D.04

Validating StatNet EEG as a reliable and effective tool in the Diagnosis of Non-Convulsive Status Epilepticus after hours

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Background: NCSE dramatically increases morbidity and mortality. It is clinically subtle, which makes it difficult to diagnose without EEG. StatNet EEG provides a quick alternative to conventional EEG, which is often unavailable after hours. *Methods:* Each patient received a StatNet EEG by a neurology resident and a conventional EEG by an EEG technologist. Blinded studies were compared for delay between the studies, setup time, artifact, and abnormality detection using conventional EEG as controls. Nonparametric Mann-Whitney two-sample T-test was used. Results are expressed in mean minutes +/- SD. Kappa score assessed inter-observer reliability. *Results:* 19 patients were collected. Two StatNet EEGs were not interpretable and excluded. The mean delay between studies was 26h53min. Electrode placement is significantly shorter: 13:14±5:24 StatNet EEG vs 18:07±5:35 conventional EEG (p=0.02).

Table 1.

Results: Interobserver Reliability

	Conventional Kappa (SD) N=19	**StatNet Kappa (SD) N=17 *2 uninterpretable**
Abnormality	0.73 (0.18) CI: 0.37-1.0	0.54 (0.18) CI: 0.19-0.89
Epileptiform Discharges	0.76 (0.16) CI: 0.43-1.0	0.76 (0.22) CI: 0.33-1
NCSE	0.64 (0.32) CI: 0.0031-1.0	1.00 (0) CI: 1.00-1.00
Seizures	1.00 (0) CI 1.00-1.00	1.00 (0) CI: 1.00-1.00
Slowing	0.36 (0.25) CI: 0.14-0.86	0.45 (0.17) CI: 0.11-0.79

Conclusions: There is high inter-rater reliability between the conventional and StatNet EEG groups (table 1) demonstrating that StatNet is a reliable and effective tool, aiding in early recognition and management of NCSE.

D.05

SREDA-like temporal EEG seizure pattern in LGI1-antibody mediated encephalitis

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Background: Leucine-rich glioma inactived-1 (LGI1) antibodies are associated with limbic encephalitis and distinctive seizure types, which are typically immunotherapy-responsive. While non-specific EEG abnormalities are commonly seen, specific EEG characteristics are not currently understood to be useful for suspecting the clinical diagnosis. Based on initial observations in two patients, we analyzed the EEG recordings in a larger series of patients and describe a novel ictal pattern that can suggest the diagnosis of LGI1-antibody mediated encephalitis, even in the absence of common clinical features. *Methods:* Clinical and EEG data were collected in nine patients with LGI1 antibodies. *Results:* Psychiatric and cognitive symptoms were common, as were tonic seizures associated with EEG electrodecremental events (often with the so-called faciobrachial dystonic semiology). A rarity or absence of interictal epileptiform discharges contrasted with frequent subclinical temporal lobe seizures in some patients, which at times showed characteristics similar to subclinical rhythmic electrographic discharges of adults (SREDA), including sensitivity to hyperventilation. *Conclusions:* LGI1-antibody mediated encephalitis may be associated with tonic seizures and corresponding electrodecremental events, as well as an unusual SREDA-like pattern of frequent subclinical temporal lobe seizures, which may be triggered by hyperventilation, all in the setting of rare interictal epileptiform discharges.

D.06

Insular involvement in intractable epilepsy: results of invasive EEG data

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Background: Exploration of the insular cortex is now commonly considered in patients with refractory epilepsy requiring invasive EEG investigations. The safety and yield of routine insular exploration is uncertain. *Methods:* All patients (pediatric and adult) who had invasive EEG (iEEG) with insular depth electrode placement, either through SEEG or open implantation, were reviewed. Ictal insular involvement was characterized as primary, secondary or not involved. Results of insular resections were recorded. *Results:* A total of 173 patients had iEEG of which 26 included insular electrodes (SEEG-18, Open - 8). No complications of placement were identified. Insular involvement was seen in 20 (76%) patients. Primary ictal involvement was identified in 9 (33 %) patients, while secondary spread was noted in 11 (42 %) patients. Six patients went on to have resections including the insular cortex of which 5 patients achieved good seizure control (Engle class I/II). *Conclusions:* Insular depth electrode placement is a safe and effective adjunct to invasive EEG investigations. Ictal involvement of the insular cortex was commonly identified in our series leading to inclusion of the insula in cortical resections with good seizure control, which may not have been considered without iEEG evidence.

