Progressive Neural Hearing Impairment: Case Report

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Abstract

This report describes a patient whose ability to understand speech had so deteriorated over 20 years time that she was being considered for a cochlear implant, despite the fact that pure-tone sensitivity loss had not deteriorated proportionately. An unusual combination of otoacoustic emissions (OAEs) and auditory evoked potential (AEP) results are described. Click-evoked and distortion product emissions were present and normal-appearing. Auditory brainstem and middle latency responses were totally absent but the N1–P2 complex of the long-latency response was present. The case illustrates the contribution of otoacoustic emissions to site of lesion testing. It also illustrates that the manifestations of pathology can appear in certain epochs of the family of AEPs without affecting the others.

Key Words: Auditory brainstem response (ABR), evoked otoacoustic emissions (EOAEs), long-latency evoked response (LLR), middle latency response (MLR), N1–P2 complex, progressive hearing loss

The case we report was interesting to us for several reasons: its initial referral to us (a cochlear implant candidate with considerable residual hearing), the unusual complement of results from otoacoustic emission (OAE) and auditory evoked potential (AEP) testing, and the documented progression of the loss over the past 13 years. The addition of emissions to the test battery gives an unusual insight to the possible site of lesion that has not been possible in the past.

CASE REPORT

R is a 35-year-old white female who was referred to the University of Kansas Medical Center specifically for otoacoustic emissions testing. R has been a patient of a local otologist since 1981. Her ability to understand speech has gradually deteriorated to the extent that she has abandoned the use of amplification altogether. She relies on lipreading for communication. From our perspective, she manages remarkably well. Her speech is very good, though it has a typical deaf quality. Her latest visit to the otologist was to discuss her candidacy for a cochlear implant. Otoacoustic emissions testing was requested to try to further delineate the site of lesion, specifically to separate sensory from neural components of the hearing loss.

Background

R reported that she first noticed difficulties with her hearing at 15 years of age. She assumes that she passed elementary school hearing screening tests as no one ever suggested that there might be a problem. No one else in her family has hearing impairment. The following historical information was provided by R or gleaned from chart notes. The audiometric results were obtained from different audiologists in different settings. As a result, each evaluation was not equally thorough and the details of test administration were not always noted.

At her 1981 otologic visit when R was 23 years of age, she reported that she had no understanding of speech in her left ear. She had always heard better from her right ear and had been unable to use her left ear for the telephone for several years. She reported occasional tinnitus
lasting for a minute or two, more frequent at the left than the right, and denied dizziness or headaches. Otoscopic examination was normal. Audiologic exam confirmed her report of poor speech understanding at the left ear.

The first set of audiograms dated February 25, 1981 and displayed in Figure 1 show pure-tone hearing thresholds in the mild loss range. For the right ear, a speech reception threshold (SRT) of 30 dB HL was in general agreement with pure-tone findings and the word recognition score at 40 dB re SRT was 88 percent. However, for the left ear, an SRT was not obtained until a level of 70 dB HL was reached and monosyllabic word recognition was not possible. A speech detection threshold (SDT) was obtained at 30 dB HL. Tympanometric findings were normal with contralateral acoustic reflex thresholds reported at 90 to 100 dB HL for each ear at 500 and 1000 Hz (apparently higher frequencies were not tested). Auditory brainstem responses (ABR) showed poor waveform morphology but were essentially symmetrical. When a slow click rate (9.3 per second) was used, the I–V interwave interval for each ear was calculated at 3.96 msec. CT scans revealed normal internal auditory canals. Chemistry profile was normal and FTA-Abs nonreactive.

Audiometric assessment on May 11, 1983 (see Fig. 1) showed little change in pure-tone sensitivity. Word recognition score for the right ear had gone from 88 percent to 76 percent (details of administration such as word lists, taped or live voice, etc. are not available). Acoustic reflex test results were not given.

At her August 17, 1987 visit to the otologist, R reported that her hearing aid, a BTE purchased 2 years earlier for the right ear, seemed to distort voices and that she relied heavily on lipreading. She was unsure if her hearing had worsened but had been encouraged by coworkers and family to return to the otologist to see if anything could be done. She asked about cochlear implants. The most notable changes are a marked reduction in word recognition ability for the right and a reduction in low-frequency pure-tone sensitivity for the left ear. Although pure-tone thresholds for the right ear remained essentially unchanged from her 1981 audiogram, acoustic reflex thresholds were elevated in comparison. The otologist and audiologist apparently discussed implants with her but recommended hearing aid evaluation and possible assistive devices for TV, stereo, etc.

