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Case Report

Neuropsychological evaluation in American Sign Language: A case study of a deaf patient with epilepsy



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ABSTRACT

In high-stake cases (e.g., evaluating surgical candidacy for epilepsy) where neuropsychological evaluation is essential to care, it is important to have culturally and linguistically appropriate and accessible neuropsychological instruments and procedures for use with deaf individuals who use American Sign Language (ASL). Faced with these ethical and professional issues, clinicians may be unable to provide equitable services without consulting with other psychologists and collaborating with the patient and interpreter. This is a case report describing a 43-year-old male with bilateral sensorineural deafness and a lifelong history of drug-resistant temporal lobe epilepsy who presented as a candidate for a comprehensive neurological workup to determine surgical candidacy. He was bilingual (ASL and written English). We describe all aspects of the evaluation, including functional magnetic resonance imaging (fMRI) and Wada testing, using an ASL interpreter. Results from the neuropsychological evaluation were not clearly lateralizing, but suggested greater compromise to the non-dominant right hemisphere. fMRI and Wada test results revealed language and verbal memory functions were lateralized to the left hemisphere. The patient was deemed to be an adequate candidate for surgical resection of portions of the right hemisphere. Comprehensive assessment of neuropsychological functioning in deaf persons who use ASL is feasible. This case report illustrates the important considerations relevant to neuropsychologists providing culturally and linguistically informed assessments to deaf ASL users with epilepsy. Additional research in this area will support future efforts to develop effective and efficient models that could be implemented across different settings. Moreover, clinical guidance is warranted to guide professionals interested in promoting access to high quality neuropsychological services.

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Introduction

In the United States (U.S.), it is estimated that over 38 million people have some form of deafness or hearing loss in at least one ear, and the majority stems from age-related changes in hearing status [1]. Nevertheless, accurate demographic data for Deaf people who use American Sign Language (ASL) as their primary language is scarce and based on a limited available literature [2]. Several factors interfere with our ability to quantify the demographic characteristics of individuals living in the U.S. who identify as culturally Deaf and/or who use ASL as their primary language, versus individuals who view their deafness from an audiological or medical perspective and likely identify with the hearing culture and/or use spoken English as their primary mode of communication. Such data has been infrequently collected and the only source of national data on people with hearing loss developed by the U.S. Bureau of the Census does not explicitly ask about Deaf culture, the nature and extent of ASL use, or characteristics related to hearing loss.

Deaf people experience cognitive difficulties at five times a higher rate relative to the general public, but this generally results not from the experience of being deaf or having a hearing loss, but rather comorbidities such as congenital syndromes, birth trauma,

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or acquired illnesses [3]. The prevalence and presentation of neurological conditions among deaf adults is largely unknown outside of studies including small sample sizes and case studies [4–6]. The available literature specific to describing the demographic, clinical, and cognitive characteristics of deaf people with epilepsy is almost nonexistent. This is concerning given some of the primary etiologies of deafness are also those contributing to the onset of epilepsy (e.g., meningitis, prematurity, Rh factor, rubella, etc.) [7].

Epilepsy is the most common, chronic, neurological disease, worldwide, and affects at least 65 million people [8]. The lack of current scientific knowledge about the characteristics at the intersection of epilepsy and deafness limits clinical care. This is particularly true in the context of clinical neuropsychology which relies heavily on language-based instrumentation to determine cognitive functioning and assess language lateralization. Patients with epilepsy are at risk for drug-resistant epilepsy, a debilitating chronic disorder that emerges when epilepsy no longer responds to pharmacotherapy. Drug-resistant epilepsy is associated with increased economic and psychosocial burden, and surgical intervention becomes an option for a selected group of patients [9]. When surgery is an option, the role of a presurgical neuropsychological evaluation assists in establishing baseline level of functioning for future comparisons, lateralization/localization of brain injury, and informing post-operative rehabilitation recommendations. Traditionally, the intracarotid amobarbital procedure (Wada testing) has been considered essential as it allows for assessment of language lateralization, provides information on risk for global amnesia and post-operative verbal memory loss, and provides information on side of seizure onset [10]. More recently, however, emerging evidence suggests that functional magnetic resonance imaging (fMRI) of language lateralization is less invasive and may be as effective as Wada testing in identifying typical lateralization among hearing individuals [11]. In some epilepsy centers and surgical sites, fMRI has become the predominant approach to presurgical lateralization of language and memory functions.

