Clinical Forum

Cortical Deafness: A Longitudinal Study

Linda J. Hood*  
Charles I. Berlin†  
Prudence Allen‡

Abstract

We have studied a patient with MRI-confirmed bilateral absence of considerable portions of her temporal lobes resulting in cortical deafness. Although physiologic measures demonstrate normal peripheral hearing sensitivity, this patient’s speech has the inflection and prosodic characteristics associated with profound peripheral hearing loss, and she is unable to understand spoken communication. Behaviorally obtained pure-tone thresholds taken over nearly 20 years range from normal to moderate hearing loss with normal middle ear muscle reflexes and normal ABRs; however, we consistently found abnormal middle latency and cortical evoked potentials. Because of her total inability to communicate auditorily, this patient was ultimately taught American Sign Language and educated at the Louisiana School for the Deaf. This rare case highlights the importance of using multiple audiologic measures sensitive to abnormalities at various levels of the auditory system.

Key Words: Auditory evoked potentials, cortical deafness, bilateral temporal lobe dysfunction, otoacoustic emissions

True cortical deafness is a clinical rarity that presents unique evaluation and management challenges. Classifying cortical deafness is often difficult since patients generally exhibit inconsistent responses to sound and inordinately poor understanding and production of speech, even though objective measures of peripheral auditory function such as acoustic reflexes and auditory brainstem responses are normal. The literature contains reports of a number of cases of cortical deafness in children and adults, some congenital and others occurring as a result of disease or cerebral infarcts (e.g., Landau et al., 1957; Jerger et al., 1969). Over the years, the limits of each of these studies have been set by contemporary audiologic and radiologic technology.

In this report, we present a patient who lost much of her temporal lobes bilaterally following an extended high fever at 1 year of age. She was left with an inability to utilize auditory information that was variously diagnosed over the first 11 years of her life as (in alphabetical order) aphasia, brain damage, deafness, emotional disturbance, mental retardation, severe central auditory processing disorder, and severe language disorder. Diagnosis of cortical deafness was not possible without objective measures of auditory function at various “levels” of the auditory system coupled with accurate radiologic testing. These measures provided information regarding areas of the auditory system that were compromised and allowed a diagnosis based upon the site of the lesion, rather than the simple behavioral manifestations of deafness.

With the perspective afforded by study over nearly 20 years, we adapted and applied a battery of auditory tests designed to delineate her cortical hearing loss. We trace longitudinally the evaluation and diagnosis of this case, emphasize the need for objective measures of auditory function at various levels of the auditory system, discuss the relationships among...
audiologic and other measures, and address educational and management strategies.

PATIENT HISTORY

The patient, who for purposes of this paper will be referred to as "WL" (not her real initials), was born at full term in rural, northern Louisiana on June 10, 1970. Pregnancy, labor, and delivery were normal, birth weight was 5 lbs., 3 oz., and the neonatal period was normal. She is one of three female siblings. Between 6 and 7 weeks of age, WL was readmitted to the hospital for a viral illness and diagnosed with congenital nonspherocytic anemia, caused by a deficiency of the enzyme glucose 6 phosphate dehydrogenase (G6PD), apparent in the family pedigree. She also had significant jaundice at that time.

WL's development followed normal milestones for sitting, walking, early social skills, and the onset of single words up until 1 year of age. At 1 year of age, she developed a fever of 106 degrees that reportedly persisted, along with vomiting, for 5 to 7 days. This illness was originally diagnosed as a "viral infection without meningitis," although some question has remained regarding its meningitic nature. Following this fever, her mother reported that WL "stopped walking and talking." Her mother suspected that she might not hear, because she appeared to stop listening and seemed not to understand verbal instructions.

Otolologic evaluations over the years have indicated an absence of middle ear problems, although several bouts of otitis media were reported between 1 and 3 years of age.

PATIENT BEHAVIORAL PRESENTATION

WL is right-handed, rather quiet, and carefully attends to the many social cues provided by family members who accompany her. She exhibits some weakness of the lower extremities particularly on the left side, her feet toe inward, and her heels do not touch the floor. WL acts as if she has a profound peripheral hearing loss, and her speech production, similar to that of individuals with profound hearing loss, is characterized by lack of intonation, poor intensity control, poor articulation of speech sounds, and little or no ability to monitor her own speech. She is reluctant to try to lipread and depends on gestured or signed input in an effort to avoid failure. WL strives to perform well on tasks and will sometimes avoid a task rather than risk failure. She has, according to reports from her mother and others, shown inconsistent responses to auditory stimuli over the years. For example, in the history taken in 1988, her mother indicated that WL sometimes responded to familiar auditory stimuli at home such as the baby crying, the family dog barking, and the buzzer on the clothes dryer sounding. These responses reportedly are not always consistent, and it is difficult to determine how they might be situationally versus acoustically cued.