The next visit took place on November 2, 1989 when the fourth in the series of audiograms...
Figure 2  Click-evoked otoacoustic emissions records for the right and left ears, respectively. The top panels for each ear display the ILO record for click stimulation at approximately 80 dB pSPL, the middle panel for 70 dB, and the lower panel for 60 dB. (On the records, the stimulus waveforms and spectra differ for the two ears simply because the reference test gains were set differently for each ear; stimulation for each ear was the same.)

was done (see Fig. 1). Word recognition ability could not be tested with monosyllabic stimuli for either ear, although an SRT could be obtained for the right ear.

The audiogram obtained on July 20, 1993 showed a further reduction in pure-tone sensitivity (see Fig. 1). R could not understand spondees presented at levels from 50 to 90 dB HL in
either ear. It was at this time that she was referred to the Midwest Ear Institute for comprehensive pre cochlear implant evaluation to determine candidacy.

The audiometric results from the subsequent evaluation are shown as the sixth and final of the series in Figure 1. (The discrepancy between SDT and pure-tone thresholds for the right ear was unexplained in the reports we received.) Immittance testing revealed normal Type A tympanograms with absent acoustic reflexes bilaterally (0.5, 1, 2, and 4 kHz and broadband noise, up to 110 dB HL, ipsilaterally and contralaterally).

Speech perception test results using the Cochlear Corporation/University of Iowa Revised Cochlear Implant Test Battery are shown in Table 1. All testing was performed in sound field at 70 dB SPL under best aided conditions. On the three closed-set tests (four-choice spondee, Iowa vowels, and Iowa medial consonants), R performed at chance or better. She had a few isolated correct responses on the open-set items.

![Right ear](image1.png)
![Left ear](image2.png)

**Figure 3** Distortion product emissions recordings for right and left ears, respectively. The amplitude of the emissions (circles connected by lines) is shown as a function of frequency. The shaded area represents noise and its variance. The top panels for each ear show the results for F1 and F2 primaries of 70 dB SPL; the lower panels show the results when the level of F1 was 65 dB SPL and F2 was 50 dB SPL.

<table>
<thead>
<tr>
<th>Table 1 Speech Perception Test Results</th>
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<tbody>
<tr>
<td>Four-choice spondee (closed set)</td>
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<tr>
<td>IOWA vowels (closed set)</td>
</tr>
<tr>
<td>IOWA medial consonant (closed set)</td>
</tr>
<tr>
<td>NU #6 List 1B (open set)</td>
</tr>
<tr>
<td>CID everyday sentences (open set)</td>
</tr>
<tr>
<td>IOWA sentences (open set)</td>
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<td></td>
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</tbody>
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Table 2  AEP Recording Parameters

<table>
<thead>
<tr>
<th></th>
<th>ABR</th>
<th>MLR</th>
<th>LLR</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Electrode Configuration</strong></td>
<td></td>
<td>Vertex (Cz)</td>
<td>Ipsilateral and contralateral ear canals</td>
</tr>
<tr>
<td>Noninverting (+)</td>
<td>Ipsilateral and contralateral ear canals</td>
<td>Ipsilateral ear canal</td>
<td>Ipsilateral and contralateral ear canals</td>
</tr>
<tr>
<td>Inverting (−)</td>
<td></td>
<td>Nasion</td>
<td></td>
</tr>
<tr>
<td><strong>Ground</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Signal Averaging</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Timebase (msec)</td>
<td>15</td>
<td>50</td>
<td>500</td>
</tr>
<tr>
<td>Samples</td>
<td>2000</td>
<td>1000</td>
<td>50</td>
</tr>
<tr>
<td>Filter bandpass (Hz)</td>
<td>150-3000</td>
<td>15-1000</td>
<td>1-100</td>
</tr>
<tr>
<td>(12 dB/octave slope)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Stimuli</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transducer Type</td>
<td>Broadband click</td>
<td>Tubal insert (ER-3)</td>
<td>1000-Hz toneburst</td>
</tr>
<tr>
<td>Level (dB HL)</td>
<td>95 and 105</td>
<td>95 and 105</td>
<td></td>
</tr>
<tr>
<td>Repetition rate</td>
<td>11.3/sec</td>
<td>0.7/sec</td>
<td></td>
</tr>
<tr>
<td>Polarity</td>
<td>Rarefaction, condensation, and alternating</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Prior to further consideration of an implant, additional testing was recommended to delineate site of lesion. It was at this point that she was referred to our clinic for otoacoustic emissions testing.

