranial hypotension (IH) is typically characterized by an orthostatic headache. There have been limited case reports describing iatrogenic IH presenting with seizures. **Methods:** Case report. **Results:** A 71-year-old woman with chronic back pain developed convulsive status epilepticus (SE), characterized by generalized clonic seizures, immediately following scoliosis surgery. She had no history of seizures or seizure risk factors. Despite treatment with Midazolam, Phenytoin and Lacosamide, seizures recurred five times over three hours. Thus, Propofol and Midazolam infusions were initiated. An electroencephalogram revealed burst suppression and bilateral hemispheric epileptiform discharges. MRI brain was consistent with IH without cortical vein thrombosis. Fluid from the surgical drains was positive for Beta-2 transferrin, indicating cerebral spinal fluid. Her intracranial hypotension was likely due to an intraoperative dural tear causing SE. Over two weeks, she remained on bedrest, sedation was weaned, and Phenytoin and Lacosamide were tapered and discontinued. She had no further seizures. **Conclusions:** IH is an under recognized cause of seizure following spinal or cranial surgery, lumbar puncture, or spinal anaesthesia. Proposed mechanisms include traction on cortical structures, increased cerebral blood flow and cortical irritation secondary to subdural hygromas.

## P.014

### Survey of epilepsy and seizure awareness in Manitoba: an evaluation (SESAME)

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**Background:** Epilepsy/seizure awareness is improving across Canada. With the formation of a Comprehensive Epilepsy Program in Manitoba (including a new Pediatric Epilepsy Monitoring Unit), a provincial strategy has been proposed outlining a path towards improved access to epilepsy care. We now sought to qualify the current state of clinician knowledge and comfort towards diagnosis and management of this condition. **Methods:** A qualitative online survey, comprised of 36 short-answer questions, was delivered to primary care and specialist physicians in Manitoba. **Results:** 108 subjects responded, across varying medical disciplines. 101 (93.5%) have previously managed epilepsy patients, and 87 (80.6%) have previously ordered an EEG. A total of 63 (59.4%) have referred to a neurologist, with a lower proportion (30, 28.3%) referring specifically to an epileptologist. 36 respondents (33.3%) have heard of the ILAE guidelines, with 43 (63.2%) reporting refractory epilepsy to be

defined by the failure of 3 (or more) medications. 61 (56.5%) were unaware of invasive EEG techniques. Most (85, 78.7%) understood a role for surgery in treating epilepsy, with 12 (11.1%) unaware of surgical therapies beyond VNS. **Conclusions:** SESAME successfully identified strong awareness towards epilepsy, with small lapses in knowledge that will benefit from a formal provincial-wide educational curriculum.

## P.015

### Mesial Temporal Sclerosis is a rare occurrence in Intractable Pediatric Temporal Lobe Epilepsies

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**Background:** Temporal lobe epilepsy (TLE) accounts for approximately 20% of pediatric epilepsy cases. Of those, many are considered medically intractable and require surgical interventions. In this study, we hypothesized that mesial temporal sclerosis (MTS) was less common in patients who had undergone surgery for intractable pediatric TLE than in adult series. We further hypothesized that there was a radiological and pathological discordance in identifying the cause of pediatric TLE. **Methods:** We retrospectively reviewed the charts of pediatric patients with TLE who had undergone surgical treatments as part of the University of Alberta's Comprehensive Epilepsy Program between 1988 and 2018. Along with preoperative magnetic resonance imaging (MRI) reports, post-surgical pathology results and seizure outcomes were studied. **Results:** Of the 83 pediatric patients who had undergone temporal lobe epilepsy surgery, 28% had tumors, 22% had dual pathologies, 18% had MTS, 11% had focal cortical dysplasia, and 22% had other pathologies. In addition, for 36% of these patients, discordance between their pre-surgical MRI reports and post-surgical pathology reports were found. **Conclusions:** This was one of the largest retrospective cohort studies of pediatric patients who had undergone surgery for intractable TLE. This study showed that tumors, and not MTS, were the most common pathology in surgical pediatric TLE.

## P.016

### A novel de novo GABRA1 mutation linked to epileptic encephalopathy: pathophysiology and potential therapeutic options

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**Background:** Epileptic encephalopathy (EE) is a severe neurological disorder characterized by treatment-resistant seizures and poor neurodevelopmental outcomes. EE is associated with mutant genes, including those that encode for  $\gamma$ -aminobutyric acid type A (GABA) receptor subunits. We identified a novel *de novo* GABRA1 mutation in a patient with EE, characterized its impact on GABA receptor function, and sought potential therapeutic options. **Methods:** We described the clinical and electrophysiological features of a patient with a novel *de novo* GABRA1 (R214C) mutation; performed

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