METHODS AND RESULTS

Audiologic Evaluations

Auditory evaluations, repeated on several occasions, included pure-tone audiometry, tympanometry and acoustic reflexes, auditory evoked potentials including electrocochleography (ECoG), auditory brainstem response (ABR), middle latency response (MLR), and cortical potentials (N1-P2). Various speech detection and recognition measures were also attempted, as were the most basic subtests of the Minimal Auditory Capabilities (MAC) Test Battery (Owens et al, 1981) and studies of gross auditory localization. Results of objective and behavioral tests completed over the years are summarized in Table 1. The details of specific procedures that vary from standard clinical protocols are indicated below.

WL was first seen at Kresge Hearing Research Laboratory of the South in New Orleans in August, 1974, when she was 4 years of age. At that time, she was unable to follow any verbal instruction, demonstrated speech quality characteristic of profound hearing loss, was essentially unresponsive to auditory stimuli in her environment, and depended upon visual cues. Behavioral audiology was attempted but resulted in grossly inconsistent responses, leading the examiners to use objective measures such as EcochG to obtain objective quantification of hearing. Results of these tests led us to conclude that "the peripheral system was functioning well enough to allow development of speech and language," and she was referred for additional speech and language, neurologic, evoked potential, and psychological evaluations (see discussion of results below). The possibility of a severe central auditory processing disorder was suggested, and recommendation for intensive language therapy was made.
### Table 1 Summary of Audiologic Test Results from 1974 through 1993

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Year and Age at Test</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pure-tone Thresholds</td>
<td>25 dB HL</td>
<td>30-40 dB HL</td>
<td>35-60 dB HL</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Speech Thresholds</td>
<td>25 dB HL</td>
<td>20 dB HL</td>
<td>10-35 dB HL</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tympanograms</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Acoustic Reflexes</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>EOAEs</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>EOAE Suppression</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>DPEs</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>ECochG</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>ABR Latencies</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>ABR Thresholds</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>ABR Rate</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>MLR</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
</tr>
<tr>
<td>Late (N, -P)</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
</tr>
<tr>
<td>Localization</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
</tr>
</tbody>
</table>

Not all tests were completed on all occasions, due to nonavailability of the procedure or weighting of test priorities against patient abilities and fatigue. Blank cells represent visits when particular tests were not completed.

*Amplitude decreases in the mid frequencies.

In subsequent years, WL was seen for repeat evaluations at Kresge Laboratory. Her increasing age and cooperation enabled us to complete more thorough evaluations. For purposes of presentation, results are presented according to test type, and findings for each particular measure are tracked over the years of evaluation.

**Pure-Tone Thresholds**

Reports of behavioral pure-tone sensitivity have varied. Pure-tone hearing reports obtained by others from the age of 2 to 4 years ranged all the way from a very mild to a severe-to-profound hearing loss. Responses were reported at screening levels of 20 dB HL (age 6) and 25 dB HL (age 10), suggesting normal peripheral hearing sensitivity. At age 18, because of improved attention, we were able to obtain threshold responses for noise and tonal stimuli between 500 and 4000 Hz at 35 to 60 dB HL (Fig. 1). These behavioral pure-tone thresholds were variable, poorer than previous tests, and most likely above her levels of best detection. Whether the apparent shifts in pure-tone thresholds over time represent any true change in sensitivity or simply shifts in reliability and attention is difficult to determine since the stimuli appeared meaningless to the patient.

**Middle Ear Measurements**

The first objective screening test was obtained at 2 years of age using acoustic reflexes. Contralateral acoustic reflexes were normal for each ear, which, when coupled with the lack of any startle reflex, led to the conclusion that the patient could not have a profound hearing loss but could have a moderate hearing loss. Tympanometry at age 4 indicated normal middle ear function (type A tympanograms bilaterally), and contralateral acoustic reflexes were present at normal levels for 500-4000 Hz for each ear. Subsequent tympanograms and ipsilateral and contralateral acoustic reflexes have been normal on all test dates.

![Figure 1 Audiometric testing completed at age 18 showed pure-tone thresholds in the mild to moderate hearing loss range. Pure-tone responses did not agree with thresholds for vowel and consonant sounds (Ling "Five-Sound Test"). Tympanograms and ipsilateral and contralateral acoustic reflexes were normal.](image-url)
Electrocochleography and Auditory Brainstem Response Testing

Electrocochleography (ECochG) was first completed at age 4 and provided the first objective quantification of cochlear function. Results suggested normal cochlear function with responses obtained at 10 to 25 dB above normal thresholds for clicks (Fig. 2). Subsequent ABRs at ages 10, 12, and 18 indicated replicable responses with normal absolute and interpeak latencies at higher intensities, as well as normal latency-intensity functions with responses to click stimuli present at levels of 25 dB HLN (hearing level for a group of normal hearing subjects) in each ear. Normal responses have also been obtained using low-frequency (500 Hz) tone bursts. ABRs have been normal on tests at all ages with no changes in absolute or interpeak latencies. Auditory evoked potentials, including ABR, MLR, and late (N1-P2) responses obtained at age 18 are shown in Figure 3.

