Auditory Neuropathy: Case Study with Hyperbilirubinemia

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Abstract

Auditory neuropathy (AN) has been described in the literature as presenting with a combination of audiometric findings that include elevated behavioral audiometric thresholds, auditory brainstem response findings that are not consistent with audiometric findings, poor speech recognition, and present otoacoustic emissions (OAEs) and/or cochlear microphonics. Since the availability of clinical OAE testing, AN has come to be identified with increasing frequency; however, incidence and prevalence figures are unavailable. There is a great deal of discussion about the accurate diagnosis of AN, its characteristics, and its treatment. Some of this discussion is occurring on the Internet and over the telephones. The need to continue to provide information in accessible peer-reviewed journals is paramount. Following a review of the literature, a case study is presented of a boy who was diagnosed with AN as a newborn. He experienced hyperbilirubinemia and other neonatal health complications. His educational intervention was managed elsewhere until recently. Information is presented about the progression of the case over a 5-year period that includes audiologic data and communication development results.

Key Words: Auditory brainstem response, auditory neuropathy, hearing disorder, hyperbilirubinemia, otoacoustic emission

Abbreviations: AN = auditory neuropathy, CI = cochlear implant, CM = cochlear microphonic, DPOAE = distortion product otoacoustic emissions, DSL = Desired Sensation Level, IHC = inner hair cell, MRI = magnetic resonance imaging, OAE = otoacoustic emission, OHC = outer hair cell, SS = standard scores, TC = total communication

With the advent of practical and reliable methods for measuring otoacoustic emissions (OAEs), audiology and hearing science professionals now have a tool that allows them to assess outer hair cell (OHC) function. This has led to the identification of a distinctive type of hearing deficit that is labeled auditory neuropathy (AN) (Sininger et al, 1995). The features of AN are detailed below. It has been suggested, however, that a more accurate name might be auditory neuropathies to reflect the fact that multiple etiologies appear to be present (Hood, 1998). Despite the fact that research into this hearing disorder is relatively recent, this is not a new disorder. There are past reports in the literature regarding cases now thought to have involved AN (Davis and Hirsh, 1979; Worthington and Peters, 1980). These cases were described at the time as paradoxical, and specific diagnoses were not offered because test findings were inconclusive or contradictory. Advances in the measurement of OAEs now allow us to further characterize and study this condition and its variations.

Although the population of patients with presumed AN is heterogeneous, they consistently exhibit a constellation of findings that suggests that OHC function is normal and that inner hair cell (IHC) and/or VIII nerve function is impaired (Starr et al, 1996; Berlin et al, 1999). In general, these patients have elevated auditory thresholds with a variety of audiometric configurations; however, Kraus et al (1984) reported individuals with AN who had normal hearing sensitivity. Most of the patients described in the literature have mild-to-moderate degrees of peripheral hearing sensitivity loss (Sininger...
et al, 1995; Starr et al, 1996). However, there are also reported cases with severe-to-profound hearing losses (Lutman et al, 1988; Prieve et al, 1991; Widen et al, 1995; Laccourreye et al, 1996; Rance et al, 1999). Hearing losses with rising audiometric configurations have been observed (Sinninger and Starr, 1997; Hood, 1998; Rance et al, 1999) and flat, sloping, and unusual configurations have also been noted. Elevated thresholds are usually observed in both ears, but there may be asymmetric findings, and there have been cases of unilateral findings reported as well (Jerger et al, 1992; Konradsson, 1996; Hood, 1998). In a number of cases, the hearing loss seems to be relatively stable (Rance et al, 1999; Sininger, personal communication, 1999), but it may fluctuate (Gorga et al, 1995; Starr et al, 1998; Rance et al, 1999) or progress (Widen et al, 1995; Starr et al, 1996; Hood, 1998). These variations lend weight to the hypothesis that different etiologies and sites of lesion may be responsible for AN (Hood, 1998).

