Recent developments in physiologic measurements of hearing in humans have allowed for diagnosis and description of a condition that Sininger and colleagues (1995) called auditory neuropathy. With this condition, patients present with complaints of hearing problems. They tend to have a difficult time understanding what is being said, even in quiet environments. Their audiograms show decreased hearing sensitivity and poorer word recognition ability than expected based on pure-tone thresholds. Both contralateral and ipsilateral middle ear muscle reflexes are absent. Auditory evoked responses are absent or are grossly abnormal. The otoacoustic emissions (OAEs), however, are present, and testing OAEs with ipsilateral, contralateral, or bilateral masking does not suppress the emission in auditory neuropathy patients as it will with patients having normal hearing (Berlin et al. 1994, 1993a, 1993b).

The patients that have been reported tend to vary in demonstration of other peripheral neuropathies. Some show no signs of any sensory or motor neuropathies other than the auditory findings, while others have very obvious involvement. There are some well-described processes with other neurologic involvement in addition to the auditory findings, such as Friedrich’s ataxia, Charcot-Marie-Tooth syndrome, and cerebellar ataxia. Patients also tend to vary in their demonstration of hearing loss. Patients have been reported with anywhere from mild-to-profound hearing loss and may have bilateral or unilateral findings (Konradsson 1996; Starr et al. 1996).

The site of the abnormality is still unknown. Some theories are that it is within the inner hair cell structure; that it is a synaptic disorder between the inner hair cells and the 8th nerve; that it is within the 8th nerve itself. Some have thought it could be a timing disorder, a dysynchrony in the neural response.

We have seen several patients at Mayo Clinic, but the cases being presented involve the first three children identified.

Assessment Findings

In June of 1995, child A was brought to the audiology section at Mayo Clinic because her parents were concerned about her speech and language development. She had just been to a 15-month well child check-up, and the physician said that if she was not talking by 18 months, then a hearing test should be done. The parents went home and did their own hearing test, and child A was in the office the next morning for evaluation.

Her parents reported a normal birth history with no complications during pregnancy, labor, or delivery. She was mildly jaundiced at birth, but required no treatment for this. There was no reported head trauma or intracranial infection. Her parents reported no physical or health concerns, and both parents reported negative family histories for hearing loss. At 15 months, all other developmental milestones were within normal limits.

The audiogram for child A may be seen in figure 1. The S represents sound field responses to warbled tones or narrow band noise using visual reinforcement audiometry. Her responses to speech, air, and bone conduction were consistent with profound hearing loss. Her tympanograms showed normal middle ear pressure and compliance, and her acoustic reflexes were absent.

Click-evoked otoacoustic emissions were assessed because they were relatively new in our clinic at that time and almost all patients were tested. The results were normal (figures 2 and 3), suggesting normal outer hair cell function for both ears. Child A was only scheduled for audiometric testing that morning, but in light of the results that were elicited, it was deemed important to obtain an
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Figure 1. Initial audiogram for child A.

Figure 2. Right ear click-evoked otoacoustic emissions for child A.

Figure 3. Left ear click-evoked otoacoustic emissions for child A.
ABR. Our sedation protocol requires evaluation by a physician and administration, and monitoring of medication by a nurse in our ENT department.

Her ENT exam that morning was negative, chloral hydrate was administered, and ABR testing was completed (figure 4). Auditory evoked responses were abnormal for both ears. This response was obtained for both ears that morning. They are abnormal, but in the first few milliseconds there is a repeatable pattern for both ears. Child A was seen about one month following our evaluation at Boys Town National Research Hospital (BTNRH) in Omaha, Nebraska. When they performed the ABR, they reversed the polarity of the clicks from rarefaction to condensation. This ABR (figure 5) was done at BTNRH. It shows rarefaction and condensation clicks at 80 dB nHL for the right ear. There are no waveforms present, but in the first few msec there is a repeatable pattern for both ears. The same repeatable pattern that reverses when the polarity of the click is reversed is present. Hood (1996) tell us that this comparison of evoked potentials can be used to differentiate the cochlear microphonic from the neural response representing the ABR. The response you see is not a neural response, but represents the cochlear microphonic. We know that the OAE measures preneural cochlear function, and that ABR assesses neural status. These tests are not hearing tests, but we can use the information to draw conclusions about the ability to stimulate the auditory system with auditory signals.

Child A's medical evaluations have yielded negative findings for ENT, neurology, and ophthalmology. Medical genetics findings were negative for any syndromes, and imaging studies were negative for any abnormalities. Her brother, child B, was 2 months old when child A was ini-

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Figure 4. Initial ABR for child A.

Figure 5. ABR for child A performed at BTNRH.

Figure 6. Right ear click-evoked otoacoustic emissions for child B.
tially evaluated. The parents were encouraged to have child B’s hearing evaluated as well, in light of his sister’s findings. At 4 months of age child B was evaluated. His parents reported a normal birth history, with no pregnancy or delivery complications. There were no concerns about his development at this age.

At 4 months he did not respond consistently to any tones, noisemakers, or speech stimuli presented. Immittance testing showed normal middle ear pressure and compliance for both ears with absent acoustic reflexes. His otoacoustic emissions (figures 6 and 7) were normal for both ears. His ENT evaluation was negative, chloral hydrate was administered, and ABR testing was completed. His responses can be seen in figures 8 and 9, and were found to be similar to his sister’s. The same repeatable pattern is present in the first few milliseconds, and it reverses as the polarity of the click reverses, representing the cochlear microphonic and not a neural response.

