Auditory steady-state response audiometry in profound SNHL: The impact of abnormal middle ear function

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Abstract
Auditory steady-state response (ASSR) audiometry is a commercially available tool that is used to predict behavioral auditory threshold levels. Its particular value stems from the technology’s ability to measure frequency-specific responses in the background electroencephalogram to auditory stimuli presented across a broad range of frequencies and sound pressure levels. It is clearly of benefit when used to assess threshold levels in infants and children with severe-to-profound hearing impairment (i.e., cochlear implant candidates). Although numerous authors have provided evidence of the usefulness of ASSR testing, their reports have concerned patients whose middle ear impedance measures were normal. We report the cases of 2 patients who, following improvement of abnormal middle ear impedance values, experienced a marked improvement in measurable thresholds by ASSR testing.

Introduction
Auditory steady-state response (ASSR) audiometry is a new, commercially available clinical tool that measures far-field electrical potentials in response to amplitude/frequency-modulated, frequency-specific auditory stimuli. Predicted behavioral thresholds on ASSR audiometry and measured behavioral thresholds are highly correlated ($r > 0.95$). The ASSR-predicted thresholds also have been found to be highly predictive of tone-burst auditory brainstem response (ABR) thresholds. Furthermore, the accuracy of threshold prediction is directly proportional to the degree of hearing impairment. A further advantage of this new assessment tool is its ability to determine frequency-specific thresholds from 250 to 8,000 Hz at stimulus levels as high as 127.8 dB HL.

The effects of middle ear disorders on the latency of evoked potentials is well established. Jerger et al studied impedance measures in approximately 300 children and found that at least 30% of those with sensorineural hearing loss (SNHL) classified as severe or worse had abnormal middle ear findings. Furthermore, the proportion of children with abnormal impedance increased as the hearing deficit increased. With universal screening programs for hearing problems, the pediatric otolaryngologist is confronted with the possibility that a fluctuating hearing loss is beyond the typical output limits of click and tone-burst ABR audiometry and well outside the output limits of standard bone-conduction ABR audiometry. With the aforementioned objective measures, the assessment and management of fluctuating hearing loss in the patient with a severe-to-profound hearing impairment have been speculative at best.

We report the cases of 2 children with bilateral severe-to-profound SNHL that was confirmed on ASSR audiometry by abnormal tympanograms. Significant changes in the predicted auditory behavioral thresholds were found on repeat ASSR measures when middle ear function was normal. Both patients were being evaluated as part of a preoperative assessment of cochlear implantation. We discuss the implications of our observations.

Case reports
To obtain impedance measurements in both patients, we used a GSI 33 immittance bridge and a 226-Hz probe tone for tympanometry. The ASSR in both cases was measured by the GSI Audera system, which determines the phase coherence between the phase of the modulating frequency of an amplitude/frequency-modulated stimulus and the phase of the background electroencephalogram (EEG) at the same modulation frequency. The probability that the
Patient 1. A 2-year-old girl presented to the Hearing Center at Texas Children’s Hospital in January 2003 for an initial evaluation of hearing loss. Behavioral audiometry, performed with visual reinforcement audiometry and insert earphones, suggested a profound sensitivity loss at 4,000 Hz in the right ear and a severe-to-profound sloping SNHL in the left ear. With impedance audiometry, a type B tympanogram was obtained from the right ear and a type A tympanogram from the left (figure 1, A). No ASSRs were measured in the right ear except at 8,000 Hz. These findings were consistent with the limited behavioral responses observed in the right ear. ASSRs in the left ear were consistent with the measured behavioral responses in the left ear.

The patient was seen for follow-up impedance audiometry and ASSR measurements 6 months later. This time, testing yielded type A tympanograms bilaterally (figure 1, B). In addition, ASSR threshold levels were present in both ears (figure 1, C).

Patient 2. A 2-year-old boy was brought to the Hearing Center at Texas Children’s Hospital in September 2002 to be evaluated for possible cochlear implantation. Initial impedance audiometry revealed a type C tympanogram from the right ear and a type B tympanogram from the left (figure 2, A). ASSR testing in the right ear found no predicted behavioral responses at equipment limits. ASSR testing in the left ear found predicted behavioral responses at only 1,000 and 4,000 Hz.