Another audiometric feature observed in cases of AN is absent or abnormal auditory brainstem responses (ABRs). This occurs in spite of the fact that behavioral thresholds are in a range that typically yields ABRs. Also, some patients have reported that they could hear the stimuli during testing even when the ABR could not be measured (Starr et al, 1991, 1996; Sininger and Starr, 1997). Because the presence of an ABR depends on the ability of auditory neurons to respond synchronously to an input signal, it has been postulated that these abnormal findings occur due to a lack of synchrony in the afferent portion of the auditory nerve (Starr et al, 1991). It is this lack of correspondence between the ABRs and behavioral thresholds that first led to AN cases being described as paradoxical.

Another finding in AN patients that supports the idea of inaccurate timing of the neural response is the number of hearing losses with rising configurations. Starr et al (1996) suggest that this may relate to the fact that time-locking of the auditory nerve discharges plays a greater role in the encoding of signals below 2000 Hz than it does for higher frequencies.

In some patients, middle and long latency evoked potentials are also abnormal, but this does not occur in all patients with AN (Starr et al, 1991; Gorga et al, 1995; Sininger et al, 1995; Widen et al, 1995). The presence of these potentials in some cases may be due to the fact that they are not as dependent on neural synchrony as the ABR.

Absence of acoustic stapedial reflexes is another reported finding in patients with AN. This has been observed even when audiometric thresholds are normal or near normal (Gorga et al, 1995; Starr et al, 1998). It has been postulated that this may be related to the same dysynchrony affecting the ABR (Berlin et al, 1993).

Patients diagnosed with AN tend to have word recognition abilities that are disproportionately poorer than would be predicted by audiometric thresholds (Yellin et al, 1989; Sininger et al, 1995; Starr et al, 1996; Doyle et al, 1998; Rance et al, 1999). Children with the disorder generally have speech-language problems and poor performance in school. This finding may also be related to the altered neural synchrony believed to be responsible for the abnormal ABR results in these patients. This hypothesis is supported by reports that individuals with AN have been shown to exhibit impaired perceptions on temporally based psychoacoustic tasks such as gap detection, lateralization, thresholds for short-duration tones, and masking level differences (Starr et al, 1991; Sininger et al, 1995; Sininger, 1998; Zeng et al, 1999). Speech perception ability depends on accurate encoding of timing information in the rapidly changing speech signal; however, based on psychoacoustic findings in some patients with AN, it would appear that the auditory system is not able to accomplish this task. Disruption of synchronous activity, however, may not be the only explanation for the observed poor speech perception. Rance et al (1999) highlight the fact that speech perception abilities can be quite good in cochlear implant (CI) recipients. These individuals, however, exhibit poor discrimination of periodicity, and their implants use coding schemes that do not mimic normal cochlear temporal patterns. Rance et al (1999) conclude that this may point to a distortion of tonotopicity of neural signals reaching higher centers of the auditory system in patients with AN.

The characteristic that most clearly distinguishes the patient with AN is the presence of OAEs and/or a cochlear microphonic (CM) despite abnormal ABRs. Both the OAEs and CM are associated with normal cochlear OHC function. Although measurement of the CM has been possible for some time (Davis, 1976), it is reportedly difficult to record in human subjects and is susceptible to artifact contamination (Starr et al, 1996). Consequently, it is the development of OAE testing that has facilitated the diagnosis and investigation of cases of AN. In
addition to being present, CMs and OAEs measured in patients with AN may appear exaggerated or more robust than those seen in individuals with normal hearing sensitivity (Berlin et al, 1993; Siningger, 1998; Starr et al 1998). Further, OAEs cannot be suppressed via contralateral presentation of noise, suggesting an interruption in the afferent-to- efferent loop (Berlin et al, 1993). It has also been reported that some patients with AN have shown OAEs that progressively diminish in amplitude over time (Siningger, 1998; Deltenre et al, 1999). The reason for this is unclear, especially in light of the fact that the CM may still be measurable. Starr et al (1996) hypothesize that it may be a consequence of the neural disorder manifested in the hair cells through a lack of stimulating factors from the auditory nerve. Others suggest that this finding might be due to a scattered loss or disruption of OHCs sufficient to cause disappearance of the OAE—which has been shown to be sensitive to even minor cochlear insult—but not of the CM (Deltenre et al, 1999; Rance et al, 1999).

