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


Getting on Your Patient’s Good Side

By Kelly Hoffard Barninger

Case History
A 34-year-old female presented to the Hospital of the University of Pennsylvania reporting sudden onset of left ear hearing loss, aural fullness, tinnitus, and mild imbalance that had begun two weeks prior. The patient had been treated with oral prednisone for two weeks but denied an improvement in hearing or tinnitus. She reported a gradual improvement in balance over time. The patient denied facial weakness or numbness, known autoimmune disease, and family history of hearing loss. She also denied recent illness but stated that her dog had been diagnosed with methicillin-resistant Staphylococcus aureus (MRSA). Her medical history was positive for migraine headaches and a recent outbreak of mouth ulcers.

Otologic Consultation
The neurotologist ordered an MRI of the brain and internal auditory canals, and the patient was treated with two sequential intratympanic injections of dexamethasone in the left ear. Repeat audiologic evaluation did not demonstrate any significant improvement in hearing (FIGURE 2), and the MRI results were unremarkable. Hearing was determined to be essentially stable, and treatment by dexamethasone was discontinued. The patient was referred to audiology for evaluation and consultation of treatment options for single-sided deafness (SSD).

Audiometric Findings
Tympanometry revealed normal middle ear pressure and admittance bilaterally. Acoustic reflexes were present at normal sensation levels in the right ear (stimulus right) and absent in the left ear (stimulus left) when assessed at 1000 Hz. Pure tone testing suggested normal hearing sensitivity in the right ear and severe to profound sensorineural hearing loss in the left ear from 250 to 8000 Hz. A pure tone Stenger test was negative at 1000 Hz. The right ear monosyllabic word recognition score was normal (100 percent at 50 dB HL). There was no measurable word recognition ability in the left ear (FIGURE 1).
What communication difficulties and “hearing handicaps” would you expect an individual with SSD to experience?

SSD Evaluation

The patient reported significant difficulties with localization, speech understanding in background noise, and sound awareness on the left side. She also reported increased stress as a result of her communication difficulties. Subjective assessment using the Speech, Spatial, and Qualities of Hearing Scale (SSQ) questionnaire (Gatehouse and Noble, 2004) revealed significant difficulty with spatial hearing (i.e., localization) and speech understanding (TABLE 1).

The SSQ consists of 14 questions related to speech hearing, 17 questions related to spatial hearing, and 18 questions related to “other” functions and qualities of hearing (i.e., segregation and recognition of sounds, naturalness, clarity, and listening effort). The patient was asked to answer each question on a scale from 0 to 10 regarding her ability to complete a task where 0 indicates not at all and 10 indicates perfect ability.

*Based on these results, what treatment options would you consider for this patient?*

The patient was informed that a conventional hearing aid (HA) would likely be of no benefit and that the approved treatment options available for SSD included an auditory osseointegrated implant system (Baha™ is the system we use), contralateral routing of the signal (CROS) hearing aid, and an intraoral bone conduction system (SoundBite™). The patient declined the CROS hearing aid option due to the significant out-of-pocket expense. It was also determined that she was not a candidate for the SoundBite due to a history of oral and dental health issues that were contraindicative for SoundBite use.
Conventional Hearing Aid Evaluation
The patient was seated in the soundfield, one meter from a speaker located at zero degrees azimuth. The left ear was fit with our clinic’s linear Oticon 380 P behind-the-ear hearing aid, and 60 dB HL of masking was presented to the right ear via an ER3-A insert earphone. Aided sound field thresholds were present between 45 and 55 dB HL from 250 to 4000 Hz and at 90 dB HL at 6 kHz. Both consonant nucleus consonant (CNC) word and AzBio sentence test results were zero percent when assessed in quiet. As expected, the patient did not demonstrate significant benefit with conventional amplification of the left ear.

Baha Evaluation
For individuals with SSD, the Baha utilizes bone conduction to route the signal from the impaired ear to the functioning cochlea. During the evaluation, the patient was positioned in the soundfield with speakers at ±90 degrees azimuth on both sides. To assess the patient in the most difficult listening situation, speech was presented to the poorer ear, and when necessary noise was presented to the better ear. Aided testing was completed with a power processor on a headband behind the left ear. Results for the speech reception threshold (SRT) in quiet, word recognition (W-22) at various presentation levels in quiet, and speech in noise (QuickSINTM) testing are shown in TABLE 2.

The patient opted to undergo a one-week headband trial with the Baha. After one week, she reported subjective benefit with speech located to her left side and minimal improvement with speech understanding in background noise. She denied any improvement with localization, which was a primary complaint as reflected by her SSQ scores.

Why did the patient report minimal improvement in background noise?
The QuickSINTM is a speech-in-noise test scored in signal-to-noise ratio (SNR) loss. SNR loss is the increase in the SNR required for the hearing impaired patient to understand speech in noise compared to an individual with normal hearing (Christensen, 2000). The patient demonstrated only a 2 dB improvement in SNR loss with use of the Baha on the headband when compared to her unaided performance. Results of the QuickSIN are consistent with the patient’s subjective report of minimal improvement in background noise with use of the Baha during the headband trial.

