

Clinical Management of a Patient with Pure Word Deafness

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Introduction

The problem of pure word deafness along with other problems that in some manner isolate language functions from other brain functions has attracted much attention from aphasia researchers (Oppenheimer and Newcomb, 1978; Shoumaker, Ajax and Schenkenberg, 1977; Goldstein, Brown and Hollander, 1975; Albert, Sparks, Von Stockert and Sax, 1972; Ernest, Monroe and Yarnell, 1977; Gazzaniga, Glass, Sarno and Pozner, 1973; Kanshepolky, Kelly and Waggener, 1973). The interest in such problems has largely centered on their value in helping to understand localization of language functions. Although patients with isolated language deficits present unusual challenges to traditional treatment, little literature is available to help the clinician. A clinical report by Kirshner and Webb (1981) is one of the few exceptions. It documents the value of sign language therapy in a case of what they describe as "selective involvement of the auditory verbal modality." However, Kirshner and Webb only briefly describe the treatment, a combination of lip reading and of teaching Amerind and Ameslan. They also do not provide generalization data beyond the actual number (127) of signs their patient was taught in a period of 8 months.

In what follows we will describe a patient who had well-documented pure word deafness and was intensively treated for a period of 6 months for a total of 124 treatment sessions. This information may be of use to other clinicians in planning treatment for such patients.

Background

The patient, (CP), a 65-year-old right-handed man was hospitalized initially at another hospital in December 1979 for arteriosclerotic cardiovascular disease with congestive heart failure. He had an 8th grade education and, prior to his retirement at age 60, had been employed as a laborer. He was single and lived with his 82-year-old father. During his hospitalization, a left posterior temporal lobe infarct was identified by computerized tomography after he experienced an abrupt decline in behavioral and linguistic functioning. However, the diagnostic workup, which included psychiatric, psychological, audiology and speech pathology evaluations, resulted in a diagnosis of organic brain syndrome with psychosis. CP was discharged in July 1980, having had no treatment for his communication disorder. One and one-half years after the onset of symptoms, in July 1981, he was admitted to our hospital because he was depressed and a suicide risk. This was CP's first psychiatric admission.

He came to our attention by accident. In September 1981 he was transferred from his psychiatric ward to a medical ward because of repeated cardiac arrhythmias. A nurse there noted in passing the "he did not seem crazy" and suggested that he might be of interest to us. He was.

His comprehension difficulties, coupled with appropriate social behavior, ability to read moderately well, rapid, disfluent and somewhat paraphasic speech, led us to undertake a comprehensive diagnostic evaluation. Part of the evaluation was directed toward documenting the nature of his language problem and toward providing a baseline against which rehabilitation gains could be measured. Part of the evaluation was directed toward justifying our working diagnosis. We will discuss each in order.

Documenting Language Disorder. Mr. P was given The Boston Diagnostic Aphasia Examination (BDAE), The Boston Naming Test (BNT), The Ravens Colored Progressive Matrices (RCPM), The LaPointe-Horner Reading Comprehension Battery for Aphasia (RCBA) and the short form of the Token Test. These tests revealed severely reduced auditory comprehension and moderately well-preserved speech, writing, and reading comprehension. (Figure 1 shows his BADE profile.) On the basis of these tests, we made the diagnosis of pure word deafness. We will discuss post-treatment changes on these tests later in the paper.

Corroborating Tests. We corroborated his diagnosis in a number of ways. Pure tone audiometry demonstrated moderate sensorineural loss above 2,000 Hz. Brain stem evoked potentials were abnormal only at the cochlear level, consistent with audiometric findings. Cortical evoked potentials were depressed, indicating higher-level central nervous system dysfunction. (Figure 2 shows a repeat CT scan done as part of our work-up.) This scan reveals an obviously large area of radiolucency in the left temporal lobe. It extends medially just inferior to the sylvian fissure and just superior to the temporal horn so that it is most probably deep enough to involve the auditory radiations in the temporal isthmus as they ascend from the medial geniculate body to Heschl's gyrus and Wernicke's area (Hanaway, Scott and Strother, 1977). In addition, the right temporal lobe shows absence of more than two gyri with corresponding atrophy of the sylvian fissure, supportive of the presence of smaller areas of cerebral infarction. This is the lesion pattern most consistently reported in pure word deafness (Earnest, Monroe and Yarnell, 1977; Goldstein, 1974; Kanshepolky, Kelly and Waggener, 1973; Parving, Salomon, Elberling, Larsen and Sassen, 1980; Oppenheimer and Newcomb, 1978; Ulrich, 1978; Nielsen, 1946).

