a 19 yo male with idiopathic LETM remained quadraparetic and ventilator dependent with active MRIs despite multiple courses of intravenous methylprednisolone, plasma exchanges, and in the NMOSD patient, IVIg and a 4-week course of rituximab. Both patients ultimately improved significantly and are now ambulatory with subsequent cyclophosphamide induction. *Conclusions:* In patients with severe LETM of presumed immune origin, who fail to respond to corticosteroids and plasma exchange, cyclophosphamide induction should be considered. This agent provides a more robust immunosuppressive response and can be induced rapidly. Cyclophosphamide effects and supportive evidence are further discussed.

P.050

Autoimmune encephalitis associated with GAD65 antibodies: brief review of the relevant literature

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Background: Recently, many cases of autoimmune encephalitis with positive GAD65 (*Glutamic acid decarboxylase*) antibodies have been described in the literature. However, it remains an understudied topic. *Methods:* We conducted a search on reported cases of anti-GAD65 encephalitis. Specific variables were identified as general characteristics, clinical manifestations, MRI and EEG findings, concomitant systemic autoimmune disorders and cancer, and outcome and autoantibodies findings. *Results:* We identified a total of 58 cases, from one to 70 years old. It most frequently presented with seizures (97%) and memory impairment (59%). It commonly occurred in association with systemic autoimmune disease, particularly diabetes (28%). Brain MRI was usually abnormal (78%); involvement of temporal lobes was more frequent than multifocal abnormalities (59% vs 16%). GAD65 antibodies were reported positive in CSF and/or serum (31% in serum only). Other antibodies such as GABABR, GABAAR and VGKC were concurrently reported positive in some cases (19%). However, we found that the vast majority of cases were not tested for all those cell-surface antibodies. Overall, no distinctive pattern of clinical and paraclinical findings was found. Persistent impairments were common. Optimal treatment remained undefined. *Conclusions:* Prospective studies recruiting patients with autoimmune encephalitis are needed to better elucidate the contributions of GAD65 autoantibodies, and to evaluate treatment and outcomes in this population.

P.051

Patient-reported adverse events on Multiple Sclerosis disease-modifying therapies in an urban tertiary MS clinic

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Background: Disease-modifying therapies (DMT) have been shown to reduce relapses and delay disability in individuals with relapsing-remitting multiple sclerosis (MS). However, these medications can cause adverse events (AE) leading to poor adherence. To better understand their clinical utility, this study examined real-life experiences with DMT in a tertiary MS clinic. *Methods:* A retrospective chart review (1999-2015) was conducted to evaluate the prevalence of AE and discontinuation rates of Health Canada approved DMT. *Results:* 445 MS patients who have used at least one DMT in their lifetime were reviewed. Among first-line injectable therapies, interferon beta (IFNβ) 1-α IM users (49.6%) were most likely to report an AE. Flu-like reactions and injection site reactions were the most commonly reported AE. Among first-line oral therapies, BG-12 users (58.5%) were most likely to report an AE. The most common AE were flushing and gastrointestinal upset. DMT that were most frequently discontinued as a result of AE were IFNβ 1-α SC (39.3%), IFNβ 1-α IM (36.8%) and BG-12 (34.6%). *Conclusions:* The prevalence of AE and discontinuation rate were congruent. In comparison with recent literature, this study demonstrated lower prevalence of AE but equivocal or higher discontinuation rates. This discrepancy could represent a more realistic depiction of the impact that DMT AE have on patients.

P.052

Late-onset adrenoleukodystrophy mimicking primary progressive multiple sclerosis

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Background: Adrenoleukodystrophy (ALD) is a peroxisomal disorder that leads to the accumulation of very long chain fatty acids in the body. Younger males typically present with a catastrophic cerebral demyelinating disease, while adult males tend to develop a progressive myelopathy and neuropathy. *Methods:* Case presentations and literature review. *Results: Case1:* A 58-year-old male with a three-year history of unsteady gait. His MRI showed multiple T2-hyperintensities most prominently in the posterior corpus collosum (which progressed over time) as well as spinal cord atrophy. Primary progressive multiple sclerosis (PPMS) was suspected. *Case 2:* The patient's bother, a 49-year-old, had a ten-year history of difficulty walking. MRI findings included a single large T2 hyper-intensity spanning the anterior aspect of the corpus collosum and an atrophic spinal cord. Given the family history, both brothers were investigated for and diagnosed with ALD. *Conclusions:* Both cases are of males presenting with a progressive myelopathy in middle age. In the first case, the history, physical exam, and imaging findings were most consistent with PPMS. However, the second case was less typical for MS prompting further investigations. These cases highlight the need to have a broad differential when confronted with atypical cases of MS and reminds the clinician of the phenotypic variability of ADL.