Consider the Facts
The patient presents with:
- Unilateral severe to profound hearing loss
- No objective benefit with a traditional hearing aid
- Limited objective or subjective benefit observed when signal is routed from poorer ear to better ear
- Primary disability of impairment in spatial hearing (localization)

The patient’s primary complaint was impaired localization, followed by poor speech perception in noise. Given the inability to improve localization and the lack of improvement on speech-in-noise tasks when the signal was routed from the impaired ear to the normal cochlea, the patient declined all of the aforementioned options. A cochlear implant (CI) could provide a patient with SSD the opportunity to take advantage of binaural cues as the CI allows for ipsilateral stimulation of the poorer ear versus contralateral stimulation as described with the Baha, CROS, and SoundBite. While the CI does not provide natural restoration of hearing, the ability to utilize binaural input may result in improved localization and speech perception abilities in noise. The patient was advised that use of a CI for SSD was not currently an approved treatment option by the Food and Drug Administration (FDA) and was considered to be off-label. She was counseled on the potential benefits and limitations of CI versus the aforementioned treatment options and chose to pursue cochlear implantation.

Why wouldn’t the patient have perceived improved localization ability with use of a Baha, SoundBite, or CROS for treatment of SSD?
The brain utilizes interaural time differences (ITDs) and interaural level difference (ILDs) between ears to identify the location of a sound. When an individual has a profound sensorineural hearing loss in one ear, they are unable to utilize those binaural hearing cues. A Baha, SoundBite, and CROS system all route acoustic information from the impaired side to the better functioning cochlea. As a result, binaural cues are not restored and localization abilities remain impaired.

Something to consider: Based on the findings above, which treatment option do you think the patient should pursue? Are there any other treatment options that you might consider to address the patient’s reported handicaps?

Why was the off-label use of the CI supported?
Successful use of cochlear implantation for treatment of SSD is
increasingly being reported throughout the literature. Traditional treatment options for SSD that utilize contralateral routing of the acoustic signal are unable to restore hearing and auditory cues in the impaired ear. Although preoperative CI testing cannot predict postoperative outcomes, cochlear implantation has the potential to restore speech understanding abilities in the affected ear. Additionally, binaural cues may be restored, which may allow for improved localization abilities and speech understanding in noise. The patient’s age, level of motivation, duration of deafness, and overall health were taken into serious consideration prior to offering cochlear implantation as a treatment option.

How would you assess postoperative CI benefit in a patient with normal hearing in the opposite ear? Would you conduct tests to assess both functional benefit and benefit from the implanted ear alone?

CI Outcomes
The patient was encouraged to utilize the CI during all waking hours. With time, she reported significant improvement with speech understanding, localization, speech perception in background noise, and tolerance for the volume of her own voice.

Speech Testing
Aided performance on AzBio and CNC tests was assessed at 60 dB SPL presentation level in quiet. In an attempt to isolate the implanted ear during testing, the normal hearing (right) ear was plugged and muffled. AzBio testing resulted in a score of 64 percent with the CI versus 75 percent without the CI, indicating contribution from the normal hearing ear and lack of adequate attenuation from plugging/muffling. This is consistent with soundfield results demonstrating that plugging and muffling reduces binaural cues.

TABLE 2. Baha Preoperative Evaluation

<table>
<thead>
<tr>
<th></th>
<th>Unaided</th>
<th>Aided (Baha left)</th>
</tr>
</thead>
<tbody>
<tr>
<td>SRT</td>
<td>15 dB HL</td>
<td>5 dB HL</td>
</tr>
<tr>
<td>W-22 at 20 dB HL</td>
<td>28%</td>
<td>76%</td>
</tr>
<tr>
<td>W-22 at 30 dB HL</td>
<td>76%</td>
<td>100%</td>
</tr>
<tr>
<td>W-22 at 50 dB HL</td>
<td>100%</td>
<td>100%</td>
</tr>
<tr>
<td>QuickSIN (signal at 50 dB HL)</td>
<td>6.5 dB (mild SNR loss)</td>
<td>4.5 dB (mild SNR loss)</td>
</tr>
</tbody>
</table>

FIGURE 3. Left graph shows unaided thresholds with the right ear plugged and muffled (no left ear CI). Right graph shows aided thresholds (left ear CI) with the right ear plugged and muffled.
muffling the better ear only allows for 40–55 dB of attenuation (FIGURE 3). As a result, a direct audio input (DAI) system (CI-BiTS Version 5.1) was utilized during testing to isolate the implanted ear for assessment. Results suggested significant benefit for word and sentence recognition with use of the CI compared to the preoperative conventional hearing aid results (TABLE 3).

How does the CI-BiTS DAI system work, and why is it beneficial?