Middle latency responses obtained at age 10 using clicks at a rate of 9.7/sec and a filter of 30-250 Hz showed little synchrony; the responses were low in amplitude with very delayed latency (65 msec), particularly for the left ear. The synchrony at about 65 msec may reflect Pb activity rather than Pa, which would normally occur at about 30 msec. At age 12, middle latency and late responses showed no replicable responses upon stimulation over either hemisphere using traditional electrode montages (Cz-Al, Cz-A2) and 100 µsec clicks presented at 70 dB HLN at 1/second using a 30-300 Hz bandpass and 100 msec post-stimulus recording time. The MLR recorded at age 18, obtained from Cz-A1 and Cz-A2 at 3.3/sec with 5-3000 Hz filters, showed some replicable activity (Fig. 3), perhaps related to better attentiveness. This activity, however, was minimal and delayed when compared to a normal control (Fig. 4). At age 23 years, minimal MLRs were again recorded centrally (Cz-A1, Cz-A2), and no responses were obtained from lateral (C3 and C4) electrode sites.

Late Cortical Responses

Long latency potentials (N1-P2) obtained at age 4 were asymmetric, with the right response approximately 20 msec slower than the left. At age 12, no N1-P2 complex was observed for stimuli presented at 70 dB HLN at 1 to 4 stimuli/second using a 1 to 300-Hz bandpass and a 1-second post-stimulus recording time. At age 18, long latency potential activity was obtained, again possibly consistent with greater attentiveness (Fig. 3). Compared to a normal control (Fig. 4), the response from the right ear showed reduced amplitude and asymmetry between the two ears, which is interpreted as abnormal. This set of observations, showing greater auditory brainstem and late cortical activity than middle latency responses, highlights the independent and parallel nature of evoked responses from the central auditory pathways.

Speech Detection and Identification

Speech detection thresholds have generally been better than pure-tone thresholds would predict, as exemplified by the detection thresholds for the "Ling sounds" (Ling, 1978) ranging from 15 to 35 dB HL (Fig. 1). These, coupled with the more reliable results of objective tests such as acoustic reflexes, ECochG, and ABR intensity functions for both clicks and tone bursts, support the conclusion of normal peripheral function.

Several attempts have been made to assess WL's ability to utilize auditory stimuli in a meaningful manner. At age 12, she showed some ability to identify and differentiate various noisemakers (rattles, squeeze toys), but speech discrimination was not possible without visual cues. At age 18, performance on the Word Intelligibility by Picture Identification (WIPI) Test (Ross and Lerman, 1971) indicated scores below chance in the auditory only condition (right ear: 16%, left ear: 8%) and a combined auditory-visual score of 60 percent. Recognition
of environmental sounds, discrimination of noises versus voices, and recognition of familiar voices (her mother and father) were all below chance. She was able to distinguish among three sounds ("ah," "oo," "ee") for a few minutes following training; however, this ability was short-lived and required retraining when another task intervened.

**Localization and Other Measures**

A gross localization task was completed at age 12, where stimuli were presented in a sound field and head movement was not restricted. Results indicated correct localization of approximately 75 percent of the stimuli (tones and noise bands). Localization was evaluated again at age 18 using narrow bands of noise presented left-right (80% correct), front-right (80% correct), front-left (68% correct), left front-left rear (64% correct), right front-right ear (60% correct), and front-rear (40% correct). Accuracy of localization on this task in normal individuals is usually 100 percent for all conditions.

Additional procedures have included a key tap test with delayed auditory feedback (DAF) and auditory-visual temporal order judgments (TOJ). Performance on both tests was below chance and was consistent with WL's inability to monitor the auditory components of her own motor activity (DAF test) and a cortical site of lesion (TOJ).

**Otoacoustic Emissions**

The recent availability of otoacoustic emissions allowed us the opportunity to further document WL's normal cochlear function as well as to evaluate the effects of contralaterally presented stimuli on cochlear emissions (Collet et al, 1990; Berlin et al, 1993a, b). In 1991, WL demonstrated normal evoked (TEOAE) and distortion-product (DPE) emissions for the right and left ears, although amplitude of the evoked emissions for an 80-dB peak sound pressure nonlinear click was 5-6 dB SPL.

