Otoacoustic Emissions as a Cross-Check in Pediatric Hearing Assessment: Case Report

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Abstract

Audiologic assessment of infants and young children can be confounded by neurologic disorders or neuromaturational delays. In some cases, this results in an inability to assess hearing sensitivity by behavioral measures or by auditory evoked potentials. This case illustrates such an audiologic challenge. Subject DF was born with hydrocephaly, which was treated with repeated shunt surgeries and resulted in seizures and pervasive developmental delays. At 9 months of age, the child was tested by auditory brainstem response (ABR) measurement and found to have no response to sound. Believing that her child had hearing, DF's mother sought a second opinion. Results of an audiologic evaluation at 11 months of age showed no measurable behavioral responses in the sound field and an ABR abnormality that prevented prediction of hearing sensitivity. In contrast, sensitivity prediction by the acoustic reflex and results of both transient-evoked and distortion-product otoacoustic emissions predicted normal peripheral hearing sensitivity. This case illustrates the usefulness of otoacoustic emissions as an additional cross-check measure in pediatric hearing assessment.

Key Words: Auditory brainstem response (ABR), hearing assessment, otoacoustic emissions, pediatric audiology

The assessment of hearing in children has been enhanced over the past 2 decades by the addition of immittance audiometry and auditory evoked potentials. This has led to the ability to predict hearing sensitivity, with considerable accuracy, in newborn infants and young children. Combined with behavioral measures, techniques such as the sensitivity prediction by the acoustic reflex (SPAR) test (Jerger et al, 1974) and the auditory brainstem response (ABR) test (Sohmer et al, 1972; Hecox and Galambos, 1974) serve as invaluable cross-checks in pediatric audiometry (Jerger and Hayes, 1976).

The nature of our challenge has changed, however, as we have extended the limit at which we expect to identify hearing loss in children to ever younger ages. The Joint Committee on Infant Hearing (1991) has now set as a goal that all infants with hearing impairment be identified and treatment initiated by 6 months of age. Simultaneously, we are being challenged to evaluate neonates who are of ever increasing fragility. Recent studies have shown dramatic increases over the past 2 decades in the survival rate of neonates with very low birth weights (Shenai, 1992). Of those who do survive, the incidence of major handicaps is substantial and inversely related to birth weight (Grøgaard et al, 1990). That means that as our goal begins to shift to identifying hearing loss by 6 months of age, we are, at the same time, being challenged to evaluate infants with increasingly complex neurodevelopmental complications.

Conventional audiologic assessment of infants and young children can be readily confounded by these neurologic disorders or neuromaturational delays. In many cases, such complex problems can result in an inability to
predict hearing sensitivity by behavioral measures or by auditory evoked potentials. It is no longer uncommon to encounter a neonate or young infant who does not have an apparent startle response, or other visible reaction, to auditory stimulation. Nor is it uncommon to encounter infants who have abnormal ABRs as a result of neurologic impairment. In such cases, information about hearing sensitivity may be limited to predictions based on acoustic-reflex measures and clinical intuition.

Recent advances in techniques for measuring otoacoustic emissions (OAEs) hold promise as an additional cross-check in pediatric hearing assessment. First described by Kemp in 1978, OAE measures have been found to be exquisitely sensitive indicators of cochlear function and, as a consequence, useful predictors of peripheral hearing sensitivity. OAEs are robust in infants with normal middle ear and cochlear function and can be measured routinely in newborns (Bonfils et al., 1988; Kok et al., 1993; White et al., 1993). Since OAEs reflect the integrity of cochlear structures and are preneural in nature (Martin et al., 1987), they are unlikely to be compromised by neurologic dysfunction (Lutman et al., 1989), which might render behavioral hearing assessment impossible and auditory brainstem response interpretation difficult.

The present paper provides an illustrative example of the application of OAEs to the audiologic challenge presented by a neurologically impaired child. Subject DF was born with severe hydrocephaly, resulting in seizures and pervasive developmental delays. At 9 months of age, he was reported to have no measurable ABRs. Results of an audiologic evaluation at 11 months of age showed no measureable behavioral responses in the sound field and an ABR abnormality that prevented prediction of hearing sensitivity. In contrast, sensitivity prediction by the acoustic reflex and results of both transient-evoked and distortion-product OAEs predicted normal peripheral hearing sensitivity. This case illustrates the usefulness of OAEs as an additional cross-check measure in pediatric hearing assessment.

**CASE REPORT**

**Description of Subject and Clinical Course**

Subject DF was the second of two children, the first being a healthy, normal-hearing, 7-year-old girl. The pregnancy was complicated by maternal hypertension, which was treated with Aldomet. At 35-weeks gestation, a routine ultrasound revealed severe hydrocephaly. DF was delivered at 36-weeks gestation, by cesarean section, on March 5, 1992, with Apgar scores of 5 and 7. Birth weight was 2.76 kilograms. He had mild respiratory distress syndrome, which required intubation, but was weaned to room air and extubated by day 4 of life. A sonogram and computerized tomography (CT) scan of the head, on day 1, demonstrated significant ventriculomegaly with only a small rim of cortical tissue. A shunt was placed immediately, and DF was treated with vancomycin and cefotaxime postoperatively. He progressed well initially and was discharged at 2 weeks of age. During the first 4 months of life, he was readmitted three times for shunt malfunction and revision. One of these admissions was complicated by a shunt infection, meningitis, and the development of a seizure disorder. At the time of the audiologic assessment, DF was undergoing treatment for pervasive neurodevelopmental delays and was on phenobarbital to control his seizures.