**Evoked Otoacoustic Emissions**

The ILO92 Otodynamic Analyser was used to test for both click-evoked and distortion product otoacoustic emissions while R rested in a reclining chair in a quiet room. Click-evoked OAEs were recorded for both right and left ears using the "nonlinear" click stimuli at 50, 70, and 60 dB peak SPL. The records from those recordings are shown in Figure 2. The top record in each series shows her responses to 80 dB peak SPL click stimulation, which is the default condition of the ILO equipment. Emissions were present across a broad frequency range for both ears, from 390 to 4300 Hz at the right ear and from 585 to 4400 Hz at the left. Good response reproducibility was maintained even when click level was reduced, as shown in the succeeding records. Likewise, distortion product emissions were recorded for both ears from 700 through 6000 Hz. DPEs were recorded with equal level primaries (70 dB SPL) and unequal and lower level (65/50 dB SPL) primaries, as shown in Figure 3.

The presence of emissions indicated that there were functioning outer hair cells (OHCs) throughout the cochlea. They were recorded in the frequency range from 400 to 6000 Hz, which reflects the frequency limits of the equipment. They were recorded at a range of stimulus levels, which also is typical of normal OHC function. Thus, R's OAE results were similar to those recorded from normal-hearing individuals. With sensory hearing impairment greater than 30 dB HL, emissions are generally absent. Since they were present in R's case, the site of lesion is presumably central to the OHCs, involving either the inner hair cells (IHCs), eighth nerve, and/or brain stem. For this reason, a comprehensive AEP study was recommended to assess the integrity of the central auditory pathway.

**Auditory Evoked Potentials**

AEPs were recorded using standard, gold-plated scalp electrodes on the vertex (Cz) (non-inverting input) and nasion (ground), and TIP-trodes seated in the ear canals (inverting inputs). Two-channel ABR and long-latency response (LLR) (N1-P2 complex) were recorded using Cz (+)-to-ipsilateral and contralateral ear canal (-) configurations. Middle latency response (MLR) to click stimuli was recorded using a Cz(+)-to-ipsilateral ear canal(−) array. Acoustic stimuli for ABR and MLR were broadband clicks (100 microsecond electrical pulses). The N1-P2 complex was evoked by 1000-Hz tonebursts (9 millisecond linear rise/fall, 50 millisecond plateau). Stimuli were presented at 95 and 105 dB HL (0 dB HL = average behavioral threshold to clicks/ tonebursts for a normal population) in rarefaction, condensation, and alternating polarities. The specific parameters used to record the ABR, MLR, and LLR are listed in Table 2.
Figures 4 to 7 are representative tracings of the ABR (Fig. 4), MLR (Fig. 5), and LLR (Figs. 6 and 7) recorded from this patient. As can be seen from these tracings, no identifiable ABR or MLR components were recordable from either ear at maximum levels of stimulation. However, a well-defined N1-P2 complex was evoked from right-ear stimulation. Component latencies were slightly delayed, but this may have been due to the patient's state of arousal (i.e., she fell asleep during the examination). Left-ear stimulation also produced an N1-P2 complex, but the components were shallower and less defined than those evoked from stimulation to the right ear.

The absence of ABR and MLR components in the presence of OAEs and an N1-P2 complex suggests a site of lesion somewhere between the IHCs and the auditory cortex. It is highly unusual to observe the N1-P2 complex of the LLR in the total absence of ABR and MLR components in adults. Squires and Hecox (1983) reported cases wherein cortical responses were intact despite the absence of ABR and MLR components. This pattern was attributable to central auditory disorders brought on by multiple sclerosis, hepatic encephalopathy, or other undiagnosed conditions. Now that we can measure OAEs in addition to AEPs, and given the findings for our patient, peripheral dysfunction at the level of the IHCs also must be included as a possible factor.

