CLINICAL NOTE

"Pure Word Deafness": Implications for Assessment and Management in Communication Disorder—A Report of Two Cases

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ABSTRACT. Wee J, Menard MR. "Pure word deafness": implications for assessment and management in communication disorder—a report of two cases. Arch Phys Med Rehabil 1999;80:1106-9.

In "pure word deafness" after acquired brain injury, the auditory comprehension of words is much more impaired than other aspects of communication or cognition. Two cases are presented, one early and one late presentation. The key to diagnosis of communication disorders is to remember to assess all six basic aspects of language function and to be vigilant for coexisting diagnoses that can complicate such assessment (especially psychiatric diagnoses). Rehabilitation management of impaired communication should emphasize the teaching of specific coping mechanisms to the patient and to all others who are involved.

"PURE WORD DEAFNESS" (PWD) is not a hearing impairment, but an impairment in central auditory processing: an auditory agnosia primarily affecting recognition of spoken language. It is a rare presentation after stroke (or other acquired brain injury) in which impairment of auditory comprehension of words is more prominent than that of other aspects of communication or cognitive function.[1] The term "pure word deafness" was coined by Kussmaul in 1877.[2] The actual localization and neuropsychology of disorders of auditory processing continues to be debated.[1]

Communication impairments are important in clinical practice for two reasons: they challenge the affected individuals in adapting psychologically to their acquired impairments, and they challenge caregivers and significant others who are attempting to help the patient attain the highest possible degree of psychological and social reintegration. The interesting and uncommon syndrome of pure word deafness is important more generally in physiatry because it offers insight into the way that communication and cognition impairments interact to influence an individual's global functioning (ie, ability). In other words, it is difficult to separate cognitive and communicative aspects of a person's functioning, either clinically or in systematic quantitative analyses of data.[3,4]

Caregivers say that they value preservation of communication more highly than continence, mobility, and self care.[4] Communication disorders also are important in the medical-legal area; it has been suggested that failure of institutions to address such barriers to the medical care of affected patients might be a violation of the Americans with Disabilities Act provisions concerning equality of access to services.[5]

We present two cases of pure word deafness. One patient had pre-existing psychologic and social features that in themselves had not prevented her from living independently, but which interacted with a new communication disorder to pose a formidable threat to independence. This case illustrates the problems in initial diagnosis and early management. The other patient had a similar overall presentation, but a valid diagnosis of her communication disorder was not made for more than a decade after the causative events. Her case illustrates the long-term consequences to the patient and her significant others.

CASE 1: EARLY PRESENTATION

The patient was a 46-year-old left-handed divorced woman who, before this health episode, lived in a small house in a relatively remote area. She shared custody of two children with her ex-husband. Her elderly father lived in a nearby city.

Her medical history indicated that she had had a depressive disorder since her teenage years. She had at least one inpatient psychiatric admission for "depression," although she did not receive electroconvulsive therapy. At age 28 years, she was involved in a motor vehicle accident in which she suffered cervical fractures (C1 and C2). Afterward, she reported daily headaches and neck pain, and she subsequently habitually used prescription narcotic analgesic medication. The duration of unconsciousness and posttraumatic amnesia are unknown, but acquired brain injury was suspected. She regained sufficient independence to get married and raise a family, however, and to participate in school and employment, though never consistently. She participated in a drug rehabilitation program at age 39 years.

Her main physical health problem was mitral valve dysfunction; a murmur had been detected when she was 28 and was presumed to have resulted from previous rheumatic heart disease. Mitral valve replacement was performed when she was 31 years old, after which she was prescribed lifelong anticoagulation with Coumadin. Periodic international normal ratio testing showed that she was compliant. Four years later, she was admitted to her local hospital with an embolic stroke, despite this anticoagulation therapy. Motor deficit was minimal, but she required outpatient therapy for a communication disorder that was dominated by word-finding difficulties. Three months later, she was admitted for lysis of a clot that had developed in the mechanical heart valve.

