(55%). Most conditions were under-observed in training environment. Many noted a need for more independent practice development and community neurology. *Conclusions:* Although our training was found to be very good, some identified needs included advocacy training, and more training in general neurology in the longitudinal outpatient/community settings.

B.04

Distal and asymmetric myasthenia gravis: a case series of 54 patients

AG Florendo-Cumbermack (London) MW Nicolle (London)*

doi: 10.1017/cjn.2016.63

Background: Distal/asymmetric presentations of myasthenia gravis (MG) are uncommon and occur in 3-7% of patients with MG. This pattern of weakness is often not recognized as a manifestation of MG, leading to inappropriate investigations, delayed diagnosis and potentially missed opportunities for treatment. Our knowledge about this atypical presentation is limited to small case series and individual case reports. This study therefore aims to expand our understanding by describing the clinical course, diagnosis and treatment of a larger series of patients with this presentation. *Methods:* We conducted a retrospective chart review of patients with definite MG (either acetylcholine receptor [AChR] or MuSK antibody positive or clear evidence of postsynaptic neuromuscular junction dysfunction on electrodiagnostic studies), who attended the MG Clinic in London. Details of the clinical course, electrodiagnostic studies, antibody testing and response to treatment are reported. *Results:* 5.9% (54/921) of patients with definite MG had distal/asymmetric limb involvement, 56% at onset and 4% developing more than 10 years later. Males predominated (2:1). Finger extensors were most affected. 83% were AChR antibody positive. 7% had thymomas. On repetitive nerve stimulation most patients showed the most significant decrement distally on the more affected side. Almost all patients improved with treatment. *Conclusions:* This study expands our understanding of distal/asymmetric presentations of MG.

B.05

Optimizing IVIg utilization for neuromuscular disease in BC: high user project

KM Chapman (Vancouver) R Mosewich (Vancouver) T Trewern (Vancouver) C Li (Vancouver) G Gibson (Vancouver)*

doi: 10.1017/cjn.2016.64

Background: In British Columbia, neuromuscular disease accounts for 31% of IVIg use, at a cost of $10.1 M. In addition to the new screening pathway, the BC Neuromuscular IVIg Program developed the Chronic High User Project to identify areas for improvement in utilization. *Methods:* Utilizing CTR data, all patients on IVIg maintenance therapy for approved neuromuscular conditions between April 1, 2013 and March 31, 2014 were identified. Patients receiving higher than usual IVIG treatments (CIDP and MG >1110 grams/year, MMNCB > 1400 grams/year) were evaluated. Following panel review, utilization data was compared with a second cohort (2014 to 2015) to determine impact. Following review, appropriateness of treatment was determined by consensus from a 3-member panel, and recommendations were made. *Results:* Of 377 patients, 38 "High Users" were identified. 29 cases were determined to be appropriate; 9 were not. There was a reduction in mean grams/episode in CIDP (1135g to 990g) and MG (1099 g to 1022g) between cohorts. The mean grams/episode for MMNCB did not change. *Conclusions:* In specific cases, the IVIg High User Program identified patients in whom the treatment could be optimized. However, the vast majority of use of IVIg for Neuromuscular Disease in BC is appropriate, including in patients requiring higher that "usual" doses.

B.06

CNS André Barbeau Memorial Prize

Two definite sudden unexpected deaths in epilepsy in a family with a DEPDC5 mutation

FA Nascimento (Toronto) F Borlot (Toronto) P Cossette (Montreal) B Minassian (Toronto) D Andrade (Toronto)*

doi: 10.1017/cjn.2016.65

Background: DEPDC5 gene, mapped to 22q12.2-q12.3, has been associated with a variety of familial epilepsies, including FFEVF, autosomal dominant nocturnal frontal lobe epilepsy, and familial TLE. Notably, *DEPDC5* has never been linked to increased risk of sudden unexpected death in epilepsy (SUDEP). *Methods:* Cases review. *Results:* We studied a three-generation, non-consanguineous, French-Canadian family with nine clinically affected individuals. The index case is a 39-year-old man who started having seizures (as 2rily GTCS) at the age of 13 years. EEGs showed interictal discharges over the right anterior-temporal region. Brain MRI was unremarkable. Two individuals in this family suffered definite autopsy-confirmed SUDEP, at the ages of 58 and 50 years, respectively. Overall, seizure-history in this family can be summarized by an onset before reaching adulthood followed by subsequent progressive decrease in seizure frequency. Seizures were predominantly nocturnal 2rily GTC. Genetic analysis revealed a pathogenic heterozygous variant in the *DEDPC5* gene (p.Gln216, c.646C>T), which results in a premature stop codon, in all affected family members plus on heatlhy relative. Importantly, all the subjects were cognitively intact, and there was no history of cardiac symptomatology/cardiovascular risk factor. *Conclusions:* The finding in this family suggests that DEPDC5 mutations may be a risk factor for SUDEP.

