CSI: Audiology

A Case of Hysteria?

By Amy Ariss and Ingrid McBride

Case History
A 47-year-old female was referred for a cochlear implant by vocational rehabilitation. Audiologic evaluation performed three years prior suggested a significant hearing loss in both ears (FIGURE 1).

The patient arrived wearing a power hearing aid in the left ear. She reported hearing loss during childhood following a traumatic brain injury at age seven, although she denied awareness of any hearing loss until three years prior when she experienced sudden onset of left-ear hearing loss. Right-ear hearing loss was reported as gradual onset, resulting after a hearing test that caused “buzzing in the ear.”

Otologic history was positive for right otalgia, aural fullness, tinnitus, dizziness, and bilateral ear infections. She described “dropped hearing” episodes that occurred two to three times per week, followed by buzzing in the right ear. Lying in bed resulted in the sensation of “falling in an airplane.” Medical history was positive for diabetes managed by diet, and multiple sclerosis diagnosed at 24 years of age with associated symptoms of blurred vision, weakness, and headaches.

The patient reported that she was employed as a parking attendant in a parking garage for 11 months. She cited difficulty in hearing communication broadcast over the intercom in her parking booth, which reportedly was creating friction with her supervisors. She said that she was reprimanded for speaking too loudly and she disliked having to ask for repetition due to her hearing loss.

She indicated that she was able to communicate on a cell phone using the telecoil in the hearing aid worn in her left ear. She expressed a desire to learn American Sign Language (ASL) and to use alerting/signaling devices for the phone, door, and when driving, in order to hear sirens.

Consider the Facts
Report suggests fluctuating hearing loss, tinnitus, and imbalance.
- Significant communication difficulties that were affecting job performance.
- Reported history is inconsistent regarding when the hearing loss first started and how rapidly it progressed.
- Previous audiograms were inconsistent, lacked sufficient cross-checks.
- Audiograms were inconsistent with ability to recognize speech and use the telephone with left ear.


Welcome Back

to an ongoing series that challenges the audiologist to identify a diagnosis for a case study based on a listing and explanation of the nonaudiology and audiology test battery. It is important to recognize that a hearing loss or a vestibular issue may be a manifestation of a systemic illness. Being part of the diagnostic and treatment “team” is a crucial role of the audiologist. Securing the definitive diagnosis is rewarding for the audiologist, and enhances patient hearing and balance health care and, often, quality of life.

—Hillary Snapp, Investigator-in-Chief
Something to consider: Based on the history, what diagnostic approach would you take with this patient?

Immittance audiometry was completed first, due to its powerful predictive value regarding what to expect during audiology. Immittance results were consistent with normal middle-ear function bilaterally (Figure 2). Based on these results, no more than a severe degree of hearing loss would have been expected. Additionally, speech recognition thresholds (SRT) could not be established due to inconsistent responses, and word recognition testing (Table 1) was better than expected provided the patient’s threshold responses (Figure 3).

Based on the test results, what are the next steps? What other diagnostic, objective testing would be helpful in this case? Going back to the case history, consider the patient’s symptomology and test results:

- Possible sudden hearing loss in the left ear; gradual hearing loss in the right ear; etiology is currently unknown
- TBI at age 7 years; Multiple sclerosis; Diabetes
- Blurred vision; overall weakness; headaches
- “Buzzing” in the right ear and fluctuating hearing loss occurring 2-3 times per week
- Sensations of falling when lying down in bed
- Severe to profound hearing loss in both ears
- Full-time hearing-aid use in the left ear only

### PREVIOUS AUDIOMETRIC EVALUATION

**FIGURE 1.** Results from outside audiometric evaluations performed in 2007 and 2009 suggesting profound sensorineural hearing loss bilaterally.

**FIGURE 2.** Immittance results from audiomotor re-evaluation revealing normal middle ear function bilaterally. Admin note: No bone conduction results.

<table>
<thead>
<tr>
<th>IMMITTANCE</th>
<th>Right</th>
<th>Left</th>
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<tbody>
<tr>
<td>Peak Ytm (mmhos)</td>
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<td>0.9</td>
</tr>
<tr>
<td>Peak Pressure (daPa)</td>
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<td>20</td>
</tr>
<tr>
<td>Volume (cc)</td>
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<td>1.6</td>
</tr>
<tr>
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<td>95</td>
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<tr>
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<table>
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<tr>
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<th>2k</th>
<th>4k</th>
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<th>1k</th>
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<th>4k</th>
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<tbody>
<tr>
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<td>1k</td>
<td>100</td>
<td>2k</td>
<td>100</td>
<td>4k</td>
<td>105</td>
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<tr>
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Inconsistent report obtained during the case history
Pure-tone results suggestive of a severe to profound hearing loss, but inconsistent with the immittance and speech audiometry findings

