Movement Disorders

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Benign spasms of infancy - a mimicker of infantile epileptic disorders

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Background: Benign spasms of infancy (BSI), previously described as benign non-epileptic infantile spasms or benign myoclonus of early infancy, are non-epileptic movements manifesting during the first year of life and spontaneously resolving in the second year of life. BSI are characterized by spasms typically lasting 1-2 seconds, involving to varying degrees the head, neck, trunk, shoulders and upper extremities. Ictal and interictal EEG recordings are normal. BSI are not associated with developmental retardation and do not require treatment. Distinction between BSI and infantile epileptic disorders, such as epileptic spasms or myoclonic epilepsy of infancy, can be challenging given the clinical similarities. Moreover, interictal EEGs can be normal in all conditions. Epileptic spasms and myoclonic epilepsy require timely treatment to improve neurodevelopmental outcomes. **Methods:** We describe a 6-month old infant presenting with spasm-like movements. His paroxysms as well as a positive family history for epileptic spasms were in keeping with a likely diagnosis of West syndrome. **Results:** Surprisingly, ictal video EEG did not reveal epileptiform activity, and suggested a diagnosis of BSI. **Conclusions:** We emphasize that ictal EEG is the gold standard for classification of infantile paroxysms as either epileptic or non-epileptic, thereby avoiding overtreatment of BSI and facilitating timely targeted treatment of infantile epilepsies.

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Whole-genome sequencing identified a frameshift mutation at LMNB1 in a family with early-onset dystonia

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Background: Dystonia is a hyperkinetic condition that produces abnormal movements or postures. Its diagnostic procedure is often challenging and time consuming. Genetic testing provides an effective approach for diagnosis, but currently only very few dystonia genes have been identified. We propose that studying early-onset forms of dystonia with the use of whole-genome sequencing (WGS) will improve the identification of dystonia-relevant genes and mutations. **Methods:** We performed deep WGS using the Illumina HiSeq X technology in a mother-proband pair with dystonia. The mother has generalized dystonia (age of onset: 15) and the proband has myoclonic dystonia (age of onset: 11). **Results:** No pathogenic mutation was identified in any of the known dystonia genes. However, we identified a rare heterozygous frameshift mutation (p.K342fs*7) at *LMNB1* that was shared between the mother and the proband. Duplication of *LMNB1* is known to cause Adult-onset Demyelinating Leukodystrophy. A heterozygous deletion of *LMNB1* has been reported in a patient with microcephaly and global developmental disorder. **Conclusions:** Further characterization of phenotypes in the participants and their family members is needed to confirm the relationship between mutation in *LMNB1* and dystonia. This work provides a proof-of-principle that novel disease-relevant genes can potentially be identified using the proposed approach.

Neurocritical Care

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Electroconvulsive therapy and epilepsy: a case report

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Background: Electroconvulsive therapy (ECT) involves the induction of a generalized seizure with an electrical current and has been used worldwide when treating medically refractory psychiatric illness. Here we describe a patient with no prior history or risk factors for epilepsy who developed temporal lobe epilepsy after chronic treatment of ECT. **Methods:** A 16-year-old right-handed boy with severe refractory depression received ECT treatment every 10 days for 8 months. Six months into his ECT treatment, the patient developed seizures and was admitted to a pediatric epilepsy monitoring unit. **Results:** Initial clinical events included lightheadedness, diaphoresis, and nausea with associated kaleidoscopic vision changes. Seizures progressed to confusion, fear and paranoia by the time the patient was admitted for monitoring. Long-term video EEG captured many focal seizures with impaired awareness, all originating from both temporal lobes. MRI was normal. ECT was terminated and the patient started on carbamazepine. He has been seizure free for the past 2 years on medication **Conclusions:** While rare, we present a case of a patient with no prior risk factors for epilepsy who developed temporal lobe epilepsy after chronic ECT treatment. Although ECT is an indispensable treatment for many medically refractory psychiatric illnesses, we suggest caution in young patient undergoing ECT.

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A clinical pathway of combined EEG monitoring in high-risk critically ill neonates

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Background: overtreatment of neonatal seizures may result in neurological morbidity. aEEG, despite low sensitivity, is widely used, for ease of bedside interpretation. vEEG, is a limited resource needing expert interpretation. We hypothesize that using aEEG combined with vEEG will increase the sensitivity and specificity of seizure detection and reduce anti-convulsants use compared to aEEG alone. **Methods:** Prospective cohort of neonates admitted to CHEO NICU with suspected seizures between April 1st 2018 to present. Seizures (clinical/aEEG) were documented by bedside clinicians and compared to the vEEG. Bedside clinicians could call a neurologist for remote review of the vEEG. Outcomes include concordance of aEEG and vEEG events and number of episodes where management was changed based on both readings **Results:** 27 patients had both