**COMMENT**

A few cases with similarities to R have been reported in the literature. Individuals with OAEs in the presence of severe to profound sensorineural hearing loss have been reported by Lutman et al (1989) and Prieve et al (1991). A case reported by Robinette (1992) was also being considered for cochlear implant. The absence of AEP components in the presence of OAEs has been reported in children (Sininger et al, 1993; Gravel and Stapells, 1993). In these cases, however, behavioral pure-tone thresholds were normal or near normal. In some of these children, wave I of the ABR was the only recordable AEP component. The appearance of wave I, however, would suggest intact peripheral function and thereby justify the presence of OAEs. In our patient, ABR and MLR were totally absent. In addition, pure-tone sensitivity was in the moderate-to-severe range bilaterally, except at 3000 to 4000 Hz in the left ear, where the loss was mild.
Berlin et al. (1993) described a patient with audiometric configuration similar to R's left ear who had no ABR or MLR, but LLRs were not reported. At this time, we can only speculate as to how OAEs and LLRs could be recordable in the total absence of brainstem and middle latency AEPs. One possible explanation is that the neural synchrony needed to produce the components of the ABR and MLR was reduced or absent in this particular patient, possibly due to the absent/abnormal output from the IHCs (Starr et al., 1991; Berlin et al., 1993; Sininger et al., 1993). The altered afferent output from the cochlea, however, may have been sufficient to produce the LLR, which is less dependent on neural synchronization (Hecox et al., 1976; Durrant and Wolf, 1991; Hyde, 1994). In the right ear, afferent cochlear output in our patient may be coming from a small population of functional IHCs in the region along the basilar membrane where 1000 Hz is coded. The patient's latest audiogram (October 12, 1993) indicates that her hearing in the right ear is most sensitive at 1000 and 1500 Hz. Pure-tone thresholds at these frequencies is 50 dB HL, which actually underestimates her SDT of 35 dB HL. In the left ear, thresholds at 3000 and 4000 Hz are in the normal range. IHC output in the region coding for these frequencies may have been sufficient to evoke the reduced $N_1 - P_2$ complex seen from stimulation to the left ear.

Figure 6 $N_1 - P_2$ complex evoked by stimulation to right ear. Tracings in left panel recorded using an ipsilateral reference ($C_5 - A_2$); tracings in right panel recorded using a contralateral reference ($C_7 - A_7$). Stimuli presented at 95 dB HL (upper tracings) and 105 dB HL (lower tracings). Values above/below arrows indicate approximate latencies for $N_1 - P_2$.

An additional explanation for the presence of the LLR relates to the innervation of the OHCs. Although their function is still controversial, a comparatively small population of afferent neurons do communicate with the OHCs (Spoendlin, 1966, 1972, 1978). The limited afferent input from the OHCs may be sufficient to evoke the LLR but not the ABR or MLR. Regardless of the rationale for our findings, this case clearly illustrates that the absence of components in early epochs may not be accompanied by absent responses/abnormal patterns in later time periods.

Our case also illustrates an interesting approach to patient evaluation. That is, few laboratories/clinics combine OAEs, ABR, MLR, and LLR in the evaluation of adult cochlear implant candidates. The otologist reports that R is currently in a "holding pattern" until more can be learned about the nature of her loss and/or appropriate treatment for it. At this point, it is assumed that a cochlear implant is

Figure 7 $N_1 - P_2$ complex evoked by stimulation to left ear. Tracings in left panel recorded using an ipsilateral reference ($C_5 - A_2$); tracings in right panel recorded using a contralateral reference ($C_7 - A_7$). Stimuli presented at 95 dB HL (upper tracings) and 105 dB HL (lower tracings). Values above/below arrows indicate approximate latencies for $N_1 - P_2$. 
contraindicated since its implantation would destroy presumably normal OHCs. Electrical stimulation of the promontory has been suggested as a possible next step in estimating if some functionally useful hearing might be achieved by cochlear implantation. Meanwhile, R communicates as best as she can via speechreading with her family and friends and is grateful for her TDD.

Acknowledgment. The authors wish to thank Shery-sue Parker for the referral, Sarah Jo Mediavilla and Dr. Charles Luetje at the Midwest Ear Institute for assistance in compiling history information, Lisa Stover for the plot consult, and Eva Callahan for help with the manuscript. Thanks is extended especially to R herself.

REFERENCES