Regarding presurgical evaluation for deaf signers with epilepsy, there is limited available information and almost no documented standards of care with respect to neuropsychological procedures. Advances in the considerations for neuropsychological assessment of cognitive functioning among deaf signers have been increasingly reported during the course of the past two decades [12–16]. However, the majority of these publications have focused on how to approach a neuropsychological protocol and the limitations in the use of existing measures, as well as how to work with spoken and sign language interpreters. Some earlier work discussed the ethical challenges in conducting research with people who are deaf, and in the selection process of psychological testing [17]. More recently, several research-based measures have been developed for use with deaf adult signers, but include very small samples with limited descriptions of their demographic and background characteristics, and at times, limited analyses of psychometric properties, especially applied to clinical samples. For example, in the U.S., the ASL Comprehensive Test [18] and the ASL Sentence Reproduction Test [19] have been developed as research measures for use with signing adults, but have almost no normative data among large samples of healthy deaf signers or those with clinical diagnoses, and are not readily available for clinical use. Two ASL verbal memory tests were developed for psychological assessment, the Signed Paired Associates Test and the ASL Stories Test, that parallel the Paired Associate Test and the Logical Memory Test of the Wechsler Memory Scale, respectively 20-22] and include small sample data for healthy and clinical adult groups. However, the ASL Stories Test requires use of a CD/DVD available from the author and has almost never been used in research or clinical studies to date. Verbal fluency (phonemic and semantic) in ASL and English also have been explored among various samples of deaf individuals [23,24]. Morere and Allen (2012) [25] also included several psychological and neuropsychological tests as part of a larger study examining literacy among a small group of healthy deaf bilingual university students and provided some data regarding their performance on the various tasks.

In light of this emerging evidence, available guidance on how to administer a culturally and linguistically informed Wada test to deaf individuals is minimal [12,13]. While the validity of conducting Wada tests administered in sign language has been explored, findings have limited generalizability as the patients in these studies were able to hear to some extent, as well as sign [12]. Moreover, existing studies have focused on the results as opposed to the process for the evaluation itself [12,13]. To our knowledge, there have been no documented studies of fMRI administration among deaf adults who have epilepsy and use ASL as their primary language. The literature regarding left-hemisphere lateralization of language among right-handed deaf signers suggests that the results of neuropsychological, Wada, and fMRI testing should be consistent with that of hearing individuals [26,27], but there is very little evidence regarding lateralization of language-based memory functions. However, these findings and assumptions should be considered in the context of emerging evidence suggesting increased righthemisphere involvement in the processing of certain aspects of ASL (e.g., ASL spatial descriptors that require classifier constructions) among deaf signers [26,28,29].

Finally, although there are an increasing number of U.S.-based psychologists trained to work with deaf and hard of hearing individuals, only a handful of neuropsychologists have the linguistic and cultural competencies, as well as the clinical training experience, to provide services to deaf individuals using ASL. The majority of these clinicians are practicing in pediatric settings where opportunities for access to high quality services and resources are different relative to the adult population. Therefore, there is a need for more guidance on conceptualizing and performing neuropsychological evaluations and related procedures with deaf adults with epilepsy.

Given the factors mentioned above, presurgical neuropsychological batteries, fMRI studies, and Wada testing with deaf patients communicating via ASL require a careful and thoughtful approach that mitigates health disparities, and promotes equity to ensure the highest level of validity to inform surgical outcomes [17]. The following case study highlights the intricacies of conducting a comprehensive neuropsychological evaluation and subsequent fMRI and Wada testing of a Deaf adult using a certified and qualified ASL interpreter.

Methods – case report

Developmental history

This case study involves a 43-year-old, right-handed, Deaf Caucasian male, who uses ASL as his primary language, but also has knowledge of written English. His medical history was significant for medically refractory focal seizures with and without impaired awareness, status post vagal nerve stimulator placement, prior history of pneumococcal meningitis, severe bilateral sensorineural deafness, hypertension, hyperlipidemia, obstructive sleep apnea, and anxiety who presented for a neurological workup to determine surgical candidacy. Results from the magnetic resonance imaging (MRI) with quantitative analysis suggested that the right hippocampus is qualitatively decreased in size with increased FLAIR signal, and quantitatively measures more than 2 standard deviations below the mean for his age. These findings were consistent with mesial temporal sclerosis. Additionally, the positron emission tomography (PET) scan indicated decreased metabolic activity associated with the right temporal lobe, including the anterolateral, anterior and anteromedial portions, when compared to the left side. Results from the three-day video EEG indicated that the ictal onset and temporal intermittent rhythmic delta activity (TIRDA) specifically supported the right inferolateral-anterior temporal as the region of epileptogenicity.

