suggests that these adults tend to have higher rates of depression and anxiety, lower rates of medication compliance, and lower education. Methods: To better understand this population and their struggles, a retrospective chart review of 58 patients transferred from pediatric to adult care was done. Results: 39.7% of participants were lost to follow-up; 12 were temporarily lost (average 1.3 years) and 11 were permanently lost. Twenty-three participants admitted to medication non-compliance, with fifteen having break-through seizures. Of the 45 patients that filled out mental health assessments at initial visit, 28.9% met the threshold for major depressive disorder, and 56.6% of patients had symptoms of anxiety. Data found that at one-year follow-up, 60% of these patients had similar or worsened depression scores, and 64% had similar or worsened anxiety scores. Conclusions: The findings of this study are concerning and highlight the need for greater education and support for these adolescents. Specifically, patients need more education on the importance of consistent follow-ups and consistently taking medication. Findings also suggest the importance of assessing and addressing mental health concerns.

P.033

Electrophysiological signatures of sedation in pediatric patients

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Background: Sedation in PICU masks physical exam findings, leading to diagnostic challenges. In adult models, electroencephalography can evaluate the brain's response to sedation using feedforward connectivity and anteriorization of alpha hubs, proving useful for prognostication. Feasibility of model translation into pediatric population was assessed, with the hypothesis that the same markers of adaptive reconfiguration would correlate with a higher potential for recovering consciousness. Methods: Electroencephalograms from children undergoing sedation were analyzed for strength and direction of functional connectivity using the weighted and directed phase lag index. Target population was refined with an iterative inclusion criteria. We examined relationships between hub location reconfiguration, directed phase lag index, baseline Glasgow Coma Scale, and 3-month post-treatment Glasgow Outcome Scale-Extended. Results: Evaluation of 14 subjects showed promise in children aged 5-18 undergoing sedation with midazolam, dexmedetomidine, and propofol. Further analysis of five subjects revealed a correlation between adaptive reconfiguration during anesthesia and both higher baseline Glasgow Coma Scale and Glasgow Outcome Scale-Extended scores post-treatment. Conclusions: The findings indicate that the functional brain network connectivity model may have diagnostic and prognostic potential regarding children's consciousness levels. While the initial data is promising, further analysis of six additional cases is pending and deemed essential to thoroughly evaluate the model's efficacy.

P.034

Cost-effectiveness of treatment strategies for medically refractory pediatric epilepsy: a systematic review

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Background: Medically refractory pediatric epilepsy is a disorder that can cause significant financial and physical burden. Although multiple treatments exist, cost-effectiveness remains unclear. We conducted a systematic review to assess cost-effectiveness of treatments for medically refractory pediatric epilepsy and to summarize key issues and areas for further inquiry. Methods: We searched MEDLINE and 6 other databases up to July 2022. We included partial and full economic evaluations (EEs) on treatments for medically refractory pediatric epilepsy. Pairs of reviewers independently screened the literature, extracted data, and assessed quality using the 24-item Consolidated Health Economic Evaluation Reporting Standards (CHEERS) checklist. We extracted data on study characteristics, health outcomes, model design, costs, and treatment characteristics. Results: We identified 37 eligible studies for analysis, 19 of which were partial EEs and 18 were full EEs. Study quality, outcomes reported, treatment comparators, and factors included in cost calculations were common influential factors in study results. Vagus nerve stimulation and cannabinoid oil were the most consistently cost-effective, in 6 of 7 and 1 of 2 studies, respectively. Other treatments were inconsistently cost-effective. Conclusions: The cost-effectiveness of treatments for medically refractory pediatric epilepsy was not definitive. Consistency in study design and inputs is necessary for future comparison of epilepsy treatment.

P.035

Cardiac screening in children with genetic epilepsy at risk for sudden unexpected death in epilepsy

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Background: People with epilepsy experience higher rates of cardiac arrhythmia and sudden death than the general population, with the highest risk in genetic epilepsies. Despite growing evidence of a possible cardiac contribution, routine cardiac screening for epilepsy patients is rarely performed. Methods: We performed a single center, retrospective review of patients with developmental epileptic encephalopathies caused by genetic variants expressed in the heart and brain. Clinical history, medications, age, and cardiac evaluation data were extracted. Results: Among 67 patients (56% female), 54 (81%) had at least one ECG. Twenty (37%) had an abnormal ECG. Forty-one had a repeat ECG: 8 showed persistent abnormalities, 7 resolution of abnormalities, and 7 a new abnormality. Five patients with an abnormality did not receive a follow up ECG. Two patients each

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

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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

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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

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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.