When using this DAI system, signals are delivered to either ear acoustically via a calibrated insert earphone and/or electrically via calibrated signals to the auxiliary input of the CI speech processor. The signals have been preprocessed with calibrated head-related transfer functions (HRTFs). This flexibility allows for reliable assessment of CI recipients with SSD, as the implanted ear can be completely isolated.

Localization
Localization abilities were also assessed through use of the DAI system. In the unaided condition, an ER3-A insert earphone was used to deliver the stimuli to the right ear only. In the aided condition, stimuli were delivered to the right ear via the insert earphone and to the left ear simultaneously via DAI. Root mean square (RMS) error significantly improved from 70 degrees in the unaided (unilateral) condition to 33 degrees in the CI (bilateral) condition (P < 0.0001).

Speech-in-Noise Testing
Speech-in-noise performance was assessed using the QuickSIN in the sound field with speech presented to the poorer ear and noise to the better ear (±90 degrees azimuth). The patient demonstrated significant improvements in SNR loss with the CI over the unaided and Baha head-band conditions (TABLE 4).

FIGURE 4. Preop and postop (six months postactivation) SSQ scores (0 = unable to complete the task, 10 = can complete the task perfectly).

### TABLE 3. AzBio and CNC Scores via DAI

<table>
<thead>
<tr>
<th></th>
<th>Preoperative</th>
<th>Nine Months Postactivation</th>
<th>18 Months Postactivation</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Left Ear: Oticon 380 P HA Right Ear: 60 dB HL masking</td>
<td>Left Ear: DAI into CI</td>
<td>Left Ear: DAI into CI</td>
</tr>
<tr>
<td>AzBio Score (Quiet)</td>
<td>0%</td>
<td>64%</td>
<td>85%</td>
</tr>
<tr>
<td>AzBio Score (Noise: +10 dB SNR)</td>
<td>Could not test</td>
<td>Did not test</td>
<td>43%</td>
</tr>
<tr>
<td>CNC Words Score (Quiet)</td>
<td>0%</td>
<td>34%</td>
<td>44%</td>
</tr>
</tbody>
</table>

### TABLE 4. QuickSIN Scores

<table>
<thead>
<tr>
<th></th>
<th>Preoperative Unaided</th>
<th>Baha on Headband</th>
<th>CI Nine Months Postactivation</th>
</tr>
</thead>
<tbody>
<tr>
<td>QuickSIN (signal at 50 dB HL)</td>
<td>6.5 dB (mild SNR loss)</td>
<td>4.5 dB (mild SNR loss)</td>
<td>−2 dB SNR loss (within normal limits)</td>
</tr>
</tbody>
</table>
SSQ
Postoperative assessment indicated subjective improvements in all categories of the SSQ, with the most significant improvement noted in spatial hearing (FIGURE 4). These subjective outcomes were consistent with the behavioral speech understanding, speech-in-noise, and localization results.

How did we assess aided benefit and address the patient’s complaints?

Complaint: Speech understanding for sound arriving to the impaired ear
Assessment: CNC words and AzBio sentences with left ear isolated via DAI

Complaint: Localization of speech and sounds
Assessment: Localization via DAI (monaural versus binaural)

Complaint: Understanding speech in background noise and in group settings
Assessment: QuickSIN in the sound field (monaural versus binaural)

Complaint: Increased stress due to communication difficulties
Assessment: SSQ (preoperatively and postoperatively)

Discussion
Progress for this case study was slower than what is typically observed with patients who have bilateral hearing loss and a cochlear implant; however, the results suggest that cochlear implantation in patients with SSD can be an effective treatment option. There is considerable variation in CI outcomes from patient to patient, and this variation will likely continue in CI patients with SSD. Additionally, traditional prognostic indicators for successful CI use (etiologies of deafness, duration of deafness, HA use) may not apply in cases of SSD.

Effective preoperative and postoperative testing of SSD/CI patients requires removing the contribution of the nonimplanted ear. Based on limited experience, masking and plugging/muffling the better ear do not accomplish this adequately. Testing via DAI allows the implanted ear to be isolated, which is essential for assessment when the nonimplanted ear has normal or near-normal hearing. Speech testing in the sound field (with and without the CI) and subjective questionnaires (preop and postop, with and without CI) allow for comprehensive assessment of hearing handicap and posttreatment benefit. Impaired localization is often a primary complaint in patients with SSD and may be the mitigating factor in pursuing CI versus other more traditional treatment options. While localization testing may not be standard in the clinical assessment of traditional CI candidates, this case demonstrates its usefulness in evaluating benefits in patients with SSD. In this case, the unique diagnostic workup was valuable in assisting the patient as she navigated through the treatment selection process, and modification of the test battery to address the auditory complaints was also beneficial. In short, it helped us to get on the patient’s “good side.” Case closed until the next issue of AT! ©

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Note
1. CI-BiTS Version 5.1 DAI system is not commercially available at this time. The CI-BiTS is currently configured for use with the Cochlear Freedom, N5, or N6 CI speech processors.

References