Contralateral suppression of TEOAEs was measured using 80-dB peak sound pressure nonlinear clicks and contralateral narrow bands of noise centered at 250, 500, 1000, 2000, and 4000 Hz. The effect of the contralateral noise at the five center frequencies showed similar configurations, and data were thus collapsed across frequency (Berlin et al, 1993b). Data obtained from WL ("patient") are compared to a group of normal subjects in Figure 5. The upper portion of the figure shows overall echo amplitude without contralateral stimulation (the "zero" condition) and for contralateral narrow bands of noise at 20, 40, 60, and 80 dB HL. While WL's overall emission amplitude was below the normal range, the configuration of suppression (decrease in emission amplitude with increasing contralateral noise level) remained similar. This is further demonstrated in the lower por-
tion of Figure 5, where the actual emission amplitude is ignored and only the amplitude shift, or the amount of change in overall amplitude of the emission with and without contralateral stimulation, is plotted. When emission amplitude is ignored, WL's results are identical to the average of the normal group.

In 1993, TEOAEs were obtained using 70-dB linear clicks, and results were consistent with previous data. Suppression of TEOAEs by introduction of contralateral white noise at 60 and 70 dB HL was present, again consistent with previous observations. Distortion-product emissions were also present for each ear, although minimal emission amplitudes were noted in the mid-frequency range for each ear.

It is important to recognize that the normal contralateral suppression shown here was accomplished without conscious awareness of sounds in either ear. The reason for the reduced emissions amplitude is unknown, although possibilities may include WL's history of otitis media or her use of a hearing aid prior to correct diagnosis of her auditory disorder.

**Summary of Audiologic Tests**

Results of objective and behavioral auditory tests show that auditory function at the level of the cochlea and brain stem was essentially normal, while measures above the brainstem level were abnormal (refer to Table 1). Behavioral thresholds were better for speech than for nonspeech stimuli (Fig. 1). These behavioral results were supported by click and 500-Hz tone-burst ECochG and ABR latency-intensity functions that showed responses to stimuli presented at intensities near normal threshold levels for each ear (Fig. 2). ABR absolute and interwave latencies were also within the normal range bilaterally (Fig. 3, top). Results of these measures were essentially unchanged from early tests to 1993. Middle latency responses were grossly abnormal across multiple electrode recording sites (Fig. 3, middle) while late potentials were present, although asymmetric (Fig. 3, bottom). Some changes in the late potentials from the earlier tests may be related to maturation and improved attentive-ness.

Audiologic test results suggest a normal peripheral auditory system with severe degen-
eration of central function essential to the processing of auditory information. WL evidences very limited conscious awareness of sound with essentially no usable auditory capabilities for the perception of speech. However, how much she is aware of and to what extent she can use that minimal awareness to monitor and learn from her surroundings is unclear. Responses to pure-tone audiometry show that she can respond to tonal stimuli, although perhaps not near her cochlear threshold levels. She has been reported to make distinctions among tones and noises in the past, although she did not do well on this task on subsequent testing. WL is unable to localize the sources of sounds normally, which is consistent with the cortical site of the lesions, and she exhibits neurologic abnormalities that are thought to stem from the same etiology as her deafness. Auditory tests at 18 and 21 years of age were essentially the same as earlier tests and not suggestive of any retrograde degeneration at or below the brainstem level.

Radiologic Evaluations

The first radiologic evaluation was obtained in 1982 at age 12. Computed tomography (CT) scans, obtained without contrast, indicated a "bilateral prominence of the Sylvian fissures with the left more prominent than the right." No mass lesions were observed, and the significance of the prominence was questioned in the absence of associated abnormal findings. Thus, the CT scan results were initially reported as within normal limits. Reinterpretation of these CT scans indicated severe temporal lobe damage extending to the left frontal cortex near the precentral nucleus and the frontal-parietal operculum. No mass effect, shift in midline structures, or mass in the posterior fossa or CPA regions was reported.

In 1988, MRI studies of the coronal, axial, and sagittal planes confirmed the absence of considerable portions of the temporal lobes bilaterally. Decreased signal in the axial sections (Fig. 6A) and increased signal in the T2-weighted coronal sections (Fig. 6B) were seen along the Sylvian fissures bilaterally. The anterior tip of the right temporal lobe was also rather atrophic. Elsewhere, no abnormal area of increased or decreased signal was observed to suggest the presence of other lesions of brain parenchyma or shift of midline structures or other mass effect. Findings suggested atrophy or degeneration along the superior surfaces of the temporal lobes, including the auditory cortex on each side, with this process extending further anteriorly along the lateral aspect of the right temporal lobe to the apex.