We also pursued some additional behavioral indications for pure word deafness. A full neuropsychological evaluation conducted by writing all instructions contradicted the earlier evaluation; no evidence of generalized neuropsychological dysfunction was noted. Mr. P. was also able to identify correctly 40 of the 50 taped DLM environmental sounds, ruling out more generalized auditory agnosia. Finally, using written instructions and removing them before he carried out the commands, Mr. P's Token Test performance was significantly improved.

Treatment

Rationale. With regard to treatment, our approach was essentially compensatory. That is, rather than approach the patient's disproportionately severe auditory comprehension disorder directly, the decision was made to bypass the auditory modality. The promising Kirshner-Webb results with sign language furnished the rationale for the decision to teach sign. However, we used a total communication approach, pairing Amslan and idiosyncratic signs with their verbal referent. Because the patient was institutionalized and

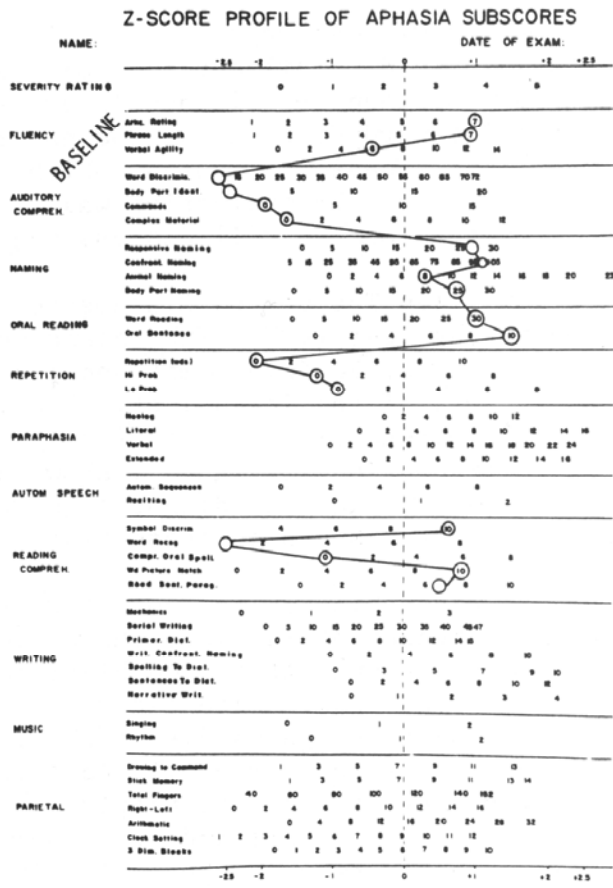


Figure 1. Boston Diagnostic Aphasia Examination Z-Score Profile: Baseline Data.

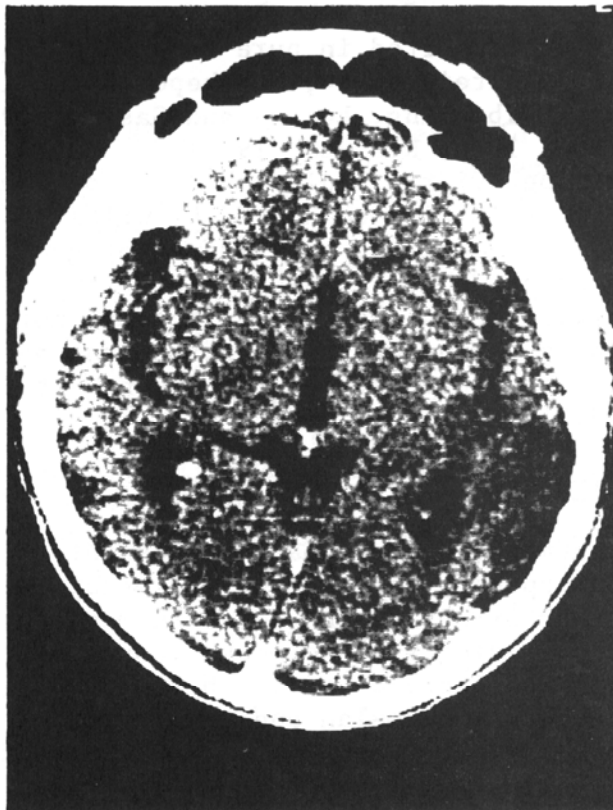


Figure 2. CT scan without contrast revealing bilateral temporal lobe lesions, the left much larger than the right.

on a supportive rehabilitation unit, idiosyncratic signs were justified by the relative ease of teaching each learned sign (regardless of system) to the staff with whom he was in daily contact. In addition to our total communication program, we also taught self monitoring of verbal output.

