

A CASE HISTORY OF ACQUIRED AUDITORY VEBRAL AGNOSIA

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ABSTRACT

A young girl had been developing speech and language normally until about the age of three and a half. Over a period of weeks, her communicative ability deteriorated to the point where she was simply pointing and using gestures. She was diagnosed as having suffered bilateral temporal lobe seizures. The seizure activity has been under control since December, 1977. For the last three years, this girl has been attending a school which uses the Aphasic Method. This approach had helped her in terms of communication to some extent, but she was auditorily and orally virtually nonfunctional. It was decided to teach her sign language. Interestingly, although the prognosis was guarded, she has developed a significant degree of auditory-oral language. Sign language appeared to help her close the verbal system. Her auditory-oral repertoire has continued to grow, both with and independently of continued sign language exposure.

Acquired auditory verbal agnosia in children is an impairment in reception and/or expression of previously acquired language without loss of hearing. Children with this disorder respond to nonverbal, environmental stimuli to varying degrees. There have been a number of reports in the literature in recent years concerning acquired childhood aphasia (Campbell and Heaton, 1978; Cooper and Ferry, 1978; Gascon et al., 1973; Landau and Kleffner, 1957; Rapin et al., 1973; Shoumaker et al., 1974; Worster-Drought, 1971). The syndrome is characterized by seizures preceding or following the loss of previously acquired language. Loss of varying degrees of communication skills occurred as much as six months (Shoumaker et al., 1974) to three years (Gascon et al., 1973) before the presence of seizures. In fact, while EEG's were abnormal, in three cases (McKinney and McGreal, 1974; Rapin et al., 1973) clinical seizures were never observed. The most comprehensive article concerning this disorder is Cooper et al. (1978), which provides a summary and insights into 45 cases of acquired childhood aphasia. They reported that language abilities improved in some children with medication alone, in others with medication plus speech and language therapy, and in some with no particular intervention. Most studies reported that poor comprehension of language was followed by expressive difficulties. Cooper and Ferry (1978) stated that "children become mute, use jargon or produce odd sounds, exhibit misarticulations, inappropriate substitutions of words, and anomia, or resort to gestures and grunts" (p. 177). Landau and Kleffner (1957) noted that some children acquire a vocal quality and pitch which is similar to that of the deaf child.

Recovery from the disorder is variable. Of the 45 cases which Cooper and Ferry (1978) reviewed, "42% were left with severe residual deficits, 24% had mild-to-moderate residual deficits, and 33% made complete recovery" (p. 182). Complete recovery can require from several weeks to years. However, most children suffer some degree of aphasia ranging from auditory verbal agnosia with no functional verbal skills to varying degrees of learning disabilities (Gascon et al., 1973).

Speculation as to the cause of the disorder ranges from some type of subacute viral encephalitis (Harel, Walsh, and Menkes, 1973) to vascular anomalies (Rapin et al., 1973).

CASE HISTORY

I met Lori and her mother November 8, 1978. The mother wanted me to assess Lori, who was nine, to determine whether I could help improve her comprehension and use of speech. Lori's speech and language production appeared to have been within the broad-normal range of development until she was about three and a half. At that time, over a period of weeks, her use of speech deteriorated from sentence level, to phrases, to words, to finally pointing to objects instead of calling them by name. Also, she no longer responded to oral language. However, she continued to react to environmental, nonverbal auditory cues, such as a phone ringing.

An EEG report administered when Lori was four indicated bilateral temporal lobe epileptiform activity which had been present for some time. Lori was placed on medication to control the seizures. An audiogram performed at about this time indicated that hearing was normal, as was a brain scan. Lori's mother reported in November of 1973 that she had observed a series of petit mal seizures over a period of weeks. She observed these seizures after Lori's speech had deteriorated to simple sounds and after the above-mentioned EEG report indicated bilateral temporal epileptiform activity. The doctors at the Hospital for Sick Children in Toronto speculated that Lori must have exhibited petit mal seizures perhaps before her speech deteriorated and certainly during the deterioration. Lori's mother became cognizant of the seizures only after she was made aware of what to look for (i.e., a series of eye blinks).

A speech and language report in December of 1973 indicated that there was no apparent comprehension of spoken language. Lori used only a few sounds and words: "ow" for hurt, "ssss" to name matches, and occasionally "no" and "that". It was also reported that Lori exhibited an impairment of pre-language abstract abilities, i.e., sorting, associations, etc. In 1974 the Speech and Language Department recommended that visual reading and sign language be tried.