The current episode began 2 years later, when she presented to her regional hospital with dyspnea. Prosthetic valve malfunction was suspected, and she was transferred to the regional cardiac center for management. Transesophageal echocardiogram confirmed the provisional diagnosis. A course of tissue plaminogen activator infusion was initiated, during which she sustained a recurrent embolic stroke. She experienced two

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Submitted for publication January 19, 1999. Accepted March 19, 1999.

No commercial party having a direct financial interest in the results of the research supporting this article has or will confer a benefit upon the authors or upon any organization with which the authors are associated.

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0003-9993/99/8009-5343$3.00/0

episodes of seizure activity and was started on phenytoin. Initial computed tomography (CT) of the head showed no hemorrhage or other acute process. Repeat CT 5 days later showed a large right middle cerebral artery territory infarction. Areas affected included the temporoparietal cortex, basal ganglia, corona radiata, and gray and white matter. Evidence remained of the previous left temporoparietal infarction. Coumadin was reintroduced. She was not believed to be a good candidate for cardiac reoperation.

First seen by a physiatrist 6 days after admission, she was alert but indifferent to the examiner's presence. Spontaneous verbal output was clear but bore little relation to the immediate situation. She did not follow one-step verbal commands, even with gestural demonstration and cueing. Left hemiparesis was worse in the upper extremity, and she had decreased response to threat in the left visual field. The corneal reflex was decreased on the left. She swallowed water without difficulty. Impairments included left-sided weakness, left-sided inattention, apraxia, impulsivity, concrete thinking, paranoid ideas, and difficulty in understanding the spoken word. She required significant assistance with all activities of daily living.

Because of her "alert but indifferent" clinical presentation and the possible combination of organic brain syndrome (new stroke, seizures, phenytoin therapy, previous stroke, suggested previous traumatic brain injury) with psychiatric disorder (delirium or psychosis, chronic depression, substance abuse), the Psychiatry Service was consulted. Formal speech-language assessment was also requested.

The speech-language pathologist found verbal expression, reading comprehension, and written expression to be relatively intact, consistent with pre-existing mild deficits. Moderate cognitive language impairments were typical of right hemisphere cerebrovascular accident, including decreased attention, concentration, and inferencing abilities. She had a profound deficit in auditory comprehension. Though no significant dysarthria was observed, she reported that her voice had changed pitch, that voices sounded distorted and cartoonlike, and that music sounded like unpleasant noise. It was concluded that she had pure word deafness with cognitive language impairments.

The patient was transferred to the acute inpatient rehabilitation unit, where the rehabilitation team worked out a routine of communicating with her exclusively by writing on a dry-erase board. Care was taken to make eye contact and to gesture in a friendly manner frequently. Speech therapy sessions emphasized lip reading. Audiometry and auditory brain stem evoked potentials were normal. The electroencephalogram was normal, so phenytoin was discontinued after 1 month, by which time she had become independent in transfers and, with the assistance of one person and a cane, could ambulate while wearing a Swedish knee cage on her left knee, which was prone to hyperextension. She became independent in propelling her manual "hemi" wheelchair throughout the hospital. Basic self-care activities required only minor assistance.

Approximately 8 weeks after admission, she was transferred to a freestanding university-affiliated rehabilitation hospital for better access to a "communications technology" resource so that she could explore augmented communication. Staff at the rehabilitation hospital later reported that she was initially cooperative with the prescribed program, but became increasingly distractible and tangential, "muttering to herself and perseverating on imagined topics." She began to refuse food and drink. Delusional thoughts were attributed to auditory sensory misperception. Risperidone was begun, with no noticeable benefits. We arranged for transfer back to the acute hospital. On return to our unit, she was disorganized in presentation and reported auditory hallucinations. When reestablished on the previous regimen of care, she settled clinically within a few days.