Otologic history included two episodes of otitis media since birth. Although there was no family history of hearing impairment, the positive medical history led to a referral for auditory

**Figure 1** Auditory brainstem responses from the right and left ears of Subject DF, a 9-month-old male, to click stimuli presented at 85 dB nHL. Responses were recorded with reference electrodes that were ipsilateral (ipsi) and contralateral (contra) to the stimulated ear. Testing was carried out at an independent neurophysiologic laboratory.
brainstem response testing at 9 months of age. On December 24, 1992, ABR testing was carried out at an independent neurophysiology laboratory. Results are shown in Figure 1. ABRs were recorded in response to rarefaction clicks presented at 85 and 65 dB nHL at a rate of 10/sec. No observable ABRs were recorded from either ear at either stimulus intensity. The neurophysiologic report stated, "No response from either ear at 85 or 65 dB HL, suggesting a severe hearing loss bilaterally. This could also be due to muscle and movement artifact. Retesting in three months is recommended if clinically indicated."

DF's mother was not convinced that he had a significant hearing loss. Although she was aware that he had many developmental problems, she was adamant that hearing impairment was not among them. As a result, she sought a second opinion, and an audiolingual evaluation of DF was carried out, at 11 months of age, on February 17, 1993, at the Georgetown University Medical Center Division of Audiology in Washington, DC.

**DATA AND OBSERVATIONS**

**Audiologic Test Battery**

The test battery used for pediatric assessment at the Georgetown University Medical Center includes behavioral audiometry, immittance audiometry, auditory brainstem response testing, and otoacoustic emission measurements.

Behavioral testing was carried out by conventional behavioral observation audiometry in a sound field. Warbled tones, narrow-band noise, and speech stimuli were presented via an audiometer (Grason-Stadler, GSI-16) through loudspeakers in a sound-isolated test booth. One audiologist controlled signal presentation, and another monitored behavioral responses.

Immittance audiometry included measures of tympanometry, static immittance, and acoustic reflex thresholds. A conventional immittance meter (Grason-Stadler, GSI-33) was used. Values for the SPAR measurements were calculated as the difference between the crossed acoustic reflex threshold to broad-band noise and the average acoustic reflex threshold to pure-tone signals at 500, 1000, and 2000 Hz, plus a correction that yielded a score of 20 dB in normal-hearing young adults. A SPAR value of greater than 15 is considered to be consistent with normal hearing sensitivity.

ABR testing was carried out with conventional signal and recording parameters (Stach et al., 1993) on a Nicolet Spirit instrument. Gold-cup electrodes were affixed in a conventional manner, with electrode impedance maintained at levels below 3000 ohms. A vertex electrode

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**Figure 2** Results of immittance audiometry in Subject DF, an 11-month-old male. Uncrossed and crossed acoustic reflex thresholds were measured from the right (RE) and left (LE) ears to broad-band noise (BBN) and to pure tones at frequencies of 500, 1000 (1K), 2000 (2K), and 4000 (4K) Hz. The sensitivity prediction by the acoustic reflex (SPAR) is calculated from the pure-tone average (PTA) of reflex thresholds at 500, 1000, and 2000 Hz, minus the BBN threshold, plus a correction factor (Corr).
ABR to clicks at 95 dB nHL.

Figure 3A Auditory brainstem responses, with component peaks I and III, from the right and left ears of Subject DF, an 11-month-old male, to click stimuli presented at 95 dB nHL. Responses were recorded with reference electrodes that were ipsilateral (ipsi) and contralateral (contra) to the stimulated ear.

OAE testing was carried out with an IL092 Otoacoustic Analyzer. Transient-evoked OAEs were recorded in response to click stimuli of approximately 85 dB SPL. Responses were signal averaged and analyzed according to the conventions delineated by Kemp (Kemp et al., 1990). Distortion-product OAEs were recorded with the same apparatus. Briefly, two tones (F1 and F2) were presented simultaneously at an intensity level of 70 dB SPL and a frequency ratio (F2:F1) of 1.2. Amplitudes of the cubic distortion product were computed and plotted as a function of F2. Three complete frequency sweeps were made for each ear, and the results were averaged.

Audiologic Results

No consistent responses to any auditory stimuli could be identified at signal intensity limits by behavioral audiometry. It was unclear whether the lack of responsiveness was attributable to hearing impairment or to the pervasive neurodevelopmental delays.

Results of immittance audiometry, shown in Figure 2, were consistent with normal middle ear function, characterized by normal tympanograms, normal static immittance, and normal thresholds for crossed and uncrossed reflexes, bilaterally. SPAR values predicted normal hearing sensitivity in both ears.
Figure 4A Transient-evoked otoacoustic emissions from Subject DF, an 11-month-old male. Inset is the spectral analysis of the waveform (open) and the noise (shaded).

