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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doi: 10.1017/cjn.2015.162

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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doi: 10.1017/cjn.2015.163

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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doi: 10.1017/cjn.2015.164

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

https://doi.org/10.1017/cjn.2015.162 Published online by Cambridge University Press