Child B returned when he was 6 months old to obtain a behavioral audiogram (figure 10). VRA was used in sound field, and results were similar to his sister’s: profound hearing loss for speech and tones. No bone-conduction response could be obtained; child B would not tolerate it. All his medical evaluations were negative. No imaging studies were obtained at the time of his early visits.

Although the majority of patients reported have bilateral neuropathy, the first pediatric case identified at Mayo Clinic was actually a unilateral neuropathy. Child C was seen in 1994 at 3 years old. His medical history was negative. His pregnancy and delivery were normal, and all his
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Figure 10. Initial audiogram for child B.

Figure 11. Initial audiogram for child C.

Figure 12. Left ear click-evoked otoacoustic emissions for child C.
developmental milestones were within normal limits. He had had no head trauma or intracranial infection. His parents were suspicious about a unilateral hearing loss because of child C's behavior. For example, child C would turn his head in favor of his right ear and would change the telephone from the left to right ear when talking. Child C had no other abnormalities. His family history was positive for a paternal grandmother with unilateral hearing loss of unknown etiology.

His audiogram (figure 11) showed thresholds for his right ear within normal limits, but his left ear (unmasked) was in the severe hearing loss range. His speech reception thresholds were in agreement, and word recognition scores could not be obtained for his left ear. Tympanometry was normal for both ears, and acoustic reflexes were absent when the stimulus was presented to the left ear.

Otoacoustic emissions were relatively newly available for clinical use in 1994, and it was not uncommon for us to try to test anyone who would sit still. OAEs were normal for both ears. Figure 12 shows his left ear results. His first ABR was done for diagnostic purposes, and four-
channel recordings were done. Figure 13 shows normal tracings for the right ear, but abnormal tracings for the left ear.

The most current results, figures 14 through 17, show that the audiogram remains unchanged, the left ear OAEs are present, and the ABR is still abnormal for the left ear. The results of the 1997 ABR testing (figure 17) show the cochlear microphonic reverse as the click polarity reverses. All child C’s medical evaluations have been negative, and his imaging studies were negative for any abnormalities.

Management

Because child C’s speech and language, motor skills, and all areas of development were normal, we worked with the school district to help with in-servicing and educating teachers and assistants in the classroom about unilateral hearing loss. When he was in preschool, a sound field FM system was placed in the room. He was seated near the speaker in the classroom. He was managed the same as any child with a unilateral hearing loss. He is very bright and has always done well in the four years we have followed him.

Management strategies for children A and B have been a little more complicated. The first areas discussed...
and initiated were those of communication and amplification. The family was referred to the local school district the day child A was identified, and the school district began services within a few days after identification. The family chose cued speech and quickly became “fluent” and were utilizing cues within a month of identification. Approximately one week after identification, child A was fitted with a Tactaid II vibrotactile instrument. She used the device several times during the day while doing activities (such as presence vs. absence). The goal was to teach child A to make the connection between vibration of the oscillators and sound.

Child B was fitted with the Tactaid II two months later, and child A was fitted with a Tactaid VII. Child B was simply too small to wear a Tactaid VII device. The children were fitted with Resound BT2 hearing aids with 25 to 30 dB of gain and 95 to 100 dB output. The trial went for about six weeks with no benefit observed. A Sonovation LRR 200 LD was also used for an extended trial with no observed benefit.

The public schools were very involved from the start with the siblings. Team members included the audiologist, teacher of persons who are deaf and hard of hearing, speech-language pathologist, and early childhood special education. They were involved in center-based programs and one-on-one language programs, and the parents became actively involved in the teaching process.

At the end of the first school year the parents re-evaluated the progress of the children. Though the parents were pleased with the children’s receptive language skills (which were age appropriate), they were not pleased with the expressive language skills, in which the children were grossly behind. The parents decided to consider some alternative programs.

They enrolled their children in the Moog Oral School in St. Louis in 1997. While they were there, they underwent a trial with power hearing aids under careful OAE monitoring. For example, the audiologist used a power hearing aid on one ear while child A was involved in intense one-on-one therapy sessions, but changed to the mild gain aid when in group sessions. Otoacoustic emissions were monitored daily, and it was found that power amplification could be utilized daily without destroying the OAE response.

Her aided sound field thresholds were present at 50 to 60 dB through 1500 Hz. In addition, she received a score of 29% on the pattern perception (7/24) portion of the Early Speech Perception test administered at the Moog School.

At the end of the school year the parents were starting to pursue the possibility of a cochlear implant. In August 1998 child A had surgery to implant a CI 24. Her postoperative testing (Neural Response Telemetry) was positive, her hookup went well, she is responding well, but it is a bit early to make specific comments about her performance at this stage. Child B has also received a cochlear implant.

Summary

In our experience with a very small sample, we have found that our patients with profound hearing loss having auditory neuropathy (only auditory findings) did not receive any measurable benefit from mild gain hearing aids, but did receive minimal benefit from power hearing aids.
aids. At present, we recommend that patients with auditory neuropathy (only auditory findings) who have mild, moderate, or severe hearing loss should be fit appropriately with amplification. They should be closely monitored for any tolerance problems at the office. Further, monitoring with OAE is important to make sure the response remains.

In addition, we have found that patients with only auditory findings and no other peripheral neuropathies are able to be stimulated with CI 24. While the site of lesion remains unknown, but based on the fact that both child A and B are able to be stimulated with an implant, it seems more likely that these patients with only auditory findings have a problem within the inner hair cell structure or perhaps with the inner hair cell/8th nerve junction.

References