Description. The patient was seen daily for 45-minute sessions. The two major goals of therapy were first, to establish a receptive sign vocabulary and second, to provide a means for monitoring the parameters of rate and fluency during spontaneous verbal expression. Throughout the course of therapy, instructions and most messages were communicated in writing, the patient responding verbally. In order to present our approach to therapy clearly, I will discuss each goal separately. However, keep in mind that both the progressive elaboration of the signed communication system and the progression from a one-word pacing strategy to self-monitoring of verbal expression occurred simultaneously.

To establish an initial core vocabulary we utilized only objects that were present and actions that occurred in the immediate environment. Thus, some of our first signs were iconic gestures for words like drink, pen, write, look, listen, etc. Subsequently, we expanded the vocabulary to include objects and actions that were not available within the immediate context of treatment. Most of these were hospital-related people, places and activities and included signs for nurse, doctor, medicine, shower, toilet, etc. They were taught by writing the word for the patient and providing him with the sign. Eventually, pronouns, prepositions, modifiers and WH words were incorporated.

The transition from recognition of single signs to recognition of sentences consisting of two or more signs was achieved in a similar manner. The sentences were first written on paper, then signed and spoken simultaneously. The patient's task was to respond verbally and to check the accuracy of his response against the written sentence. Subsequently, yes/no questions were signed, the patient's task being to decode the questions and provide the appropriate answer. The WH questions and 1 and 2 stage commands were presented in a similar manner.

As mentioned earlier, our patient also presented with rapid, dysfluent speech. The first stage of treatment directed toward monitoring verbal expression consisted of the imposition of a strict rate control program. Initially, a pacing board was utilized and the patient was required to speak one word at a time pairing each word with one move on the board (Helm, 1979). This resulted in an immediate and almost magic reduction of word and phrase repetitions as well as a significant reduction in the number of verbal and literal paraphasias. However, because this method, as well as the actual paceboard itself, was too cumbersome, we moved to a less rigid method of rate control following one month of this technique. In principle, the next stage was identical to that of the pacing strategy. However, in this stage we required the patient to click the top of a ballpoint pen at constituent junctures. This provided him with a prosodically more acceptable manner of speech while maintaining a reduced rate of repetitions and paraphasias. Eventually, the ballpoint pen was replaced with a signed cue from the clinician, initially at constituent junctures and subsequently only occasionally as reminders to slow down during the course of a conversation.

Pre-treatment to Post-treatment Comparisons. Figure 3 shows a composite of CP's Boston profile at time of initiation of treatment and at treatment termination. It is obvious that the pattern of his deficits in no way changed following treatment. That is, Mr. P was as purely word deaf when he left the hospital as he was when he arrived.

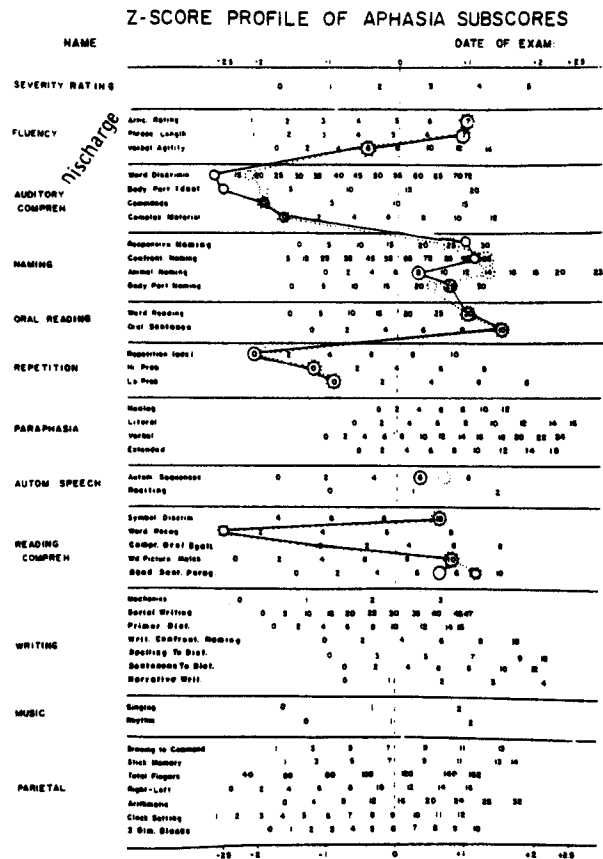


Figure 3. Boston Diagnostic Aphasia Examination Z-Score Profile Composite Scores. Baseline = — Discharge = ···

However, at the time of discharge CP had acquired a receptive sign vocabulary of 152 words. He responded in a functional manner (with 80% accuracy) to signed simple declarative statements, yes/no questions, and WH questions, both within the context of treatment and in other hospital situations. He also learned to increase the comprehensibility of his speech by maintaining deliberate control of his speaking rate and reducing paraphasic errors.