Differential diagnosis at this time could include:
- Auditory Neuropathy Spectrum Disorder (ANSD)
- Non-organic hearing loss (NOHL)

ANSD is an auditory system disorder, post outer hair cell, that disrupts auditory function because both afferent and efferent regulation of the cochlea is impaired. According to Sininger and Oba (2001), to be considered as having ANSD, a patient must have all of the following: (1) evidence of poor auditory function, regardless of pure-tone thresholds; (2) evidence of poor auditory neural function, such as absent acoustic reflexes and abnormal auditory brainstem response (ABR); and (3) evidence of normal hair cell function as measured by otoacoustic emissions (OAEs) or the cochlear microphonic.

NOHL can be defined as “responses to a hearing test indicating a deficit greater than can be explained by organic pathology” (Austen et al., 2004).

Objective Findings
The transient-evoked OAE and distortion-product OAE (TEOAE/DPOAE) results were consistent with normal to near-normal outer hair cell function in each ear (FIGURES 4A, 4B). The ABR resulted in robust synchronous responses at 80dBnHL, bilaterally (FIGURE 5). Threshold responses could not be obtained due to high artifact levels resulting from patient restlessness during testing.

### TABLE 1. Results for word recognition in quiet, measured using recorded Computer Aided Speech Perception Assessment (CASPA) to evaluate performance based on phonemic and whole-word scoring.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Speech Materials</th>
</tr>
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<tbody>
<tr>
<td>Unaided</td>
<td>CASPA-Word</td>
</tr>
<tr>
<td>Unaided</td>
<td>CASPA-Phoneme</td>
</tr>
</tbody>
</table>

### TABLE 2. Results for aided speech and sentence recognition in quiet, measured using CASPA monosyllabic words and AzBio sentences for soft (32dBHL) and moderate (47dBHL) input levels.

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<tr>
<td>Aided</td>
<td>CASPA-Word</td>
</tr>
<tr>
<td>Aided</td>
<td>AzBio</td>
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### FIGURE 3. Pure-tone results from audiometric re-evaluation suggesting severe-profound sensorineural hearing loss bilaterally. Bone conduction could not be reliably obtained.
Hearing Aid Assessment

An electroacoustic analysis of the patient’s hearing aid revealed it to be functioning within the manufacturer’s specifications. An average gain of 67 dB SPL with a full-on volume setting, 40 dB SPL at a volume setting of two, and 30 dB SPL at a volume setting of 1 was recorded.

![Figure 4A](image1.png)

**FIGURE 4A.** Distortion product otoacoustic emissions (DPOAEs) demonstrating present responses at 1.4 kHz through 6 kHz in the right ear and at 1.4 kHz through 2.8 kHz in the left ear.

![Figure 4B](image2.png)

**FIGURE 4B.** Transient-evoked otoacoustic emissions (TEOAEs) demonstrating present responses at 1 kHz through 4 kHz in the right ear and at 1.4 kHz through 2 kHz in the left ear.

Monaurally-aided sound field warble tone thresholds with the hearing aid in the left ear revealed that the hearing aid provided exceptional gain across the frequency range (FIGURE 6). This was inconsistent with the amount of residual hearing in the left ear in conjunction with the maximum gain of the hearing aid, even if a full-on volume setting was used. At a volume setting of two or lower, which is where the hearing aid was worn, the aided thresholds measured would be impossible, based on the admitted unaided hearing thresholds in the left ear.

Additionally, monaurally-aided, the patient CASPA and AzBIO scores improved significantly, indicating excellent aided benefit and speech-recognition ability. However, the aided speech measures in quiet were inconsistent with the unaided and aided audiometric thresholds.

Based on a profound hearing loss with no measurable hearing thresholds (except for 125 Hz and 250 Hz), achieving 100 percent word
recognition and 95 percent sentence recognition, even with a hearing aid, would be impossible. Based on the aided thresholds, the speech level of 47 dB HL would be close to threshold and the speech level of 32 dB HL would be inaudible. However, the patient heard enough to repeat 60 percent of phonemes at a level that was below her aided thresholds, and 100 percent of phonemes and words and 95 percent of sentences at a level that was close to threshold.

**Diagnosis and Discussion**

The concept of a test-battery approach and implementation of the cross-check principle has always been integral to differential diagnosis in audiology (Turner, 2003; Jerger and Hayes, 1976).

A battery of tests that assess function at different levels of the auditory system and provide cross-checks to other tests in the test battery facilitates differential diagnosis. Each test contributes information about auditory processes that are normal, as well as those that are abnormal, and the clinician can determine if these findings are congruent or incongruent. The cross-check principle reminds us that the results of a single test should never be accepted as conclusive proof of the nature or site of an auditory disorder without support from at least one additional independent test (Jerger and Hayes, 1976).

