

# Neuropsychological Sequelae of Anti-N-Methyl-D-Aspartate Receptor Encephalitis in a 15 Year-Old Hispanic Female

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## REVIEW OF LITERATURE

Anti-N-methyl-d-aspartate receptor encephalitis (Anti-NMDAR-E), is a rare life threatening paraneoplastic disease.

It was discovered in 2007 and its incidence is unknown. It mostly affects women with ovarian teratomas; however, it can also affect men and women without tumors.

Abnormalities on EEGs, cerebrospinal fluid, and vital signs are commonly seen. The presence of slow wave activity on EEGs occurs in approximately 80% of the cases.

NMDA receptors are found in the forebrain, hypothalamus, and limbic system structures (e.g. hippocampus).

These receptors are crucial for adequate synaptic transmission, hippocampal long-term potentiation, and dendritic sprouting. NMDA receptors are involved in learning related plasticity.

The overall recovery process for this condition varies. For some patients no major improvements are seen at 6 and 12 months post-diagnosis. However, at 72 months significant improvements are seen in attention, memory, and problem-solving.

### Symptoms of Anti-NMDAR Encephalitis

Cognitive	Psychiatric/Behavioral	Emotional
Short and Long-term Memory Impairments	Hallucinations	Apathy
Inattentiveness	Catatonia	Lack of Emotion
Executive Dysfunction	Agitation	Depression
	Language Impairments	Anxiety
	Paranoid Ideations	
	Social Withdrawal	
	Stereotypical Behaviors	

## BACKGROUND

This case study examines A.G, a 15 year-old, right-handed Hispanic female who reportedly tested gifted during Elementary school. Acute behaviors included incoherent speech alternating with catatonia. Lip smacking, agitation, and aggression were observed. A.G. was diagnosed with Anti-NMDAR-E based on EEGs showing diffuse slowing with delta brushes and an ictal pattern. No tumor was found.

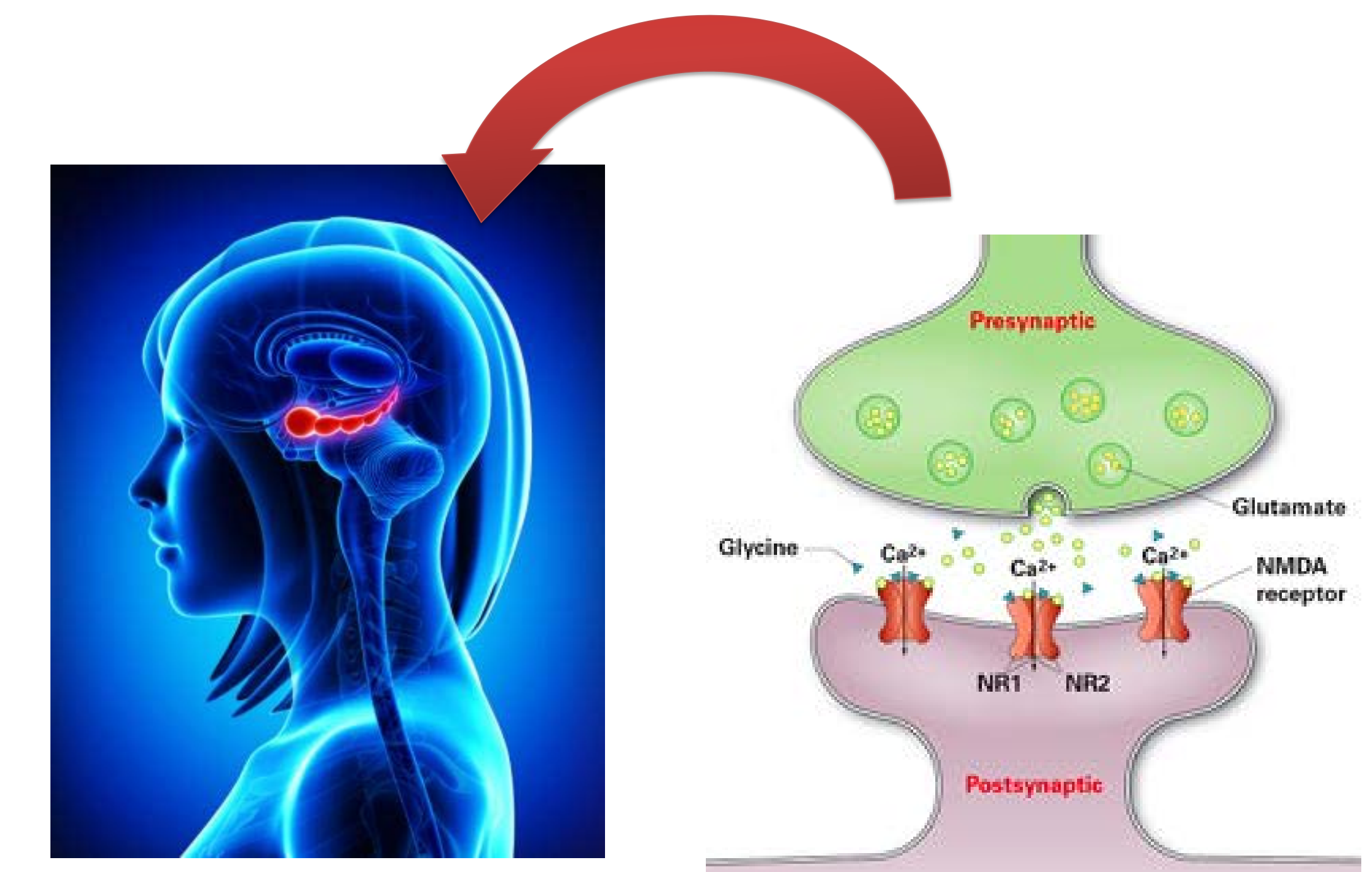
A.G. did not respond to Solumedrol, IVIG, or plasmapheresis treatment, but did respond to Rituximab. A.G. received a Menactra booster vaccination against meningococcal disease two weeks prior to symptom onset.