Neurocritical Care/ Neuro Trauma

P.054

The use of robotic technology to define post-operative neurological dysfunction in patients undergoing coronary bypass surgery: a feasibility study

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Background: Cognitive dysfunction following coronary artery bypass surgery is a regular occurrence, but its cause is still unknown.

In order to devise strategies to mitigate this acquired disability, a precise and quantitative description of the post-operative neurocognitive phenotype is necessary. This study is designed to assess the feasibility of using the KINARM robot to quantify the changes in the neurological function after cardiac surgery. *Methods:* Patients without prior history of cognitive dysfunction were recruited from the pre-operative cardiac surgery clinic, and underwent pre-operative assessment with the KINARM. The KINARM provides a quantitative assessment of the neurocognitive control of the upper limbs. During bypass surgery, brain tissue oxygen levels were measured with near-infrared spectroscopy. Patients were reassessed with the KINARM post-operatively at 3 months. *Results:* To date, 12 participants have been recruited (mean age = 65 years, all male). On straightforward tasks, such as visually guided reaching, the majority of patients scored within the normal range, both pre- and post-operatively. In more complex tasks, required visuospatial and executive functioning, post-operative deficits were more pronounced. *Conclusions:* It is feasible to use the KINARM robot to provide a quantitative measurement of the neurocognitive phenotype of patients after cardiac surgery.

Neuromuscular

P.055

Bilateral facial nerve gadolinium enhancement and GBS

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Background: Facial diplegia with parasthesia (FDP) is a rare variant of Guillain–Barre syndrome (GBS), and has been reported in less than 1% of GBS cohorts. Here we describe a case of FDP with novel imaging findings and discuss the differential diagnosis. *Methods:* Case: A 39-year-old man referred to the emergency department with a 2 week history of right facial palsy progressed to bilateral facial palsy. His exam demonstrated severe, complete facial diplegia with only very mild limb weakness and present but diminished deep tendon reflexes. *Results:* Cerebrospinal fluid analysis showed albuminocytologic dissociation. Electromyography was consistent with a demyelinating process. MRI with contrast revealed bilateral enhancement of the facial nerves in the intracanalicular portion and in the region of the geniculate ganglion. A diagnosis of GBS was made and the patient was treated with IVIG. Over the course of several weeks the patient improved. *Conclusions:* Although nerve root enhancement of the spinal cord is described with GBS, nerve root enhancement effecting cranial nerves has only rarely been described. In addition, the relative limb-sparing with complete facial paralysis in this case is also an unusual phenotype. The Gadolinium enhancement of the bilateral facial nerves is thought to represent blood brain barrier breakdown due to GBS.

P.056

Survey of Canadian myotonic dystrophy patients' access to computer technology

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Background: Myotonic dystrophy is an autosomal dominant condition affecting distal hand strength, energy and cognition. There is a neuromuscular patient portal under development that has the potential to give voices and resource access to patients, regardless of location via the internet and social media. The current state of access to technology and underlying factors affecting use and interest were explored. *Methods:* Surveys were mailed to 156 participants with myotonic dystrophy type 1 (DM1) through the Canadian Neuromuscular Disease Registry. The survey questions touched on demographics, technology use, internet use, and interest in the portal. *Results:* Seventy-two participants (43 female) responded so far and 50% were younger than 46 years. Most (62/72) used the internet and 94% of participants had access to an internet-connected device. Almost half of the responders (34/72) used social media. The complexity and cost of technology were commonly cited reasons not to use technology. The majority of responders (79%) were interested in a myotonic dystrophy patient portal. *Conclusions:* DM1 patients have access to and use technology such as computers and mobile phones. They expressed interest in a portal that would provide them with access to relevant information such as guidelines, self-management modules, educational videos, and support groups.

P.057

Distal hereditary motor neuropathy type I due to the GARS: c.1415A>G, p.His472Arg mutation

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Background: The distal hereditary motor neuropathies (dHMN) have been characterized through case reports and family studies. Their genetic characterization remains a work in progress. An appreciation of the genetic underpinnings may lead to treatment options in the future. Hence reports, like this one, which add to this understanding, remain extremely important. *Methods:* The clinical presentations, electrophysiology and genetics of two patients with the dHMN I phenotype are described. *Results:* A mother and son presented with slowly progressive distal weakness of the lower extremities with onset in the first and second decades. Distal weakness of the upper extremities developed later. Examination 20 and 50 years after onset revealed wasting and weakness of distal upper and lower extremity muscles with absent distal and preserved proximal deep tendon reflexes. Sensory examination was normal. Electrodiagnostic studies demonstrated a motor axonopathy. Genetics testing revealed a missense mutation on chromosome 7, exon 11 of the GARS gene: c.1415A>G (p.His472Arg). *Conclusions:* GARS mutations have been identified in patients with the dHMN I (juvenile onset distal weakness and wasting) and dHMN V (upper limb predominant) phenotypes. However, this mutation has not previously been directly linked to the dHMN I phenotype.