

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

BK Steven (Hamilton)*

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 hyperostoses, 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

V Karnik (Edmonton) T Roberts (Edmonton) W Johnston (Edmonton)*

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

BK Steven (Hamilton)*

doi: 10.1017/cjn.2015.164

Background: Progressive encephalomyelitis, rigidity, and myoclonus (PERM) is a variant stiff-person plus syndrome consisting of brainstem and pyramidal 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

but evidence of direct glycinergic inhibition of the external urethral sphincter in Onuf's nucleus may be responsible for urinary retention.

NEUROLOGY (STROKE)

P.055

Distinguishable distribution of cerebral artery stenoses: ultrasonographic evidence from a northeast Chinese cohort

H Liu (Changchun) X Wu (Changchun) Y Xing (Changchun) K Liu (Changchun) H Zhang (Changchun)*

doi: 10.1017/cjn.2015.165

Background: Cerebral artery stenosis is an important risk factor for ischemic strokes. This study aims to explore intracranial and extracranial artery stenosis in a large northeast Chinese cohort. **Methods:** We recruited 14793 outpatients and hospitalized patients to identify cerebral artery stenosis. Artery stenosis screening was done with transcranial Doppler (TCD) for intracranial arteries and carotid duplex sonography for extracranial arteries. **Results:** More intracranial than extracranial artery stenoses were identified (4255 versus 2809, i.e. 28.8% versus 19.0%, $P < 0.05$). Similarly, more intracranial stenosis was significantly more common than extracranial artery stenosis in this population (2632 versus 1186, i.e. 17.8% versus 8%, $P < 0.05$). Among all identified intracranial arteries stenoses, the proportion of middle cerebral artery (MCA) stenosis was the highest. More intracranial than extracranial artery stenoses was seen within each age group, and rates of both increased with age. Intracranial and extracranial artery stenosis was more frequently identified in males than females. **Conclusions:** Incidence of cerebral artery stenosis in the population increases with age. Intracranial artery stenosis is more common than extracranial artery stenosis and the MCA stenosis accounted for the highest proportion, within each age group. More males suffer from intracranial or extracranial artery stenosis than females.

P.056

Noninvasive assessment of ischemic penumbra by using MR-SWI during the acute phase of cerebral infarction: a comparison to PWI

S Luo (Changchun) X Wu (Changchun) F Deng (Changchun) Y Zhang (Changchun) J Miao (Changchun) Y Chen (Changchun) L Wang (Changchun) J Feng (Changchun) H Zhang (Changchun)*

doi: 10.1017/cjn.2015.166

Background: Assessment of ischemic penumbra during the acute stage of cerebral infarction is crucial for a decision to initiate thrombolytic therapy and for predicting stroke evolution. Although controversial as a perfect equivalence to penumbra, perfusion weighted imaging (PWI)-diffusion weighted imaging (DWI) mismatch may predict the response to thrombolysis. Due to the reliance on contrast agents in PWI, noninvasive alternatives remain an unmet need. **Methods:** We herein investigate the potentials of SWI as an alternative to PWI in defining ischemic penumbra and in predicting

stroke outcome. A multimodal magnetic resonance imaging work-up which includes conventional magnetic resonance imaging sequences (T1WI, T2WI and FLAIR), DWI, PWI and SWI was performed. The Alberta Stroke Programme Early CT Score (ASPECTS) was used to evaluate the changes in DWI, SWI and PWI. **Results:** The mismatch of SWI-DWI was comparable with that of PWI-DWI ($p > 0.05$). Furthermore, the grade of prominent vein and the cerebral blood volume in the ipsilateral brain tissue were positively correlated. **Conclusions:** SWI can be used as a noninvasive alternative to identify occlusive arteries and to evaluate the ischemic penumbra. The susceptibility vein sign may represent thrombosis in arteries whereby being helpful to identify responsible blood vessels in ischemic stroke.

P.057

Different strokes for different folks: epidemiology of cerebrovascular diseases amongst Chinese-Canadians residing in Toronto

JY Chu (Toronto)* JK Chu (Atlanta) DK Chu (Hamilton) S Lam (Toronto)

doi: 10.1017/cjn.2015.167

Introduction: It has been recognized in the past few decades that different ethnic groups living in Canada may have different stroke epidemiology. This presentation is focused on the stroke patterns of Chinese-Canadians living in the Toronto area. **Methods:** Two retrospective case-controlled studies were carried out between 1990-2000 to study the stroke characteristics of Chinese-Canadians living in Toronto. Statistical analysis was carried out by the Institute of Clinical Evaluative Sciences. A further retrospective study was also carried out in 2011 to look at the relationship between stroke and diabetes mellitus amongst this population. **Results:** Chinese-Canadians were found to have 1/6 the prevalence of extracranial vascular stenosis. They have a higher frequency of intracranial vascular disease which may be due to the higher frequency of hypertension and diabetes mellitus. Higher incidence of intracranial hemorrhage was found compared to Caucasian controls which may be due to the lack of awareness and optimal treatment of their hypertension. Details of the results of these three studies will be presented. **Conclusions:** This is the first long term retrospective study of the stroke patterns and epidemiology for Chinese-Canadians residing in Toronto. Further prospective population-based study will be vital to study the important interactions between genetics and environment in the pathogenesis of different strokes for different folks.

P.058

Bilateral thalamic infarction due to artery of percheron occlusion with corresponding CT perfusion abnormalities

A MacLellan (Toronto)* K Boyle (Toronto)

doi: 10.1017/cjn.2015.168

Background: The Artery of Percheron (AOP) is a rare anatomic variant that supplies the bilateral medial thalami from a common origin; occlusion results in a characteristic pattern of bilateral thalamic infarction seen on neuroimaging. To date, we have not identified any cases in the literature describing corresponding ischemic changes seen on hyperacute CT Perfusion imaging. We aimed to characterize perfusion abnormalities seen in AOP occlusion by describing a case