D.07

Clinical electromyography training in Canada: The experience of neurology and physiatry residents

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Background: There are currently no national standards for clinical electromyography (EMG) training for residents in neurology and physiatry in Canada. The purpose of this study was to obtain demographic and qualitative data pertaining to EMG residency training

in Canada, with the goal of facilitating discourse that could lead to national standards for EMG training. *Methods:* An online survey was distributed to senior neurology and physiatry residents (post-graduate years 3-5), at seven tertiary Canadian centres. The study authors, who are trainees and consultants with a broad range of EMG expertise (junior and senior resident, clinical neuromuscular fellows, senior physiatrist and neuromuscular neurologists), developed pertinent demographic and qualitative questions. *Results:* Thirty-eight residents completed the survey (23 neurology, 15 physiatry). There was inter-program variation in quantity of the training experience, content of the curriculum, access to expertise (including technologists) and goals for future training and practice. Similarly, differences were identified between the training experiences of neurology and physiatry residents. *Conclusions:* Inter-program variability in EMG training was identified. Additionally, differences were identified between neurology and physiatry resident training. This data provides evidence of training discrepancies across the country and can be used to establish national training standards for EMG in Canada.

D.08

ACT DMD (Ataluren Confirmatory Trial in Duchenne Muscular Dystrophy): effect of Ataluren on timed function tests (TFT) in nonsense mutation (nm) DMD

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Background: Ataluren is the first drug to treat the underlying cause of nmDMD. *Methods:* ACT DMD is a Phase 3, randomized, double-blind study. Males 7-16 years with nmDMD and a screening six-minute walk distance (6MWD) ≥150m and <80%-predicted were randomized to ataluren 40 mg/kg/day or placebo for 48 weeks. A pre-specified subgroup included patients with baseline 6MWD 300-400m. A meta-analysis of the overall ACT DMD population and the 'ambulatory decline phase' subgroup of the Phase 2b study (those patients meeting ACT DMD entry criteria) was pre-specified in the statistical plan. *Results:* In the overall ACT DMD population (N=228), changes in TFTs favored ataluren over placebo: 10-meter walk/run, -1.2s (p=0.117); 4-stair climb, -1.8s (p=0.058); 4-stair descend, -1.8s (p=0.012). In the pre-specified subgroup (n=99), these differences increased to -2.1s, -3.6s, and -4.3s, respectively, and were statistically significant (p<0.01) for 4-stair climb and descend. Results are supported by the meta-analysis (N=291), which demonstrated significant differences (p<0.05) in 10-meter walk/run, 4-stair climb, 4-stair descend. *Conclusions:* TFT results showed a benefit for ataluren in ACT DMD, and a larger treatment effect in the pre-specified baseline 6MWD 300-400m subgroup as well as the pre-specified meta-analysis of ACT DMD and the Phase 2b study decline subgroup.

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D.09

Pharmacological therapy for the prevention and management of cardiomyopathy in Duchenne muscular dystrophy: a systematic review

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Background: Improved respiratory care of Duchenne muscular dystrophy (DMD) patients has unmasked cardiomyopathy as a major source of morbidity and mortality. There is currently no consensus regarding the management of DMD-associated cardiomyopathy (DMD-CM). The objective of this systematic review was to evaluate the efficacy of pharmacological therapies for prevention and management of DMD-CM, and determine the optimal timing to commence these interventions. *Methods:* A systematic search was conducted in October 2015 and updated in January 2016 using MEDLINE, EMBASE and CINAHL databases and 9 grey literature sources for studies evaluating the use of angiotensin-converting enzyme inhibitors (ACEi), angiotensin receptor blockers (ARB), beta-blockers (BB) or aldosterone antagonists (AA) in DMD patients. References of retrieved records were searched. Quality assessment was conducted using the Downs and Black Quality Assessment Checklist. PRISMA reporting guidelines were used. *Results:* The 11 included studies were of low methodological quality. However, the use of an ACEi, ARB, BB and AA tended to improve or preserve left ventricular systolic function and delay the progression of cardiomyopathy. *Conclusions:* While there is evidence supporting the use of heart failure medication in patients with DMD-CM, data regarding these interventions for delaying the onset of DMD-CM and when to initiate therapy is lacking.

D.10

Acute flaccid paralysis in Canadian youth, 1996 to 2014

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Background: Acute flaccid paralysis (AFP) is notifiable in Canada with a differential diagnosis that includes a number of conditions. This analysis describes the epidemiology of AFP in Canadian youth less than 15 years old. *Methods:* Monthly active surveillance for AFP was conducted as part of the Canadian AFP Surveillance System. *Results:* From 1996 to 2014, 850 cases of AFP were reported, representing an average annual crude incidence rate of 0.77 cases per 100,000 youth less than 15 years old. The mean age of cases was 6.8 years (median 5.9 years). Nine percent had an abnormal neurological history and 53% had an acute respiratory illness within 30 days of onset. Fever occurred in 23% of cases, 96% experienced bilateral weakness, 21% had respiratory muscle involvement, and 26% had cranial nerve involvement. The average hospital length of stay was 13.5 days. The most common diagnoses were Guillain-Barré Syndrome (GBS) or a variant (70%), and transverse myelitis (TM, 14%). At the time of the initial report, 14% had fully recovered. *Conclusions:* Our AFP surveillance system provides a baseline for AFP and its causes in the Canadian paediatric population. While rare, AFP is associated with