In terms of other more explicit measures, however, some of the generalized effects of his treatment can be observed. (Figure 4 shows pre- and post-treatment gains on the Token Test, The Boston Naming Test (BNT), The Reading Comprehension Battery for Aphasia (RCBA) and the Ravens Colored Progressive Matrices (RCPM). (Figure 5 shows our pre- and post-treatment analysis of CP's paraphasic speech errors on the Boston Naming Test and reflects the general improvement in speech production obtained through his treatment.)

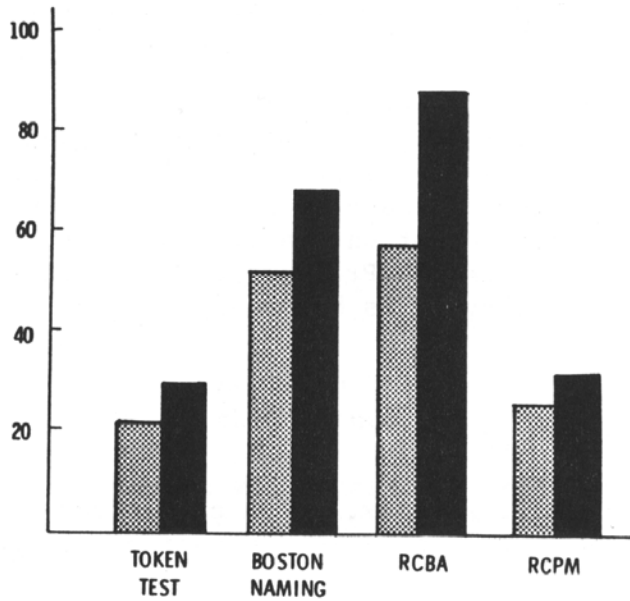


Figure 4. Total raw scores for the Token Test, the Boston Naming Test, The Reading Comprehension Battery for Aphasia, and the Ravens Colored Progressive Matrices. Baseline = ■ Discharge = ▨

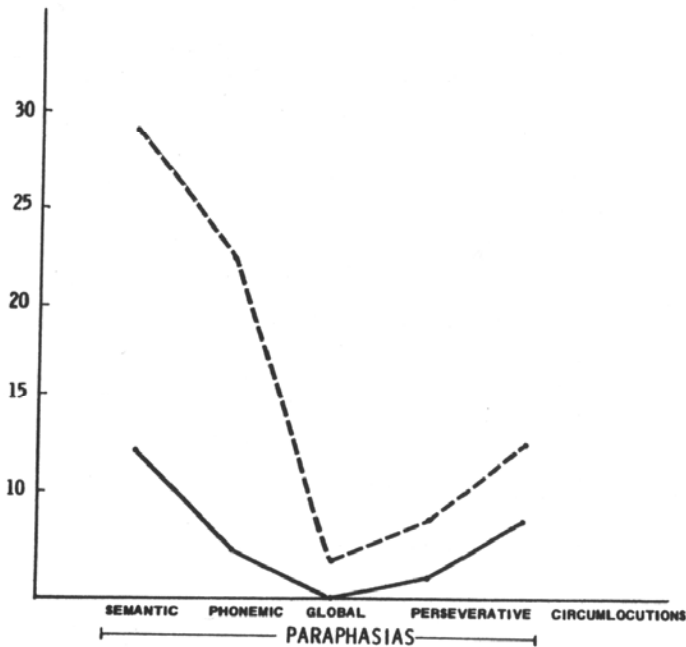


Figure 5. Total number of paraphasias by type on the Boston Naming Test. Baseline = ---- Discharge = —

We believe that these scores indicate that CP increased his ability to focus on and appreciate linguistic and paralinguistic cues and to modify his behavior as a result of that increased sensitivity. That is, we taught him to use all information available within the context of a communication event rather than attempting to retrain him on those very skills devastated by his disorder.