The focus of each of our interventions was to help the patient to deal with her comprehension deficit in each setting. Exercises aimed at improving auditory attention included vigilance tasks and minimal pair dictation and discrimination. In trying to interpret what people were saying around her, she frequently arrived at erroneous conclusions about her situation and tended to develop paranoid ideas. These could easily be corrected, and care was taken to do so promptly. To prevent misunderstandings, she was asked to request that all visitors write down what they wanted to say. She was reminded that she could not rely on her initial impression of what she heard. She was provided with a communication book in which to record all important information. A quiet environment was provided. She displayed a good sense of humor, but expressed to caregivers a profound sense of grief for her situation.

Reassessment approximately 20 weeks after her initial admission to the acute care hospital showed an improvement in auditory comprehension from "severe" to "moderate-severe." She could comprehend simple sentences in context and 50% of two-step commands for predictable material. Out-of-context material remained extremely difficult for her.

The main goal of discharge planning was to find for her a protective and supportive setting, preferably a group home. Meanwhile, she was transferred from the acute rehabilitation ward to a quieter transition ward, but was overseen by the same clinicians (there are no skilled nursing facilities in Canada). She reported that the quieter environment was easier for her. If people tried to talk to her, she told them that she was deaf, so they would not continue. She improved in lip-reading skills. After 3 months, she was transferred to a group home near her home community. The family physician to whom her medical care was transferred was notified of her special care needs related to communication. The local speech-language pathologist who had treated her after her previous stroke monitored communication issues in the group home. The group home operator was advised about the communication issues, and subsequent progress reports were quite positive.

CASE 2: DELAYED PRESENTATION

A 71-year-old woman was admitted to our rehabilitation hospital for musculoskeletal rehabilitation after a hemiarthroplasty for fracture of the femoral neck from a fall. She could read, but she was unable to comprehend speech; her physiatrist, upon assessment, recognized elements of pure word deafness. A speech-language assessment was completed. Her husband indicated that she had not been "deaf" until a recurrence of stroke 10 years previously.

Her first stroke had occurred at age 60 years, with infarction in the territory of the right middle cerebral artery. She was reported to have fully recovered from "temporary weakness on the left side of the body." Three months later, she sustained a second infarction, this time in the left frontal lobe. Her husband noted her inability to comprehend speech or to monitor her own speech. Two months after the recurrence of stroke, she was assessed by a physiatrist from a freestanding rehabilitation hospital, who judged her not able to cooperate with treatment. An audiologist was unsuccessful in attempting to gain her cooperation within her own home. An otolaryngologist arranged for complete audiology testing and found intact and normal eardrums. He obtained auditory brain stem evoked potentials, as she was unable to comply with standard testing. He concluded that she had adequate hearing bilaterally and did not require amplification of hearing.

Thereafter, she was cared for at home by her husband. Because it was obvious to him that she could understand his written but not spoken messages, he communicated to her by writing. He noticed a significant change in her speech in that she tended to speak very quickly and "babble." He found, however, that when he reminded her to speak slowly, she was easily understood. The patient recalled that she had been having occasional difficulty registering certain environmental sounds. When reassessed by the same physiatrist 8 months later, her husband reported that she was still having difficulty monitoring her speech and was failing to register sounds such as the whistling of a kettle. She had just begun to recognize the sound of the telephone, however. At this assessment, the patient was more cooperative and was able to read simple sentences coherently.

Nine months poststroke, she experienced a single seizure, and was started on phenytoin therapy, which has continued to the present day. No further seizures were noted. She had been receiving antihypertension therapy, but her blood pressure normalized after the seizure, and medication was discontinued because of side-effects.

Hearing aids did not help her. Her husband purchased a type telephone for the deaf for her, and she also used this to type messages. She attended a community center program for the deaf for 6 months in an unsuccessful attempt to learn lip-reading. The patient and her husband became socially isolated, largely because of her difficulties with communication. More recently, he has encouraged her to learn how to send messages via computer electronic mail.