The patient was the product of a full-term and uncomplicated pregnancy. Early language and motor developmental milestones were reportedly achieved within typical time expectancies. By 7 months of age, he experienced pneumococcal meningitis that caused a high fever which in turn caused subsequent seizures and bilateral deafness. He lost his previous developmental gains. When he recovered from the pneumococcal meningitis, his seizures subsided. At 10 months of age, he was enrolled in an early intervention parent/infant program for the deaf that focused on the acquisition of spoken rather than signed language. In the summer before kindergarten, he was enrolled in a communication disorders program where he continued to learn how to speak. When he started kindergarten, he was placed in a "failure program," as reported by his mother, because he was unable to communicate. A student in the communication disorders program suggested that they should use sign language. He began learning Signing Exact English (SEE) and in a short time span, he reportedly was able to better communicate. SEE is considered a manual system of communication that incorporates signs for vocabulary that follow English grammar. It is not considered a true sign language like ASL, which has unique linguistic features that differ from spoken and written English.

At 7 years his seizures returned, and were described as staring and having sialorrhea, but he was able to continue using SEE. At approximately 8 or 9 years of age, he was diagnosed with epilepsy and started on anti-epileptic medications, which were frequently changed as his seizures persisted. During this time, he continued to communicate using SEE and it was not until he was 12 years of age that he began learning ASL, as this is when the first educational program in the area allowed for this language of instruction.

History of presenting concerns

His seizures continued and increased in severity as he began experiencing generalized convulsions. His last generalized seizure occurred in the late '90s, but he continued to experience focal seizures with and without impaired awareness. In 2001, a vagus nerve stimulator (VNS) was implanted and he began experiencing an aura of a déjà vu sensation and a strange taste in his mouth. After the VNS placement, he only experienced auras that lasted about 30–60 s without loss of awareness. He continued to drive and would stop on the side of the road when he felt an aura. Of note, he once was involved in a motor vehicle accident due to having a seizure with loss of awareness.

He was admitted for continuous video electroencephalography (EEG) monitoring for three days to adjust medications, obtain information on seizure localization, and to assess for surgical candidacy. EEG results recorded nine focal impaired awareness seizures characterized by aura, staring, loss of awareness, lip smacking, right-handed motor automatism, left-hand dystonic posturing, and ictal fleeing behavior. These features suggested a right temporal focus. The ictal onset and the temporal intermittent rhythmic delta activity (TIRDA) supported a right inferolateralanterior temporal region epileptogenicity. His MRI results indicated right mesial temporal sclerosis, and mild decreased metabolic activity associated with the right temporal lobe and right thalamus.

Neuropsychological evaluation

Neuropsychological evaluation was performed to evaluate baseline cognitive functioning and to provide information on any lateralization and treatment outcomes. The neuropsychological evaluation was informed by the American Psychological Association (APA)'s ethics code and multicultural guidelines for assessments to ensure a culturally-informed evaluation [30]. As the primary author does not have the cultural and linguistic competence to provide services to this patient population, attempts were made to find an appropriate referral source in the geographic area. Given the lack of such a provider and to promote timely provisions of services, the primary author reviewed the limited available research for providing a neuropsychological assessment for Deaf individuals using ASL. Consultation with the co-authors, including a bilingual and multicultural neuropsychologist trained in working with Deaf individuals using ASL, occurred to identify the most appropriate test battery for this specific patient. A certified and qualified ASL interpreter, provided by the patient's hospital, was used throughout the evaluation. A pre-conferencing meeting was held with the interpreter before the formal evaluation to discuss the process, train them in test procedures, translate all test stimuli and instructions, and answer any questions. Additionally, for the prose memory test, the interpreter video recorded administration of the story. Given the types of tests administered and the use of the ASL interpreter, the results were interpreted as a fair, but possibly lower estimate of the patient's true functioning.