Neurologic Evaluations

Electroencephalograms obtained between 1 and 4 years of age showed normal waking and sleep records. WL began to show signs of grand mal seizures between 1 and 3 years of age and was placed on phenobarbital to control the seizure activity. This was discontinued later due to adverse reactions. She did not experience seizures again until age 20, when she was again placed on medication (Tegretol). Follow-up at age 21 indicated that she was doing well with no further seizures, no ataxia, and intact ocular and facial movements. She continues to take Depakote to control seizure activity.

Neurologic evaluation at 4 years revealed a mild left hemiparesis with abnormal motor reflexes and clumsiness apparently with CNS damage precipitated by the same event that produced the high fever at 1 year of age. Additional neurologic testing at 12 and 18 years of age indicated widespread motor problems with coordination difficulties in upper and lower extremities, more accentuated on the left. WL is right handed and has mildly dysmorphic facies characterized by slight antimongoloid slants to the eyes and a slightly open-mouthed facial expression. Visual acuity was normal up until age 18, at which time she began wearing glasses.

In summary, WL evidences a static encephalopathy manifested by bilateral motor coordination difficulties with evidence of moderate athetosis, hyperreflexia (left greater than right), impaired proximal strength, seizures beginning early in life, and a cortical sensory deficit for graphesthesia and for auditory signals.

Neuropsychological Evaluations

Psychological testing has generally been consistent with function in the average range of intelligence when auditory-verbal limitations are taken into account. Delays in diagnosis of the nature of the auditory problem resulted in performance well below age level. For example, neuropsychological testing at age 12 years indicated a WISC-R Performance Scale IQ within the normal range with overall achievement at the first grade level and reading comprehension when signing at the mid-second grade level.
Figure 6  Magnetic resonance images obtained in the axial (A) and coronal (B) planes showed absence of considerable portions of the temporal lobes bilaterally. The axial view is T1-weighted with abnormal areas darkened (abnormalities marked by arrows; anterior upward), and the coronal view is T2-weighted with spaces bright (see arrows indicating abnormalities).

LANGUAGE DEVELOPMENT AND EDUCATIONAL MANAGEMENT

WL had no school experiences up to the age of 4. According to her mother, at age 4 she had no behavioral problems and was easily managed, although she exhibited fear of loud sounds. After 1 year of age, she had no meaningful speech other than reports by the mother of verbalizations resembling "mama" and "no" and uncontrolled pitch of vocalizations. Linear hearing aids were tried for about 1 month and rejected. Tactile stimulation was not tried.

At 4 years of age, WL was managed as a child with a severe central auditory processing disorder with normal hearing, was placed in a highly structured classroom environment for severely language-impaired children, and continued in this environment from 1974 to 1980. At age 6, she continued to be diagnosed as "aphasic" and continued in intensive speech and language therapy. She tested at a language level of 4 years, and her spontaneous verbalizations reported at home consisted of four words ("mama," "no," "coke," "water"). She was enrolled in a speech and language program where she showed little progress. Emphasis was placed on speech therapy and auditory training. Reports indicated that she could speechread limited sets of stimuli but had inconsistent responses to auditory stimuli.

Spoken language development was negligible. At ages 6 and 7, WL went to an oral school for the deaf in a neighboring state where she reportedly showed improvement in speech, language, motor development, and behavior. The approach at that school was primarily verbal language and, while attending this oral school up until about the age of 8, she achieved a speaking vocabulary of about 70 words. At the time of discharge, auditory responses were inconsistent, intelligible spontaneous speech was limited, spontaneous utterances were distorted by articulation errors, voice control was poor, and she seemed to depend on visual cues. WL responded inconsistently to her mother's voice but not at all to others. A classroom for severely language-impaired children was recommended with a primary emphasis on speech therapy and attention to sound production. A class for
hearing-impaired children was not considered appropriate at that time since she had normal peripheral hearing.

From 8 to 9 years of age, reading and writing skills progressed, spontaneous speech was very limited, and she demonstrated dependence upon visual cues and information. Lipreading skills were poor, with greater reliance on written cues. Based upon her lack of progress in classes for language-impaired children, WL was placed in a class for hearing-impaired children in 1978 and began to learn some sign language. Reports have indicated that she "seemed to hear" on some days, made little progress in speech therapy, and responded to visual but not auditory cues. Following additional auditory testing and confirmation of the disorder with CT scans in 1982, auditory input was completely abandoned, manual communication was emphasized as her primary mode of communication, and full attention was directed to signing both at home and at school. Thus, once the nature of the problem was recognized, emphasis was placed on language development as the most important issue to address, best accomplished through manual communication and visual channels.

From 1982 to 1989, WL attended the Louisiana State School for the Deaf, using American Sign Language (Ameslan) as her primary method of communication. She was enrolled in a vocational/technical program and showed particular interest in computers, drawing, and sewing. Following completion of the program at the State School for the Deaf, she attended a residential program in a neighboring state that emphasized independent living skills and vocational development. She then returned to her home town.

