had histories of cardiac arrest, syncope, and sudden death in a family member. Cardiac phenotypes differed in patients who experienced generalized tonic-clonic seizures and patients with epilepsy for 3+ years. Conclusions: Almost 1/3 of our high-risk epilepsy cohort had history of cardiac events or abnormalities on cardiac testing. Seizure type and epilepsy duration were associated with altered cardiac phenotypes. Since some findings were potentially clinically significant, routine cardiac screening of high-risk epilepsy patients may be warranted.

P.036

Vagal nerve stimulation in three cases of continuous spike and wave in slow-wave sleep

D Bourcier (Halifax) W Stewart (Saint John)*

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Background: Continuous spike and waves during slow-wave sleep (CSWS) is a childhood-onset epileptic encephalopathy that is characterized by clinical seizures, electrical status epilepticus during sleep (ESES), and neurocognitive regression. Early intervention can preserve neurocognitive development, and vagus nerve stimulator (VNS) therapy had positive outcomes in the few previously reported case reports. We present three patients with intractable CSWS unresponsive to medications, who had a positive response to VNS therapy. Methods: Review of clinical records of three pediatric patients diagnosed with CSWS were compared for selected clinical outcomes and electrographic data both prior to and in the years following the initiation of VNS therapy. Results: Three patients now aged 13, 16 and 20 years, were treated with VNS following intolerance and a lack of response to multiple medications (5-9) for CSWS. The ketogenic diet was not an option. The CSWS resolved in all three patients, resulting in improved cognitive function. Patient 3 had resurgence of CSWS on EEG when the VNS settings inadvertently reset to the factory settings and improved with adjustment in the cycling. Conclusions: In patients who are unresponsive to medication, VNS provides an alternative option for resolving CSWS to preserve and, in some cases, potentially restore neurocognitive function.

Metabolic Disease

P.037

Refractory status dystonicus and hypotension after cardiac arrest in a child with AADC deficiency post gene therapy: a case report

D Peacock (Vancouver) G Horvath (Vancouver)*

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Background: Aromatic l-amino acid decarboxylase (AADC) deficiency is a metabolic disorder that causes deficient serotonin, dopamine, and catecholamine synthesis. How children respond to neurological insult post intracranial gene therapy remains underreported. We present a 10 year old girl with profound neurological injury after a brief in-hospital cardiac arrest, secondary to viral infection-induced respiratory failure, 4 years after gene therapy. Methods: Patient's chart review included brain imaging, clinical notes, laboratory results, and treatment. Results: MRI showed symmetric abnormalities in the basal ganglia, thalami, cortex, and cerebellar hemispheres. CSF analysis showed homovanillic acid 27 nmol/L (reference range 167-563) and 5-hydroxyindoleacetic acid 7 nmol/L (reference range 67-189). She developed generalized dystonia and oculogyric crises which were not seen since before gene therapy. There was poor catecholamine production causing refractory hypotension. She required a one-month stay in ICU for hypotension and status dystonicus. Dystonia was controlled with high doses of 6 agents. Conclusions: We describe a patient with AADC deficiency post gene therapy who experienced disproportionately severe neurological injury and decreased AADC activity after hypoxic neurological insult. There may be unique considerations of dopaminergic neuron integrity, AADC gene promoter sensitivity, and cerebrovascular autoregulation in children with AADC deficiency post gene therapy.

Movement Disorders

P.038

Exploring alternative deep brain stimulation targets for movement disorder in children – a systematic literature review

H AlGethami (Toronto) AT Sulistyo (Toronto) S Breitbart (Toronto) A Fasano (Toronto) G Ibrahim (Toronto) C Gorodetsky (Toronto)*

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Background: Deep Brain Stimulation (DBS) has become increasingly prevalent in the management of paediatric movement disorders, with the globus pallidus interna (GPi) serving as the most utilized target. However, limitations exist, including variable responses in genetic versus acquired forms of movement disorder and structural damage in the GPi would preclude its use as a target. Given these limitations, there is a pressing need to explore alternative targets. We investigated the application of non-GPi targets in paediatrics through a systematic review. Methods: Individual data points were gathered from references identified through a systematic electronic search and analysed descriptively. We included paediatric patients (0-18 years) with movement disorders who underwent non-GPi-DBS. We excluded adults and other indications. Results: Preliminarily, 64 patients were identified from 40 references. Dystonia was the most common movement disorder type, followed by tremor and chorea. The subthalamic nucleus was the frequent DBS target for dystonia, yielding promising outcomes of improvement as measured on the Burke-Fahn-Marsden movement scale ranging from 43% to 95%. The ventral intermediate nucleus was the second most employed target, demonstrating favourable results. Conclusions: Non-pallidal DBS targets hold promise as potentially efficient and safe. However, to further validate their effectiveness and safety, larger multi-centre randomized studies are required.