

Language Remediation in a Case of Wernicke's Aphasia Post Herpes Simplex Virus Type 1 Viral Encephalitis

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Abstract

Objective: This report aims to disseminate a successful language remediation in a case of Wernicke's aphasia following encephalitis caused by herpes simplex virus type 1.

Methods: A female patient was referred two months post onset of Herpes Simplex Encephalitis (HSE) with severe Wernicke's aphasia, confusion, environmental disorientation, and dependence in activities of daily living. Non-standardized portions of the Porch Index of Communicative Ability (PICA) was administered in speech-language assessment. A treatment program following the Context-Based Therapy was applied on an intensive schedule, with emphasis on the auditory system. PICA's multidimensional scoring system was used to monitor patient's progress.

Results: Sixteen months post onset of Wernicke's aphasia, patient was able to perform auditory comprehension/memory tasks with gestural/ visual augmentation and demonstrate no deficits on a non-standardized PICA, other than occasional delays and self-corrections. Patient became independent and functional in activities of daily living.

Conclusion: There is no proven method for rehabilitation of the Wernicke's aphasia. The successful rehabilitation including language remediation in this case suggests that a treatment program following the Context-Based Therapy may be helpful in rehabilitation of Wernicke's aphasia caused by HSE when administered on an intensive schedule.

Keywords: Wernicke's aphasia; Herpes simplex virus; Viral encephalitis; Speech-language rehabilitation; Context-based therapy

Abbreviations: HSV-1: Herpes Simplex Virtual Type 1; HSE: Herpes Simplex Encephalitis; PICA: Porch Index of Communicative Ability

Introduction

Herpes simplex virus Type 1 (HSV-1), along with HSV-2, are neurotropic members of the Herpesviridae family. HSV-1 most commonly causes cold sores but can also lead to genital herpes infections, while HSV-2 predominately causes genital herpes. Transmission occurs via mucosal surfaces including the oral and respiratory surfaces or through compromised skin, such as by sharing drinking vessels or utensils, kissing, and other high-risk skin-to-skin contact. HSV-1 is usually contracted during infancy or childhood through exposure to an infected adult. Transmission does not require an active infection or visible sores and can be spread through asymptomatic viral shedding [1,2]. There is a high seroprevalence in the general population, with an estimated 70-90% of asymptomatic individuals harboring the HSV-1 [3,4]. There is currently no cure for HSV but symptoms can be managed with antiviral medications.

During transmission, HSV-1 first enters the body through epithelial cells. HSV can then infiltrate sensory neurons that innervate the infected epithelial cells. Unlike many viruses, HSV has the ability to evade the host's immune system by sheltering inside neurons. HSV can also affect multiple organs via viral dissemination, although this is mostly seen in neonatal cases [1,2]. If left untreated, HSV can lead to Herpes Simplex Encephalitis (HSE), which has a high mortality and high morbidity rate, causing 10-20% of viral encephalitis cases [5]. Exactly how HSV-1 infiltrates the central nervous system to cause to HSE is greatly debated, with the olfactory bulb and trigeminal ganglia implicated in mice models [6]. The olfactory pathway is the most likely avenue of infection, as recurrent herpes labialis, which occurs in the trigeminal ganglia, rarely leads to HSE [7]. HSE normally affects the

temporal lobe, which is responsible for retention of visual memory, language comprehension, processing of sensory input, and emotion. Therefore, symptoms of HSE include aphasia, confusion, and behavioral changes there can also be extra temporal involvement, including the frontal and parietal lobes, with an estimated 16% of patients with HSE having extra temporal infections [8]. This localization of infection is thought to be caused by the proximity of the temporal lobe to the olfactory bulb or trigeminal nerve [9] or preference of HSV for limbic cortices. Briefly, HSV-1 causes degeneration of cell nuclei and loss of plasma membranes, leading to multi-nucleated giant cells. This in turn causes inflammation, hemorrhaging, and eventual tissue necrosis and liquefaction [10].

