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Acute Flaccid Myelitis: Treatment Outcomes From a Tertiary Referral Center

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Background

- Acute Flaccid Myelitis (AFM) has effected hundreds of children throughout the world.
- Recognized as a variant of transverse myelitis in after an outbreak in 2014 there has been significant controversy about both the cause and the recommended approach to treatment.
- Most data supports a relationship between Enterovirus D68 and this paralytic condition.
- Since it's recognition there has been a controversy about whether the syndrome is infectious or post infectious and whether it's potential infectious etiology would be a contraindication to the use of corticosteroids or PLEX.
- Early recommendations from the CDC supported the use of IVIG in children diagnosed with AFM and cautioned against the use of steroids or PLEX without outcomes data to reference.
- Given the lack of therapies available for AFM patients a review of outcomes data could help guide clinicians.

Objectives

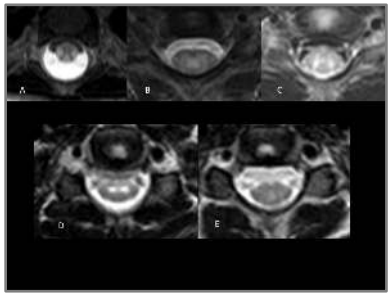
- To determine the outcomes among children diagnosed with acute flaccid myelitis who were treated with steroids, IVIG and plasma exchange.

Methods

- Outcome data from pediatric TM patients with an AFM phenotype was collected under an IRB approved protocol.
- Cases were retrospectively defined based on the clinical and radiographic evidence of significant gray matter involvement.
- Treatment regimens were recorded and motor outcomes were recorded.

Results

- Sixty-nine patients with TM were identified, of which 44 were included in this analysis.
- The average age of the patient at diagnosis was 8.9 years of age (range 7 months to 17 years) and 61% of the patients were female (n= 27).
- Of the 39 patients included in the follow-up analysis, the mean time to last follow-up was 17 months with a median of 10 months. Eight patients had follow-up of less than 6 months, leaving 31 patients in the outcome analysis cohort.
- Of the 31 patients included in the outcome analysis, 19 (61%) had excellent, very good or good outcomes as defined in this study (MRS < 3).
- The most common MRI pattern was a mixed involvement pattern with involvement of both gray and white matter. This was present in 73% of patients. 14% of patients had a central cord pattern. There were 3 patients (7%) each in the lateral and central cord pattern categories.
- 17 AFM patients had therapeutic PLEX as part of their treatment. 10 had good outcomes (MRS 0,1,2) and 7 had poor outcomes (MRS 3,4)
- In our cohort, the recovery of patients with predominantly gray matter damage was similar to patients with lateral and central cord injury patterns (p=0.18) despite a comparable use of anti-inflammatory interventions (e.g. corticosteroids and PLEX).



- Examples of T2-weighted axial MR images of the spinal cord from patients included in this analysis. (A)White matter restricted pattern; (B) Central cord pattern; (C) Gray matter restricted; (D and E) Evidence of gray matter and white matter involvement from a single patient.

Conclusions

- Children with AFM and evidence of white matter involvement appear to benefit from the use of steroids and/or PLEX therapy.
- This suggests that the anterior horn cell damage in AFM is likely mediated by viral infection while upper motor neuron damage may be mediated by an inflammatory pathophysiology.
- Anti-inflammatory therapies may be most beneficial at preventing long term upper motor neuron sequelae

Discussion/Future Directions

- Treatment recommendations should be based on data/experience when possible.
- Early recommendations based on theoretical concerns should be studied in a controlled fashion.
- Future studies of TM/AFM should work to subtype patients and track associations between phenotypes and outcomes. This is necessary to create meaningful treatment algorithms.

Disclosures

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