Introduction to the Audiological Evaluation: Case-Based Applications to Patients with Skull Base Disease

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J Neurol Surg B 2019;80:111–119.

Abstract

Objectives To provide an introduction to the role of audiological evaluations with special reference to patients with skull base disease.

Design Review article with case-based overview of the current state of the practice of diagnostic audiology through highlighting the multifaceted clinical toolbox and the value of mechanism-based audiological evaluations that contribute to otologic differential diagnosis.

Setting Current state of the practice of diagnostic audiology.

Main Outcome Measures Understanding of audiological evaluation results in clinical practice and value of contributions to interdisciplinary teams to identify and quantify dysfunction along the auditory pathway and its subsequent effects.

Results Accurate auditory information is best captured with a test battery that consists of various assessment crosschecks and mechanism-driven assessments.

Conclusion Audiologists utilize a comprehensive clinical toolbox to gather information for assessment, diagnosis, and management of numerous pathologies. This information, in conjunction with thorough medical review, provides mechanism-specific contributions to the otologic and lateral skull base differential diagnosis.

Keywords

- hearing
- assessment
- hearing loss
- audiology
- normal hearing

Introduction

More than 48 million Americans are estimated to have hearing loss in at least one ear, with 30 million of those having hearing loss in both ears.¹ Many factors may contribute to hearing loss, including but not limited to: aging, ear disease, surgical interventions, genetic disorders, exposures to ototoxic agents, infection, tumors of the temporal bone and lateral skull base, and preventable hazardous noise exposures. Because of the heterogeneous nature of hearing loss etiology, a patient-centered and team-based care approach for identification and management of hearing loss is critical. The process of identifying and quantifying dysfunction along the auditory pathway and its subsequent effects may be best facilitated by combining

otologic evaluation and audiology assessment. Although there are individual variations in the degree and scope of impact hearing loss can have on an individual's life, there are some universal challenges associated with hearing loss and decreases in health-related quality of life.^{2–4} Increased understanding of the impact of hearing loss on quality of life has led to improved methods of identifying hearing loss, assessing hearing handicap, improving hearing assistive technologies and communication strategies, as well as auditory rehabilitation. For further consideration, refer to companion articles in this special issue exploring advancements in rehabilitative options. Here, we will focus our attention on providing a case-based overview of the current state of the practice of diagnostic audiology by highlighting the multifaceted clinical

received

April 21, 2018

accepted after revision

December 31, 2018

published online

February 4, 2019

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Stuttgart · New York

DOI <https://doi.org/10.1055/s-0039-1678564>.
ISSN 2193-6331.

toolbox and the value of mechanism-based audiological evaluations that contribute to differential diagnosis.

Audiological Evaluation

To identify hearing loss and quantify status of the auditory system, a thorough audiologic evaluation is necessary. Current audiometric assessment techniques do not permit a “one-size-fits-all” approach; rather, they provide different components of the larger whole to help distinguish the presence, type, and degree of hearing loss. Due to this, the clinician can prioritize both a mechanism-based assessment approach as well as employ team-based care which can improve clinical decision making, access, cost efficiency, and the patient experience.⁵ ▶ **Table 1** provides an overview of the key assess-

ment tools and the potential clinical implications of the outcomes. The audiometric assessment is used to quantify hearing loss, to assist in the differential diagnosis of ear and balance disorders, and to monitor auditory function over time. This is of particular importance when considering the onset of the hearing loss (i.e., sudden, progressive, or fluctuating), as a comprehensive audiologic evaluation provides valuable insights into the underlying damage to auditory mechanism and opportunities for prevention and management of hearing loss. Of note, ▶ **Table 1** consists of 11 evaluation tools with no single assessment tool used in isolation for the diagnosis of an auditory disorder, as a test should be cross-checked by one or more additional measures given the varied underlying mechanisms reflected in each outcome.^{6,7}

Table 1 Primary audiologic assessment tools with associated clinical application and summary of clinical implications of abnormal findings