The major success of Mr. P's treatment program must surely be that these strategies allowed for functional encoding and decoding of language. To insure that these systems continue to be functional, Mr. P's discharge plans included a referral to a home health care agency in his community that had a certified speech/language pathologist on staff. Our last follow-up report indicated that our Total Communication Program is being reinforced at home and that his improved language behavior is being maintained in this way.

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DISCUSSION

- Q: Tell me the sequence of these lesions, how they occurred in time?
A: We are not sure how they occurred.
- Q: You don't know if damage occurred to both sides simultaneously?
A: No, we don't. It is possible that he had a right hemisphere lesion first that was not apparent clinically or that they occurred simultaneously. One of the problems with the first scan was that it was artifact-laden and was essentially useless to us.
- Q: He seemed to have a lot of aphasia. Did you do any comparison of tests administered in their standardized way with tests presenting all the stimuli in writing?
A: Yes, we gave the Token Test in this manner and a full neuropsychological evaluation in this manner.
- Q: So this person was aphasic in addition to having pure word deafness?
A: That is a matter of interpretation. A close look at the symptoms reported for most cases of pure word deafness reveals a pattern very similar to Mr. P. That is, their output is typically paraphasic, possibly because of inability to monitor their own speech and some general effects of brain damage are usually apparent as well. The truth is that "pure" in this syndrome and any other "pure" syndrome is not to be interpreted as "pure as the driven snow." Rather it seems to be a monster modality deficit in comparison to relative sparing of other language abilities.
- Q: Since he was reading at the sentence level why didn't you choose some visual linguistic augmentative system rather than gestural communication?
A: He communicated verbally. He also carried a magic slate for others to write on if that is what you mean by visual augmentative system.
- Q: Could this patient lipread at all? Did you make any comparisons between testing him with and without visual facial cues?
A: Our observations indicated that lipreading was too complex. Lipreading tasks confused the patient.
- Q: Did you decide he had no potential for improvement in auditory comprehension?
A: Yes we did.
- Q: Do you think that is fair? Why were you so absolute in that position?
A: This patient could not repeat at the phonemic level. He could not repeat at the word level. He did not understand the most simple of social utterances. Throughout the six month course of therapy he demonstrated no capacity for understanding verbally presented material in an isolated manner. However, we did not deny him access to auditory stimulation since every one of our signs was paired with their verbal referent.
- Q: Did you speculate as to what was happening with regard to the tie between the lexical and semantic systems and improving his fluency?

Is it just perhaps that he was buying some time and able to retrieve better?

A: We felt that the results reflected an overall increased sensitivity to the communication event itself. I think probably that because he was doing something deliberately he may well have been buying some time.

Q: What were his verbal problems like? Were they mostly word retrieval or phonological problems?

A: When he initially presented to us his speech was very rapid and consisted of many word and phrase repetitions. Within these repetitions he had more semantic paraphasias than he did literal paraphasias, although clearly he had both.

Q: Did you test his oral reading? Was this better?

A: Yes, when he was reading he seldom had disfluencies or paraphasias. We found that whenever there was concurrent visual information he was able to monitor his verbal output much better.

Q: Did you test his ability to match rhyming words presented in writing?

A: Yes, using two different paradigms. Initially, we presented him with a field of six words consisting of the rhyme pair and semantic, vowel change, final consonant change and unrelated foils, and asked him to "find the two words that rhyme." On this task he performed poorly. However, when given one of the rhyme words and asked to "find the word that rhymes with this one" he performed with 100% accuracy. We also did some other informal investigations that we did not report in the paper such as recognition of melody, pitch perception and identification of voices.

Q: How did you rule out hysteria or malingering?

A: On the basis of his obvious lesions, his depressed cortical evoked potentials and his normal behavior. It did not occur to us to consider malingering.

C: But aphasic patients will also have depressed cortical evoked potentials and left hemisphere lesions and they are not called pure word deaf.

Q: You commented that he did well on taped environmental sounds. Did you do anything with taped voices where he had to identify whose voice was on the tape?

A: Yes we did. He could distinguish my voice from his voice and he could distinguish between his family members' voices. He could also recognize different musical instruments. He distinguished between a violin, a piano, a horn, etc.

Q: Do you happen to know if the left temporal lesion extended into the angular gyrus?

A: I don't believe that it did.

Q: Was there ever a time post-onset that he reported auditory hallucinations

A: There is an indication in the medical records that he reported auditory hallucinations. However we question the reliability of these reports from the previous hospital.

Q: Was he concerned about this problem?

A: Yes very much so.