Lori was placed in prekindergarten and kindergarten in 1974 and 1975, respectively. At the invitation of Lori's School Board in June of 1975, a psychiatrist from the Clarke Institute was asked to consult with her teachers and parents regarding language training and future placement. When she was four, a report from the Hospital for Sick Children indicated that her general behavior was typical of brain dysfunction — distractible, impulsive, and over-reactive, but also reported that she was cheerful and socially responsive when her communication difficulties were not being brought into evidence. The 1975 report confirmed that behaviorally Lori was adapting very well to her communication difficulties, indicating that her unusual alertness and intelligence may well have been attributable to the excellent and warm home environment provided. From school reports and my observations, Lori's excellent emotional adjustment to her language difficulties and her strong motivation to improve her communication abilities has continued to this day. The 1975 report also indicated that Lori was able to comprehend about 50 signs which her teacher had made up. It was noted that in a teaching experiment, Lori showed a startling ability to acquire and subsequently recall signs shown to her. Though no formal intelligence testing was attempted, the report indicated that other than in the area of conventional communication skills, Lori exhibited unusual alertness and intelligence. It was suggested that Lori would be an excellent candidate for exposure to gestural communication based on the Ontario sign language combined with verbal training.

Because of Lori's auditory-oral difficulties, Lori's kindergarten teacher felt that Lori would soon become frustrated in a regular classroom. At about this time, Lori's mother became aware of classes for aphasic children in Belleville, Ontario. In September of 1976, Lori was enrolled in an aphasic classroom in Belleville. She is still attending this school. At the school, the Hisky-Nebraska Test of Learning Aptitude was administered. Lori demonstrated average or better than average ability for her age in visual areas, but exceedingly poor auditory capabilities.

Lori's last seizure occurred on December 31, 1977. In a medical discharge summary in September of 1977, it was reported that EEG studies still showed seizure activities in both temporal lobes. A follow-up EEG study in September of 1978 indicated that seizure activity was significantly reduced, and that temporal lobe functioning was much more normal. The most recent EEG study in November of 1979 showed no abnormal seizure activity; brain functioning was normal. She has not been on any medication for the last two years.

When I saw Lori first, she had been attending a school for aphasic children for two years. The method used to teach Lori was the Association Method (DuBard, 1976). The method is somewhat similar to that reported by Lea (1965) in that visual presentation is coupled with auditory information. However, the main focus of the Association Method is verbal rather than visual, as in Lea's. As with the expressive speech program for children with acquired aphasia discussed by Campbell and Heaton (1978), the procedure for teaching speech with the Association Method involves incremental sequencing of phonemes into words and then connected speech. The general structure of the method is to see it, say it, write it, and recall it at every stage of production. The procedure for developing connected speech with the Association Method is similar to the method advocated by Campbell and Heaton (1978) in that one language structure is used in a variety of situations and expanded until mastery is assured. While she was at the school for aphasic children, on weekends when she didn't go home, she stayed in the junior deaf children's residence. Here she learned some sign language and to finger spell, which, of course, improved her communicative abilities. These skills combined along with visual aids such as mirrors and diagrams and placement and tactile stimulation, assisted Lori in developing articulatory mastery because they helped her to obtain a mental image of what was required.

When Lori was brought to my Department in November of 1978, both Lori's mother and the school director and her teachers were concerned about her progress. Lori was able to read and comprehend at a very rudimentary level: She recognized only a number of basic words and her level of reading comprehension was, therefore, not at a grade one level. Her speech was virtually unintelligible. Lori's voice quality, which was hypernasal and lacked natural quality and inflection, compounded her intelligibility problems. With her inability to understand speech, Lori was essentially auditorily-orally nonfunctional. Thus, it was impossible to communicate to her such ordinary things as that the family was going shopping, going on a trip, etc.

The assessment of Lori's speech and language capabilities confirmed that Lori was verbally nonfunctional. On the Goldman-Fristoe Test of Articulation, Lori's speech was characterized by extensive omissions for which she was not stimuable. Her free speech was essentially unintelligible, except where she was reading material that she had learned at school. Even with this material, vocal quality, as discussed above, and articulation were poor. She was able to discriminate only the simplest items auditorily on the Peabody Picture Vocabulary Test. Even with visual cues, she was able to discriminate accurately only the first 15 items. On the Reynell Vocabulary Comprehension Scale "B", she was able to comprehend only the most rudimentary words and concepts, and even with these items, she often needed repetition. She was not able to discriminate among the objects by function. However, on the Raven's Coloured Progressive Matrices, Lori performed at the 50% level, corroborating previous impressions and tests that other than in the auditory modality, Lori's performance capabilities were within the broad-normal range for her age.

Considering Lori was nonfunctional auditorily and orally, and considering Lori's age of nine years, three months, I felt that an alternate mode of communication had to be considered. It was now more than five years since Lori had lost her ability to comprehend auditory language. The prospects of her developing functional verbal skills were certainly poor (Cooper and Ferry, 1978).

At a meeting in January of 1979 with Lori's parents, her teachers, and the school director, I recommended that Lori be taught to sign. There were concerns expressed that adding another mode of communication to what Lori was already learning at school might be overwhelming; however, it was decided to try teaching Lori sign language for six weeks and then to evaluate her progress.

I recommended that Lori use signing because she had demonstrated some facility with signs in kindergarten. Also, her school was able to provide the appropriate teaching necessary, and Lori's parents and family were willing to learn sign along with her. The sign dictionary used was published by Gallaudet College Press (Bornstein et al., 1975). Lori received instruction in sign for a half an hour a day, five days a week, from a student teacher at Sir James Whitney School who had just completed a course in sign.