Premorbid intellectual functioning was obtained using an abstract visual reasoning test and was scored in the high average range. Neuropsychological test results did not reveal a clear lateralization, but there was a trend indicating greater compromise of the non-dominant hemisphere. This was due to primary deficits with graphomotor constructional praxis in the copying of a complex figure that affected his memory of the figure, and confrontation naming. There were also secondary weaknesses with memory of simple figures, simple auditory attention, and aspects of processing speed. His language and learning and memory of verbal material were intact. He exhibited a strengths in phonemic fluency (using ASL phonological cues: handshapes), learning of a story, and learning and memory of a word list as these were in the High Average range.

fMRI procedure

A Functional Magnetic Resonance Imaging (fMRI) was ordered by the interdisciplinary team to obtain further information on language and memory lateralization. Tasks performed in the fMRI were completed in both English and ASL at the request of the team. English tests were administered first during the initial imaging session. The ASL tests were administered subsequent to this session. The tasks included reading sentence completion, silent word generation, story reading, verbal animacy encoding, spatial size encoding, ASL movie, ASL picture naming, thumb movement, tongue tapping, and lips pucker. On the reading story, reading sentences and word generation tasks administered in English, there was activation in the left inferior frontal gyrus in the expected region of Broca's area. There was no significant activation in the right inferior frontal gyrus. All three tasks produce activation in the language pre-supplementary motor area. On the ASL picture naming task, the patient was shown a series of images of common objects and imagines signing their names. For this task, there was robust activation in the left greater than right pre-supplemental motor area, left premotor cortex, left dorsolateral prefrontal cortex, and left Broca's area. There was also activation in the left frontal eye field in addition to the premotor area. On the ASL movie task, the

patient watched a movie clip of an individual giving a speech in ASL. The control condition was a static picture of an individual signing. For this task, there was relatively symmetric activity within the bilateral superior temporal sulci, as well as increased activation in bilateral premotor cortices, left expressive speech area, and left dorsolateral prefrontal cortex. Both the ASL picture naming and the ASL movie tasks produce activation in the medial superior and middle temporal areas. For the verbal memory encoding tasks in English there were significant clusters of activation in the left and right hippocampi. For size memory encoding tasks in English, there was significant activation in the left hippocampus. Therefore, the fMRI found bilateral receptive and expressive speech activation that was slightly greater in the left hemisphere and mixed hippocampal activation during memory tasks. The fMRI did not provide information on lateralization of memory.

Wada test procedure

Based on the findings from the neuropsychological evaluation, the neuroimaging results, some lack of clarity regarding laterality of language and memory functions, and the unique language experiences of the patient, the interdisciplinary team ordered a Wada test that was administered based on the standard procedures set by the Medical College of Georgia [31]. Additional consultation with co-authors occurred to identify any available literature to inform the viability of conducting a Wada test in ASL, identify the most valid culturally and linguistically appropriate and accessible process using ASL instructions and stimuli, and determine how best to work with the interpreter in the evaluation suite given the specific logistics of Wada testing.

A different certified and qualified ASL interpreter was included in the Wada testing due to lack of availability of the interpreter used during the neuropsychological evaluation and fMRI procedure. As part of a pre-conferencing session, the results of the neuropsychological evaluation were discussed with the new interpreter in an effort to share the experiences with the patient to date, their specific language needs and provide training by the primary author in the Wada procedures. The provider and interpreter met one week before the Wada evaluation to review and adapt the test procedures, and again for an hour before the evaluation to practice the procedures and for the interpreter to meet and consult with the patient. At the first meeting, the Wada procedure was explained, including the objective of assessing receptive and expressive language, as well as assessing memory. As the test procedures were reviewed, the interpreter served as the cultural liaison and helped to inform the provider when to adapt items to ensure that the objects presented for the memory portion could either be fingerspelled or signed using the functioning hand. Given ASL is a visuospatial language and vocabulary and grammar are produced at times using both hands, the sentences used for repetition and reading were revised to ensure that they could be meaningfully produced by this patient using either the left or right hand. The instructions for a modified token test to assess receptive language were also revised due to grammatical differences in English and ASL (e.g., "show me" versus "point to"). The interpreter was asked to pay attention to linguistic parameters of the patient's language as these could represent errors in lexicon or grammar. On the day of the evaluation, the provider and interpreter met with the patient to discuss procedures and what they should expect. The patient was informed to always look at the interpreter, to stay awake during the procedure, and to remember the objects he would be shown. Questions were answered and the patient was prepared for the Wada. Fig. 1 shows the timeline for the test procedures.