At follow-up 10 months later, ASSR audiometry yielded type A tympanograms bilaterally (figure 2, B). ASSR responses were measurable at 500 and 2,000 Hz in the right ear (figure 2, C). ASSR responses in the left ear were noted at approximately 90 dB between the frequencies of 250 and 2,000 Hz.

Discussion
A key issue in the management of hearing loss in children who are identified at birth with severe-to-profound SNHL is the determination of appropriate target gain levels for amplification, which is based on auditory thresholds. Prior to the availability of ASSR audiometry,
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the setting of target gains in the “no response by ABR” child was problematic at best, and gain targets were only educated guesses. With ASSR, predicted behavioral threshold levels in the profound range can be established.

The 2 cases described here illustrate the importance that middle ear function has on ASSR testing:

- After patient 1’s tympanogram had changed from type B to type A on the right, she displayed measurable threshold responses from 250 to 4,000 Hz. Previous ASSR testing had yielded a measurable response only at 8,000 Hz.
- Patient 2 had initially displayed near-equipment-limit thresholds at 1,000 and 4,000 Hz on the left. After a change in middle ear compliance from type B to type A tympanograms, he went on to display near-90-dB thresholds from 250 to 2,000 Hz in the left ear.

These improvements in threshold are clinically significant and would have an impact on target gain levels; they may even affect the clinical decision to pursue amplification rather than cochlear implantation.

It is not surprising that middle ear dysfunction appears to affect the ASSR threshold response. In a recent analysis of children with moderate-to-severe hearing impairment, Cone-Wesson et al demonstrated attenuation of the ASSR response in relation to abnormal middle ear impedance levels. Their patients, in contrast to our 2 patients, had lesser degrees of hearing loss, and bone thresholds were therefore established. Although the ability to obtain a “bone line” with a bone-conducted to amplitude/frequency-modulated, steady-state signal is technically feasible, the technique is not typically seen in standard pediatric audiologic practice. The level of hearing impairment in our 2 patients, however, may preclude the use of even the modified sensorineural acuity level procedure with auditory steady-state audiometry.

In the patient population with severe-to-profound hearing loss, assessment of hearing thresholds by ASSR testing may lead to accurate predictions in an ear with normal middle ear function, but a word of caution needs to be sounded with regard to the assessment of a patient with abnormal middle ear compliance. The determination of hearing threshold by ASSR testing in the presence of abnormal middle ear findings may lead to elevated predicted

Figure 2. Patient 2. A: At the initial evaluation, tympanograms (226-Hz probe tone) show the type C result from the right ear with negative pressure (–180 daPa) and a type B result from the left ear. B: At the 10-month follow-up tympanometry, type A results are seen from both ears. C: Initial ASSR measurements (open circles) reveal no predicted auditory behavioral thresholds at equipment limits in the right ear and responses at equipment limits only at 1,000 Hz and 4,000 Hz in the left. At follow-up (shaded circles) predicted behavioral thresholds in the left ear are significantly improved to approximately 90 dB HL with normal middle ear function.
Figure 3. Illustration shows our proposed algorithm for otologic evaluation of infants and young children who are possible candidates for cochlear implantation.
behavioral thresholds, which could lead to inappropriate levels of prescriptive gain for the patient when the middle ear disorder resolves. Furthermore, the possible elevation of behavioral thresholds from the effects of abnormal middle ear findings could significantly affect the clinical management of the younger pediatric patient who is under consideration for a cochlear implant.

We wish to issue two caveats regarding our findings:

• First, although research to date has clearly shown that ASSR audiometry is a reliable predictor of behavioral thresholds, especially in patients with severe-to-profound hearing loss, test-retest statistics compiled over extended periods of time have not been reported. Therefore, we cannot exclude the possibility that ASSR measurements will vary over an extended period of time.

• Second, in both of our patients, no independent otologic evaluation of the ear was conducted to confirm the objective findings of abnormal middle ear function or normal middle ear findings on retest.

Our preliminary findings regarding the effect that an assumed conductive component (as measured by ASSR testing and impedance audiometry) has on a profoundly hearing-impaired child suggest to us the importance of the management of middle ear disorders in the infant or child with a severe-to-profound hearing loss who is a possible candidate for cochlear implantation. We take this opportunity to propose an algorithm for the otologic evaluation of the pre cochlear implant infant/child (figure 3). Our emphasis in devising this protocol is to attempt to stabilize auditory thresholds to maximize the effectiveness of the amplification trial period.

References