When damage occurs to Brodmann area 22 (Wernicke's area), located in the superior temporal gyrus, such as by HSE, problems with auditory and written comprehension occur. This is known as Wernicke's aphasia (e.g. fluent or receptive aphasia), and is characterized by fluent, albeit nonsensical verbal and written language that conveys little useful information. Individuals affected by Wernicke's aphasia are usually unaware of any errors in their speech or writing. Those with Wernicke's aphasia can string sentences together with real words paired in nonsensical combinations, neologisms (newly synthesized non-words), or both [11]. Depending on the location of brain lesion,

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Received September 25, 2014; **Accepted** November 16, 2014; **Published** November 18, 2014

Citation: McMicken BL, Kunihiro A, Wang L, Rogers K (2014) Language Remediation in a Case of Wernicke's Aphasia Post Herpes Simplex Virus Type 1 Viral Encephalitis. J Clin Case Rep 4: 441. doi:10.4172/2165-7920.1000441

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auditory and reading comprehension can be affected to varying degrees. For example, in alexia with agraphia, individuals cannot understand written language but have limited ability to understand words spelled out loud and can generate written output [12].

The likelihood of positive outcomes are greatly increased when appropriate interventions are implemented without delay. Acyclovir, an antiviral medication, is the standard pharmacological treatment method and is recommended to begin even before a positive diagnosis is made [13]. Despite reduced mortality with acyclovir [14], dose has not been associated with HSE outcomes [13] nor has there been an appreciable reduction in morbidity [15,16]. Even those who receive prompt treatment still a chance of experiencing detrimental side effects, including behavioral modifications, amnesia, emotional changes, aphasia, and other cognitive deficits [17-24]. A variety of rehabilitation methods have helped to improve recovery in activities of daily living and semantic processing, such as positive reinforcements or extinction techniques [25], associations (motor imagery strategy) [26], and community-based programs [27]. Other pharmacological interventions also seem to improve psychiatric issues, including carbamazepine [28] and risperidone [29]. There have also been reports of improvements in HSE-aphasics, both spontaneously [30-32] and through interventions [33].

Case Reports

A female patient in her late 60's, was referred 2 months post onset of HSE with Wernicke's aphasia. This individual was a left handed lifetime author and teacher, who's MRI had demonstrated encephalitis involvement of the left temporal lobe and left inferior frontal lobe. She was treated with antiviral drugs within 72 hours of onset.

At the time of initial speech-language pathology assessment the following factors were noted:

- Bodily functions and structures: presented with language, memory, confusion and disorientation impairment; no physical deficits.
- Activities limitations: unable to be independent in activities of daily living (ADL) due to confusion and disorientation, required assistance or supervision.
- Participation restrictions: none, as long as there was assistance or supervision in ADL and speech activities.
- Environmental factors: lived with long-time partner in a supportive environment with frequent visits from friends.
- Personal factors: A nearly life-long writer and "intellectual worker" she was extremely frustrated with her situation, her lack of independence, and her dependence literally for ADLs in nearly every situation due to the confusion. "My life crashed" as she said.

Symptoms at the time of speech-language and cognitive evaluation were of a severe Wernicke's aphasia, confusion and environmental disorientation. Non-standardized portions of the Porch Index of Communicative Ability (PICA) were administered in the speech-language assessment. Assessment began with auditory comprehension of nouns through identification of objects, and then proceeded to naming, sentence formulation, word imitation, matching a written word with an object, following written directions, writing names of objects, writing to dictation, copying, and finally, as a facilitating technique, out loud reading of her books. Results are as follows:

Verbal expression

The patient demonstrated a fluent aphasia characterized by some appropriate automatic phrases, perservative responses, occasional jargon and run-on non meaningful speech. Patient demonstrated zero success when repeating words. None of the objects were named correctly on a spontaneous basis or with cues. Errors typically reflected verbal paraphasias, perseveration and neologisms. Some responses were intelligible but in error, some were not intelligible. During conversation speech was fluent, but contained word finding and meaningless run-on sentences. Patient would have instances of saying "that's wrong," "I don't know," but this behavior was mixed with jargon and apparent general lack of awareness of content.