The patient was brought into the interventional radiology room and prepared for an internal carotid angiography through the femoral artery. The right hemisphere was examined first, and a

large crossover of the posterior communicating artery was found through the angiography. The interpreter stood on the right side, next to the patient's head and explained the start of the procedure. The provider stood above the patient's head, next to the interpreter and informed the interpreter of the instructions. The patient was asked to raise both arms up with their hands open and palms facing up. He was asked to sign from 1 to 10 using both hands and then to continue counting until one arm drops. Three milligrams of methohexital sodium were injected into the right hemisphere, and complete flaccid hemiparesis was not attained; therefore, an additional milligram was provided. The patient demonstrated the expected flaccid hemiparesis of the left arm and face, he remained alert, and there was no affective response. Using his right arm and hand, he was asked to state his name and the day of the week. He was then asked to open his mouth and snap his fingers. The patient was able to respond appropriately and follow these simple verbal and motor commands. He was then presented with and asked to name eight objects, for which he chose to fingerspell (i.e., representation of the English written alphabet using specific handshapes) except for one item that was produced via sign. He then was asked to squeeze the provider's hand using his left hand to test for grip strength. The patient was provided a second dose of methohexital sodium at about two minutes after the first injection, since his grip strength had returned. At this time, his language was assessed. Receptive language was assessed using a modified token test during which he was asked to follow a simple one-step verbal command, which he was unable to complete. To ensure that this behavior was not an error due to interpretation or related language issues, the question was repeated six times, but he was unable to comply until the sixth attempt. This may have been due to the posterior communicating artery crossover. The patient was then asked to name a different object and repeat short sentences, which he was able to do. At four minutes and fifty seconds after the first injection, he was able to comply with a simple two-step command. The patient was able to continue communicating using clear ASL produced by the left-hand throughout the evaluation. At ten minutes and thirty-eight seconds, his EEG returned to baseline. His memory was then assessed, and he exhibited episodic recall for the objects by spontaneously recalling three of the objects. When provided with a recognition format, he accurately identified all eight of the items with one false positive error.

After 30 minutes, the test was repeated for the left hemisphere. The internal carotid angiography revealed no crossfilling. He was again asked to raise both of his arms and count from 1 to 10 using both hands. Four milligrams of methohexital sodium were injected and he demonstrated the expected signs of flaccid right hemiparesis involving the face and arm. He initially had difficulties focusing on the interpreter due to drowsiness from the anesthetic, but this subsided quickly. He remained alert and there was no affective response. The patient was aphasic and unable to respond to any questions or comply with simple motoric commands. He was then asked to name eight new objects but was unable to perform the task and fixed his gaze towards the interpreter as each object was named for him. At two minutes and twenty-nine seconds after the methohexital sodium was injected, he was able to fingerspell his last name and the name of the hospital. His grip strength on the right hand was tested and had returned. A second dose of methohexital sodium was injected and he remained aphasic, and was unable to comply with simple commands, name objects, read short sentences, or repeat short phrases. At four minutes and fiftyfive minutes after the first injection, he was able to read a short sentence in English and comply with simple one- and two-step commands, name objects, and repeat a short sentence provided in English and interpreted to ASL. His EEG returned to baseline at eight minutes and six seconds. His memory was assessed after ten minutes. He did not exhibit episodic recall for any objects



Fig. 1. Preparation and Process of Wada Testing.

and was unable to spontaneously recall the stimuli. When a recognition format was used, the patient was able to correctly identify six of the eight objects with two false positive errors.

Results

In summary, the results of the neuropsychological evaluation suggested a trend of greater compromise in the non-dominant hemisphere. Please see tests and results in Table 1. The test results indicated greater dysfunction of the non-dominant hemisphere, due to deficits in learning and memory of a complex figure, and secondary weaknesses in memory of simple figures. However, there was also a select area of impairment in naming.

