use to make this decision. *Results:* Responses from 10 of 17 clinics were received. One clinic had written guidelines. Most used decline in respiratory function, dysphagia, weight loss or some combination of all three. Six clinics reported dropping FVC, ranging from 70% to 50% as prompting tube insertion. Five clinics reported weight loss as part of their criteria. Dysphagia was reported as the most important factor by 7 clinics. Psychological readiness for tube placement was a key factor in 3 clinics. Some clinics comment they place tubes in advance of dysphagia. *Conclusion:* Criteria for tube insertion varies between clinics. Practices generally reflect published recommendations, but vary on the emphasis of specific criteria. The lack of strong scientific evidence to guide decisions may contribute to management variability. Further study is needed to guide practice.

P.048

Auto-antibodies against gangliosides in patients with Charcot-Marie-Tooth disease

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Background: In Charcot-Marie Tooth (CMT), vital components of either the myelin sheath or axon are abnormal, slowing nerve conduction and causing functional disability. Recently, there has been speculation the CMT may have an autoimmune component resulting from abnormal protein expression. *Methods:* Custom autoimmune neuropathy-focused microarray panels were printed in-house using antigens from Sigma, Abnova, Fitzgerald and Matreya according to Cambridge Life Science instructions. Antigens including Myelin Protein Zero, Peripheral Myelin Protein 22, and 20 other well-known gangliosides were tested for IgM and IgG antibodies. Students T-Test and Bonferroni correction factor were used to determine statistical significance between groups and in post-hoc subgroup analysis. *Results:* Plasma was tested from 17 patients with CMT and 25 young healthy individuals. CMT population consisted of 9 CMT-1a, 1 CMT-1b, 4 CMT-2a, 2 CMT-2f and 2 undetermined CMT type 2 patients. No ganglioside reached statistical significance under a Bonferroni Correction factor ($p>0.01$) nor were any gangliosides notably raised compared to normal. Sub-group analysis did not reveal theorized peak auto-antibodies levels depending upon their CMT subtype compared to normal. *Conclusions:* Although previously shown to have increased auto-antibodies to myelin or axonal proteins, CMT individuals did not demonstrate increased autoantibody levels for the proteins tested at our centre.

P.049

Intravenous Immunoglobulins (IVIG) 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 whether patients who do not respond to 2 IVIG treatments or those whose condition stabilizes (ICE Trial) would benefit from additional treatments remains unclear. We aim to identify time period required to reach maximal strength gains from IVIG treatment (plateau). Furthermore, we will assess nerve conduction studies (NCS) changes over time with IVIG treatment. This will help in establishing a time course for treatment of CIDP with IVIG to maximize recovery. *Methods:* We performed a retrospective chart review of 27 patients with CIDP, with diagnosis confirmed by European Federation of Neurological Societies/Peripheral Nerve Society Guidelines (EFNS/PNS). Each patient's strength response including: grip strength, knee extension, elbow flexion and dorsiflexion (using JAMAR Dynamometer) and NCS changes over time during IVIG treatment were analyzed. *The primary outcome* is duration of IVIG treatment, in months, required to reach a plateau in strength. *Secondary outcome* is NCS change including: Terminal Latencies, Conduction Velocities, Compound Sensory and Motor action potentials in nerves of upper and lower extremities over treatment time (emerging trends). *Results:* Pending (available by April 2015) *Conclusion:* Pending (available by April 2015)

P.050

Acute combined central and peripheral demyelination: a case report

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Aim/Background: We report a case of Acute Combined Central and Peripheral Demyelination (ACCPD). This rare disease presents with features of both peripheral and central demyelination. *Methods:* Case Report *Results:* A 24 year-old Iraqi female presented with acute onset of ascending paralysis, numbness and areflexia over the course of a few days. Systemic examination was negative. She responded to IVIG. She suffered two severe relapses over the next three months, which resolved rapidly with PLEX/corticosteroid. CSF was normal after first and second relapses. Brain and cord MRI revealed multiple T2/FLAIR hyperintensities consistent with multiple sclerosis. There were no longitudinally extensive cord lesions. Aquaporin 4 antibody assay is pending. ANA was strongly positive; anti-DSDNA and SS antibodies were negative; complement4 was low and serum cyroglobulins were positive. Hepatitis C was negative. Ganglioside antibody assay was negative. Anti-neurofascin is pending. Neurophysiology confirmed features of an acquired demyelinating neuropathy with profound secondary axonal denervation. *Conclusions:* The underlying etiology of ACCPD is presumed autoimmune likely secondary to auto-antibody targeting of central and peripheral myelin epitopes. Low complement component 4 and cryoglobulinemia in this patient supports an autoimmune pathogenesis. Neurofascin has been previously reported as one such auto-antibody in ACCPD.