Assessment tool	Primary clinical application	Possible implications of abnormal findings by primary location			
		Outer ear	Middle ear	Inner ear	Retrocochlear
Air conduction pure-tone audiometry (including conventional behavioral audiometry, conditioned play, and visual reinforcement may be obtained with supra-aural or circumaural headphones, insert earphones, or sound field speakers)	–Behavioral measure of the outer, middle, and inner ears; retrocochlear disorders	–High-frequency conductive loss with headphones may indicate collapsing ear canal –Cerumen impaction –Tympanic membrane perforation	–Otosclerosis –Cholesteatoma –Ossicular discontinuity –Glomus tumor –Otitis media with effusion	–Sensorineural hearing loss –Presbycusis –Noise-induced hearing loss –Ototoxic hearing loss –Meniere’s disease –Enlarged vestibular aqueduct –Superior canal dehiscence	–Vestibular schwannoma
Bone conduction pure-tone audiometry (including same techniques as air conduction)	–Behavioral measure primarily of the inner ear; retrocochlear disorders	–Cross-check for conductive hearing loss	–Otosclerosis (Carhart’s notch: dip in score at 2,000 Hz in bone conduction)	–Sensorineural hearing loss –Presbycusis –Noise induced –Ototoxicity –Meniere’s disease	–Vestibular schwannoma
Speech threshold audiometry (includes SRT and “speech detection threshold/ speech awareness threshold”)	–Behavioral measure to primarily cross-check reliability of PTA –Valuable tool for children or adults who may be suspected of malingering			–If SRT and PTA differ by more than ± 10 dB, the reliability of the pure-tone test results may be questioned –Employ additional measures to cross-check results	
Suprathreshold speech recognition in quiet (includes monosyllabic “word recognition scores,” phoneme recognition, and sentence recognition)	–Behavioral measure to assess ability to understand speech at a suprathreshold level in a quiet environment			–Sensorineural hearing loss –Meniere’s disease	–Vestibular schwannoma –Central auditory disorders
Suprathreshold speech recognition in noise	–Behavioral measure to assess ability to understand speech at a suprathreshold hearing level in background noise	–More severe degrees of conductive hearing loss	–Mixed hearing loss	–Sensorineural hearing loss –Cochlear synaptopathy or hidden hearing loss	–Central auditory disorders
Tympanometry (standard probe tones of 226 or 1,000 Hz depending on age)	–Objective measure of the outer and middle ears (pressure, compliance, and ear canal volume used for differential diagnosis)	–Cerumen impaction –Otitis externa –Tympanic membrane perforation –Retracted tympanic membrane	–Middle ear effusion –Otosclerosis –Cholesteatoma –Glomus tumor –Ossicular discontinuity –Monomere or tympanosclerosis		

Table 1 (Continued)

