P.007

Autoimmune Encephalitis Timing and Incidence: the Manitoba Experience

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Background: Early treatment of autoimmune encephalitis (AE) can improve outcomes. Despite expert recommendation, it remains unclear if suspected AE patients consistently receive empiric treatments prior to availability of antibody results. **Methods:** Retrospective chart review of patients referred for AE testing in Manitoba. Primary outcomes were the proportion of patients treated empirically prior to the availability of antibody results. Incidence, clinical presentation, investigations, complications, mortality rates, and hospital course were secondary outcome measures. **Results:** We identified 151 patients from 2012-2018. 43 patients met inclusion criteria. The annual incidence of AE in Manitoba was 0.37/100,000. 28/43 (65%) patients were treated prior to availability of antibody results ("Early group"). 15/43 (35%) patients did not receive treatment ("Late group"). Significantly more Early group patients had repeat immunotherapy (p=0.001), abnormal MRI (p=0.027), and investigations for malignancy (p=0.015). Durations of hospital and intensive care admission, complication rates, and mortality rates were not different between the two groups. **Conclusions:** This is the first-ever AE incidence, timing, and management study of a comprehensive Canadian geopolitical and medical catchment area. Just over 1/3 of suspected AE over seven years were not treated prior to antibody results becoming available. Patients treated earlier did not experience greater complication rates.

Dementia and Cognitive Disorders

P.008

Surgical Procedures are Common in Patients with Prion Disease

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Background: Surgical instruments used in patients with prion disease must be decontaminated or decommissioned to prevent iatrogenic transmission. This is only done when the diagnosis of prion disease is known. To assess the potential for iatrogenic transmission, we determined the prevalence of surgeries and use of precautions in patients with prion disease at two academic medical centers. **Methods:** Clinical details, results of investigations, and surgical interventions (performed within one-year of symptom onset) were extracted for patients with probable/definite prion disease at Mayo Clinic (n=107; 1-2014 to 12-2020) and Washington University School of Medicine (n=14; 2-2015 to 12-2019). **Results:** Twenty-six patients (21.5%) underwent 32 procedures, including 2 high-risk procedures involving the brain. Most procedures (17/32, 53%) occurred in the 1-year period preceding the onset of symptoms attributed to CJD. History of arthritis (OR: 7.4, 95%CI: 1.05-51.8), lack of behavioral symptoms (OR: 3.0, 95%CI 0.97-9.1), and greater time (months) from symptom onset to first MRI (OR: 1.1, 95%CI 1.03-1.2) were independently associated with odds of undergoing an invasive procedure. Prion disease precautions were observed in one case (diagnostic brain biopsy). **Conclusions:** Procedures were common in patients with diagnosis of prion disease; precautions were not. Coordinated approaches to screening and reporting are needed to prevent iatrogenic transmission.

P.009

Improving Detection of Creutzfeldt-Jakob Disease Mimics in Clinical Practice

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Background: Assays capable of detecting prions in CSF (e.g., RT-QuIC) have greatly improved the antemortem diagnosis of Creutzfeldt-Jakob disease (CJD) yet take time to conduct and are not widely accessible. There is a need to identify clinical features and common tests that identify mimics at presentation. **Methods:** Mimics were identified within longitudinal studies of rapidly progressive dementia at study sites. Mimics met clinical criteria for probable CJD but did not have CJD. Clinical features were compared between mimics and patients with CJD assessed at Mayo Clinic Enterprise (n=79) and Washington University in St. Louis (n=10; Jan-2014 to Oct-2020). **Results:** Mimics (10/155; 6.5%) were diagnosed with autoimmune encephalitis (n=7), neurosarcoidosis, frontotemporal lobar degeneration with motor neuron disease, and unknown dementia. Age-at-symptom onset, gender, presenting symptoms, and EEG and MRI findings were similar between mimics and CJD patients. Focal motor abnormalities (49/93, 10/10), elevations in CSF leukocytosis (4/92, 5/10) and protein (39/92, 9/10) were more common in mimics ($p<0.01$). Neural-specific autoantibodies associated with autoimmune encephalitis were detected within the serum (4/9) and CSF (5/10) of mimics, but not CJD cases. **Conclusions:** Autoimmune encephalitis, neurosarcoidosis and neurodegenerative diseases may mimic CJD at presentation and should be considered in patients with early motor dysfunction and abnormal CSF studies.