In some individuals with AN, other peripheral neuropathies may be present. Starr et al (1996) reported this finding in 8 of 10 of the patients identified in their study. Some of the patients have been diagnosed with hereditary motor-sensory neuropathy (Charcot-Marie-Tooth type I), Friedreich's ataxia, or Guillain-Barré syndrome (Starr et al, 1996; Siningger and Starr, 1997). Other patients show sporadic neuropathies of unknown type and origin. Cevette et al (1995) reported two cases of sudden unilateral hearing loss and present OAEs associated with multiple sclerosis. In almost all of the cases reported above, however, hearing loss seems to have been the initial complaint with other symptoms of peripheral neuropathy surfacing later.

The prevalence and incidence of AN are unknown. Davis and Hirsch (1979) estimated that AN is responsible for 1 in 200 cases of hearing impairment (0.5%). Kraus et al (1984) reported finding 7 instances of AN among 48 cases examined (15%). Rance et al (1999) found an incidence of 1 in 9 (11%) in their population of children with permanent hearing loss. Berlin (personal communication, 1999) has estimated that AN is present in at least 4 percent of children with hearing loss.

Although this condition has been labeled as a neuropathy, it is not certain that the VIII nerve is the locus of the disorder or if it is the only site of lesion in patients with AN. Some researchers have suggested that AN may be associated with loss or lack of myelin resulting in pathologic changes in neural conduction properties and could therefore be localized to the type I afferent fibers of the auditory nerve (Kraus et al, 1984; Starr et al, 1996, 1998). This hypothesis is based in part on the fact that a number of patients with AN have also been diagnosed with peripheral neuropathies. Siningger and Starr (1997) reported that a sural nerve biopsy from one of their patients with AN showed axonal neuropathy and myelin sheath thinning, and they assumed that the auditory nerve possessed a similar pathology. There are two reports in the literature of patients meeting the profile of AN who suffer transient hearing loss and impaired speech perception ability when fever is present (Gorga et al, 1995; Starr et al, 1998). Starr et al (1999) suggest that this results from a myelinating disorder of the auditory nerve.

The locus of abnormality has also been hypothesized as being at the synapse between the IHC and the auditory nerve dendrites, the nerve dendrites themselves, the spiral ganglion, or the auditory nerve axons or any combination of these sites (Siningger et al, 1995; Starr et al, 1996). These are considered to be likely loci due to the OAE and CM findings in combination with abnormal temporally dependent auditory perceptions and the absence of neural components of the ABR. A disconnected or abnormal tectorial membrane has been suggested as a possibility (Berlin et al, 1998). Absence or pathology of the IHCs themselves has been postulated as a primary etiology (Konradsson, 1996; Berlin et al, 1998; Harrison, 1998). Harrison (1998) arrived at this conclusion based on the fact that OAE/ABR characteristics paralleling AN were induced in chinchillas with carboplatin, an anticancer agent known to produce selective IHC lesions. However, Rance et al (1999) state that an IHC lesion by itself cannot fully account for the inconsistency between audiometric and ABR findings. Finally, findings revealing problems in the efferent portion of the auditory nerve as well as the afferent portion have prompted the theory that the root of the problem may involve neurotransmitters (Berlin et al, 1993). Unfortunately, however, the site(s) of lesion remains undetermined because there are no procedures currently available that allow the status of IHC or the synapse between IHCs and VIII nerve fibers to be assessed in vivo.

The variability of audiometric findings, as well as the possibility of multiple sites of lesion, suggests that AN may result from a number of different etiologies. As mentioned above, in many
cases, AN has been linked with the occurrence of peripheral neuropathy (Cevette et al, 1995; Starr et al, 1996, 1998; Sininger and Starr, 1997). A number of reports associate AN with hyperbilirubinemia (Kraus et al, 1984; Katona et al, 1993; Deltenre et al, 1997; Berlin et al, 1998; Gupta and Mann, 1998; Rance et al, 1999). Excessive amounts of bilirubin have been shown to cause insult to both the peripheral and central nervous systems, including the cochlear nucleus (Chisin et al, 1979; Kraus et al, 1984; Vohr et al, 1989). It is of note, however, that, in some cases, these effects seem to have been temporary and improvements in ABR responses have been noted over time (Perlman et al, 1983; Gupta and Mann, 1998; Rance et al, 1999). Sustained cochlear hypoxia is another proposed factor in AN (Deltenre et al, 1997; Harrison, 1998; Rance et al, 1999). Two studies have described patients with AN apparently resulting from mitochondrial disorders (Deltenre et al, 1997; Corley and Crabbe, 1999). Prieve et al (1991) reported a patient with present OAEs and a severe hearing loss following childhood measles and/or mumps. Siblings with AN have been identified, raising the possibility of genetic factors (Robinette and Durrant, 1997; Starr et al, 1998). Other proposed causes of the pathophysiology seen in cases of AN include prematurity (Rance et al, 1999), uremia, diabetes, and platinum compounds (i.e., cisplatin) toxicity (Starr et al, 1996).