Assessment tool	Primary clinical application	Possible implications of abnormal findings by primary location			
		Outer ear	Middle ear	Inner ear	Retrocochlear
Wideband reflectance tympanometry	<ul style="list-style-type: none"> –Objective measure primarily of the middle ear, across a wider bandwidth of frequencies than standard tympanometry (226 Hz) –Demonstrates amount of sound energy being absorbed into middle ear space relative to normative data 		<ul style="list-style-type: none"> –Otitis media with effusion –Otosclerosis –Ossicular discontinuity –Tympanic membrane perforation –Cholesteatoma 	<ul style="list-style-type: none"> –Superior canal dehiscence 	
Acoustic reflex threshold testing	<ul style="list-style-type: none"> –Objective measure of the outer, middle, and inner ears; retrocochlear disorders –Primarily used to identify the site of lesion of hearing loss –If present, should be 85 dB HL (± 10 dB) with normal hearing 	<ul style="list-style-type: none"> –Conductive hearing loss, prohibiting the signal to reach the cochlea with sufficient intensity 	<ul style="list-style-type: none"> –Conductive hearing loss or other pathology which prohibits the signal to reach the cochlea with sufficient intensity 	<ul style="list-style-type: none"> –Hearing loss greater than ~60 dB HL 	<ul style="list-style-type: none"> –Vestibular schwannoma –Auditory neuropathy spectrum disorder –Facial nerve disorders (e.g., Bell's palsy) –Intra- and extra-axial brainstem tumors –Demyelinating disorders
Acoustic reflex decay	<ul style="list-style-type: none"> –Objective measure of the outer, middle, and inner ears; retrocochlear disorders –If reflex thresholds cannot be obtained, decay cannot be evaluated 	<ul style="list-style-type: none"> –Cross-check for conductive hearing loss 	<ul style="list-style-type: none"> –Cross-check for conductive hearing loss 	<ul style="list-style-type: none"> –“Negative” acoustic reflex decay anticipated 	<ul style="list-style-type: none"> –If reflex decay is “positive,” this indicates a 50% reduction in the magnitude of acoustic reflex –Positive is associated with retrocochlear disorders
OAEs	<ul style="list-style-type: none"> –Objective measure of the outer, middle, and inner ears –Most likely to be measured in a patient with no greater than a mild loss (30–40 dB HL) –May be quantified as “present/absent” for screening applications (e.g., newborn hearing screening) –May be quantified per frequency for diagnostics 	<ul style="list-style-type: none"> –Conductive hearing loss, prohibiting the signal to reach the cochlea with sufficient intensity or the OAEs to be measured as they return from the cochlea 	<ul style="list-style-type: none"> –Conductive hearing loss or other pathology which prohibits the signal to reach the cochlea with sufficient intensity or the OAEs to be measured as they return from the cochlea –May or may not be recorded with ear tubes –Pressurized OAEs may be applied in negative middle ear pressure 	<ul style="list-style-type: none"> –Sensorineural hearing loss –Early identification of noise-induced hearing loss –Early identification of ototoxic hearing loss 	<ul style="list-style-type: none"> –OAEs can also be measured in patients with an intact cochlea, even when the auditory nerve may be compromised (e.g., ANSD, vestibular schwannoma)
ABR evaluation	<ul style="list-style-type: none"> –Objective measure of the outer, middle, and inner ears; retrocochlear disorders –Differentiate conductive, sensorineural, mixed, or retrocochlear hearing loss –May be used to estimate degree of hearing loss –Air or bone conducted stimuli 	<ul style="list-style-type: none"> –Abnormally prolonged waveform latencies are typically associated with a conductive hearing loss 		<ul style="list-style-type: none"> –Morphology of waveforms, absolute, and interpeak latencies can be used to quantify sensorineural hearing loss 	<ul style="list-style-type: none"> –Prolonged interpeak waveform latencies may be associated with vestibular schwannoma –Absent or abnormal ABR response in conjunction with normal OAEs is typically associated with ANSD –Demyelinating disorders

Abbreviations: ABR, auditory brainstem response; ANSD, auditory neuropathy spectrum disorder; OAE, otoacoustic emission; PTA, pure-tone average; SRT, speech reception threshold.

Note: Abnormal findings are delineated by chief location of dysfunction to highlight opportunities of outcome validations by behavioral and objective measures. Abnormal findings assume accurate equipment calibrations, troubleshooting, and cross-checks or confirmation with multiple pieces of test equipment have been employed. It is to be noted that for the scope of this article, ► **Table 1** is not comprehensive of all audiometric evaluation tools excluding questionnaire measures, full scope of auditory evoked potentials (e.g., auditory steady state response), auditory processing disorder screenings, Stenger test, etc.

Source: Information in this table was adapted and summarized from American Academy of Audiology⁸ and American Speech-Language-Hearing Association.⁹

Behavioral hearing thresholds obtained in a controlled, audiometric sound booth at conventional test frequencies (0.25–8 kHz) is often the initial, “gold-standard” assessment tool.^{8,9} The goal is to obtain audiometric threshold, or the lowest intensity the listener can identify the presence of the test signal at least 50% of the time. Audiometric thresholds are obtained through both air and bone conductions with differences between air- and bone-conduction responses indicating the type of hearing loss (sensorineural, conductive, or mixed). This serves as the most global snapshot of ear-specific auditory comprehension and function at primary speech frequencies. More detailed measures add to this initial assessment to fully define the auditory dysfunction as illustrated in the cases later.