A test-battery approach and the adherence to the cross-check principle were essential in the proper diagnosis of this case. Numerous inconsistencies were noted among the test results to suggest that the admitted thresholds were inaccurate and did not reflect the actual hearing levels of the patient.

Specifically, the aided and unaided behavioral speech measures were obtained at levels much lower than admitted pure-tone levels; that is, the patient heard and repeated words at levels that, based on her pure-tone thresholds, would have been inaudible. Her ability to hear speech when aided was inconsistent with a profound hearing loss in the left ear.

The patient could repeat 100 percent of words and 95 percent of sentences at average speech levels with the hearing aid in the left ear even though, based on her aided thresholds, a level of 47 dB HL would barely be audible. These findings were consistent with the observation of the patient communicating easily using her cell phone with her hearing aid.

The objective measurements of immittance audiometry, otoacoustic emissions, and ABR audiometry confirmed that her hearing acuity actually was much better than reflected on her pure-tone audiogram. These results also helped to rule out ANSD as a diagnosis.

Although more testing to obtain ABR thresholds was needed to indicate her thresholds in each ear, the obtained objective measures suggested essentially normal hearing.

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**FIGURE 5.** ABR results demonstrating normal absolute, inter-wave, and inter-aural latencies recorded at 80 dBnHL and 70 dBnHL in the right ear and at 90 dBnHL and 80 dBnHL in the left ear.
sensitivity in the right ear and no more than a mild hearing loss in the left ear. Based on these test results, the patient certainly did not meet the FDA criteria for a cochlear implant, and the hearing aid worn in the left ear was much too powerful for the minimal hearing loss in that ear.

Based on the test findings, the current audiometric thresholds documenting hearing thresholds in the severe to profound hearing-loss range in the right ear and profound range in the left ear were not accurate and suggested NOHL with functional overlay.

NOHL has a plethora of commonly accepted terminology such as functional hearing loss, pseudohypoacusis, psychogenic hearing loss, conversion hearing loss, feigning and malingering. There is always a psychological component in NOHL; the challenge is finding where on the continuum it falls.

In this case, it was determined that the patient suffered from a conversion disorder. She truly believed she had a hearing loss and gained psychological benefit from her symptoms, which appeared to be unconsciously generated. With the exception of the recent assistance provided by Vocational Rehabilitation, she did not have any external benefit to be gained.

A conversion disorder resides under the broad spectrum of somatoform disorders in DSM-IV-TR. The term conversion refers to somatic symptoms representing an individual’s attempt at resolving an unconscious psychological conflict. In other words, a medical condition is absent but symptoms suggestive of that condition are produced unintentionally. Patients with conversion disorder often are suggestible and their symptoms may be more common following extreme psychosocial stress. Conversion disorders occur more commonly in women, those of lower socioeconomic status, and more commonly on the left side of the body.

**FIGURE 6.** Aided threshold responses in the sound field to warble tone stimuli, obtained with the patient’s Unitron Unison Essential HP BTE worn monaurally in the left ear at a volume setting of 2.
It has been noted that people exhibiting NOHL who are requesting cochlear implantation display a consistent response pattern more common at the conversion disorder end of the continuum than seen with malingerers (Austen and Lynch, 2004). This consistency in assessment, which arises from the patients’ greater self-belief in their deafness, makes those with conversion NOHL difficult to detect without the use of objective assessment. It is important to recognize the need for professional counseling and to provide referring providers and patients with appropriate recommendations and referrals. In this case, the patient was referred for psychological assessment and counseling, as well as discontinuation of hearing aid use.

Final Thoughts
This case highlights the importance of obtaining a thorough case history and the importance of the cross-check principle. A basic test battery lacking in cross-checks or the selection of appropriate tests typically will result in missed or incorrect impressions. As is often the case with many potential diagnoses, if you do not look for them, you are unlikely to find them.

The audiologist should be keenly aware of potential discrepancies among test results, observed behaviors, and reported history, and in tune to possible motivating factors for NOHL. Considerable consequences may result from failing to identify NOHL, such as improper treatment and prolongation of suffering, poor patient outcomes, inappropriate referrals, and unjust medical costs.

It is best practice to exhaust all efforts to obtain accurate audiometric results and to solicit information on a potential etiology by taking a detailed case history. It is important to recognize when your test results do not agree and to rule out equipment or tester error.

A good diagnostic detective performs a thorough investigation, and understands the importance of test agreement and the implications of test findings when securing a diagnosis are essential to directing appropriate patient care. Another “case closed” until the next issue of AT!

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References