The literature reflects differing opinions regarding remediation and management of AN. It has been suggested that intervention typically used for sensory hearing loss, such as hearing aids, is generally not beneficial because the primary problem relates to the synchrony of the auditory nerve (Sininger et al, 1995; Sininger and Starr, 1997; Berlin et al, 1998). In addition, possible adverse effects from amplification have been considered given the apparent preservation of OHC function (Starr et al, 1996). However, there have been reported cases where hearing aid use has been of significant benefit to the patient. Rance et al (1999) report that of the 15 children in their study who had used amplification, 8 showed significant benefit on speech perception testing or in general auditory responsiveness. They also found, however, that success of amplification could not be predicted by the results of any of the assessments in their test battery. Deltenre et al (1999) present a patient with AN whose unaided and aided word recognition scores demonstrate significant benefit from amplification. The authors hypothesize that the timing of the initiation of amplification with respect to the critical period for language development may be a major influence on success or failure.

In addition to hearing aids, the question of the appropriateness of cochlear implantation in cases of AN has been raised. Rance et al (1999) present a case of a child with AN who received a CI. Relatively poor results were obtained, and after a year of device use, auditory-only speech perception scores remained at or near chance levels. Alternatively, Sininger et al (1999) report a child with AN who received a CI and has shown progress in speech perception abilities. It has been suggested that successful use of a CI may be dependent on the site of lesion in a given case. If the lesion is located medial to the IHCs in the auditory nerve itself, it would seem that the transmissions from the CI would be subjected to the same conduction block experienced by the cochlear mechanisms. If the locus of the disorder is restricted to the IHCs or the synapse with the afferent nerve fibers, a greater chance of success with the CI might be realized (Hood, 1998; Rance et al, 1999).

Manual communication training has been suggested as an important part of remediation in cases of AN, particularly with young children (Sininger et al, 1995; Sininger and Starr, 1997; Berlin et al, 1998). This is the case not just when severe-to-profound hearing loss is present but also when part or all of the speech spectrum might be assumed to be audible to the individual. This is because it is not known whether the auditory system is capable of obtaining usable information from that speech signal. Berlin et al (1998) reported that all of the 19 cases of children with AN that they examined showed limited development of auditory abilities and language delays when they were allowed only auditory access to verbal information with no lip reading or signing support. Some authors have specifically recommended a visual communication system that follows the grammatical structure of English (i.e., signed English or cued speech) for children with AN (Berlin et al, 1998; Hood, 1998). The rationale for this is based partly on the possibility that the ability to use auditory information may improve. If so, it is suggested that the child might more easily assimilate spoken language into a language system that already has English structure. In addition, these systems may be easier to learn than American Sign Language for some parents who are English speakers and allow earlier instillation
Figure 1 Auditory brainstem response waveforms obtained at 2½ months of age. Click stimuli were delivered at a rate of 17/sec via insert earphones. Left ear responses are shown on the left side of the figure. The upper two tracings were in response to 90 dB HL rarefaction clicks, the next two in response to 90 dB HL condensation clicks, and the lower two tracings represent the sum (R + C).

