(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)

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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)

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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)

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Background: DEPDC5 gene, mapped to 22q12.2-q12.3, has been associated with a variety of familial epilepsies, including FFE-VF, 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)

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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 paraclinical variables were analyzed against a historical cohort. Binary logistic regression analysis was performed to predict impact of dichotomized