

# NMDAR Encephalitis in a Patient with NMO: A Double Hit without Cognitive Sequelae Following Treatment

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## Objective

To describe the clinical, behavioral, and cognitive outcomes of an adolescent with Neuromyelitis optica (NMO) who later developed N-methyl D aspartate (NMDAR) encephalitis.

## Background

Reports of NMDAR encephalitis and NMO occurring successively in the same patient have been rare in adults with even fewer cases described among the pediatric age group.<sup>1</sup> In adults with NMDAR encephalitis it was shown that cognitive impairment may persist despite treatment.<sup>2</sup>

## Methods

Retrospective chart review and prospective data collection of an 11 year old evaluated over a 3 year period at the Lourie Center for Pediatric MS.

## Results

An 11 year old girl, born and raised in Anguilla, developed acute disseminated encephalomyelitis (ADEM) manifested with acute onset of weakness, facial droop, slurred speech and seizures. MRI showed multiple lesions in right periventricular and bilateral cerebellar nuclei involvement without enhancement. She was brought to the USA, treated with intravenous (IV) steroids, anticonvulsants, and fully recovered. She returned home, did well socially and returned to school.

Four years later, she had transverse myelitis (TM) and improved with IV steroids. A few months later, she presented to Stony Brook Pediatric MS Center for a diagnosis. She had residual lower extremity increased tone and hyperreflexia. She had a + anti-aquaporin serology. Mental status was normal; symbol digit modality test (SDMT) raw score=79 (WNL). She again home to Anguilla.

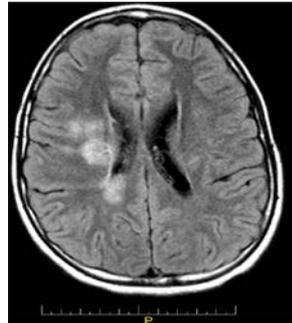


Figure 1. MRI of initial episode of ADEM. T2 FLAIR of brain showed multiple "fluffy" lesions in right periventricular and also cerebellar area (not shown).



Figure 2. Thoracic MRI STIR sequence showing lesion along T5-T8 consistent with episode of transverse myelitis.

At 16 years old, one year after her episode of (TM), she returned to the ED with new acute symptoms of disorientation, confusion and psychosis. She was hospitalized and developed catatonia. Her neuroimaging abnormalities were unchanged. NMDAR encephalitis was confirmed with positive CSF antibody.

	During NMDAR encephalitis	One month post encephalitis	Two years post encephalitis
<b>SDMT</b>	14 (Severely impaired)	43 (Mildly Impaired)	84 (WNL)
<b>General Cognition</b>	MoCA= 17 (Moderate to Severe)	MoCA=25 (Mildly impaired)	IQ=100
<b>Clock drawing</b>			
<b>Draw a person</b>			

At the height of her illness, she had a raw SDMT score of 14 (severely impaired) and Montreal Cognitive Assessment score (MOCA) of 17. Following plasmapheresis x 7 exchanges and IV Rituximab her mental status improved.

One month into symptoms her raw SDMT score=43 (mildly impaired); MOCA=25.

Patient was continued on Rituximab 1 gm infusions every 6 months.

Two years later, (9/2015); her mental status and behavior were back to her normal baseline: raw SDMT score=84 (WNL), verbal memory on the Rey Auditory Verbal Learning Test raw score=70 (WNL) and visual learning on the Brief Visual Memory Retention Test-Revised raw score=30 (WNL).

## Discussion

- NMDAR encephalitis recovering cognitively has been described but there is no objective data supporting this. Presented is a case of NMDAR encephalitis cognitive testing in a patient with NMO, who was improved fully with treatment.
- In conclusion, in children NMDAR can follow NMO with cognitive and behavioral dysfunction improving with treatment pediatric patient with NMO.

## References

- Titulaer, Maarten **Overlapping demyelinating syndromes and anti-NMDA receptor encephalitis.** *Ann Neurol.* 2014 Mar; 75(3):411-428
- Finke C, Kopp UA. Cognitive deficits following anti-NMDA receptor encephalitis. *J Neurol Neurosurg Psychiatry.* 2012 Feb;83 (2);195-8