Figure 2 Results of distortion product otoacoustic emission (DPOAE) testing at 2½ months (upper panel) and at 4 years of age (bottom panel). The upper hatched area represents amplitude levels from the 90th to the 95th percentile in the distribution of impaired ears. Values plotted in this area and above are considered to reflect normal outer hair cell (OHC) function. The lower hatched area represents the 5th to the 10th percentile in the distribution of normal ears. Values plotted in this area and below are considered to reflect impaired OHC function. The shaded area shows the overlap between the two distributions where peripheral auditory function is uncertain (Gorga et al, 1997). Amplitude levels for left ear (x) and right ear (O) responses at signal-to-noise ratios ≥ 6 dB are plotted. In the upper panel, DPOAEs are in the normal range for the entire frequency range tested at the time. In the lower panel, emissions were not present below 6000 Hz for either ear.

no ABR components. OAEs were present for both ears (Gorga et al, 1997) and are shown in Figure 2. Given these findings suggesting AN, a referral was made to pediatric neurology for evaluation and to the schools for early intervention services. The child's family reported that he seemed to respond to sounds at home. There were no concerns for his development.

CASE REPORT

The boy who is the focus of this report is now 5 years old and presents with findings that are consistent with AN. He was first seen for hearing screening when in the neonatal intensive care unit just prior to hospital discharge at 5 weeks of age. ABRs to click stimuli were abnormal bilaterally, with no identifiable wave V up to 70 dB HL; however, distortion product OAEs and transient evoked OAEs were present.

Birth history includes delivery at 35 weeks gestational age, with a birth weight of 6 pounds, 8 ounces. The infant experienced respiratory arrest at birth. He was diagnosed with persistent pulmonary hypertension of the newborn and hyaline membrane disease. Prolonged mechanical ventilation (22 days) was necessary. His neonatal course was further significant for hyperbilirubinemia requiring three exchange transfusions and phototherapy. Also, he received medications that included gentamicin, vancomycin, and furosemide. A cranial ultrasound was normal when he was 1 month old.

At 2½ months of age, he was seen for follow-up testing with ABRs and OAEs. The findings were similar to those obtained in the nursery. As shown in Figure 1, the ABR waveforms for click stimuli presented at 90 dB nHL show CM but of language into the children by the parents. Regardless of the particular form, however, some type of visual communication training appears important to maximize use of available auditory information, and relying on auditory input alone does not generally appear to be productive.
A multidisciplinary team evaluation at 6 months resulted in "no verified disability." Behavioral audiometric testing initially was completed when he was 6 months of age using visual reinforcement audiometry (VRA). Test reliability was limited, and thresholds were in the mild-to-moderate hearing loss range at 500 and 1000 Hz. At this point, the family felt that he was responding to sound well.

By the time he was 1 year old, he had experienced approximately five episodes of otitis media with effusion and was being followed elsewhere by an otologist. It was difficult to obtain reliable audiometric data, and it was suspected that the otitis media with effusion was a contributing factor, although it was recognized that AN played a role as well. Repeat ABR and OAE testing during this time yielded the same results as those initially obtained. Behavioral audiometric threshold measurements remained unreliable until he was 14 months old, at which time his pure-tone audiometric thresholds were in the mild-to-profound hearing loss range, as displayed in Figure 3. At that test, the distortion product otoacoustic emissions (DPOAEs) were measured and were present only at 6000 and 8000 Hz for the right ear. The left ear presented with middle-ear dysfunction, and OAEs could not be assessed reliably. His mother reported that he responded consistently to sounds at home unless they were high-frequency sounds. Also, she stated that he was saying some single words (e.g., daddy, hello) and that he understood spoken words. During this time, the pediatrician reported that the child was beginning to produce some word approximations and was exhibiting mild gross motor delays.

Although a trial period with amplification was considered, the complication of otitis media with effusion and AN findings precluded amplification use. At 1½ years of age, bilateral myringotomies with placement of tymanostomy tubes were completed elsewhere. He was enrolled in educational services for speech and language delays with verification of hearing as the primary impairment and speech/language impairment as the secondary impairment. Test data from that time were not available to us.

Tymanostomy tubes were replaced at 3½ years, in conjunction with an adenoidectomy and tonsillectomy, once again elsewhere. His mother reported some improvements in his speech following this surgery, but concerns were increasing for speech and language delays. Magnetic resonance imaging (MRI) of the neck and soft tissues was normal when he was 3½ years old. MRI and computed tomography of the temporal bones were done at 4 years of age and also were reported as normal.

