

Hallee Mingus

Speech-Language Pathology

Neuromyelitis Optica Spectrum Disorder: Performance on Language and Cognitive Testing

Introduction

- Neuromyelitis Optica Spectrum Disorder (NMOSD) is a relapsing inflammatory demyelinating disease that affects the central nervous system and attacks the brain, resulting in severe physical &/or visual disability (Pfeuffer et al., 2017).
- Persons with NMOSD can present with fatigue, pain, & stiffness, and report anxiety, depression, bowel/bladder problems, and excessive drowsiness (Eaneff et al., 2017).
- Unknown cause; may result from infection or other autoimmune disease (Papp, 2021).
- Limited information exists on the communicative-cognitive characteristics in persons with NMOSD. One report describes a man with spinocerebellar ataxia type 31 who, after 14 years, developed NMOSD and slurred speech (Takahashi et al., 2017). Researchers believed the co-occurring diagnoses were unrelated, but they speculated underlying inflammation could be a plausible unifying factor.
- Of 30 adults with NMO given a neuropsychological battery: 9 (30%) had impaired long-term memory, 1 had impaired short-term memory, 6 (20%) had slowed processing speed, 8 (~27%) had executive dysfunction, and 4 (~13%) had language impairments (Blanc et al., 2008).
- The purpose of this case study was to describe the performance of a young adult with NMOSD on the the Quick Aphasia Battery (QAB)-Remote (Wilson et al., 2018) and the ALS Cognitive Behavioral Screen (ALS-CBS) (Woolley, 2014), both delivered via teleconference.
 - ALS-CBS was used to establish how persons with NMOSD could score on this screening and if data could be applicable to future treatment planning.
 - QAB-Remote and ALS-CBS were given remotely to determine viability of testing.

Research Questions

- What are the communicative-cognitive characteristics in a person with NMOSD when using the QAB-Remote (Wilson et al., 2018) and ALS-CBS (Woolley, 2014)?
- What are the challenges of remote test administration of these two tests?

Methods

- **Participant:** Data was collected from H. H., a 32-year-old female with a complex medical history who was diagnosed with NMOSD in 2018.
- **Procedures:** The participant completed the QAB-Remote and the ALS-CBS via an approximately one-hour Zoom session while she was alone at her home. The QAB-Remote was administered using the remote-adapted stimulus book using Form 1.

Results & Discussion

- H. H. demonstrated varied severity levels throughout the two formalized assessments. Her scores on the ALS-CBS rated her as having suspected frontotemporal dementia. However, several challenges occurred during the remote administration of the ALS-CBS (Please see next section). H. H. **DOES NOT** present with dementia. She is at home alone by herself much of the day, can independently complete activities of daily living, and can log onto her computer and Zoom into her various individual and group therapy sessions held across two different clinics multiple times a week. She has a strong sense of humor and can give complex answers during treatment sessions.
- The QAB-Remote classified H.H. as having mild aphasia. She had the greatest difficulty on the speech motor programming section.

ALS Cognitive Behavioral Screen (ALS-CBS)

Section	Score
Attention	3/5
Concentration	2/5
Tracking/Monitoring	3/5
Initiation and Retrieval	0/5
Total Score	8/20
Severity Rating	Suspected Frontotemporal Dementia

Quick Aphasia Battery (QAB) - Remote

Subtests	Score	Severity Level
Word Comprehension	10.00	No Aphasia
Sentence Comprehension	10.00	No Aphasia
Word Finding	9.50	No Aphasia
Grammatical Construction	8.63	Mild
Speech Motor Programming	5.00	Moderate
Repetition	8.75	Mild
Reading	8.75	Mild
QAB Overall	8.83	Mild

Challenges with Remote Test Administration

- **ALS-CBS:**
 - Difficult to remotely assess certain aspects, particularly the saccades and antisaccades eye movements. It was hard to see where H. H. was looking through the camera. She then attempted to verbalize her eye movements. It was unclear if instructions involving saccades and antisaccades were clearly understood or if the directions should be repeated.
 - When testing required pointing in response to commands given, it was difficult to observe H. H.'s responses due to her arm being outside the camera's view. Additionally, clinicians were unsure if the cameras were mirrored during testing, potentially skewing H. H.'s responses as either correct or incorrect.
 - **Recommendations:** The ALS-CBS should not be given remotely due to the difficulties stated above. Adaptations and/or alternate tasks will need to be made in order to make this a useful screening if used remotely.
- **QAB-Remote:**
 - Easy to understand and administer remotely with no obvious challenges. H. H. also had the verbal abilities needed if instances of confusion arose. If assessing a person without such verbal abilities, this exam may have been more difficult to administer.
 - **Recommendations:** QAB-Remote appears to adequately capture a higher-level client's abilities. Clinicians will need to consider other clients' verbal abilities.

Future Research

- Complete additional assessments on more persons with NMOSD, ideally in person. Could re-attempt ALS-CBS in person.

Selected References

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Acknowledgements & Disclosures

The researchers wish to thank H. H. and have no financial or nonfinancial relationships to disclose.