Diagnostic ABR results are shown in Figure 3A. Responses were recorded to click stimuli of alternating polarity, presented at an intensity level of 95 dB nHL and at a rate of 11.1/sec. Waveform morphology of responses recorded from both ears was abnormal. Although component waves I and III were identifiable and repeatable, wave V was not clearly definable in either ear. Neither changing stimulus polarity nor reducing stimulus rate served to provide a better definition of the later waves. Because a wave I response was present, we were reassured that DF did not have a profound hearing sensitivity loss. Nevertheless, lack of a repeatable wave V precluded a conventional ABR threshold search. Responses down to 55 dB HL are shown in Figure 3B. As anticipated, wave I diminished in amplitude as intensity was decreased.

Results of the OAE measurements suggested normal or near normal cochlear function. Transient-evoked OAEs are shown in Figure 4A. The right ear emission had an overall amplitude of 27.3 dB and a reproducibility value of 98 percent. The left ear emission was not as large, with an amplitude of 13.7 dB, and had a reproducibility value of 72 percent. In both ears, the presence of transient-evoked OAEs was consistent with good cochlear function and normal hearing sensitivity or, at most, no more than a mild hearing sensitivity loss. Amplitude-frequency functions of the distortion-product OAEs are shown in Figure 4B. These results suggest good cochlear function across the middle and high frequencies of the audiometric range. Their amplitudes also suggest normal hearing sensitivity or no more than a mild hearing sensitivity loss.

In summary, results of the audiologic evaluation showed normal middle ear function, no response to sound by behavioral measures, and abnormal auditory brainstem responses, bilat-
erally. Hearing sensitivity could not be predicted from the ABR, since component wave V could not be measured below 75 dB in the right ear and could not be measured in the left ear. One other conventional measure, the SPAR test, predicted normal hearing sensitivity. Results of OAE measurements corroborated the SPAR results, suggesting normal cochlear function.

Based on these results, our best estimation was that DF had normal hearing sensitivity and abnormal brainstem function. We concluded that hearing sensitivity was most likely adequate for aural language development and that intervention by conventional hearing aid amplification was not indicated. We also concluded that the abnormal brainstem function placed this child at risk for a communication disorder, and we recommended periodic monitoring of communication function.

COMMENT

This case illustrates an increasingly common audiologic challenge. An 11-month-old child, who had pervasive neurodevelopmental problems, was referred for a hearing consultation. He did not respond behaviorally to sound, and his auditory brainstem response was abnormal to a degree that made threshold prediction difficult. Although the SPAR predicted normal hearing, the lack of an independent cross-check resulted in uncertainty with regard to the audiologic diagnosis. The addition of OAE testing provided the necessary cross-check and permitted a much more confident diagnosis of normal hearing sensitivity.

Without the OAE results, decisions related to audiologic intervention would have been deferred until more definitive testing was completed. Thorough evaluation with behavioral and auditory evoked potential measures would have awaited maturation of neurologic function. However, because the OAE measures suggested normal hearing sensitivity and provided a cross-check to the acoustic-reflex results, questions related to audiologic intervention became clearer.

To be sure, we cannot conclude, on the basis of these results, that DF does not have a hearing impairment. All that we can conclude is that he probably does not have a substantial hearing sensitivity loss. In other words, although his cochlear function appears to be normal, the presence of an abnormal auditory brainstem response places this child at risk for hearing impairment. How much of a risk and for what type of hearing impairment, we really do not know. It may be that children who have abnormal ABRSs at this age are at risk for being among those who are diagnosed later in life as having auditory processing disorders. At this point in time, there is little basis for predicting the outcome of communication skill development in this type of child. We can conclude, however, that DF did not have a significant hearing sensitivity loss of cochlear origin and that conventional intervention with hearing aid amplification would not be appropriate. Since he is likely to be at risk for communication disorders, we were careful to recommend periodic communication assessment.

One could argue that the presence of ABR component wave I at an intensity level of 55 dB nHL is consistent with no greater than a mild hearing sensitivity loss in the higher frequencies of the audiometric range. In the case of DF, the combination of normal middle ear function, normal reflex thresholds, and a measurable wave I at 55 dB could, in fact, lead the intuitive audiologist to conclude that there is not likely to be a hearing loss of a degree sufficient to preclude learning language through hearing. Nevertheless, the careful audiologist is left with little clinical comfort in an audiologic diagnosis based on such limited information. In this case, the addition of the OAE data served the clinician well in providing a reassuring cross-check of the SPAR and ABR results. In addition, it provided an indication that hearing sensitivity was likely to be normal throughout the audiometric frequency range.

It seems important to emphasize that OAE results, like those of behavioral, immittance, and evoked-potential audiometry, should not be the sole basis for an audiologic diagnosis. That is, OAE results also require an independent cross-check. One need only understand that OAEs are compromised by middle ear disorders to realize that this newer technology will be most useful in audiologic assessment as part of a larger test battery.

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REFERENCES