Case 1: Hearing Loss Secondary to Vestibular Schwannoma

The first case (►Fig. 1) illustrates the importance of team-based care for patients with multiple auditory complaints. The patient presented with primary complaints of unilateral hearing loss and tinnitus, accompanied by transient nonvertiginous dizziness. Audiologic assessment demonstrated an asymmetric sensorineural hearing loss (►Fig. 1). This hearing loss is classified as sensorineural, as hearing thresholds obtained via air-conducted signals are equal to those obtained via bone-conducted signals. Specifically, Case 1 demonstrates hearing within normal limits for the right ear 250 to 8,000 Hz and a moderate sloping to severe sensorineural hearing loss 3,000 to 8,000 Hz in the left ear. This hearing loss is classified as asymmetric, as there is a more than 15 dB difference in at least two frequencies when comparing the ears.¹⁰ Given the interaural asymmetry, further evaluation was warranted beyond audiometry and case history to delineate the origin of the damage as cochlear or retrocochlear (eighth nerve and beyond in origin).

Acoustic reflex threshold testing was performed to assist with differential diagnosis of the site of lesion.^{11,12} Acoustic reflex threshold testing involves the presentation of an intense sound to the ear to elicit a contraction of the stapedius muscle in the middle ear, causing an increase in stiffness that can be easily detected at the tympanic membrane. The response to the acoustical stimulation may occur in the ipsilateral (probe ear) or contralateral (opposite) ear with the complete pattern of results obtained in each probe ear. Stimulating and recording in both modes allow for isolation of the location of an abnormality within the acoustic reflex pathway. Specifically, contralateral testing evaluates the contralateral eighth nerve, crossover pathways of the brainstem, ipsilateral seventh nerve, and middle ear system. Ipsilateral testing targets the ipsilateral eighth nerve, brainstem connections, ipsilateral seventh nerve, and middle ear. These key structures and testing pathways are demonstrated in ►Fig. 1C adapted from Emanuel.¹³ Acoustic reflexes are present in normal hearing individuals when the stimulus is 85 dB HL (± 10). In Case 1, the left ear ipsilateral and right ear contralateral acoustic reflexes were absent, isolating the location of abnormality to the left ear eighth nerve consistent with hearing thresholds (►Fig. 1). If facial paralysis on the affected side was noted, the left ipsilateral acoustic reflex would be absent as well. Given presumed localization to the

Fig. 1 (A) Audiogram legend represented. (B) Pure-tone audiometry with contralateral acoustic reflex thresholds represented for Case 1 (vestibular schwannoma; left ear). Specifically, air-conduction hearing threshold level (dB HL) as a function of frequency (250–8,000 Hz) for the right ear (O) and left ear (X or square illustrated when masking was used) are demonstrated here. Symbols displayed in the gray-shaded region at 25 dB HL or better demonstrate hearing is within the normal sensitivity range, while those thresholds outside of the region with corresponding bone conduction results are consistent with hearing loss. Acoustic reflex threshold testing demonstrates that right contralateral reflexes are within the normal range (bands at bottom of graph), while left contralateral reflexes are elevated (500 Hz) or absent (1,000 and 2,000 Hz). (C) Schematic illustration of acoustic reflex pathways demonstrating the pathway of a signal that travels through the outer, middle (ME), and inner ear (IE), along the eighth nerve to the brainstem. There, the signal arrives first at the cochlear nucleus (CN), both superior olivary complexes (SOC) and facial nerve (VII) nuclei to result in a contraction of both stapedius muscles.

