CNS / C SCN Chair’s Select Abstracts

B.01
CNS Francis McNaughton Memorial Prize
Predictors of dysphagia screening after acute ischemic stroke: Who gets tested?
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Background: Dysphagia is a devastating complication of stroke and can lead to malnutrition, immobility, aspiration pneumonia, and death. Guidelines advocate screening all patients with acute stroke for swallowing impairment. However, previous research suggests only 60% are screened, and it is unclear what factors contribute to receiving dysphagia screening. Methods: We used the Ontario Stroke Registry to identify patients who were admitted to Regional Stroke Centres from 2010-2013. We used multivariable regression to identify predictors of receiving a dysphagia screen within 72 hours. Results: Among 7172 patients with acute ischemic stroke, 1705 patients (23.8%) did not undergo screening. Factors increasing the odds of being tested were: Stroke unit admission (adjusted odds ratio aOR 6.5), presenting with speech deficits (aOR 1.9) or weakness (aOR 1.5), or receiving thrombolysis (aOR 1.9). Seizure (aOR 0.49) and mild stroke (aOR 0.59 vs moderate stroke) decreased the odds of being tested. Among those with mild strokes who received a swallowing screen, 33% failed. *All p<0.0001. Conclusions: Patients with mild stroke are at risk of not being screened for dysphagia, despite a significant fail rate among those tested. This may expose untested patients to a higher risk of complications from dysphagia, and suggests a gap in process of care that should be addressed.

B.02
CSCN Herbert Jasper Prize
Burst-suppression EEG is reactive to photic stimulation in comatose children with acquired brain injury
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Background: Burst-suppression is an electroencephalographic pattern observed during coma and reflects severe encephalopathy. We investigated the reactivity of burst-suppression to photic stimulation in children with acquired brain injury. Methods: Intensive care unit electroencephalographic monitoring recordings containing burst-suppression were obtained from 5 comatose children with acquired brain injury of various etiologies. Intermittent photic stimulation was performed at 1 Hz for 1 minute to assess reactivity. We quantified reactivity by measuring the change in the burst ratio (fraction of time in burst) following photic stimulation. Results: Photic stimulation evoked bursts in all patients, resulting in a transient increase in the burst ratio, while the mean heart rate remained unchanged. The regression slope of the change in burst ratio, referred to as the standardized burst ratio reactivity, correlated with subjects’ Glasgow Coma Scale scores. Conclusions: Reactivity of the burst-suppression pattern to photic stimulation occurs across diverse coma etiologies. Standardized burst ratio reactivity appears to reflect coma severity. Measurement of burst ratio reactivity may represent a simple bedside tool to monitor coma severity in critically ill children.

B.03
The Canadian neurology graduate survey
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Background: Planning for neurology training necessitated a reflection on the experience of graduates. We explored practice characteristics, and training experience of recent graduates. Methods: Graduates from 2010-2014 completed a survey. Results: Response rate was 37% of 211. 56% were female. 91% were adult neurologists. 65% practiced in an outpatient setting. 63% worked in academics. 85% completed subspeciality training (median 1 year). 36% worked 3 days a week or less. 82% took general call (median 1 night weekly). Role preparation was considered very good or excellent for most; however poor or fair ratings were 17% in advocacy and 8% in leadership. Training feedback was at least “good” for 87%. Burnout a few times a week or more was noted by 5% (6% during residency, particularly PGY1 and 5). 64% felt overly burdened by paperwork. Although most felt training was adequate, it was poor or fair at preparing for practice management (85%) and personal balance.
(55%). Most conditions were under-observed in training environment. Many noted a need for more independent practice development and community neurology. Conclusions: Although our training was found to be very good, some identified needs included advocacy training, and more training in general neurology in the longitudinal outpatient/community settings.

