

ively (mean change, standard deviation [SD]: MG Activities of Daily Living score [MG-ADL], -5.3 [4.0] vs -2.1 [2.8]; Quantitative MG score [QMG], -4.1 [6.1] vs -1.3 [3.5]). More patients receiving eculizumab (7/9) had clinically meaningful responses (MG-ADL ≥ 3 and/or QMG ≥ 5 points) than those receiving placebo (3/9). Eculizumab safety was consistent with previous reports. Interim data from the open-label extension of REGAIN will be presented. **Conclusions:** In patients previously receiving chronic IVIg, eculizumab showed a trend toward meaningful clinical improvements and fewer exacerbations compared with placebo. (NCT01997229, NCT02301624).

P.027

Incidence of amyotrophic lateral sclerosis in Newfoundland and Labrador

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Background: There is a paucity of research regarding ALS epidemiology in Canada. Previously published data from Newfoundland and Labrador (NL) demonstrate an average incidence of 2.4/100,000 from 2000-2004 (peak 3.3 in 2001, the highest reported in Canada). Local neurologists believe that the incidence has continued to increase. **Methods:** Clinicians affiliated with the electromyography (EMG) lab at the Health Sciences Centre in St. John's compiled a list of patients diagnosed with ALS from 2012-2016, based on recall. Their medical records were reviewed and demographic information collected. This was cross-referenced with new referrals to the ALS Society NL per year. **Results:** Based on new referrals to ALS Society NL the average incidence between 2012-2016 was 2.81/100,000 (peak 3.6 in 2015). Average age-adjusted incidence from the EMG lab was 1.33 (peak 1.73 in 2016). The EMG lab documented a crude incidence of 3.97 in 2018. **Conclusions:** The incidence of ALS in NL is increased compared to the usual incidence of 1-2/100,000 per year. After the preliminary study, the EMG lab maintained more thorough records and an incidence of 3.97/100,000 was found in 2018. This makes a compelling argument for future research which could explore potential genetic or environmental causes for the increased incidence in this population.

P.028

A milder congenital myopathy in the french Canadians caused by a novel TNNT1 homozygous missense mutation

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Background: Mutations of the slow skeletal muscle troponin-T1 (*TNNT1*) gene are a rare cause of nemaline myopathy. The phenotype is characterized by severe amyotrophy and contractures. Death from respiratory insufficiency occurs in infancy. We report on four French Canadians with a novel congenital *TNNT1*-related myopathy. **Methods:** Patients underwent MRI of leg muscles, quadriceps biopsy and genetic testing. Wild type or mutated human *TNNT1* mRNAs were co-injected with morpholinos in a zebrafish knock-

down model to assess their relative abilities to rescue the morphant phenotype. **Results:** Three adults and one child shared a novel missense homozygous pathogenic variant in the *TNNT1* gene. They developed from childhood slowly progressive limb-girdle weakness with spinal rigidity and contractures. They suffered from restrictive lung disease and recurrent episodes of infection-triggered rhabdomyolysis, which were relieved by dantrolene in one patient. Older patients remained ambulatory into their sixties. MRI of leg muscles showed symmetrical atrophy and fatty infiltration in a proximal-to-distal gradient. Biopsies showed multi-minicores, while nemaline rods were seen in half the patients. Wild type *TNNT1* mRNA rescued the zebrafish morphants but mutant transcripts failed to rescue the morphants. **Conclusions:** This study expands the spectrum of *TNNT1*-related myopathy to include a milder clinical phenotype caused by a functionally-confirmed novel missense mutation.

P.029

Facial onset sensorimotor neuronopathy syndrome (FOSMN) associated with Non-Hodgkin Lymphoma (NHL)

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Background: FOSMN is a recently describe neurological syndrome, characterizes by slow onset of facial sensory abnormalities and motor deficits. The initial description showed a very uniform clinical presentation. Since the initial description there are clinical cases describe in literature with subtle phenotype variations. **Methods:** We describe a clinical case associated with NHL. We will report clinical data, laboratory and neurophysiological findings. **Results:** Patient initiated with left perioral and mental sensory symptoms on her left side. It spread up to include left V2 area and spread to the right side. After 2 years she developed sensory symptoms on her right hand. Progressed to weakness and atrophy on the right upper limb. Also developed dysarthria, dysphonia, dysphagia, as well as photophobia, anisocoria and double vision. Had thorough work-up and everything unrevealing. Except for Spem that showed increased free kappa. Bone marrow biopsy showed evidence of a clonal cell expansion consistent with indolent lymphoma **Conclusions:** This case provides evidence of FOSMN associated with NHL. To our knowledge this is a first case describe with NHL. There had been reports with motor neuro diseases phenotype with lymphoma that may represent a paraneoplastic disorder. Our patient expands the clinical presentation. This finding should not lessen the diagnosis of FOSMN.