eighth nerve, acoustic reflex decay testing was also performed, and proved a valuable contribution to the test battery. Acoustic reflex decay testing is performed by presenting a sustained pure-tone stimulus at 10 dB above the acoustic reflex threshold for 10 seconds to the ear contralateral or ipsilateral relative to the probe tip. The ear receiving the stimulus is the ear being evaluated with a present (abnormal) reflex decay determined by the reduction in magnitude of the reflex to one-half of its original size within 10 seconds of stimulation. In a normal ear, the reflex should stay contracted the full 10 seconds, as was measured in the patient's right ear; reflex decay, or the inability to stay contracted, was measured in the left ear. The presence of reflex decay is associated with retrocochlear disorders and the combined use of acoustic reflex thresholds and reflex decay results in 85 to 95% sensitivity depending on criteria used.^{14,15}

Speech audiometry may also provide important information for diagnosis (refer to historic review by Olsen¹⁶). Specific to retrocochlear disorders is the presence of "rollover" during suprathreshold speech audiometry. Rollover is the phenomenon of reduced word recognition scores with increasing intensity.¹⁷ In Case 1, the word recognition score at 30 dB sensation level (SL) relative to pure-tone average (PTA) demonstrated 45% correct which declined to 27% at 40 dB SL and 4% at 50 dB HL, consistent with the rollover effect found in patients with retrocochlear pathology. Given the reported unilateral symptoms combined with the audiological evaluation findings for Case 1, the patient was referred to otology for medical evaluation including radiological imaging leading to a confirmation of a vestibular schwannoma in the left ear.

Case 2: Conductive Hearing Loss from Otosclerosis

►Fig. 2 illustrates the audiologic assessment of a patient with a primary complaint of gradually (over ~10 years) worsening hearing loss greater in the right ear than the left ear. Behavioral hearing thresholds demonstrated bilateral conductive hearing loss, characterized by the normal bone conduction thresholds and elevated air conduction thresholds (air-bone gap). Thresholds obtained using a masking signal to the contralateral ear to ensure that the level of the signal to the test ear does not "crossover" to elicit a response from the nontest ear. This masking would also be necessary during speech audiometry to ensure isolation of the test ear. Although suprathreshold word recognition scores are not plotted, they were judged to be in good agreement with hearing thresholds (scores > 90% correct bilaterally) without evidence of rollover. Excellent scores are not uncommon in the presence of conductive hearing loss, as patients typically do not have the same difficulties with speech distortion as exhibited in those with cochlear losses.

►Fig. 2B, C demonstrates tympanometry, which combined with acoustic reflex testing encompasses the acoustic immittance assessment. Immittance measures contribute to differential diagnosis, as these measures specifically contribute to the determination of the type of hearing loss (conductive, sensorineural, or mixed) and quantify middle ear dysfunction.^{8,9,16} Tympanometry is especially helpful in identifying the possibility of a conductive component through measures of ear canal volume, static compliance, and middle ear pressure as measured

Fig. 2 (A) Case 2 representing otosclerosis with results represented in the same format as ►Fig. 1 (legend—►Fig. 1A). The bracket symbols at the top of the audiogram reflect masked bone conduction for the right ear (I) and the left ear (J) with interoctaves removed for clarity. Results demonstrate a bilateral moderate to moderately severe conductive hearing loss. Immittance testing demonstrates ipsilateral and contralateral acoustic reflexes are absent bilaterally. (B) Tympanometry demonstrates normal middle ear pressure, compliance, and physical volume for the right ear and (C) left ear.