B.04
Distal and asymmetric myasthenia gravis: a case series of 54 patients
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doi: 10.1017/cjn.2016.63

Background: Distal/asymmetric presentations of myasthenia gravis (MG) are uncommon and occur in 3-7% of patients with MG. This pattern of weakness is often not recognized as a manifestation of MG, leading to inappropriate investigations, delayed diagnosis and potentially missed opportunities for treatment. Our knowledge about this atypical presentation is limited to small case series and individual case reports. This study therefore aims to expand our understanding by describing the clinical course, diagnosis and treatment of a larger series of patients with this presentation. Methods: We conducted a retrospective chart review of patients with definite MG (either acetylcholine receptor [AChR] or MuSK antibody positive or clear evidence of postsynaptic neuromuscular junction dysfunction on electrodiagnostic studies), who attended the MG Clinic in London. Details of the clinical course, electrodiagnostic studies, antibody testing and response to treatment are reported. Results: 5.9% (54/921) of patients with definite MG had distal/asymmetric limb involvement, 56% at onset and 4% developing more than 10 years later. Males predominated (2:1). Finger extensors were most affected. 83% were AChR antibody positive. 7% had thymomas. On repetitive nerve stimulation most patients showed the most significant decrement distally on the more affected side. Almost all patients improved with treatment. Conclusions: This study expands our understanding of distal/asymmetric presentations of MG.

B.05
Optimizing IVIg utilization for neuromuscular disease in BC: high user project
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Background: In British Columbia, neuromuscular disease accounts for 31% of IVIg use, at a cost of $10.1 M. In addition to the new screening pathway, the BC Neuromuscular IVIg Program developed the Chronic High User Project to identify areas for improvement in utilization. Methods: Utilizing CTR data, all patients on IVIg maintenance therapy for approved neuromuscular conditions between April 1, 2013 and March 31, 2014 were identified. Patients receiving higher than usual IVIg treatments (CIDP and MG >1110 grams/year, MMNCB > 1400 grams/year) were evaluated. Following panel review, utilization data was compared with a second cohort (2014 to 2015) to determine impact. Following review, appropriateness of treatment was determined by consensus from a 3-member panel, and recommendations were made. Results: Of 377 patients, 38 “High Users” were identified. 29 cases were determined to be appropriate; 9 were not. There was a reduction in mean grams/episode in CIDP (1135g to 990g) and MG (1099 g to 1022g) between cohorts. The mean grams/episode for MMNCB did not change. Conclusions: In specific cases, the IVIg High User Program identified patients in whom the treatment could be optimized. However, the vast majority of use of IVIg for Neuromuscular Disease in BC is appropriate, including in patients requiring higher that “usual” doses.

B.06
CNS André Barbeau Memorial Prize
Two definite sudden unexpected deaths in epilepsy in a family with a DEPDC5 mutation
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Background: DEPDC5 gene, mapped to 22q12.2-q12.3, has been associated with a variety of familial epilepsies, including FEFV, autosomal dominant nocturnal frontal lobe epilepsy, and familial TLE. Notably, DEPDC5 has never been linked to increased risk of sudden unexpected death in epilepsy (SUDEP). Methods: Cases review. Results: We studied a three-generation, non-consanguineous, French-Canadian family with nine clinically affected individuals. The index case is a 39-year-old man who started having seizures (as 2rily GTCs) at the age of 13 years. EEGs showed interictal discharges over the right anterior-temporal region. Brain MRI was unremarkable. Two individuals in this family suffered definite autopsy-confirmed SUDEP, at the ages of 58 and 50 years, respectively. Overall, seizure-history in this family can be summarized by an onset before reaching adulthood followed by subsequent progressive decrease in seizure frequency. Seizures were predominantly nocturnal 2rily GTC. Genetic analysis revealed a pathogenic heterozygous variant in the DEPDC5 gene (p.Gln216, c.646C>T), which results in a premature stop codon, in all affected family members plus on healthy relative. Importantly, all the subjects were cognitively intact, and there was no history of cardiac symptomatology/cardiovascular risk factor. Conclusions: The finding in this family suggests that DEPDC5 mutations may be a risk factor for SUDEP.

B.07
Evaluating the single seizure clinic model: findings from a Canadian centre
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Background: The effect of the single seizure clinic (SSC) model on patient diagnosis, work-up, wait-times, and clinical care is poorly characterized. This study assesses patient characteristics and evaluates the impact of a SSC model on wait-times and access to care. Methods: A prospective study of all patients (n=200) referred to our SSC for first-seizure evaluation. Demographic, clinical, and paraclinical variables were analyzed against a historical cohort. Binary logistic regression analysis was performed to predict impact of dichotomized