FMRI results indicated bilateral receptive and expressive speech activation that was slightly greater in the left hemisphere and mixed hippocampal activation during memory tasks. These results were further supported and clarified by that of the Wada test that revealed left hemisphere representation for ASL. The results for verbal memory suggest that there was adequate functional memory reserve in the contralateral hemisphere. The functional adequacy of memory in the ipsilateral hemisphere was not optimal. This may be due to the impact of seizures and corroborates neuropsychological data of lateralization to the right hemisphere. The results of the neuropsychological evaluation, fMRI, and Wada testing suggested optimal surgical candidacy irrespective of the risk for further decline in visual memory after surgery. The patient underwent a right anterior temporal lobectomy and right amygdalohippocampectomy. At eight months post-surgery, he was reported to be seizure-free with no cognitive or functional complaints.

Discussion

This case study fills a gap in the existing literature and may serve as a guide for neuropsychological evaluation, fMRI imaging, and Wada testing in deaf patients with epilepsy who use ASL as their primary language. While we are aware that further research is needed to validate existing measures used with deaf signers and to create a battery that is truly culturally and linguistically sensitive for deaf neurological patients, the current work provides a comprehensive, well-thought-out, and collaborative approach illustrating modifications and accommodations incorporated into a traditional presurgical evaluation that integrates an ASL interpreter. Our goal was to highlight that in the absence of expertise in working with deaf people with ASL fluency, successful evaluation and treatment can occur through consultation with trained peers, an extensive review of validated instruments and procedures, careful selection of a culturally- and linguisticallysensitive battery and related procedures, and close collaboration with qualified interpreters and the patient.

It is important to note that ASL has unique linguistic features and syntax that are different from spoken or written English. It is not directly translated from English given the visuospatial modality, the sequential and simultaneous features, and that there often are no sign equivalents for many English words [11]. Therefore, tests that are heavily verbal and depend on knowledge in English are generally not considered appropriate [14].

For professionals with no ASL proficiency, certified and qualified interpreters must be involved prior to the evaluation to ensure that test instructions and content are understandable, as well as linguistically and culturally relevant following the patient's language preferences. Throughout this process, the provider and the interpreter should be collaborating with the patient to determine their individual needs and provide accessible and appropriate language and related test instructions and stimuli. It also is helpful to score tests and discuss behavioral observations with the interpreter after testing to discuss nuances of the experience, any misunderstandings on the part of the neuropsychologist, and any unique aspects of the testing process that potentially affect test results and related interpretations. Our findings illustrate that cross-cultural evaluations with ASL interpreters require significantly more time, but are vital when evaluations cannot be conducted in the language of the patient, especially for fMRI and Wada testing, given the

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Table 1

Neuropsychological Tests and Results.