A language evaluation was completed in 1993 to gain some insight into WL's current level of language function. Results indicated extremely limited reading comprehension, although she can read and use single words. She appears to have great difficulty integrating single words into slightly more complex linguistic units and relies heavily on others to demonstrate concepts to her. WL demonstrates naming failures both in Ameslan and in written English that are interpreted as a consequence of vocabulary deficits. The examiner concluded that the patient "... exhibited very severe deficits in linguistic functioning [and] that she suffers not only the effects of being introduced to a language system late in her childhood but also a rather significant degree of aphasia."

It was further observed that WL's signing is distorted by apparent fine motor dysfunction and that she generally uses broad movements of the arms when signing and fingerspelling rather than just the fingers and wrists.

**DISCUSSION**

The subject of this report is a patient who displays evidence of acquired speech and language disorders associated with the onset of seizures following a relatively normal period of growth and development. Cases of cortical deafness derive from a number of etiologies, including congenital causes (Landau et al, 1957), meningitis (Lechevalier et al, 1984), and cerebral infarcts (e.g., Jerger et al, 1969).

Landau et al (1957) reported a case of a child diagnosed in the absence of CT scans or MRI as having congenital aphasia. This child had a normal birth history complicated by cyanosis at 10 days, a heart murmur, and pneumonia several times from 3 months to 3 years. Development was delayed, with walking commencing at 5 years, no speech production or comprehension of spoken language at age 6 years, and communication via gestures and facial expression. This child could imitate short words but did not know their meaning and at 8 years was reported to have an IQ score of 97 and functional vocabulary (reading, writing, speaking) of about 175 words. Pure-tone audiometry resulted in inconsistent responses. Shortly after this, the child suddenly died at age 10 of cardiac complications. Examination of the brain showed the gyri of posterior portions of the parietal, temporal, and occipital lobes reduced in size, and histologic analysis showed that the normally distinct medial geniculate structures were difficult to identify and appeared severely degenerated. The severe damage to the primary auditory projection pathway bilaterally and inability to utilize auditory stimuli are consistent with the present case study and an example of true thalamo-cortical deafness.

Jerger et al (1969) provided documentation of deafness due to clearly demonstrated cortical lesions without evidence of a peripheral lesion in an extensive report of an adult who sustained two sequential cerebral hemisphere infarctions with maximal damage in the temporal lobes. While the first episode produced no salient auditory deficit, the second resulted initially in a severe bilateral hearing loss that gradually recovered in the speech frequencies, leaving a low- and high-frequency hearing loss. This is con-
sistent with the observations of hearing ability of experimental animals with bilateral cortical lesions (Heffner and Heffner, 1986). Although speech detection was good, speech recognition was poor in one ear and nonexistent in the other ear. Localization ability was impaired. An objective measure used to study this patient was the late (N1-P2) cortical potential. No clearly defined responses were obtained to any auditory stimuli while visual potentials were normal. Middle latency responses were not studied, and the ABR was not yet in clinical use. This case differs from the current case in that a central lesion appeared to affect behaviorally tested pure-tone thresholds. Indeed, over the years, our patient has shown variability in pure-tone thresholds that are not substantiated by EOChG or ABR. She could not be tested at the time of the insult, so any early shift in thresholds cannot be documented.

In addition, cortical potentials, while abnormal, did show some synchrony in the 100–300 msec time period. However, functionally, neither patient could communicate auditorily and depended upon visual input for communication.

Other case studies of cortical deafness have been reported where the peripheral and brainstem pathways have remained intact and function has been assessed both behaviorally and electrophysiologically. For example, Graham et al (1980) reported a patient who suffered embolic lesions of cardiac origin affecting both temporal lobes. This patient, despite some apparent peripheral age-related loss, showed no response to pure tones behaviorally, but had stapedius reflexes at elevated levels, normal ABR waveforms at higher intensities, but no middle latency or cortical responses. This is similar to a patient reported by Earnest et al (1977) with bitemporal lesions documented by CT scan who, after 3 years, showed variable pure-tone thresholds but normal acoustic reflexes, and a persistent inability to consistently identify the presence or nature of sounds. In this patient, cortical potentials were recorded, although only at elevated levels. Another case reported by Adams et al (1977) showed discrepancies between pure-tone, speech, and electrophysiologic measures. While pure tones and cortical potentials were consistent with a severe hearing loss and speech understanding was nonexistent, acoustic reflexes and ABR thresholds were normal.

These and other cases (e.g., Özdamar et al, 1982; Lechevalier et al, 1984; Woods et al, 1984; Nakayama et al, 1986; Ho et al, 1987; Bahls et al, 1988; Hasegawa et al, 1989; Tramo et al, 1990) document several common characteristics of bilateral cortical lesions. Pure-tone thresholds may show decrements or variability and generally do not support the total inability to understand speech. Further, objective measures of peripheral and brainstem function are normal, while evoked potentials that putatively originate in the cortex or auditory radiations to the cortex show abnormalities along with localization and abilities of cortical origin.