B.07

Evaluating the single seizure clinic model: findings from a Canadian centre

SA Rizvi (Saskatoon) L Hernandez-Ronquillo (Saskatoon) F Moien-Afshari (Saskatoon) G Hunter (Saskatoon) JF Tellez-Zenteno (Saskatoon) K Waterhouse (Saskatoon)*

doi: 10.1017/cjn.2016.66

Background: The effect of the single seizure clinic (SSC) model on patient diagnosis, work-up, wait-times, and clinical care is poorly characterized. This study assesses patient characteristics and evaluates the impact of a SSC model on wait-times and access to care. *Methods:* A prospective study of all patients (n=200) referred to our SSC for first-seizure evaluation. Demographic, clinical, and paraclinicial variables were analyzed against a historical cohort. Binary logistic regression analysis was performed to predict impact of dichotomized

variables on diagnosis of epilepsy. Diagnostic concordance between SSC nurses and epileptologists was also assessed. *Results:* Predominant referral sources were emergency department physicians and general practitioners. Mean wait-time for first assessment was significantly reduced by 70.5% employing the SSC model versus historical usual care. A diagnosis was established at first-contact in 80.5% of cases while 16.0% of patients required a second visit. Eighty-two patients (41.0%) were diagnosed with epilepsy. The most common non-seizure diagnosis was syncope (24.0%). An abnormal EEG was found in 93.9% of patients diagnosed with epilepsy. Sixty-three patients were started on anti-epileptic drugs. In 18% of cases driving restrictions were initiated by the SSC. There was moderate correlation between SSC nurses and physicians (kappa=0.54; p<0.001) diagnoses. *Conclusions:* The SSC model reduces wait-times, streamlines assessments, and impacts clinical care decisions.

CNSS Chair's Select Abstracts

C.01

CNSS K.G. McKenzie Memorial Prize in Clinical Research

Intrathecal morphine following lumbar fusion: a randomized, placebo-controlled trial

D Yavin (Calgary) P Dhaliwal (Orlando) T Whittaker (Calgary) GS Hawboldt (Calgary) GA Jewett (Calgary) S Casha (Calgary) S du Plessis (Calgary)*

doi: 10.1017/cjn.2016.67

Background: Despite the ease of intraoperative injection, intrathecal morphine (ITM) is rarely provided in lumbar spine surgery. We therefore sought to demonstrate the safety and efficacy of ITM following lumbar fusion. *Methods:* In this double-blind trial, 150 patients undergoing elective instrumented lumbar fusion were randomly assigned to receive a single injection of ITM (0.2 mg) or placebo (saline) prior to wound closure. Primary outcomes were postoperative pain on the visual-analog scale during the inital 24 hours after surgery and respiratory depression. Secondary outcomes included related adverse events, opioid requirements, and length of stay. Outcome curves were estimated in an intention-to-treat, repeated-measures analysis. *Results:* Age, disability, operative times, and pre-operative pain were similar in both groups. ITM was associated with less pain both at rest (p<0.002) and with movement (p<0.02) during the initial 24 hours following surgery. ITM did not increase the cumulative incidence of respiratory depression (hazard ratio 0.86, p=0.66). While ITM reduced postoperative opioid requirements (p<0.03), there was no significant difference in length of stay (p=0.67). Adverse events did not significantly differ between groups. The early benefits of ITM on postoperative pain were no longer apparent after 48 hours. *Conclusions:* A single ITM injection safety reduces postoperative pain following lumbar fusion. (ClinicalTrials.gov NCT01053039)

C.02

CNSS K.G. McKenzie Memorial Prize in Basic Neuroscience Research

Whole genome expression profiling of blood-brain barrier endothelial cells after experimental subarachnoid hemorrhage

MK Tso (Calgary) P Turgeon (Toronto) B Bosche (Toronto) J Ai (Toronto) P Marsden (Toronto) RL Macdonald (Toronto)*

doi: 10.1017/cjn.2016.68

Background: The pathophysiology of subarachnoid hemorrhage (SAH) is complex and includes disruption of the blood-brain barrier (BBB). We freshly isolated BBB endothelial cells (BECs) by 2 distinct methods after experimental SAH and then interrogated their gene expression profiles with the goal of uncovering new therapeutic targets. *Methods:* SAH was induced using the prechiasmatic blood injection mouse model. BBB permeability studies were performed by administering intraperitoneal cadaverine dye injections at 24h and 48h. BECs were isolated either by sequential magnetic-based sorting for CD45-CD31+ cells or by fluorescence-activated cell sorting (FACS) for Tie2+Pdgfrb- cells. Total RNA was extracted and analyzed using Affymetrix Mouse Gene 2.0 ST Arrays. *Results:* BBB impairment occurred at 24h and resolved by 48h after SAH. Analysis of gene expression patterns in BECs at 24h reveal clustering of SAH and sham samples. We identified 707 (2.8%) significant differentially-expressed genes (403 upregulated, 304 downregulated) out of 24,865 interrogated probe sets. Many significantly upregulated genes were involved in inflammatory pathways. These microarray results were validated with real-time polymerase chain reaction (RT-PCR). *Conclusions:* This study is the first to investigate in an unbiased manner, whole genome expression profiling of freshly-isolated BECs in an SAH animal model, yielding targets for novel therapeutic intervention.

C.03

CNSS K.G. McKenzie Memorial Prize in Clinical Research (2nd place)

Progressive contralateral hippocampal atrophy following surgery for medically refractory temporal lobe epilepsy

CA Elliott (Edmonton) D Gross (Edmonton) B Wheatley (Edmonton) C Beaulieu (Edmonton) T Sankar (Edmonton)*

doi: 10.1017/cjn.2016.69

Background: It remains difficult to predict which patients will experience ongoing seizures or neuropsychological deficits following Temporal Lobe Epilepsy (TLE) surgery. MRI allows measurement of brain structures, such as the contralateral (non-resected) hippocampus (cHC) after TLE surgery. Preliminary evidence suggests that the cHC atrophies following surgery, however, the time course of this atrophy, relation to cognitive deficits and seizure outcome remains unclear. *Methods:* T1-weighted MR imaging and hippocampal volumetry in 26 TLE patients pre- and post-TLE surgery (and 12 controls) as: 1) two-scan group (TSG) (pre- and post-operatively at 5.4 years) and 2) longitudinal group (LG; pre- and on post-operatively on day 1,2,3,6,60,120 and at an average 2.4 years. Seizure outcome and