During her current rehabilitation admission, she reported that she still could not hear music. Her memory for events was good. She repeatedly asked for another opportunity to learn lip-reading. She indicated that she has become used to "being deaf." She watched closed-caption television and spent most of her time reading books.

The origin and nature of the communication disorder were explained to the patient and her husband. Resources for learning lip-reading were explored. An approach to music that emphasized rhythm rather than tone was suggested to her husband.

DISCUSSION

Neuroanatomy and Neurophysiology

Both patients had elements of pure word deafness in addition to their other impairments. The syndrome has been described as a disorder associated with auditory linguistic stimuli, analogous to the acquired dyslexias, which are disorders associated with visually presented linguistic stimuli.[1] The disability lies in the inability to interpret all types of sounds, and patients frequently lose the ability to recognize melodies, but the effect on auditory comprehension of words is most prominent. It is not a problem of hearing perception, but rather an auditory agnosia for speech. It is thought to result from acquired impairment involving certain areas of the brain that are more central than the areas responsible for auditory perception, and brain stem auditory evoked responses are usually normal (although abnormalities have been reported when certain techniques are used[6]).

Localization of the lesion causing this entity has long been debated. For a clinically recognizable disorder of auditory processing to occur, it appears that either the temporal lobes must be affected to a substantial degree on both sides (because the auditory cortex is located laterally in the temporal lobes[7] or that the subcortical area of the left posterior temporal lobe must be affected.[2] It is conceivable, however, that a single lesion might disrupt essential connections to these regions or that a generalized degenerative disorder could affect both regions.[8]

Patients in whom the right hemisphere is affected often have more difficulty with interpreting prosody or affective intonation of language and interpretation of tone of voice, while patients with left hemisphere lesions are reported to present with more linguistic errors.[9] These clinical features often overlap. Other causes of deficits of auditory comprehension should always be ruled out, of course. For example, auditory inattention can exist in right-hemisphere-damaged patients,[10] causing an exaggerated attentional bias towards the right. Lesions of the insula can cause difficulties in understanding speech presented to the contralateral ear, despite maintenance of hearing sensitivity peripherally. This may be due to an amplification deficit and can be corrected by rerouting stimuli to the other ear with a hearing aid.[11] The peripheral auditory system should always be checked though audiograms and brain stem auditory evoked responses.

In both cases presented here, the syndrome of pure word deafness arose after the second recognized episode of stroke. There had been substantial recovery of functional independence after the first stroke, with at worst only a mild communication disability. Both patients were women. Both developed seizures after the second stroke, which affected the contralateral hemisphere to the first infarction in both cases. Both reported persisting altered perception of sound, including voices and music. That is, both had distinguishable disorders of auditory processing, not solely a disorder of linguistic processing. Both were able to interact with others by means of nonverbal methods of communication.

Communication Versus Cognition

Cognitive impairment is reported to be more disabling than motor impairment in published analyses of the structure of the leading disability measures.[12] Communication is often grouped with cognition in such analyses.[12,13] It is our clinical experience that communication disorders can be subtly devastating to the person afflicted and to the person's significant others. The patients presented here, in whom both cognition and communication were affected, reported their situations to be frustrating and isolating. It seems plausible to hypothesize that a "focal" deficit in one or another aspect of communication tends to make it more challenging for the patient to come to terms psychologically with his or her loss. It could also present more of a challenge for the rehabilitation team to arrive at a valid diagnostic formulation, and the communication disorder may not be detected when a primary or secondary cognitive disorder is also present.

Clinical Screening for Communication Disorder

In both cases presented here, diagnosis and management of the communication disorder were problematic even for some of the specialist physicians who were involved. How should the physiatrist screen clinically for communication disorder? Popular clinical handbooks recommend that the clinician assess functional communication in six areas: auditory comprehension and verbal expression, written expression and reading comprehension, naming, and repetition.[14] Consensus guidelines for clinical practice recommend that complete assessment of speech and language functions include instruments that are well constructed, well normed, and in wide use. These specialized tests include the Boston Diagnostic Aphasia Examination, the Porch Index of Communicative Ability, and the Western Aphasia Battery,[15] and are usually performed by a certified speech-language pathologist.

