



FIGURE 1: BAMF and GMFCS scores illustrate motor decline in a cross-sectional analysis of patients with MLIV. Abstract 234 [Color figure can be viewed at www.wileyonlinelibrary.com]

extrapyramidal signs (rigidity, dystonic posturing, bradykinesia) in a subset of patients that have not been reported in MLIV. Among atypical patients, identical symptoms with milder severity were appreciated and a genotype-phenotype relationship with partial preservation of TRPML1 channel function was suggested. **Conclusions:** Though previously considered a static condition, functional decline with age is a clinically assessable feature of MLIV. Extrapyramidal signs are variably present but can be prominent. Genotype explains a portion of variance in symptom severity. Our findings expand the clinical description of MLIV and can guide future gene therapy trials which are currently in the preclinical phase.

Keywords: Rare Diseases, Genetics, Movement Disorders

STROKE

235. Retrospective Chart Review: Pediatric Demographics in Children Presenting with Acute Neurological Deficit Concerning for Acute Ischemic Stroke: An Evaluation of the Stroke Alert Process

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Objective: To describe the demographics and characteristics of patients who present with stroke-like symptoms to Children's Mercy Hospital

Methods: Descriptive retrospective chart review of patients who presented to CMH from September 1, 2016-August

31, 2018 with concern for acute stroke who have the Stroke Alert Process and Powerplan activated.

Results: There were a total of 61 stroke activations between September of 2016 through August of 2018. 14/61 patients (23%) met final diagnosis of ischemic stroke or TIA. Of the patients that met the final diagnosis of ischemic stroke, the most common presenting symptom was unilateral weakness. Two were candidates for intervention with mechanical thrombectomy and none received tPA. Average age of all activations was 14years, while average age of patients who had final diagnosis of ischemic stroke or TIA was 4years. 61% of (37/61) activations were female and the most common racial demographic was Caucasian. Ischemic stroke/TIA was the most common diagnosis of all activations. Seizure/Todd's paralysis (12/61, 20 %) and migraine (12/61, 20 %) were the second leading diagnoses. Other common diagnoses included psychogenic/conversion disorder (15%), complications of meningitis/encephalitis (6.6%), and oncologic process (5.0%). No intra-cranial hemorrhages were identified in this patient population.

Conclusions: Ischemic stroke or TIA comprised nearly one-fourth of all pediatric stroke activations and is the leading diagnosis in all activations obtained in this study. These findings are consistent with current reported literature. This data in conjunction with previous studies highlights the importance of developing protocols for early recognition and evaluation of children who present with stroke like symptoms.

Keywords: Stroke