Because he made limited progress in language, his general special education preschool introduced a few signs to him when he was 4 years old. Although it was the school's impression that signs helped him, consistent sign models were not available at school or home. Consequently, he rarely used signs spontaneously.

At this point, his mother switched to this center for primary provision of audiologic services. Because of her concerns for her son's limited intelligible speech and her desire for him to communicate orally, discussions about amplification were reinitiated.

Numerous audiologic evaluations were completed during the next few months in an attempt to verify pure-tone audiometric thresholds. However, he was active and inattentive during testing, and in some test sessions only one or two thresholds could be established before responses became highly unreliable. The audiogram in Figure 4 is a composite of the testing that was completed during a 4-month time frame. Note its general resemblance to the audiogram obtained at 14 months of age. Audiometric thresholds were in the moderate-severe hearing loss range for the right ear and mild-severe hearing loss range for the left ear. Consistent with the observations at 14 months, the DPOAEs
were measurable at 6000 and 8000 Hz for the right ear but not at other frequencies. For the left ear, responses were present at these frequencies as well. These OAE findings are shown in Figure 2.

A trial with a loaner hearing aid for the left ear was initiated when he was 4 years, 2 months old. The left ear was fitted because more reliable and complete information was available for that ear at that time. He was fitted with a linear compression limiting behind-the-ear hearing aid that was set using Desired Sensation Level (DSL) (Seewald et al, 1997) targets for gain and output. Because of the diagnosis of AN, the maximum output was set below the DSL targets in an effort to safeguard against levels that might exceed his tolerance. This resulted in less than 105 dB SPL maximum output, except for a peak of 111 dB SPL at 2400 Hz. On the day that he was fitted with the hearing aid, he commented that it was too loud at the recommended settings. After a trial of 2 weeks and a reduction in the initial gain and output settings, he wore the hearing aid at preschool but would not wear it at home for more than an hour a day. Despite the fact that the DSL results suggested that the amplified long-term average speech spectrum was audible at most frequencies, reports from the family and teacher indicated that there was no difference in his auditory behaviors when the hearing aid was in place. Because of the lack of observable improvements in auditory responses with the hearing aid and of his family's impression that wearing amplification seemed to be almost painful or punishing, hearing aid use was ultimately discontinued in favor of alternate remediation strategies.

A speech and language evaluation was completed by his school district when the child was 4 years, 3 months. His articulation was characterized by multiple omissions and inconsistent substitutions with inappropriate voicing. On an intelligibility rating scale of 0 (adequate) to 4 (unintelligible), he was rated at 3. Voice resonance was hyponasal and his speech was described as having a hollow, muffled quality with a low pitch. Vocal prosody and breath control were judged as poor, with prolonged production of vowels. Receptive vocabulary was below the 0.1 percentile and expressive vocabulary was at the 4th percentile. He exhibited limited lexical diversity and consistently used only a few verbs.

Classroom observations completed when he was 4 years, 5 months old revealed that he responded inconsistently to auditory stimuli and that his responses were always poorer when there was background noise. He exhibited extreme variability in his auditory performance, experiencing good and difficult days. Expressively, he tended to be highly orally oriented, but spontaneous expressive language contained a high proportion of intonation-inflected jargon and stereotypic phrases. He had difficulty comprehending spoken messages when only auditory cues were present, and it appeared that he processed intonation contour and syllable number but not spectral details. This latter effect was reflected in poor fine discrimination ability (i.e., chance performance on minimal pairs tasks, such as discriminating between cat vs hat or ball vs bell). Also, during testing, it was noted that when he experienced difficulty comprehending the speaker's utterance, he sometimes turned his unaided ear (which had poorer thresholds) toward the sound source. The addition of visual cues dramatically improved his receptive performance. This same improvement in performance was also observed during the audiometric testing at this time. It was noted that he subjectively appeared to have better word recognition abilities when stimulus words were presented with both visual and auditory cues as opposed to auditory only. Further, he seemed much less likely to become frustrated or disinterested in test tasks when visual input accompanied auditory input.
Performance on the Goldman-Fristoe Test of Articulation revealed extremely variable productions. He had a limited consonantal repertoire, and /w/ was the only consonant used correctly at the word level on a consistent basis. He exhibited postvocalic deletion in 84 percent of the words he produced. Some of his errors were not consistent with hearing loss, suggesting a possible motor component to his speech. His lack of tongue tip elevation was evident and caused a major impact on his speech intelligibility. This raised some concerns for oral-only methods.