Bedside cognitive testing should also be done when possible. The most familiar test to clinicians is the Folstein Mini Mental Status Examination, although this simplified test in itself may

be insufficient to determine full extent of cognitive difficulties, particularly in regards to insight, judgement, and problem-solving ability. The patient's facility with gestural communication is also a useful test of cognition.[16] The guidelines of the Agency for Health Care Policy and Research[15] caution that the effect of any language disorder on overall function and daily living must be assessed separately before the rehabilitation intervention is formulated.

Rehabilitation Management

Rehabilitation management can conveniently be remembered in the traditional biological, psychological, and social domains. In the biological domain, it is important to delineate any impairments in hearing and vision as well as any problems with vocalization or movement. "Biomedical workup" usually involves performance of brain stem auditory evoked potentials, which are widely available. In the psychological domain, in addition to assessing cognition and mood it is important to give the patient and significant others an understanding of the nature of the deficit and the expected course of recovery. Ideally, the correct diagnosis should be made early in the acute period and appropriate education of the patient begun then, rather than being deferred to the postacute period, so distress and secondary problems are reduced. In the social domain, it is important to teach the patient the skills he or she will inevitably need for dealing with people who do not understand the impairment and to aim for a living situation that does not overstimulate, yet is not isolating for the patient.

Pragmatically, we try to emphasize effective communication during the early poststroke period, when the patient is trying to make sense of the impairments and when caregivers are most inclined to accept the change in the patient's interpersonal presentation. We introduce nonverbal interpersonal skills. For example, one should provide practical methods for enhancing face-to-face communication (dry-erase board, magic slate, Bliss-type board, electronic augmentative communication device—the choice taking into account the resources available to the particular patient and any impairments of hearing, vision, motor, cognition, and mood). One should arrange the environment (home or care-facility) to be conducive to the patient's particular needs for communication (such as low background noise). For quality-of-life issues, it is important to assess and address the patient's perceived needs. For example, our second patient loved music. Concentrating on rhythms rather than tones may bring some semblance of music back into her life. She also uses the closed-captioning on television.

Compensatory strategies could be provided, especially when environmental features cannot be changed (our first patient found earplugs to be helpful in coping with unwanted noise in the hospital). Also, the first patient found lip-reading to be helpful. In trying to avoid social isolation, it is important to provide means of distance communication (type telephone for the deaf, electronic mail through the internet) for those who are able to use them. Similarly, ongoing social contact can be facilitated in the form of stroke clubs, trained volunteers, or therapists. It is prudent for patients to keep information about their condition on their person (aphasia wallet card, medical alert bracelet or necklace) in case of emergencies to prevent misinterpretation of clinical signs by caregivers. The particulars are necessarily unique to each patient, and interventions need to be individualized.

Our society is so strongly oriented to language that it is ill prepared to recognize that independence of function is feasible when language is impaired, much less to adapt interpersonal behavior and societal structures to reduce the disability and handicap of the person with language impairment. However, as the oft-quoted judgement of American lexicographer Daniel Webster points out: "If all my possessions were taken from me with one exception, I would choose to keep the power of communication, for by it I would soon regain all the rest."[17] That is, communication is the important thing for successful rehabilitation, not specific language per se.

CONCLUSION

It is important that any acquired communication disorder be detected as soon as possible after its onset so that therapy for it can be incorporated into rehabilitation planning; effective engagement of the patient in the interdisciplinary team effort is essential for successful rehabilitation. As for other clinical areas, accurate diagnosis in communication is essential to determine what specific interventions would be the most useful for individual patients. The key to detection of communication disorders during the initial examination is to remember to use the methods that traditionally are taught to clinicians. Though such disorders may appear to be "pure," the impairment almost invariably is mixed, and the less prominent features can add substantially to the distress felt by the patient.

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