At the next meeting in mid-March to evaluate Lori's progress with sign, a number of interesting observations were reported. Lori had learned to use signs for approximately 140 nouns, plus a number of adjectives. She was now also combining these nouns with adjectives and using the few basic questions she had been taught. With the advent of sign, Lori's comprehension and use of verbal material increased significantly and markedly both in the home and at school. Interestingly, when Lori learned a sign, she seemed then able to comprehend and use the equivalent verbal symbol. In other words, signing appeared to provide the vehicle to enable Lori to decode auditory symbols and use oral speech. As Lori's auditory comprehension and verbal output increased, so did her intelligibility and her vocal quality and prosody improve to the point where now all people involved with her feel that her speech clarity is good, and that the quality of her speech and vocal melody is essentially normal. Since Lori had made so much progress, it was decided to continue signing instruction to the end of the school year, with the focus being to teach verbs and to sign in complete sentences.

By the end of the school year, Lori was able to speak in two and three-word utterances. She was attempting to communicate orally in more and more situations. For example, she now was ordering her hamburger at McDonald's, and was more willing to try to communicate with strangers. It was decided at the June meeting to continue sign instruction because of its communicative function, and because of its mediating value in teaching auditory-oral language.

Lori has continued signing instruction to date, and her verbal facility has continued to grow. Lori is able to answer the phone and relay simple instructions. She phones her mother at work to tell her what is happening at home. At school, she is able to follow most simple auditory instructions, and communicates orally with her peers and teacher effectively and routinely. She is speaking in three and four-word utterances now. Occasionally, she will use a complete sentence, especially if it corresponds to a form taught at school.

In April of this year, I did some formal speech and language testing. On the Goldman-Fristoe Test of Articulation, Lori demonstrated only a few misarticulations, these being predominantly among the blends. She was stimulable for all her misarticulations, except for four of the blend sounds. None of her misarticulations were omissions, the misarticulation which characterized her speech in November and December of 1978. As mentioned earlier, now Lori's vocal quality and pitch were comparatively normal, whereas in the original assessment, her voice was hypernasal and lacked inflection. On the Peabody Picture Vocabulary Test (form B), Lori's ceiling score was 54, whereas in my earlier assessment, she was unable virtually to take the test. Also, it was interesting to note that when Lori did not know a word, she was able to make a correct discrimination with auditory cues (i.e., simpler verbal descriptors), something which she could not do earlier. On the Carrow Test for Auditory Comprehension of Language, Lori performed at the four year old level, missing only 34 of 101 items. Most of the items Lori missed, as expected, involved more complex syntactical structures, such as the past and future tenses.

DISCUSSION AND IMPLICATIONS FOR TREATMENT

Lori made significant gains in verbal communication with the introduction of sign. It should be noted that a number of studies have reported there was a high correlation between the improvement in speech and language functioning as electroencephalogram abnormality decreased (Campbell and Heaton, 1978; Landau and Kleffner, 1957; Worster-Drought, 1971; Shoumaker et al., 1974). Undoubtedly, this fact partially explains Lori's marked improvement in terms of auditory-oral language. However, her EEG report in September of 1978 indicated that brain activity was much more normal, and this was a full five months before the introduction of sign. Also, Lori's verbal improvement coincided directly with the advent of the teaching of sign. Therefore, the introduction of sign language helped Lori develop functional verbal abilities.

Cooper and Ferry (1978) noted that manual communication cannot be considered a panacea for children with acquired aphasia. They recommended that should previous language functioning not recover, then a decision must be made about the direction of the therapy, whether it be manual communication, writing, or a communication board. However, implicit in their discussion is the idea that first, attempts should be made to elicit and improve verbal communication skills before any alternate system of communication is considered.

I philosophically disagree with this position. Symbolic communication is certainly an inherent human attribute. It is my feeling, therefore, that these children should be provided with a mode of communication immediately. It is unconscionable to prevent these children from developing some mode of communication, probably even if it can be shown that a nonverbal communication system might ultimately reduce a child's potential for verbal communication. However, there is no evidence to indicate that the introduction of a communication system other than a verbal one decreases eventual auditory-oral language recovery. In fact, in the case of Lori, it definitely expedited her awareness of auditory cues, and her facility for oral communication. The use of any symbol system will not only enhance one's cognitive awareness, but one can speculate that it may provide a syntactical awareness such that, if verbal language develops, it will be possible to sequence words in the conventionally accepted mode more easily and naturally.

Keeping in mind that enhancing communication skills, whatever the symbol system or systems used, is the primary focus, I believe that a global approach to developing verbal communication skills is the procedure of choice. By this, I do not mean that the child should be bombarded with alternate modes of nonverbal communication. I believe first some exploration should be done to determine the best nonverbal mode of communication for the child. In the case of Lori, signing seemed to be the best symbol system to use. However, once an alternate communication system is selected, I think ancillary methods of enhancing communication skills are necessary. With Lori, finger spelling, visual and tactile feedback assisted more accurate articulatory placement. A global approach would certainly seem to be the best procedure to help children with acquired auditory verbal agnosia.

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