phenomenology, etiology, and outcome is heterogenous and poorly characterized, making a standardized management approach challenging. We characterized demographics of children with status dystonicus in British Columbia admitted to the pediatric intensive care unit (PICU), management patterns, and outcomes. Methods: Clinical records at our PICU were searched via ICD-10 codes. We included cases admitted 2014-2024 who had dystonia severity grade 3-5, dystonia worse than baseline, and age >30 days old. Results: Seventy-nine records were screened; 41 admissions from 19 unique patients were included. Mean age was 7.6±4.2 years; 53% were female. Most unique patients had a genetic etiology (n=8, 42%). The presenting complaint per admission was often not dystonia (n=24, 59%); infection was the most common trigger (n=23, 56%) followed by pain (n=6, 15%). Patients received several anti-dystonia medications (mean 6.9±2.5), including clonidine, benzodiazepines, ketamine, and others. Mean PICU stay was 11.0±10.8 days; 37% had multiple PICU admissions. Two patients (4.9%) died from status dystonicus complications. Conclusions: Status dystonicus is a life-threatening emergency commonly triggered by pain and infection in patients with dystonia. Given the considerable morbidity and mortality, multi-disciplinary teams should consider standardized treatment guidelines for these complex patients.

## P.064

### MOXIe clinical trial overview of omaveloxolone for patients with Friedreich ataxia

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Background: We summarize the efficacy and safety data for omaveloxolone in patients with Friedreich ataxia from the MOXIe clinical trial (NCT02255435, EudraCT2015-002762-23) and post hoc analyses. Methods: In MOXIe Part 2, patients aged 16-40 were randomized 1:1 to receive omaveloxolone 150 mg or placebo. The primary outcome was change in modified Friedreich Ataxia Rating Scale (mFARS) from baseline to Week 48—patients could roll over into an open-label extension (OLE). A post hoc propensity-matched analysis compared treated and untreated patients over 3 years. Results: Treatment with omaveloxolone significantly improved mFARS relative to placebo at Week 48, with a difference of -2.41 points for the full analysis set (n=82 [excluding severe pes cavus]; p=0.01) and -1.93 points for the all-randomized population (n=103 [including severe pes cavus]; p=0.03). Transient and reversible changes in aminotransferase levels were observed with omaveloxolone without other signs of liver injury. Headache, nausea, and fatigue were among the more common adverse drug reactions in omaveloxolone-treated patients. In a post hoc propensity-matched analysis, omaveloxolone-treated patients in the OLE progressed by 3 points at Year 3 versus 6.6 points in an untreated matched cohort. Conclusions: Patients who received omaveloxolone showed a significantly stabilized neurological function and slowing of FA progression, as measured by mFARS.

# Neurocritical Care

## P.067

### Transcranial doppler use in pediatric endovascular thrombectomy post large vessel obstruction secondary to infective endocarditis

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Background: Transcranial doppler ultrasound (TCD) in a pediatric neurocritical setting can determine cerebral hemodynamics by assessing the blood flow velocity in main cerebral arteries. In large vessel occlusions (LVO) that require endovascular thrombectomy (EVT), TCD can monitor recanalization and arterial re-occlusion. We describe one case in a previously healthy 13-year-old girl with a right M1 middle cerebral artery occlusion. Methods: Analysis was done via a retrospective case review. Results: Our patient underwent a successful endovascular thrombectomy (EVT) six hours after symptom onset. Follow up TCDs done at 4, 8, and 24 hours showed stable peak systolic velocities (PSV) on the narrowing of right M1 ranging from 245 to 270 cm/s with stable pre-stenotic PSV around 110 cm/s, indicating focal and stable narrowing of M1 without reocclusion. No high transient signals (HITS) were identified on sub 10 minute TCDs. An urgent echocardiogram revealed a bicuspid aortic valve with vegetations, with later confirmation of infective endocarditis. The patient made an impressive recovery with only mild deficits. Conclusions: TCD can be an effective tool in a pediatric neurocritical setting in guiding initial recanalization after EVT and monitoring for arterial re-occlusion, HITS and hyperperfusion. TCD monitoring also decreases the amount of radiation exposure via CTA.

## P.068

### A proposed protocol for treatment of acute necrotizing encephalopathy of childhood at Stollery Children's Hospital

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Background: Acute Necrotizing Encephalopathy of Childhood (ANEC) is an illness characterized by rapidly progressive encephalopathy, typically associated with a precipitating viral illness such as influenza. It is diagnosed clinically and through neuroimaging showing symmetric multifocal lesions involving the deep grey matter, especially the thalamus. Morbidity and mortality in ANEC are high, so prompt recognition and treatment are key, but treatment protocols vary. We propose a management protocol based on a consensus approach and available evidence. Methods: A rapid literature review was conducted. Studies included were meta-analyses, case series, and expert consensus