Auditory comprehension

Comprehension of spoken information was severely impaired at the word and phrase level. She did not demonstrate consistent auditory comprehension other than some social phrases. Directions required the modeling use of a body part. Self-monitoring was not displayed.

Visual processing

There was no ability demonstrated to match a word with an object or follow a simple written direction. However and quite unexpectedly, patient could read her books out loud with about 60% accuracy with no comprehension of the material. When a line was taken out of the book context, she did not appear to be able to transfer the activity of reading it out loud.

Graphic processing

Patient was able to accurately write her name. She could not write the name of an object, nor could she copy without direct assistance, otherwise her attempts were jargon (meaningless combination of letters) and perservative (repetition of the same meaningless word).

Treatment Philosophy

According to Altschuler et al. [34], no proven method exists for rehabilitation of the Wernicke's aphasia, and there is little direction in the literature for speech-language pathology treatment of Wernicke's aphasia patients. In this case, it was decided to follow the suggestions of Marshall using Context-Based Therapy [35]. It was recommended that the patient participate in a language treatment program on an intensive schedule, with emphasis on combining elements of the PICA multidimensional scoring system [36], and Marshall's description of Context-Based Therapy [35], an offshoot of the Context Centered Therapy concept of Wepman [37]. Emphasis was on the auditory system, after Schuell's treatment philosophy [38], thus attempting to increase the functional nature of communication across all language modalities. Also, exploration of all patient's language abilities using her previous writings, hobbies and editorial work as stimuli were used to assist with both receptive and expressive language modalities. Any communication means possible was explored, from pointing to a picture of a food on a menu, to use of a clock or calendar, or facial expressions, and also capitalizing on residual words to achieve meaning in a message [34].

Duffy summarized treatment based on Schuell's approach by stating that aphasia is a deficit that crosses all language modalities and the auditory system is critical due to its role in both the acquisition and continuous processing of language [39]. Treatment was therefore focused on the auditory system rather than each language modality

with the underlying hypothesis that improvements will spread to the other language areas [38].

According to Marshall [40], some advantages of a Context-Based Approach with the Wernicke's patient are:

- "Helps the patient cope with immediate demands to communicate in a real-life context (at home).
- Capitalizes on the patient's strengths (speaking, preserved syntax, pragmatics, mobility, speed of responsiveness).
- Promotes compensation (using all modalities of communication) and provides a scaffold for caregiver education.
- Provides immediate success in communication and sets the patient up for later deficit-specific treatment".

The goal of the context-based approach is to improve listening comprehension and information exchange in valid communicative frameworks. The patient's long term partner agreed with this approach and sat in on each session to provide background, feedback, and to continue with treatment "life oriented" tasks in the home setting.

Treatment Goals

The PICA multidimensional language scoring system was used as a task and expectation guide to increase correct receptive and expressive skills (Appendix A). The initial PICA plan based on the scoring system was to increase responses from 4, 5, and 6 to 7, 8, 9, and 10 using patient's previous writings, editing skills and hobbies as a basis of language retrieval. Increased auditory comprehension was the underlying goal with visual skills as augmentation and correctly monitored speech as the product. The patient was an "intellectual worker" and wanted to work on her skills as an author and editor.

Short term (3 months) goals

Increase task level PICA scores from 4 (incomprehensible but differentiated) and 5 (comprehensible, perservative but not an attempt at the task item) to task level PICA scores of 6 (inaccurate attempt at task item) and 7 related, inaccurate, almost accurate, and continuing to task level PICA scores of 8 (accurate with a cue), 9 (accurate with a repetition) and 10 (accurate with self-correction). Assist patient to be functionally independent in the home through gestural or augmented communication, train her partner to assist her in a supportive manner with speech correction, modeling and writing.