Following classroom observations and diagnostic teaching sessions, his family was encouraged to use total communication (TC) as a bridge to oral language development. They were agreeable to this and proceeded to increase their signing skills.

His complex learning needs necessitated a program that included ongoing diagnostic teaching responsive to his changing needs; family involvement in decision making; reduction of classroom noise; use of visual bridging strategies (cuing, signing, and writing) to ensure receptive language acquisition, support oral training, and assist in repairing breakdowns in expressive communication due to unintelligibility; regular and individual speech instruction from a therapist experienced with deafness and motor speech needs; and an additional year in preschool to further build a foundation for academics.

At 4½ years of age, behavioral audiometric thresholds remained stable. Word recognition skills were assessed in two test sessions using the Word Intelligibility by Picture Identification test (Ross and Lerman, 1971), a closed-set, picture-pointing test. When words were presented in the sound field at a level of 60 dB HL, he scored 2 of 10 words correctly. At a presentation level of 95 dB HL under earphones, he correctly identified 1 of 10 words for the right ear and 4 of 15 for the left ear. This was the first test session where word recognition could be tested given his limited attentiveness to testing in the sound booth. These informal findings suggested that even without background noise, word recognition skills were minimal when only auditory cues were present. This further supported the decision to incorporate manual communication into his remediation plan.

After there had been a systematic introduction of a TC program at school and at home for a period of 3 months, language and preacademic tests were conducted when he was 4 years, 10 months old. Testing was repeated at age 5 years, 5 months. Comparisons between standard scores (SS) for receptive and expressive language and for vocabulary at the different times of testing are shown in Figure 5. It can be observed that significant progress occurred in vocabulary and receptive language. His learning rates in receptive language were markedly improved compared to rates observed prior to the implementation of visual communication. Changes in expressive language were slower to occur, but improvements were observed at the time of the most recent testing in comparison to baseline results. In addition, mean length of utterance improved from a pre-TC program level of 2.0 to 6.1 at the time of the latest testing. Postvocalic deletions in words were reduced from 84 percent to 9 percent. As mentioned above, prior to the implementation of a TC program, /w/ was the only consonant consistently used correctly at word level. At that time, 7 consonants were categorized as developing and 14 were undeveloped. Results of the most recent assessment showed 10 consonants used correctly, 8 emergent, and only 4 not developed.

Some additional testing was done at 5 years of age. Rotary chair assessment showed peripheral vestibular output for at least one ear, but vestibular asymmetry could not be ruled out based on this testing. (He had reached age-appropriate developmental milestones for sitting unsupported and walking without assistance.) Ophthalmology evaluation, including fundus examination, showed hyperopia with no other abnormalities. Based on the absence of any other genetic abnormalities, a genetic evaluation resulted in a preliminary diagnosis of a subtype of AN related to major neonatal illness and particularly to the hyperbilirubinemia.

He continues in preschool at this time, receiving intervention for speech/language and
hearing. Phonology continues to be a problem area. Because it is uncertain how the AN impacts his phonologic rule formation and self-monitoring abilities, future challenges include exploration of strategies that will support increased speech intelligibility. There is some question of possible fluctuation in auditory performance. Informally, his parents report that his hearing seems to be worse when the outside temperature is warmer. His own reports of changes in hearing sometimes coincide with ill health. Current plans are to fit him with an auditory trainer set to unity gain in an effort to improve his performance in the noisy listening environment of the classroom.