P.011

Standardized Processes for Addressing Driving Cessation in the Memory Clinic

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Background: Discussions around driving cessation between clinicians and dementia patients are challenging. Patients view giving up their license as losing their independence. We sought to

develop a tool that enables standardized and consistent driving messaging across clinicians working in a specialist memory clinic, across the span of cognitive disorders **Methods:** We developed a driving recommendations generator that allows clinicians to produce information handouts personalized to individual patient capabilities and needs. Clinicians select from a list of established recommendations that were developed with neurologist and geriatrician input, and consistent with provincial requirements. Recommendations cover patients' current driving ability, road safety examinations, alternate transportation, and license revocation. Early driving retirement is emphasized and encouraged, to proactively support patients' choices, safety and independence. Recommendation and handouts are printed for the patients. **Results:** Patients reported that the recommendations were easy to read and understand, and helped them to implement physician suggestions. All surveyed clients recommended continuing to provide such recommendations to future patients and families. Clinicians agreed that the tool helped them to save time, and simplified the process of finding accurate information to provide patients. **Conclusions:** Clinicians have found the system timesaving and useful for simplifying the process of providing helpful, informative resources for patients.

P.012

SketchNet: Equipping Cognitive Examinations With Neural Network Computer Vision

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Background: With the advance of technology, our capacity to assess patients with dementia is also developing. It is possible to administer cognitive examinations using technology, such as the iPad-based Toronto Cognitive Assessment, but hitherto difficult to autonomously administer them. Many of the 'inputs' from patients could be easily scored with software, but highly variable inputs such as the clock drawing are extremely difficult to score, precluding automated administration and scoring. This work focuses on the development of a neural network designed to assess cube drawings, infinity drawings, and clock drawings. **Methods:** 3200 drawings, evenly split between clocks, cubes and infinities were generated, with half being correct and half incorrect. A SqueezeNet was trained on 2000 images, validated on 800 drawings, and then tested on 400 drawings. **Results:** The SqueezeNet was able to achieve 97% accuracy on 400 images it had never seen before in categorizing images as "Cube", "Clock",

"Infinity", or "Other" (incorrectly drawn). **Conclusions:** This neural network can successfully determine the difference between correctly and incorrectly drawn images commonly used in cognitive examinations, overcoming the final barrier to autonomously administering and scoring cognitive examinations. Next steps are to clinically validate an autonomous examination program which has been modeled after the Addenbrooke Cognitive Examination-3.

P.013

Machine Learning on Drawing Tests of Cognition: A Systematic Review

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Background: Machine learning (ML) methods hold promise in allowing early detection of dementia. We performed a systematic review to assess the quality of published evidence for using ML methods applied to drawing tests of cognition, and to describe the accuracy of the methods. Methods: Embase, Medline, and Cochrane Central Library databases were searched for potential studies up to December 8, 2018 by four independent reviewers. Included articles satisfied the following criteria: 1) use of ML on 2) a drawing test in order to 3) assess cognition. The quality of evidence was then assessed using GRADE methodology. Results: The initial search yielded 4620 citations. Of these, 64 were eligible for full text review. 18 articles then met inclusion criteria. Median AUC across all models was 0.765, with certain ML algorithms performing better in terms of AUC or diagnostic accuracy. However, based on GRADE, the quality of evidence was deemed very low. Conclusions: ML has been applied by several groups to drawing tests of cognition. The quality of evidence is currently too low to make recommendations on their use. Future work must focus on improving reporting, and using standard algorithms and larger, more diverse datasets to improve comparability and generalizability.

P.014

A Novel Canadian Family with the Rare IVS10+14 Tau Mutation

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Background: The IVS10+14 mutation in the microtubule-associated protein tau gene, *MAPT,* is a rare point mutation that dysregulates tau splicing resulting in pathological aggregation. This mutation has been identified in three families with severe neurodegenerative disease. We characterized the clinicopathological features of a fourth, Canadian family with the IVS10+14 *MAPT* mutation and compared them to previously reported families. **Methods:** Clinical and neuropathological records from three family members with the IVS10+14 *MAPT* mutation were reviewed. Neuropathological section from one available case were analyzed. **Results:** Considerable interfamilial phenotypic heterogeneity is reported in all cohorts that express the IVS10+14 *MAPT* mutation, with prominent motor, cognitive, behavioural, and respiratory symptoms. The Canadian cohort also expressed profound sensory and sleep abnormalities, not reported previously. In the two siblings with available neuropathological