

NEUROPHYSIOLOGY SUBSPECIALTIES

EEG

modifiable symptom—have not been studied in DMD. Our objective was to explore risk factors for fatigue in children with DMD. *Methods:* Patients aged 4–17 years identified via the Canadian Neuromuscular Disease Registry received mailed questionnaires. Fatigue was assessed using the PedsQL™ Multidimensional Fatigue Scale (patient- and parent-report). Standardized measures for depressive symptoms, sleep disturbances, functional ability and physical activity were used. Spearman's correlations and Wilcoxon rank-sum tests were computed. *Results:* Of 194 eligible patients, 64 have responded to date. DMD patients reported greater fatigue than healthy controls from published data. Depressive symptoms were associated with greater fatigue, by patient-report ($\rho=-0.44$, $P<0.001$) and parent-report ($\rho=-0.40$, $P=0.002$). Sleep disturbances were associated with greater fatigue, by patient-report ($\rho=-0.41$, $P=0.007$) and parent-report ($\rho=-0.51$, $P<0.001$). Greater functional ability was associated with less fatigue, by parent-report ($\rho=0.30$, $P=0.02$). Physical activity and ambulatory status were not associated with fatigue. *Conclusions:* Fatigue is a significant issue in DMD. Depressive symptoms and sleep disturbances are associated with fatigue, warranting attention in therapeutic strategies to reduce fatigue.

P.074

Myopathic aspects of Mowat-Wilson Syndrome

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Background: Mowat-Wilson Syndrome (MWS) is a genetic syndrome (*ZEB2*, OMIM: 235730) that occurs in 1 in 50000 births. It is characterized by microcephaly, intellectual disability, dysmorphisms (prominent chin, cupped ears, broad nasal bridge) and Hirschsprung's disease. Although motor delay and hypotonia are common components, a myopathy has not been described in MWS literature. A childhood case with myopathic features prompted further study of this rare disease. *Methods:* Patients were recruited from the Mowat-Wilson Foundation via email or social media to complete a survey. *Results:* Thirteen surveys were returned to date. Although 54% of the patients reported motor delay, none of the patients had myopathy investigations. The index patient, presented at 1 year old, with hypotonia and developmental delay. Pregnancy and family history were unremarkable. Investigations revealed high CK levels (range 300 to 500 U/L), EMG confirmed myopathic motor units, and muscle biopsy showed type 1 fibre predominance. Single gene sequencing revealed pathogenic mutations of *ZEB2*, confirming a diagnosis of MWS. *Conclusions:* The description of myopathic features expands the spectrum of this rare syndrome and adds to the differential diagnosis of hyperCKemia in early childhood.

P.075

The trend of electroencephalograph findings after starting anti-epileptic drugs during seizure assessment in children

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Background: Few studies have explored the effects of anti-epileptic drugs (AEDs) on electroencephalograph (EEG) findings during the assessment of seizure management. Although a patient may reach seizure freedom, EEG results may continue to be abnormal. Further information is required to understand the trend of EEG findings during seizure treatment. *Methods:* This is a retrospective study based on chart reviews. Patients who had epilepsy evaluations at the Royal University Hospital in Saskatoon between January 2012 and December 2015, were selected. The relationships among time of initiating AEDs, EEG findings, and seizure outcome on follow-ups, have been evaluated. *Results:* 151 patients had first seizure clinic assessments, in which 75 patients had an EEG before starting AEDs. Among the 75 patients, 54 (72%) had abnormal EEGs. From those, 38 (70.3%) patient's EEGs became normal and 16 (29.7%) patients continued to have abnormal EEGs after the introduction of AEDs. The seizure freedom was 81.5% among those who had normal EEG on follow-up, and 43.7% of those who continued to have abnormal EEGs. *Conclusions:* Although patients with normal EEGs after starting AEDs may encounter a higher chance of seizure freedom, the seizure free patients with abnormal EEGs indicate that EEG is not completely sufficient in predicting seizure status.

P.076

Quantitative EEG in Canada: a national technologist survey

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Background: Burgeoning EEG demand has largely gone unmet with insufficient supply of manpower and equipment. Quantitative EEG (QEEG) may help compress large volumes of data for expedited review. We sought to determine the current use of QEEG in Canada through a national EEG technologist survey. *Methods:* A 10-item questionnaire was administered to participants at the 2016 meeting of the Canadian Association of Electroneurophysiology Technologists, which occurred in parallel with the Canadian Neurological Sciences Federation meeting. *Results:* A response rate of 63% (14/22) represented 12 institutions (11 adult, 6 paediatric) over six provinces with 73% of the national population. Only academic institutions (9/12) used QEEG, representing five provinces with 70% of the national population. Most institutions generated QEEG either real-time or retrospectively in the critical care and epilepsy monitoring units for long-term monitoring and automated seizure detection. The most

used trends were spectrographic, seizure detection, and artifact detection. Montage use, QEEG duration, and timebase settings were highly variable. *Conclusions:* QEEG is in surprisingly frequent use across Canada. There is no consensus on optimal QEEG use, which mirrors uncertainty in the literature. The relative ubiquity of QEEG in Canada offers promise for collaborative multicentre research into unlocking the full potential of QEEG in enhancing patient care.