## METHOD

A comprehensive neuropsychological battery was administered 15 months post-illness onset, to delineate cognitive, emotional, and behavioral functioning.

## RESULTS

Neuropsychological Test	15 months post-illness
<b>Rey-15 Test</b>	15/15
<b>WISC-V</b>	
Digit-Span	Average
Block Design	Average
Similarities	Average
Matrix Reasoning	High Average
Symbol Search	High Average
Vocabulary	Average
Visual Puzzles	Low Average
Coding	Superior
Figure Weights	Average
Picture Span	Low Average
<b>WRAML-2</b>	
Story Memory	High Average
Story Memory Recognition	Average
Story Memory Recall	High Average
<b>Woodcock Johnson-IV</b>	
Letter-Word Identification	Age >30
Applied Problems	Age >30
Spelling	Age >30
Passage Comprehension	Age 15.2
Writing Samples	Age >30
Word Attack	Age >30
Oral Reading	Age 17.9
<b>CPT-3</b>	
Detectability	Average
Omissions	Average
Commissions	Average
HRT Block Change	High Average
<b>Conners CATA</b>	
Detectability	Average
Omissions	Average
Commissions	High Average
HRT	Atypically Fast
HRT SD	High Average
HRT Block Change	Elevated
<b>CVLT-C</b>	
List A Total Trials 1-5	Average
List A Trial 5 Free Recall	15/15
Recognition	High Average
<b>ROCF</b>	
Copy	Average
Immediate Recall	Average
Delayed Recall	Average
Recognition	Average
<b>DKEFS</b>	
Inhibition	Average
Color Naming	Average
Word Reading	Superior
Inhibition	High Average
Inhibition/Switching	High Average
Trail Making	Average
Condition 1 Visual Scanning	Average
Condition 2 Number Sequencing	Average
Condition 3 Letter Sequencing	Average
Condition 4 Number-Letter Switching	Borderline
Condition 5 Motor Speed	Low Average
Verbal Fluency	Average
Letter Fluency	Average
Category Fluency	Average
Category Switching	Average
Design Fluency	Average
Condition 1 Filled Dots	Profoundly Impaired
Condition 2 Empty Dots Only	Low Average
Condition 3 Switching	Low Average
Tower Test Total Achievement Score	Low Average
<b>WCST</b>	
Total Errors	Average
Perseverative Responses	Low Average
Categories Completed	5
Failure to Maintain Set	1
<b>Grooved Pegboard Dominant Hand</b>	Average
<b>Grooved Pegboard Non-Dominant Hand</b>	Average
<b>Finger Tapping Test Dominant Hand</b>	Impaired
<b>Finger Tapping Test Non-Dominant Hand</b>	Borderline
<b>Sensory Perceptual Examination</b>	
Left Hand - Right Face	1 Error
Left Ear	2 Errors
Left Eye Level	2 Errors



## CONCLUSIONS

A.G.'s case describes chronic cognitive and emotional sequelae of Anti-NMDAR-E.

Results indicated good problem solving abilities, visual scanning, and academic achievement 15 months post-illness.

Visual memory and visuoconstructional skills were adequate. Verbal memory was not significantly impaired; however, mild consolidation inefficiencies relative to estimated premorbid status were present.

Ongoing deficits were found in executive functioning, particularly with tasks that required cognitive flexibility, attention-shifting, and sustained attention.

A.G. presented as guarded and demonstrated a perfectionistic and self-demanding disposition. Post-illness symptoms of anxiety, depression and social withdrawal were noted. These were, in part, attributed to her reaction to actual and perceived changes in cognitive and academic functioning. Organic changes related to her condition also were possible.

Another case of a 15-year-old female who developed Anti-NMDAR-E following an immunization was reported in 2011. The association between immunizations and adverse neurological events should be further studied in an attempt to understand a possible correlation.

This case study investigates relatively long-term cognitive and emotional sequelae of Anti-NMDAR-E. Factors that may influence recovery from Anti-NMDAR-E include premorbid level of functioning (e.g., cognitive reserve), areas of the brain impacted by the condition, and emotional reactions to the condition.