P.051

Geographic distribution of Multiple Sclerosis (MS) mortality rates in Canada, 1975-2009

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Background: Our study examined whether there are differences in MS mortality rates across regions of Canada, which might

suggest differences in environment or health care practice that influence outcome. *Methods:* Statistics Canada data on deaths due to MS and populations at risk, 1975-2009, were derived from the Research Data Centre, University of Alberta. Mortality rates and 95% confidence intervals (CIs) were calculated per 100,000 population for the Atlantic Provinces, Quebec, Ontario and Western Provinces (including Northwest Territories, Yukon, Nunavut), age-standardized to the 2006 population. *Results:* The average annual MS mortality rates for 1975-2009 per 100,000 population (CIs) were: Atlantic Provinces 1.09 (0.43,1.74); Quebec 1.30 (0.89,1.71); Ontario 1.08 (0.77,1.38); Western Provinces 1.39 (0.99,1.78). Female mortality rates were consistently higher than male rates but there were no differences in the female:male mortality rate ratios across regions. Trend analysis showed that rates were stable over the 35 year time span in 3 regions with non-significant average annual per cent increases/decreases of: Atlantic Provinces – 0.43%; Quebec + 0.12%; and Western Provinces + 0.27%. Only Ontario showed a slight but significant increase of + 0.81% ($p<0.05$). *Conclusions:* MS mortality rates are similar across the Canadian regions, suggesting that patients are not disadvantaged in terms of mortality by their place of residence.

P.052

Novel VCP mutation associated with CMT2 phenotype

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Background: Charcot-Marie-Tooth (CMT) neuropathy is an increasingly polygenic disorder which limits clinical diagnoses due to phenotypic overlap resulting from the common final pathway of length-dependent axonal degeneration. The proband exhibited features of an axonal neuropathy manifesting upper and lower extremity distal amyotrophy, mild sensory loss, absent upper motor neuron findings, and mild hyperCKemia. *Methods:* Whole-exome sequencing (WES) was performed after failure to identify the familial variant with sequencing of multiple discrete CMT2 genes. *Results:* A novel VCP mutation (c.511A>C;p.Ser171Arg) was identified. This mutation segregated with an affected brother and father. All manifested a CMT2 phenotype with motor predominance. Further investigation of the proband revealed 1) a muscle biopsy with no inclusions or myopathic changes, 2) a skeletal survey negative for lucencies and hyerostoses, and 3) normal cognitive function. *Conclusions:* Mutations in valosin-containing protein (VCP) are associated with distinct neurologic phenotypes including 1) inclusion body myopathy, Paget disease, and frontotemporal dementia, 2) hereditary spastic paraplegia, and 3) amyotrophic lateral sclerosis. Recently, CMT2 has been described as a VCP-associated phenotype. This is the second report of a peripheral neuropathic phenotype resulting from a mutation in the VCP gene. The clinical presentation in this family suggests a unique clinical-biochemical phenotype consisting of motor-predominant CMT2 with mild hyperCKemia.

P.053

The use of standardized order sets to optimize treatment for Guillain-Barre Syndrome: a literature review

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Background: Standardized order sets are thought to improve patient outcomes in multiple ways. They reduce costs without reducing quality of care, and improve efficiency. In both surgical and medical conditions patients benefit from order sets in various disease states. In Guillain-Barre syndrome (GBS), the use of standardized order sets may be beneficial as there are a defined set of disease-specific diagnostic tests and treatments to be implemented. Here, the primary aim was to search for, and evaluate standardized order sets for GBS, and to provide a basis for development of future pathways. *Methods:* We used the Cochrane, TRIP, and MEDLINE/PUBMED databases, searching between January 1966 and April 2014. Search terms included: "Guillain-Barre Syndrome" and its synonyms, "(standardized) order set", "clinical pathway", "neurology" and "admission bundle." *Results:* Despite anecdotal evidence of order sets, no formal data has been published showing benefit after implementation of these sets in GBS or any neurological condition. *Conclusions:* Although evidence exists for use of standardized order sets in surgical and medical settings, no published data exist in neurology. Given GBS has a defined set of disease-specific and state-specific treatment options, a standardized order set used on admission for GBS patients may prove to be beneficial.

P.054

Seropositive PERM associated with leucocytoclastic vasculitis

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Background: Progressive encephalomyelitis, rigidity, and myoclonus (PERM) is a variant stiff-person plus syndrome consisting of brainstem and pyrimidal dysfunction, muscular rigidity, stimulus-evoked spasms, and dysautonomia. Continuous motor-unit activity from ectopic anterior horn cell discharges underlies the myotomal hyperactivity. *Case Report:* A 51-year-old man with an 8-year history of "spasmodic" mid and low back pain presented with increasing stiffness, hyperekplexia-induced opisthotonus-like posturing, urinary retention, long-tract motor signs, and diplopia. He had a recent history of biopsy-confirmed leucocytoclastic vasculitis-associated diffuse maculopapular rash. He responded well to IVIg treatment manifested by improved 1) gait fluidity, 2) bowel and bladder function 3) tolerance to startle, and 4) vision. *Results:* Serological testing revealed positive anti-glycine receptor antibodies. Anti-glutamic acid decarboxylase and voltage-gated K+-channel antibodies were absent. A chest CT was unremarkable. *Conclusions:* This is the second case of seropositive PERM in Canada and the first associated with leucocytoclastic vasculitis. Pathologic findings in PERM reveal perivascular lymphocytic cuffing in the rhombencephalon and spinal cord. Glycine receptor localization correlates with neurologic dysfunction. Mid-brain involvement may underlie the autonomic dysfunction