Premorbid Intellectual Functioning	Standard Score	%ile	Description
TOPF Demographic Predicted	SS = 109	73	Average
Language and Communication	Standard Score	%ile	Description
Phonemic Fluency (5-1-U handshapes) ²⁴	0.96	83	High Average
Semantic Fluency Average (AFVC)	0.09	54	Average
Confrontational Naming (BNT)	17	<0.1	Impaired
Verbal Memory & Learning	Standard Score	%ile	Description
ASL Stories Test - Story Memory ²¹	Standard Score	70IIC	Description
Story 1 Immediate Verbal Memory	0.82	79	High Averag
Story 2 Immediate Verbal Memory	-0.05	48	Average
Story 1 Delayed Verbal Memory	0.12	55	
Story 2 Delayed Verbal Memory	-0.74	23	Average
5 5 5		23	Low Average
Percent Retained	-	=	=
HVLT (Form 5 - Translated)			
Trial 1	-	-	-
Trial 2	-	-	-
Trial 3	-	-	-
Total Recall (1–3)	58	79	High Averag
Delay Recall	59	82	High Averag
Discrimination Index	57	76	High Averag
Percent Retained	55	69	Average
Attention & Executive Function	Standard Score	%ile	Description
Digit Span (WAIS-IV)	42	21	Low Average
LDSF, LDSB, LDSS	-	-	_
Spatial Span (WMS-III)	13	84	High Averag
LSSF, LSSB	_	_	-
Trails B (cognitive shift)	42	21	Low Average
WCST, Categories	_	>16	Intact
Total Errors	51	53	Average
Perseverative Errors	50	50	Average
Reasoning Ability	Standard Score	%ile	Description
Similarities (WASI-II)	44	27	Average
Matrix Reasoning (WASI-II)	59	82	High Average
Perceptual and Spatial Function	Standard Score	%ile	Description
Perceptual Reasoning Index (PRI)	SS = 110	75	Average
Block Design (WASI-II)	53	62	Average
Hooper Visual Organization Test	54	66	Average
Rey-O Copy (Copy)	-	2-5	Borderline-Impaire
Visual Spatial Memory & Learning	Standard Score	%ile	Description
BVMT			
Trial 1	34	5	Borderline-Impaired
Trial 2	61	86	High Average
Trial 3	45	31	Average
Total Recall (1–3)	46	34	Average
	37	10	Low Average
Delayed Recall			Intact
	-	>16	
Discrimination Index	-	>16 6-10	
Discrimination Index Percent Retained		>16 6-10	
Discrimination Index Percent Retained Rey-O		6-10	Borderline-Low Averag
Discrimination Index Percent Retained Rey-O Immediate	21	6-10 <1	Borderline-Low Averag
Discrimination Index Percent Retained Rey-O Immediate Delay	21 <20	6–10 <1 <0.1	Borderline-Low Averag Impaired Impaired
Discrimination Index Percent Retained Rey-O Immediate Delay Recognition	21	6-10 <1	Borderline-Low Averag Impaired
Discrimination Index Percent Retained Rey-O Immediate Delay Recognition Percent Retained	21 <20 33	6–10 <1 <0.1	Borderline-Low Averag Impaired Impaired Borderline-Impaired -
Discrimination Index Percent Retained Rey-O Immediate Delay Recognition Percent Retained Fine Motor Dexterity	21 <20 33	6-10 <1 <0.1 4 -	Borderline-Low Averag Impaired Impaired Borderline-Impaired -
Delayed Recall Discrimination Index Percent Retained Rey-O Immediate Delay Recognition Percent Retained Fine Motor Dexterity Grooved Pegboard Dominant hand	21 <20 33 - Standard Score	6-10 <1 <0.1 4 - %ile	Borderline-Low Averag Impaired Impaired Borderline-Impaired - Descriptio
Discrimination Index Percent Retained Rey-O Immediate Delay Recognition Percent Retained Fine Motor Dexterity Grooved Pegboard Dominant hand	21 <20 33 - Standard Score 48	6-10 <1 <0.1 4 - %ile 42	Borderline-Low Averag Impaired Borderline-Impaired - Descriptio Average
Discrimination Index Percent Retained Rey-O Immediate Delay Recognition Percent Retained Fine Motor Dexterity Grooved Pegboard Dominant hand Non-dominant hand	21 <20 33 - Standard Score 48 56	6-10 <1 <0.1 4 - %ile 42 73	Borderline-Low Averag Impaired Borderline-Impaired - Descriptio Average Average
Discrimination Index Percent Retained Rey-O Immediate Delay Recognition Percent Retained Fine Motor Dexterity Grooved Pegboard Dominant hand Non-dominant hand Processing Speed	21 <20 33 - Standard Score 48 56 Standard Score	6-10 <1 <0.1 4 - %ile 42 73 %ile	Borderline-Low Average Impaired Borderline-Impaired - Descriptio Average Average Descriptio
Discrimination Index Percent Retained Rey-O Immediate Delay Recognition Percent Retained Fine Motor Dexterity Grooved Pegboard Dominant hand Non-dominant hand Processing Speed Trails A (visual search)	21 <20 33 - Standard Score 48 56 Standard Score 41	6-10 <1 <0.1 4 - %ile 42 73 %ile 18	Borderline-Low Average Impaired Borderline-Impaired - Descriptio Average Average Descriptio Low Average
Discrimination Index Percent Retained Rey-O Immediate Delay Recognition Percent Retained Fine Motor Dexterity Grooved Pegboard Dominant hand Non-dominant hand Processing Speed Trails A (visual search) Coding (psychomotor scan)	21 <20 33 - Standard Score 48 56 Standard Score 41 44	6-10 <1 <0.1 4 - %ile 42 73 %ile	Borderline-Low Average Impaired Borderline-Impaired - Descriptio Average Descriptio Low Average
Discrimination Index Percent Retained Rey-O Immediate Delay Recognition Percent Retained Fine Motor Dexterity Grooved Pegboard	21 <20 33 - Standard Score 48 56 Standard Score 41	6-10 <1 <0.1 4 - %ile 42 73 %ile 18	Borderline-Low Average Impaired Borderline-Impaired - Descriptio Average Average Description Low Average

Note. Test of Premorbid Functioning (TOPF); Animals, Fruits, Vegetables, Clothing (AFVC); Boston Naming Test (BNT); Hopkins Verbal Learning Test (HVLT); Wechsler Abbreviated Scale of Intelligence, Second Edition (WASI-II); Wechsler Memory Scale, Third Edition (WMS-III); Trails Making Test A and B (TMT A & B); Wisconsin Card Sorting Test (WCST); Wechsler Adult Intelligence Scale, Fourth Edition (WAIS-IV); Brief Visuospatial Memory Test (BVMT); Rey Osterrieth Complex Figure (Rey-O); Patient Health Questionnaire-9 (PHQ-9); and General Anxiety Disorder-7 (GAD-7).