DISCUSSION

This case demonstrates many of the attributes of the AN patients who have been described in the literature. When he was an infant, his family and pediatrician were of the opinion that he was responsive to sound. This, in combination with the measurable OAEs, resulted in delayed implementation of early intervention services. Case management was further complicated by underlying otitis media with effusion, which may have been partially responsible for any compromised auditory responsiveness. Due to the above factors, the initiation of a trial with amplification for this patient was also delayed. Eventually, educational services were provided and amplification was initiated. Unfortunately, the benefits of amplification were limited, and following a relatively short trial period, hearing aid use was discontinued. The planned trial with an auditory trainer may yield a more favorable result.

This patient has acquired language, albeit delayed, through the use of audition. His auditory comprehension is enhanced when supplemental visual information (speech reading, signing, cueing, or written) is presented. His atypical speech production hampers expressive performance; however, he remains orally oriented. A motor-speech disorder cannot be ruled out. Also, inconsistent auditory input likely contributed to a disordered phonologic development or a disordered rule system.

This case lends support to the recommendations that manual or visual communication should be included in the rehabilitation plan of patients diagnosed with AN (Sininger and Starr, 1997; Berlin et al., 1998; Hood, 1998). As mentioned above, the auditory system in an individual with AN seems to lack the accurate timing necessary to make speech intelligible. Consequently, language therapy focusing only on audition may yield limited results in overcoming communication delays. To address this problem, Berlin et al. (1998) recommend the use of some type of visual language—particularly cued speech—with children having AN to allow them to participate in the conversations of the people in their environment. Hood (1998) advises the use of signed English or cued speech because they follow the grammatical structure of the English language and can be used to supplement available auditory information and speechreading. Sininger and Starr (1997) advocate the initiation of manual communication training with children as soon as AN is suspected.

When his speech-language abilities were assessed at age 4 years, 3 months, his scores indicated marked delays in development. At age 4 years, 10 months, approximately 3 months after systematic implementation of TC, his standard scores for receptive language and vocabulary showed notable improvement. More recent observations reveal rapid improvements in both receptive and expressive language, but there are continuing concerns for speech intelligibility.

Because the majority of the AN cases that have been documented in the literature display auditory disorders that are longstanding and impede speech and language development, it is hoped that it will become increasingly easier to secure early intervention services for children with this diagnosis. Referral to early intervention specialists familiar with children who have auditory disorders should be initiated after test findings that suggest AN are confirmed. The outcome of any given case of AN cannot be predicted from ABR, OAE, or audiometric test findings, making it critical that educational services be provided that are individualized and flexible and have a diagnostic teaching emphasis. The possible presence of concurrent neuropathies necessitates a referral to a neurologist for assessment and monitoring, if indicated.

Use of amplification by patients who have AN and whose behavioral test results suggest hearing loss remains controversial. In general, it has been suggested that a trial with amplification is indicated when there is evidence of elevated behavioral audiometric thresholds and that the OAEs should be carefully monitored during the hearing aid trial. It has been recommended that the maximum output of the hearing aid be set to a “conservative” level; however, the selection of this conservative level has not been defined clearly. Regardless, amplification
outcomes have been variable. Cochlear implantation has also been suggested as an alternative for some cases of AN. Again, outcomes have been variable. The source of the variability of auditory performance, both unaided and aided (whether with amplification or implantation), is not known. However, as discussed above, it is likely that AN has multiple etiologies and possibly multiple sites of lesion. Reports of functional hearing abilities vary widely within this population, and it may be that degree of neural impairment varies with etiology. This may have an influence on the amount of benefit realized with amplification/implantation for a given individual. The further delineation of the pathogenesis of AN will guide rehabilitation decisions in the future.

The test battery approach in the diagnosis of pediatric auditory disorders is not a new concept. As this approach is applied to the diagnosis of AN, a protocol is suggested that provides ongoing monitoring of auditory status using behavioral audiometric assessment, tympanometry (to ensure that middle-ear dysfunction is not influencing the OAE results), OAEs, and evoked potentials. Following identification of AN, it is recommended that referral be made for provision of aural (re)habilitation services following a multidisciplinary team evaluation that includes any indicated medical evaluations (e.g., otolaryngology, neurology, ophthalmology). Until more is understood about the site of lesion in AN, amplification use should be approached cautiously and on an individual basis, with repeated evaluations of OAEs and behavioral thresholds. Likewise, candidacy for cochlear implantation for these patients must be carefully evaluated.

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