in the ear canal. The tympanogram (►Fig. 2B, C) is the graphic representation of the middle ear compliance in relation to changes in pressure. In Case 2, tympanometry demonstrates normal results even in the presence of a conductive component. Previous work has described that only 25% of ears with otosclerosis demonstrate abnormal tympanometry, demonstrating

the importance of the cross-check principle in diagnostic assessment of auditory and otologic pathologies.¹⁸ Newer tools, such as wideband tympanometry or wideband energy reflectance (WBR), may provide a greater description of the possible etiology, particularly in the cases of middle ear disorder. WBR evaluates the middle ear response across a greater range of frequencies than does standard tympanometry which is typically only measured for a probe tone of 226 Hz for adults or 1,000 Hz for infants 6 months or younger. Anticipated results for WBR for Case 2 include higher energy reflectance than the 90% range of results (see review by Shanks and Shohet¹⁹). Acoustic reflex thresholds (plotted as symbols in ►Fig. 2A) were absent consistent with the conductive hearing loss. While the presentation of the intense sound used for acoustic reflex assessment typically produces a contraction of the stapedius muscle in the middle ear, resulting in an increase in stiffness that can be detected at the tympanic membrane, this is not the case for conductive hearing loss. In the presence of a conductive hearing loss, acoustic reflexes are typically unable to be measured for either of two reasons: (1) conductive pathology disrupts measurement of the muscle contraction changes in the plane of the tympanic membrane; (2) signal intensity delivered to the cochlea is insufficient to elicit a response, as it is dampened by the conductive component.

For Case 2, patient was referred based on audiological evaluation to a physician for medical evaluation of the bilateral conductive hearing loss in the presence of normal tympanometry, and subsequent imaging revealed bilateral fenestral otosclerosis. Given the conductive hearing loss, an additional consideration for the otologic and lateral skull base differential diagnosis may have included nasopharyngeal or infratemporal fossa tumors, resulting in Eustachian tube blockage and resultant serous otitis media. However, if serous otitis media were present, tympanic membrane movement would be impaired resulting in abnormal tympanometry. Another consideration in the differential diagnosis in the setting of a conductive hearing loss would be jugular paragangliomas from the tumor growth in the middle ear, abutting the tympanic membrane and ossicular chain; however, again tympanometry would have been abnormal, secondary to impaired middle ear function.

Case 3: Sudden Sensorineural Hearing Loss

Case 3 (►Fig. 3A–C) demonstrates a series of audiometric assessments which monitored auditory function over time in the case of a patient with sudden sensorineural hearing loss (SSNHL; refer to published guidelines for detailed recommendations²⁰) in the right ear. SSNHL is most commonly idiopathic in nature; however, further workup is necessary to rule out less common causes, including vestibular schwannoma, cerebellopontine angle meningioma, or endolymphatic tumors. Often, the first step in the full evaluation of an SSNHL is the audiometric evaluation. Behavioral hearing thresholds quantified the impact of the hearing loss at conventional test frequencies (0.25–8 kHz). Suprathreshold speech recognition, which supplements the pure-tone audiometric findings, was unable to be assessed due to the degree of hearing loss and restricted audibility in the speech frequencies; however, a speech

Fig. 3 Case 3 representing sudden sensorineural hearing loss (SSNHL) with results represented in the same format as ►Fig. 1 (legend—►Fig. 1A). (A) Pure-tone audiometry SSNHL which demonstrated a mild to moderate sensorineural hearing loss 4,000 to 8,000 Hz bilaterally. (B) Hearing thresholds obtained approximately 6 hours after SSNHL onset with stable hearing in the left ear (X) and a significant decrease in hearing thresholds across the frequency range in the right ear (O or triangle to represent masking used), with a moderately severe sensorineural hearing loss at 250 Hz and no measurable hearing 500 to 8,000 Hz (side arrows represent no response at the output limits of the audiometer). (C) Hearing thresholds obtained approximately 1 month after the patient received a series of steroid treatments with noted improvement in the right ear 250 to 1,500 Hz and stable hearing in the left ear.

awareness threshold (SAT) was obtained to corroborate the air-conducted PTA. The SAT agreed with the PTA, which is an expected finding in a patient with reliable responses. Acoustic reflex thresholds were measured at normal levels in the (none suspect) left ear, consistent with degree of normal hearing sensitivity at the test frequencies. At the time of SSNHL (►Fig. 3B), acoustic reflexes were absent in the right ear, consistent with the profound degree of sensorineural hearing loss measured on audiometry. Following steroid treatment (►Fig. 3C), acoustic reflexes were measured within normal limits in the right ear at 500 Hz, consistent with improvement in hearing at that threshold.