**Middle Latency Responses and Temporal Lobe Lesions**

While the focus of evoked potential abnormalities in WL were in the middle latency responses, reports in the literature concerning MLR results in temporal lobe lesions have been conflicting. Parving et al (1980) reported normal MLRs (wave Pa) in a patient with auditory agnosia and documented bilateral temporal lobe lesions. In contrast, Özdamar et al (1982) presented a patient with bilateral temporal lobe lesions with MLR wave Pa missing bilaterally. Kileny and Berry (1983) also reported two cases of confirmed bilateral temporal lobe lesions with one case showing abnormalities that could have been confounded by neuromaturation and the other case showing normal MLRs. Kraus et al (1982) further studied the effects of temporal lobe lesions in a series of 24 patients and found diminished Pa amplitude over the hemisphere with the lesion. Further case reports confirming this finding have also been reported (Kileny, 1985).

Thus, latency and amplitude abnormalities of the middle latency response are observed in bilateral temporal lobe lesions (Graham et al, 1980; Kraus et al, 1982; Rosati et al, 1982; Ho et al, 1987; Kileny et al, 1987; Tramo et al, 1990), although intact responses have also been reported (Parving et al, 1980; Woods et al, 1987). Because of bilateral representation of each cochlea, unilateral lesions are less readily apparent. More subtle changes, specifically reduction in amplitude of responses recorded from the hemisphere ipsilateral to the lesion, occur in patients with unilateral temporal lobe lesions (Kraus et al, 1982; Kileny et al, 1987).

Woods et al (1987) suggest that abnormal middle latency and late responses do not simply reflect primary auditory cortex damage but are also dependent upon the degree of damage to adjacent areas, particularly thalamic projections. They suggest that only patients with
subcortical as well as cortical damage will show no MLR.

**MLR and Late Generators**

Generators of the middle latency response have not been fully defined, although the most likely sources include the posterior portions of the transverse temporal gyri, thalamo-cortical projections, and reticular activating system (see review in Kraus and McGee, 1992). The involvement of cortical as well as subcortical projections in the production of the MLR has been suggested. McGee et al (1992) recently suggested that the system generating the MLR includes the auditory pathway from the midbrain to the cortex and involves such regions as the reticular formation and nonprimary divisions of the auditory thalamo-cortical pathways, which process multimodal stimuli. Temporal and midline components of the MLR in guinea pigs have been described by Kraus et al (1988), with the temporal response representing the primary auditory pathway. McGee et al (1992) further evaluated these components and suggested that subdivisions of the medial geniculate differentially contribute to midline and temporal components. Kraus and McGee (1993) suggested that this may be related to the existence of both primary and secondary pathways. The primary or lemniscal auditory system, characterized by neurons that respond only to auditory stimuli, shows good frequency tuning, tonotopic arrangement, and a high degree of time locking to stimulus characteristics (see summary in Brugge, 1992). In contrast, the auditory cortex in the broader sense may also include cortical fields that are sensitive to stimuli outside of the main auditory lemniscal pathways. Unlike the lemniscal auditory system, these systems show little or no cochleotopic organization, exhibit relatively broad tuning and long discharge latency, and may be activated by other sense modalities (Brugge, 1992).

**Importance of Including Physiologic Measures**

Central auditory processing disorders can masquerade as peripheral hearing losses for several reasons. First, severe central auditory deficits, as in the case presented here, result in an inability to monitor one's own speech. Without this monitoring information, it is difficult to control loudness, pitch, and articulation, and speech production mimics that seen in patients with severe and profound cochlear hearing losses. Second, some behavioral auditory test results may show poor interest agreement, usually relied upon as an indicator of test reliability. Inconsistent responses to pure tones and lack of agreement with speech thresholds may reflect sharply sloping hearing losses, unusual hearing loss configurations such as ultra-audiometric hearing (Berlin et al, 1978), central hearing loss, or a functional hearing loss.

Objective measures of auditory function provide a way of overcoming the dilemma presented by inconsistent behavioral responses. Otoacoustic emissions reflect normal function of the cochlea, and acoustic reflexes and auditory evoked potentials can be used to assess integrity of the brainstem, thalamic, and cortical pathways. In addition, early evoked potentials (ECochG and ABR) can be obtained to stimuli at low intensity levels to draw conclusions regarding peripheral hearing sensitivity in patients with intact auditory nerve and/or lower brainstem function.