P.030

The journey with CIDP- a Canadian perspective

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Background: Chronic Inflammatory demyelinating polyradiculoneuropathy is a rare disorder of the peripheral nerves. A disease affecting up to 8.9 out of 100,000 people, and a yearly incidence of 1.6/100,000 people, CIDP is a condition that is treatable but still relatively unknown outside of the neuromuscular community. The purpose of this research, initiated by the GBS/CIDP Foundation, is

to better understand a patient's journey living with the disease and identify unmet needs. **Methods:** The research consists of a mix of structured interviews, digital ethnography and patient records. A total of 10 Canadian patients living with CIDP and their caregivers, 7 Canadian neurologists and 3 Canadian neuroscience nurses will be the subjects for our research. **Results:** In order to identify key interactions between patients and the healthcare system, the report will map a patient's experience on 4 distinct planes. Clinical journey (ex: first symptoms, diagnosis, disease progression), Patient emotional journey (the emotional states the patient undergoes throughout his/her journey), Caregiver emotional journey, and Outcomes (ex: delays in care, damaged relationships, commitment to therapy). The report will identify key areas along the patient journey where more intervention is possible and where more research may be needed. **Conclusions:** The research is expected to be completed by April 2019.

P.031

Intravenous immunoglobulins (IVIG) therapy in chronic inflammatory demyelinating polyneuropathy (CIDP): time to maximal recovery

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Background: The response of Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) to Intravenous Immunoglobulins (IVIG) treatment is well established. However, determination if patients not responding to 2 IVIG treatments or those whose condition stabilizes (ICE Trial) may benefit from additional doses remains unclear. We aim to identify time period required to reach maximal strength gains from IVIG treatment. **Methods:** Retrospective chart review of 14 patients with CIDP was performed. Change in Grip strength (GS), Knee extension (KE), Elbow Flexion (EF) and Dorsiflexion (DF) was analyzed with a dynamometer during IVIG therapy. Averages for: percent change from baseline (Max%Δ), cumulative grams (g) of IVIG and time in weeks (w) required for maximal strength recovery was determined per function (+/-SEM). Ancillary therapy for all patients was recorded. **Results:** Strongest improvement was observed for DF (124+/-30%, p<0.001), followed by KE (113+/-19%, p<0.01), GS (100+/-21%, p<0.001) and EF (98+/-14%, p<0.05). GS improved the fastest (19.1+/-3w) followed by DF (29.5+/-7w), KE (29.6+/-4w) and EF (31+/-6w). Cumulative IVIG dose to reach Max%Δ was highest for EF (869+/-201g) and lowest for GS (573+/-78g). **Conclusions:** Our study has demonstrated effectiveness of multiple treatments with IVIG to reach significant improvement in strength. Different muscle groups manifested different time-dependency, reflecting variable amounts of IVIG required. Improvement was identified to be present on an ongoing basis, with therapy lasting between 19.1-31 weeks, requiring between 869-573g of IVIG.

NEUROSCIENCE EDUCATION

P.032

What do elective students learn about the specialty of Neurology (and what can that teach us)?

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Background: "Neurophobia" describes a fear of Neurology on the part of medical students. This contrasts with the "neurophilia" that exists in society with increasing awareness of disorders such as stroke and multiple sclerosis. Ideally, we should take advantage of "neurophilia" to promote our specialty's strengths. One step would be to better understand what students learn from a Neurology elective. **Methods:** This was a qualitative study. Students completing an elective between September 2011 and March 2015 at the Jewish General Hospital (JGH) in Montreal completed written pre- and post-elective questionnaires. **Results:** 36 students participated; 15 from McGill, 11 from other Canadian medical schools, and 10 from International medical schools. Many students changed their opinion about Neurology, with fewer citing lack of treatments or poor patient prognoses as negatives after completing their elective. They valued knowledge acquired about the neurological exam and problem-solving, while the range of cases and subspecialties surprised them. Many would diversify the setting of their elective to better experience this variety. **Conclusions:** More diversified elective experiences could showcase the strengths of our specialty and the scope of neurological practice. Presenting Neurology as a challenging, intellectually stimulating specialty that emphasizes problem solving could increase student interest.

NEUROVASCULAR, STROKE AND NEUROINTERVENTIONAL

P.033

Awareness and knowledge of stroke and heart disease: a follow-up study of the Chinese-Canadian cardiovascular health project

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Background: This is an updated on-line survey of the awareness and knowledge of stroke and heart disease amongst Chinese-Canadians carried out in 2017. **Methods:** 1001 randomly selected Chinese-Canadians from Toronto and Vancouver area. **Results:** 46% were > 45 years old and male to female ratio was: 49.3 : 50.8, with native language being Cantonese in 40%, Mandarin 24% and 31% English. 82% were Canadian citizens and 31% had been in Canada < 10 years. 44% were from Mainland China, 37% Hong Kong, 6% Taiwan and 12% were borned in Canada. 85% were able to name at least one symptom of heart attack (p=0.005) while 80% were able to name at least one symptom of stroke (p=0.0008). 85% would call 911 in response to symptoms of heart attack or stroke compared to only