Long-term (6 months to 12 months) goals

Increase task-level PICA scores from 6,7,8,9 and 10 to 11 (accurate, incomplete, delayed), 12 (accurate, incomplete), 13 (accurate, delayed) and 15 (accurate prompt, complete) Initially the clinician and the patient alternately read patient's writings out loud and discovered she would consistently correct spelling and grammatical errors in the "readings" even though it was on a recognition, not comprehension, level. Patient had been a proof reader. This strategy was incorporated into treatment including writing the corrected sentence. We gradually increased complexity of grammar, tense, and spelling. In 6 months she increased from verbal PICA naming 4, 5 to 6, 7, and occasionally 10, could match word with object 50% on 10 items and was able to copy and occasionally write a meaningful word.

Results of Treatment

At 16 months post onset of Wernicke's aphasia, patient could perform auditory comprehension/memory tasks with gestural/ visual

augmentation and first letter for naming if needed. Her reading on the PICA tasks had become 15's (word to object, sentence description direction to of manipulating a card and object). She lacked visual processing memory for longer than two simple commands. BJ demonstrated immediate auditory comprehension of objects and sentences describing objects. Some sample therapy tasks included:

- Pick out the incorrect word in a sentence both visually and auditorily presented and correct it: "grapes grow on a brick".
- Change the tense of the incorrect verb: "the dog were playing".
- Change the tense of the incorrect noun: "the dog were playing".
- Who, what, where, when, why exercises visual and auditory, with sentence and short paragraph. Visual was always completed first.
- Confrontation naming with spelling augmentation if needed (first letter).
- 3-5 object/card memory including naming.
- Drawing of objects she could not recall.

Sixteen months post onset, patient demonstrated no deficits on a non-standardized PICA, other than occasional delays (13) and self-corrections (10). At present we continue with tasks related to focus on her intellectual and home life such as:

- Create or read a line of writing and provide the next line.
- Correct complex sentences with grammatical errors and spelling errors.
- Use familiar books and book titles, cards and objects for confrontation naming and memory.
- Play "speech" Wheel of Fortune with concept and spelling.
- Formulate sentences asking therapist and partner to follow complex directions.
- Work on tasks she does throughout the day and assign language to them.

At 16 months post onset of receptive aphasia, patient is independent and functional in activities of daily living. She has resumed some professional writing, is accepting speaking engagements in which she will read her writings, discuss them on a limited basis, and interact socially with the audience. Patient remains bothered by auditory and visual memory for language material and a lack of the extensive command of vocabulary she once enjoyed. However, the patient is committed to working the rest of her life to regain her command of language. She is optimistic and once again enjoying celebrity status.

Conclusion

With no proven method for rehabilitation of the Wernicke's aphasia and little direction in the literature for speech-language pathology treatment of the condition, the successful outcomes of this case suggest that a treatment program following the Context-Based Therapy may be helpful in rehabilitation of Wernicke's aphasia caused by HSE when administered on an intensive schedule. The PICA multidimensional scoring system can a useful tool for assessment and monitoring of progress in rehabilitation.

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Appendix A

Porch Index of Communicative Ability Multidimensional Scoring

- 1 No response no response, no awareness of task
- 2 Attention no response, but patient attends to the tester
- 3 Minimal incomprehensible and undifferentiated
- 4 Unintelligible incomprehensible but differentiated
- 5 Intelligible comprehensible but not an attempt at the task item
- 6 Error inaccurate attempt at task item
- 7 Related inaccurate, almost accurate
- 8 Cued accurate, after cue is given
- 9 Repeated accurate, after instructions are repeated
- 10 Corrected accurate, self-corrected
- 11 Incomplete-delayed accurate, responsive, incomplete
- 12 Incomplete accurate, responsive, incomplete, prompt
- 13 Complete-delayed accurate, responsive, complete/complex
- 14 Distorted accurate, responsive, complete/complex, prompt
- 15 Complete accurate, responsive, complete, prompt, efficient
- 16 Complex accurate, responsive, complex, prompt, efficient