P.077

EEG attenuations in adults: clinical correlates

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Background: Intermittent EEG attenuations have relatively clear significance in pediatric populations, but a consistent clinical correlation has not been identified in adults. While generally seen in metabolic encephalopathies, the specific clinical correlates and prognostic value have not been determined. *Methods:* We prospectively collected 22 consecutive EEGs noted to have intermittent generalized attenuations. Baseline and discharge modified Rankin Scale (mRS), diagnosis at discharge, EEG altering medications, ICU admissions, relevant imaging, mental status, the location the patient was discharged to, and pertinent lab values were assessed. *Results:* Mean patient age was 73.7 (SD = 11.0) at admission. Twelve of the twenty-two patients (55%) died during their course in hospital. Four patients (18.2%) did not have a change in mRS score from baseline to discharge, while most had an increase in their mRS scores reflecting increased disability. Twelve patients (55%) were admitted to the ICU or CCU during their time in hospital. The most common etiologies were metabolic encephalopathies, and often associated with triphasic waves. *Conclusions:* Intermittent generalized EEG attenuations in adults are associated with severe metabolic encephalopathies and poor outcome including high association with mortality. The physiologic mechanism of generalized attenuations is poorly understood. Patients with this pattern should be suspected of having a severe metabolic encephalopathy and treated accordingly.

EMG

P.078

Diaphragm ultrasound in amyotrophic lateral sclerosis: a case report demonstrating a critical role for this technique

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Background: Diaphragm pacing (DP) is an experimental ALS treatment, available through a compassionate use program. Eligibility requires forced vital capacity (FVC) between 45-50% predicted and phrenic nerve conduction study (NCS) evidence showing the diaphragm can be electrically stimulated. Diaphragm ultrasound (DU) also evaluates diaphragm function by demonstrating thickening with inspiration. *Methods:* A 63 year old man with advanced ALS requested DP as his respiratory functions worsened. He was wheelchair bound and had severe dysarthria and dysphagia. He had

exertional dyspnea and used CPAP at night for obstructive apnea. *Results:* FVC was 47% predicted. Initial phrenic NCS showed a normal response on the right but no response on the left, making him ineligible for DP. Diaphragm function was further assessed with DU. This showed normal thickening with inspiration bilaterally. The DU result prompted repeating the right phrenic NCS which then showed a normal response. He successfully completed surgical implantation of diaphragm leads for DP. At surgery both diaphragms showed good responses to electrical stimulation. *Conclusions:* Phrenic NCS can be technically challenging and yield a false positive (absent) result. In this patient, DU indicated good diaphragm function, which prompted repeating phrenic NCS. The normal phrenic NCS allowed the patient to pursue DP.

NEUROIMAGING

P.079

Brain Magnetic Resonance Imaging metallic susceptibility artifacts post cardiac surgery in children

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Background: Metallic susceptibility artifact (MSA) on brain MRI has been described rarely in adult population as an incidental finding in patients undergone cardiac surgery, catheterization or prosthetic heart valve, but has not been described before in the pediatric population. Here we present two pediatric cases with MSA on brain MRI post-cardiac surgery.

Case Series

Patient1: 13-month-old girl with Transposition of great arteries, aortic coarctation and multiple VSDs, who twice had open heart surgery requiring cardiac bypass. She presented with bilateral lower extremity paralysis secondary to spinal cord embolic infarction, Brain MRI showed an incidental finding of hundreds of diffuse foci of brain MSA.

Patient2: 5-year-old boy with Trisomy21, Atrial-ventricular septal defect repaired at age one year. His brain MRI showed incidental finding of multiple, widespread foci of MSA during investigation for gait disturbance.

Discussion: Here we present two pediatric patients post-cardiac surgery, found to have multiple scattered MSA in cerebral and cerebellar hemispheres and brain stem. Presumably, these are secondary to metallic microthrombi during cardiac surgery, however, neither had prosthetic material. Metallic microthrombi may be related to cardio-pulmonary bypass. Pediatric cardiac surgery patients should be studied, to understand the significance of these lesions and further distinguish the cause and association of these findings.