Objective testing with otoacoustic emissions (OAEs), however, demonstrated no response from the right ear, consistent with degree of hearing loss. OAEs are low-intensity sounds originating from the outer hair cells of the cochlea as described by Kemp²¹ and are a product of the nonlinear processing of a healthy inner ear. OAEs are widely recognized for providing a noninvasive measurement to assist in making clinical decisions regarding normal versus impaired cochlear function.^{22,23} The presence of OAE amplitudes within normal limits when recorded with normal middle ear function suggests normal cochlear function (i.e., hearing within normal limits). OAE levels are reduced with mild or moderate hearing losses and are rarely present when hearing thresholds exceed 60 dB HL.²²

Case 4: High-Frequency Hearing Loss Due to Ototoxicity

►Fig. 4 illustrates hearing loss at frequencies above 8 kHz in a patient undergoing ototoxic cisplatin chemotherapy for oral cavity squamous cell carcinoma. This patient denied concerns about hearing sensitivity, but did report marked difficulty understanding speech in background noise. Initial audiometric

Fig. 4 Case 4 demonstrated audiometric results represented in the same format as ►Fig. 1 (legend—►Fig. 1A). Pure-tone thresholds were obtained following a course of cisplatin chemotherapy for which baseline thresholds were < 20 dB HL 250 to 16,000 Hz. Results relative to baseline demonstrate a left (X) greater than right (O) asymmetric extended high-frequency (9,000–16,000 Hz) hearing loss consistent with ototoxicity.

evaluation at conventional frequencies (up to 8,000 Hz) revealed hearing well within the normal range; however, extended high-frequency evaluation confirmed a deficit. To further characterize ototoxic damage, distortion product otoacoustic emission (DPOAE) testing was performed in the right (►Fig. 5A) and left ears (►Fig. 5B). For Case 4, ototoxicity was demonstrated in the DPOAEs consistent with the cochlear outer hair cells being particularly susceptible to ototoxic agents.²¹

It is well established that there is value in assessing auditory function at frequencies above 8 kHz to detect age-related changes^{24,25} and ototoxic damage in the cochlea.^{23,26–28} Physiological changes in the auditory periphery due to age and ototoxicity are not only initially observed, but also more prominent at frequencies above 8 kHz corresponding to the cochlear base compared with lower frequencies corresponding to the cochlear apex.^{27–29} The underlying mechanical properties of the cochlear base differ from those of the apex, resulting

Fig. 5 Distortion product otoacoustic emissions (DPOAEs) for Case 4. OAE level (amplitude) as a function of frequency for the right (O; A) and left (X; B) ears is represented. Baseline DPOAEs obtained prior to cisplatin chemotherapy were present above the noise floor (NF) within normal limits at all frequencies 1,000 to 10,000 Hz. Shown here are the DPOAEs after treatment demonstrating cochlear outer hair cell dysfunction due to ototoxicity in the presence of normal tympanometry bilaterally. Consistent with asymmetric extended high-frequency hearing loss, there is a decrement in high-frequency OAE amplitude for the right ear (A) and reduced or absent high-frequency OAEs for the left ear (B).

in systematic variations in characteristic frequency and tuning properties across the cochlea. A full review of cochlear anatomy is outside the scope of this article (see review Robles and Ruggero³⁰); however, given the variation in cochlear base to apex mechanics, it is not surprising that hearing loss manifests in different damage patterns at different frequencies.

