P.046

Extensive leukoencephalopathy as a differential diagnosis of motor neuron disease: case report and literature review

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Background: Weakness is frequently a reason to attend neurology consultation which entails a challenge due to its wide range of diagnostic possibilities. We present the case of an elderly woman with long-standing right upper limb weakness. Methods: Patient data was obtained from medical records and an extensive neuropathological evaluation was performed. Results: Weakness started off in her right hand, which progressed insidiously to her right hemibody, with hyperreflexia, atrophy, fasciculations and foot drop. Subsequently, bulbar and left hemibody deterioration began. She died due to ventilatory failure four years after the first symptom appeared. A tigroid pattern in the neuroimaging studies suggest white matter involvement, while the neuropathological studies showed loss of motor neurons in the spinal cord. Conclusions: The available evidence does not allow us to dismiss the hypothesis of a motor neuron disease or a leukodystrophy. Cases like this one establish a diagnostic challenge due to their complexity and unusual etiology.

P.047

Implications of Gold Coast Criteria in diagnosis of amyotrophic lateral sclerosis in a large subspecialty clinic

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Background: Criteria to formalize the diagnosis of amyotrophic lateral sclerosis (ALS) and refine clinical trial populations have evolved. The recently proposed Gold Coast criteria (GCC) are intended to simplify use and increase sensitivity. We evaluated sensitivity of GCC and potential impacts on therapeutic trial enrollment. Methods: We performed a single center retrospective study including patients diagnosed with ALS between 2016 -2021. We determined criteria met at diagnosis according to revised El Escorial (rEEC), Awaji (AC) and GCC. We compared sensitivity and examined impacts GCC would have on enrollment in landmark ALS trials. Results: We included 203 people with ALS. Sensitivity of GCC (96.1%, 95% confidence interval [CI] = 92.2-98.2%) was significantly higher than rEEC (89.8%, 95% CI 84.6-93.4%, $\chi^2 = 5.3$, p = 0.01) and AC (89.3%, 95% CI 84.1-93.0%, $\chi^2 = 6.1$, p = 0.006). GCC was more sensitive than clinically definite or probable rEEC (47.6%, 95% CI 40.6-54.6%, $\chi^2 = 117.6$, $p = \langle 0.001 \rangle$ and use would result in increased eligibility in landmark therapeutic trials. Conclusions: GCC are more sensitive than rEEC and AC at time of diagnosis in ALS. Use of GCC in our population would expand clinical trial participation and make results more widely generalizable.

P.048

Objective measures of balance deficits in sensory ganglionopathy

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Background: Sensory ganglionopathy (SG) is a rare form of neuropathy affecting the dorsal root ganglia and leading to nonlength-dependent sensory abnormalities. Although balance problems are frequently reported by patients, a comprehensive balance assessment in SG has not been conducted. This study quantifies balance deficits in SG and examines their relation to patient-reported outcome measures (PROMs). Methods: Prospective data was collected from five participants with SG. Balance assessments included Fullerton Advanced Balance scale, Berg Balance scale, and 360 degree turn. Participants completed PROMs assessing balance confidence (ABC scale), pain, fatigue, quality of life (OoL), and daily activity and participation. Assessment also included neurological exam, nerve conduction studies (NCS) and posturography. Results: All participants had severe SG on NCS with normal strength and significant sensory abnormalities. Balance scores indicated severe balance deficits in all participants and aligned with posturography and truncal sway measures. PROMs revealed low confidence in balance, high levels of pain and fatigue, difficulties with daily activities, and reduced QoL. Conclusions: Although balance testing is not part of routine clinical practice, PROMs and targeted assessment may help monitor patients with SG and their response to treatment. Larger sample sizes are needed to understand the impact of balance on PROMs and optimize bedside balance testing.

P.049

Determining individual substantial response in amyotrophic lateral sclerosis: utilizing a new method on CENTAUR trial results

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Background: In ALS, determining whether individuals have a substantial response to therapy is a challenge for the field. ALS naturally progresses at variable rates and a personalized approach is required to determine if individuals have a substantial response. A new method to evaluate individual response is proposed and applied to data from the CENTAUR trial of sodium phenylbutyrate/ursodoxicoltaurine (PB/TURSO). Methods: In a post hoc analysis, CENTAUR participants whose actual rate of change from baseline in the ALSFRS-R at week 18 was ≤ their own trial baseline progression rate (ΔFS) were defined as having a substantial individual response in slowing ALS progression. Results: Substantial individual response was observed in a greater proportion of participants receiving PB/TURSO (41%, n=87) vs placebo (19%, n=48; *P*=0.0076). Conclusions: Response versus

 ΔFS provides a personalized metric to determine substantial individual response in ALS. ΔFS has been shown to be highly correlated with, but to proportionally underestimate, ALSFRS-R decline in clinical trials. Consequently, those who outperform the ΔFS may be considered to have a substantial individual response. Application to CENTAUR data demonstrates a greater proportion of participants with a substantial individual response in the PB/TURSO arm. These methods may enable greater personalization and analysis of individual response in ALS.