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visuospatial nature of the language, the physical logistics of the procedures, and the need to optimize visual access to language in the imaging and surgical spaces. Budgeting additional time for these evaluations will support efforts to make specialty services accessible to deaf patients. This is consistent with efforts to promote equitable access to high quality care for all and ethical mandates for clinical neuropsychologists. Clinically, this is important for avoiding misdiagnoses and potentially adverse outcomes.

In summary, these results indicate that the utility of a comprehensive presurgical neurological workup, which includes neuropsychology testing, fMRI imaging, and Wada testing, for deaf patients with epilepsy who use ASL can be as informative to those conducted on hearing speakers of English. However, the nuances of working with deaf ASL signers requires the naïve neuropsychologist to consult with trained colleagues, work closely in selecting and collaborating with qualified and certified ASL interpreters with experience in behavioral health settings, and collaborate closely with the interdisciplinary team in the surgical suite. Moreover, it emphasizes the great need for neuropsychologists who are able to conduct such work directly with this linguistic minority group.

Limitations

There were several limitations evident in this case study. First, there are almost no documented cases of administering neuropsychological test batteries or Wada procedures for deaf signing patients with epilepsy. Anecdotally, the authors of this paper are aware that these evaluations occur for children and adults with some frequency, but often with no guidance to the lead neuropsychologist or with minimal consultation with an experienced peer. In the current case, the neuropsychologist performing this evaluation had significant experience conducting cultural neuropsychological evaluations with hearing patients using languages other than English, as well as working with spoken language interpreters. However, they did not have the linguistic or cultural knowledge, training, and experience to work directly with members of the deaf community without additional support.

Second, interpreters were used for the neuropsychological evaluation, fMRI session and the Wada test, and as such, the results need to be viewed with some caution. There is an ethical concern for the method by which tests were translated into a different language [32]. The interpreter worked with the neuropsychologist to translate all tests before the evaluation, to be prepared for the evaluation. Care was taken to ensure that the content would be understandable to the patient while still preserving as much of the standardized language as possible; however, this cannot be verified as the neuropsychologist only consulted with peers on the various ethical and professional issues, which potentially reflects what occurs for any professional not trained to work with a linguistically diverse population. More research is needed to develop tests that are linguistically and culturally appropriate, but also to better understand the typical neuropsychological underpinnings of cognitive and behavioral functions of deaf individuals who use a formal sign language, and how this may be disrupted by epileptiform activity among deaf patients with epilepsy.

From a social justice perspective, having the opportunity to learn about the patient's history, was humbling and eye-opening. His language development was delayed not only because of sequalae from medical conditions, but because there was a bias and lack of information regarding the use of sign language, as well as limited educational opportunities in languages other than English. These stories of unjust, prejudicial treatment by hearing professionals due to a mere lack of information are all too common. While deaf signers often require specialized standards of practice, providers largely do not have the adequate training and experiences, but continue to use available instruments and procedures developed and validated for monolingual, native English hearing speakers [11]. Thus, it is important that the field continues to invest the time and resources needed to remove barriers for this underserved population by providing the highest quality and most accessible services.

Conclusion

The results from this evaluation demonstrate that a comprehensive epilepsy pre-surgical evaluation of a deaf individual who uses ASL as their primary language can result in a successful postoperative outcome. Our case highlights the importance of considering surgical management in people with drug-resistant focal epilepsies and comorbid conditions. Presurgical assessment should be completed by an interdisciplinary team who strategically collaborates with the patient, interpreters, and professional consultants.

Ethical Statement

This is a case study, therefore, IRB approval/review was not required. However, there is an umbrella IRB application for collection of all pre-surgical epilepsy data, including neuropsychological, fMRI and Wada results. IRB University of Utah Application #00137934.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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