

Treatment type and EDSS outcome of pediatric acute disseminated encephalomyelitis: A retrospective analysis of children from a US Network



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Background

• Acute disseminated encephalomyelitis (ADEM) is an inflammatory demyelinating disease of the central nervous system occurring more often in children than adults. Symptoms consist of multifocal neurologic deficits with encephalopathy. Corticosteroids are usually first line therapy. Intravenous immunoglobulin (IVIG) and plasmapheresis (PLEX) are usually reserved for refractory cases that do not respond to corticosteroids. Outcome is usually good, with the majority of patients making a full recovery¹.

Objectives

• To describe treatment type and Expanded Disability Status Scale (EDSS) outcome in a cohort of children who had ADEM.

Methods

• We performed a retrospective analysis of patients prospectively enrolled in a database by the United States Network of Pediatric Multiple Sclerosis Centers. The database is maintained at the University of Utah. The database was queried for cases of ADEM prior to age 18. Demographic information including gender, race, and age at disease onset were obtained. Median length of follow up, treatment type and EDSS at last visit were also recorded.

RESULTS

- There were 94 patients [52 males (55%), 42 females (45%)] with a diagnosis of ADEM in the database
- Median length of follow up was 43.5 months.
- The median age at ADEM onset was 5.2 years (range 0.8 – 17.1 years).
- The length of time from ADEM onset to the first EDSS recorded was 2.8 years (corticosteroids), 2.5 years (IVIG), and 5.9 years (PLEX).
- Eighty patients made a good recovery with an EDSS of 2 or less at last follow up.
- Those who required IVIG had a median follow up of 34.5 months and a median EDSS at last visit of 2
- Patients who underwent PLEX had a median EDSS of 2.5 at last follow up

• Demographic Information for all 94 patients

Age at ADEM onset (years)	Median	Range
Age of all patients	5.2	0.8 - 17.1
Age of patients receiving only steroids	5	0.8 – 17.1
Age of patients ever receiving IVIG	5.5	3.2 – 16.6
Age of patients ever receiving PLEX	7.7	3.2 - 11

• Race

Caucasian	African-American	Asian	Hispanic
71 (75.5%)	12 (12.8%)	3 (3.2%)	15 (16%)

• Gender

Male	Female
52 (55%)	42 (45%)

• Type of therapy and EDSS outcome

	Initial Therapy	Ever received	EDSS at last follow up
Corticosteroids	75	77	
IVIG	5	12	2
Plasmapheresis	1	5	2.5

Conclusions

- ADEM in children is typically treated with corticosteroids only.
- IVIG and plasmapheresis are less commonly used, and usually reserved for more severe attacks refractory to corticosteroids
- ADEM is usually associated with good recovery in terms of disability measured by the EDSS

Limitations

- EDSS scores at time of illness were not available for most patients with the first scores not being recorded until 2-3 years after ADEM onset
- This is a retrospective study

References

- ¹ Tenembaun, S.N., *Disseminated encephalomyelitis in children*. Clin Neurol Neurosurg, 2008. **110**(9); p. 928-38

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