NEUROSCIENCE EDUCATION

P.050

Understanding the neurocritical care educational needs of trainees

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Background: Patients with neurological conditions account for 15% of patients admitted to the Intensive Care Unit (ICU). Neurocritical care (NCC) has been proven to reduce mortality, improve functional outcomes and increase patient/family satisfaction. Trainees often lack the knowledge, skills, and experience needed to provide quality NCC. Consequently, timely effective care is compromised, team dynamics suffer, and trainees may experience distress. Methods: To fully understand educational needs, we surveyed University of Calgary residents from various programs who rotate through the Neuro-ICU. Results: Trainees indicated a lack of confidence in their knowledge and skills of most NCC disorders/scenarios in the ICU. While the majority expressed interest in learning NCC, 58% were not aware of the NCC-related competencies outlined for their specialties by the Royal College of Physicians and Surgeons, and 30% had no objectives of their own. Teaching modalities most preferred included patient-centred bedside teaching (96%) and easily accessible resources such as pocket-sized cards (90%) and/or a phone app (96%). Conclusions: Trainees rotating through Neuro-ICU need more accessible and improved learning resources and tools. An NCC curriculum may help improve patient outcomes, team dynamics, and relieve trainee distress.

P.051

Stroke Care and Neurological Emergency Response Simulation (SCaNERS): High-fidelity acute stroke simulation increases learner confidence in providing acute stroke care

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Background: Resident physicians often observe stroke alerts before managing them alone, which exposes patients to potential harm from trainees' lack of experience. Simulation training offers a low-risk environment for skill acquisition. This project assessed learners' confidence in leading stroke codes before and after completing a stroke simulation training program during neurology rotations at the University of Saskatchewan. Methods: Highfidelity simulation cases were developed encompassing several diagnostic and therapeutic goals of acute stroke care. Standardized patients were trained for increased fidelity. Standardized debriefing was given after each session. Pre- and post-simulation surveys captured learner confidence and cognitive load. Results: Pilot data reveal learners' confidence and comfort in providing acute stroke care, including thrombolysis treatment decisions, significantly increases after simulation training (n=8; p=0.0006-0.01). They also felt more prepared to conduct future acute stroke care (p=0.009). Skills not directly addressed in simulation did not show significant improvement (p=0.09-1.89). Learners consistently rated the session as requiring high mental effort. Conclusions: Implementation of high-fidelity simulation training leads to significant improvement in learner confidence. Future cases will capture additional objectives and ensure acceptable cognitive load. Ongoing data collection to explore residents' experiences and knowledge improvement in stroke care and assess local reductions in treatment delays is underway.

P.052

Assessing the competence of neurology residents in performing an interphysician telephone consultation

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Background: Neurology Residency training in Canada is transitioned to competence based medical education (CBME) in July 2020 and the Royal College Neurology Specialty Committee has identified "providing consultation for and managing patients at outlying centres," to be an entrustable professional activity (EPA). At Western University, neurology telephone consultations from outlying centres are attended by both the resident and the staff Neurologist. This scenario provides the ideal situation for direct observation and immediate formative feedback. The resident's performance was assessed using an entrustment scale along with a short narrative portion. Methods: This mixed methods study aims to determine the reliability and validity of the entrustment scale with narrative feedback in assessing the performance on the telephone consultation by senior neurology residents. Informed consent was obtained from residents (9) and staff (7) involved. Scores on the entrustment scale and narrative comments were analyzed. Results: Information on 37 encounters (involving 9 residents) was collected. Assessment results demonstrated higher entrustment scores in PGY4 and PGY5 levels. Overall, ratings were consistent across the 7 consultant assessors. Conclusions: The use of an entrustment scale appears to be reliable and valid in assessing competence in the telephone consultation and provide coaching feedback to help learners improve their performance.

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