Use of Immunomodulatory High-Dose Subcutaneous Immunoglobulin in Treatment of Opsoclonus Myoclonus Syndrome.

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Body of Abstract:

Rationale: Opsoclonus Myoclonus Syndrome (OMS) is a rare, autoimmune condition resulting in abnormal eye movements, involuntary twitching of muscle groups, ataxia, irritability, and speech impairment. High dose intravenous immunoglobulin is part of the standard therapy in pediatric patients diagnosed with OMS. We report a 2-year old girl who was diagnosed with OMS, initiated on intravenous immunoglobulin, and successfully converted to a high-dose subcutaneous regimen.

Methods: Case report of a 2-year old girl diagnosed with OMS who initially presented with neurologic symptoms of unsteady gait and tremors at 22 months.

Results: CSF studies, EEG, MRI of the brain and spine were normal. CT scan revealed no neuroblastoma. Blood chemistries and metabolic panels, including a carnitine panel, DNA ataxia panel, and lead test were normal. Initiated on corticosteroids (2 mg/kg daily) at age 22 months and intravenous immunoglobulin at 26 months (1 g/kg IV monthly for 9 months.) Had improvement in symptoms and successful reduction of steroids. However, severe headaches and vomiting were experienced after each intravenous dose despite pre-treatment. Converted to a high-dose subcutaneous immunoglobulin protocol (3.2 g every 6 days, weight = 16 kg) with continued improvement in gait and neurologic function with no further episodes of vomiting or headaches.

Conclusions: Current therapeutic guidelines for OMS recommend a trial of IVIg (1 g/kg of body weight monthly) should be implemented for 3-6 months with continued therapy every 3-4 weeks afterwards if successful. We suggest that a high-dose subcutaneous immunoglobulin protocol may be effective with fewer side effects in pediatric patients with OMS.