**How Otoacoustic Emissions Can Contribute to Analysis of Central Auditory Disorders**

Evoked otoacoustic emissions can be suppressed by contralateral stimuli, and such suppression requires intact connections between the two ears (e.g., Collet et al, 1990; Berlin et al, 1993a, b). In our experience, patients who display lower auditory brainstem dysfunction, demonstrated by abnormal contralateral acoustic reflexes and abnormal auditory brainstem responses, show normal otoacoustic emissions (in fact, often with higher than normal amplitude) but lack any contralateral suppression of evoked emissions measured by our computer analysis program (Berlin et al, 1993a; Wen et al, 1993). WL showed normal brainstem function on all of the above measures. Thus, the presence of cochlear emissions was consistent with normal peripheral hearing, and the normal contralateral suppression was consistent with normal acoustic reflexes and ABR. Comparison of these findings to those in other patients suggests that the presence of emissions and their contralateral suppression can be useful in eliminating deficiencies to the cochlea and caudal auditory systems. Further, the presence of contralateral suppression in WL indicates that an intact cortical system is not necessary for studies of caudal brainstem interconnections between the two ears (Berlin et al, 1993a).
What Sort of Language Can One Generate in the Absence of Both Temporal Lobes?

This case underscores an important theoretical question related to the ability to generate language in the absence of both temporal lobes; however, we cannot obtain an unequivocal answer from this case. WL showed distinct language deficits, but they are basically inseparable from her limited input in childhood, her acquisition of American Sign Language at such a late age, and her absence of auditory monitoring abilities. Thus, she was being asked to learn a "foreign" language through a defective system that offered her almost no stable input to establish a language base. There was no way that she could avoid a language disorder. Whether she is "aphasic" is, for her, a moot point, although our language consultant feels that she is.

What Can We Recommend for Children Like This?

WL is now married and is expecting a child. Her husband has a moderate hearing loss, derives effective benefit from hearing aids, was educated auditorily, and acts as her interpreter. WL seems to have made a satisfactory social adjustment as a result of committed family support.

It should be re-emphasized that hearing aids are an inappropriate answer for cases such as these. This patient's peripheral system is already delivering sound to a central system that cannot process it. To manage this patient with hearing aids is both inappropriate because of normal cochlear function and useless because any auditory signal, either at normal or high intensities, will serve no purpose in a central system that is unable to utilize it. WL's rejection of hearing aids was the first indication that amplification was not the answer; delineation of the cortical deafness and her success with only nonauditory input confirm the fact that hearing aids are not useful in managing cortical deafness.

Professionally, in retrospect, it seems clear that we should have recommended from the very beginning that WL be managed as a signing deaf child. However, knowing that she had "normal hearing" physiologically, we felt obliged to advise an auditory-oral education. Nowadays, with a better understanding of the effectiveness of objective physiologic methods in the evaluation of central auditory disorders and the MRI as a diagnostic tool, we can more readily clarify the nature of these deficits. Twenty years ago, we may have avoided a language deficit through early introduction of manual communication. However, we may not ever have been able to give WL a solid written English base if she truly was aphasic following her illness.

SUMMARY

This article reports audiologic and related test results obtained over nearly 20 years from a patient with MRI-confirmed bilateral absence of considerable portions of her temporal lobes resulting in cortical deafness. Although physiologic measures demonstrate normal peripheral hearing sensitivity, this patient's speech has the inflection and prosodic characteristics associated with profound peripheral hearing loss, and she is unable to understand spoken communication. Behaviorally obtained pure-tone thresholds were variable, ranging from normal to moderate hearing loss with normal middle ear muscle reflexes and normal ABRs to high- and low-intensity stimuli. Auditory middle latency and cortical evoked potentials were grossly abnormal, consistent with the central nature of this "deafness." Because of her complete inability to communicate auditorily, this patient was ultimately taught American Sign Language and educated at the Louisiana School for the Deaf. A combination of audiologic measures such as acoustic reflexes and auditory evoked potentials that are objective as well as sensitive to abnormalities at various levels of the auditory system are very useful in diagnosing central auditory disorders. Contralateral suppression of otoacoustic emissions was normal in this patient, which further supports a caudal brainstem locus for the emission suppression effect.

Acknowledgment. This work was supported by NIH-NIDCD P01-DC00379, Kam's Fund for Hearing Research, and the Louisiana Lions Eye Foundation.

The authors appreciate the contributions of John K. Cullen, Jr., Ph.D. and Patricia Shearer, M.D. to the early testing of this patient; Daniel Johnson, M.D. for the MRI evaluation; John Willis, M.D. for neurologic evaluations; Elizabeth Gochnour, M.A. for recent language evaluations; and Kurt Hecox, M.D., Ph.D. for consultations.

We also wish to thank the patient reported here and her family for their continuing interest in furthering our understanding of this auditory problem.

REFERENCES


