

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.

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Myopathic aspects of Mowat-Wilson Syndrome

*L Subramanian (London) Y Wei (London) C Nguyen (London) R Hicks (London) P Chitra (London) C Campbell (London)**

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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.

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

A Amiraslany (Saskatoon) A Khan (Saskatoon) F Moien-Afshari (Vancouver) PK Wong (Vancouver) S Almubarak (Saskatoon)**

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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.

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Quantitative EEG in Canada: a national technologist survey

MC Ng (Winnipeg) K Gillis (Winnipeg) J Nikkel (Winnipeg)*

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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