In addition to assessing extended high-frequency hearing for purposes of detecting age-related changes and ototoxic damage in the cochlea, extended high-frequency hearing has an important role for speech understanding in noise.^{31,32} To address concerns regarding understanding speech in noise for Case 4, word recognition was performed both in quiet and in background noise. This is an important component of the evaluation for patients with concerns regarding hearing in background noise, as it allows for a better estimate of the individual's communication needs in everyday settings. Speech-in-noise testing with sentence materials was used to determine the signal-to-noise ratio (SNR) loss. SNR loss is the dB increase in SNR required to understand speech in background noise; individuals with hearing loss have greater SNR losses than those with normal hearing sensitivity.³³ In essence, when an individual has hearing impairment or difficulty hearing in background noise, increasing the level of speech (signal) and reducing the noise (quieter) is needed for successful understanding of the conversation. Consistent with normal hearing sensitivity present in the range of speech frequencies, word recognition in quiet was 100% in both ears in good agreement with audiometric findings. Speech-in-noise testing, however, demonstrated a significant SNR loss for this patient, consistent with extended high-frequency hearing loss and primary concerns regarding ability to understand in background noise.

Additional Considerations

The clinical utility of behavioral threshold testing has been largely explored; however, factors such as patient compliance, attention, and motivation can influence outcomes (see review by Lee et al³⁴). Objective measures, such as immittance measures or otoacoustic emissions permit noninvasive measures of auditory pathways and estimation of degree of hearing loss. Additional measures include auditory evoked potentials such as the auditory brainstem response (ABR), an objective measure of auditory nerve status. The ABR consists of a waveform recording five to seven peaks that reflect the synchronous discharge of a large number of auditory neurons at progressively more central points in the pathway from the cochlear to the auditory cortex and provides an estimate of hearing sensitivity (see review by Paulraj et al³⁵). Objective measures are particularly useful when obtaining information from patients whose behavioral threshold testing is compromised by the aforementioned factors (e.g., pediatric patients). Questionnaire measures may be of additional assistance for guiding the clinical encounter and in quantifying symptoms (e.g., tinnitus, imbalance, etc.).

Additional utility of the ABR includes intraoperative eighth nerve monitoring during surgery to mitigate damage to the auditory pathway (see review by Hal Martin and Shi³⁶). Given the surgical approach during vestibular schwannoma re-section or decompression of cranial nerves, the eighth nerve may be at

increased risk for damage. Intraoperative monitoring of the eighth nerve's function during surgery can be utilized to provide early warning of potential damage incurred to the auditory pathway up to the level of the brainstem; this is particularly useful if a goal of the surgery is hearing preservation. Specifically, baseline ABR waveforms may be obtained from the patient prior to any surgical intervention. During the surgical intervention, ABR recordings can be continuously obtained and compared with the patient's baseline; any deviation in perisurgical waveforms in terms of latency (neural conduction time) or amplitude (size of the response) can provide an indication of change in the functioning of the auditory pathway that may inform the surgical team's management.

There are important considerations for interpretation of assessment findings that merit mentioning. The first consideration is test reliability. On the audiometric evaluation forms, there is an indication of reliability which is judged by the audiologist regarding the validity of the reported results and is most commonly impacted by patient-related variables, such as consistency of responses, language/cultural influences, or cognitive status during the examination (e.g., tired, off-task, etc.). Reliability can also be affected by the ease with which an audiologist is able to test a patient. Certain populations may include patients who are more challenging to evaluate with behavioral assessment due to impacts on reliability. For example, patients with tinnitus as the test stimuli may interfere with the tinnitus perception, pseudohypacusis (malingering, functional hearing loss, and nonorganic hearing loss), and auditory neuropathy spectrum disorder as the perception of sound may fluctuate, or other developmental, cognitive, and behavioral delays or disorders as the patient may not understand the task. In these instances, the use of objective measures and a thorough medical evaluation is critical to supplemental standard procedures to obtain a reliable evaluation.

Conclusion

Current audiologic evaluation techniques integrating mechanism-based considerations and team-based care approach can contribute significantly to the differential diagnosis of the patient with hearing loss. This article summarized the major components of the audiologic evaluation and monitoring of hearing which applies across the lifespan. The audiologic outcomes explored here through clinical case illustrations combined with medical evaluations complete the comprehensive clinical assessment of the patient with hearing loss and guides the management pathway to meet the needs of the patient.

Conflict of Interest
None.

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