

# AUDITORY NEUROPATHY: A POTENTIALLY UNDER-RECOGNIZED NEONATAL INTENSIVE CARE UNIT SEQUELA

JO ANN D'AGOSTINO, RN, MSN, CPNP, AND LAURA AUSTIN, MS, CCC/A

## ABSTRACT

Auditory neuropathy (AN) is a hearing disorder that affects newborns. Those with high-risk neonatal histories, family history of childhood hearing loss, and hyperbilirubinemia are at greatest risk. Current neonatal intensive care unit (NICU) hearing screening methods that rely only on otoacoustic emissions will fail to detect this disorder. Auditory neuropathy differs from conductive hearing loss and sensorineural hearing loss; a specific constellation of findings on audiologic evaluation are diagnostic of this disorder. The pathophysiology of AN is unclear; however, it may be caused by demyelination or degeneration at points along the auditory pathway. The actual incidence of AN is unknown; it is more prevalent in high-risk infants. The course of AN varies widely among patients. Current management ranges from close monitoring of the child's development to cochlear implantation. Neonatal intensive care unit nurses need to be aware of this disorder to help support and educate at-risk families and to alert them of the need to monitor hearing and language development in their infants.

**KEY WORDS:** auditory neuropathy, auditory dys-synchrony, auditory brainstem response (ABR), otoacoustic emissions (OAE), universal newborn hearing screening, hearing loss, hearing impairment, newborn intensive care, infant, newborn cochlear implant.

Estimates of moderate to profound hearing loss in newborns range from 1 in 900 to 1 in 2500.<sup>1</sup> Before the implementation of early hearing detection and intervention programs, the average age of identification of hearing loss in the United States was 30 months.<sup>2</sup> Those with a mild or moderate loss were often not identified until later.<sup>3</sup> For this reason, universal newborn hearing screening is now the standard of care in most of the United States.<sup>4,5</sup> Thirty-eight states have enacted laws and 4 states have voluntary

early hearing detection intervention programs in place.<sup>5</sup> Currently, in-hospital screening techniques and procedures for follow-up of infants who fail screening vary among institutions. An extensive discussion of newborn hearing screening has been previously published.<sup>6</sup>

Auditory neuropathy (AN) is a specific hearing disorder that affects infants; high-risk infants are at increased risk.<sup>4,7-10</sup> The identification of AN is dependent on specific screening and diagnostic hearing tests and may be under-recognized due to current neonatal intensive care unit (NICU) hearing screening practices.<sup>4,11-13</sup> A child with AN may not be identified until after discharge. In their Year 2000 Position Statement, the Joint Commission on Infant Hearing acknowledged this issue. It has recommended further evaluation of this disorder's prevalence and natural history to address the disorder with better understanding. It has also suggested that as more information becomes available, future screening protocols may need to be revised; however, to date, no changes in current screening protocol have been recommended.<sup>4</sup>

Auditory neuropathy was first named in the literature in 1996 as a way to describe an unusual pattern of

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Address reprint requests to Jo Ann D'Agostino, RN, MSN, CPNP, The Children's Hospital of Philadelphia, 34th Street and Civic Center Boulevard, Philadelphia, PA 19104-4399. E-mail: DAgostino@email.chop.edu

From the Neonatal Follow-Up Program and Center for Childhood Communication, The Children's Hospital of Philadelphia, Philadelphia, Pa.

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audiologic test results of 1 child and 9 adults.<sup>14</sup> There is evidence that these findings may have been noted by audiologists for many years before being formally named.<sup>15-20</sup> As this is a relatively newly described and poorly understood disorder, there is much to be learned, including prevalence, natural course, and a clearer definition of treatment options.<sup>4</sup>

Most information about AN has been in the audiologic literature; NICU nurses may not be aware of the problem. Even though the disorder may be diagnosed after discharge, it is important that NICU nurses be familiar with AN, which can affect their high-risk patients. It is a disorder with the potential for long-term impact on a child's development. The purpose of this article is to raise NICU nurses' awareness of AN, provide a brief overview of the risk factors for and the current understanding of this disorder, and describe how current neonatal hearing screening and diagnostic techniques are involved in the identification of this disorder. See Table 1 for common terminology related to AN.

## THE ANATOMY AND PHYSIOLOGY OF HEARING

When sound is received from the environment, it passes through the external ear, via the auditory canal, to the tympanic membrane (Fig 1). Sound is then transmitted as a vibration by the middle ear ossicles through the middle ear to the inner ear. The inner ear's primary organ is the cochlea, which holds fluid and is encased in bone in the skull. Along the inside of the cochlea is the basilar membrane in which inner and outer hair cells lie within the Organ of Corti (Fig 2). The vibration from the sound moves the fluid in the inner ear. The moving fluid results in movement of the hair cells, which in turn creates neural impulses. These impulses are then transmitted via the spiral ganglion along the auditory nerve to the brainstem.

### Types of Hearing Loss

To understand AN, it is important to differentiate it from other forms of hearing loss. Conductive hearing loss (CHL) is a deficit created by an obstruction. This may include pathology in the middle ear such as fluid or infection, malformation of or damage to the external or middle ear anatomy, or a blocked ear canal such as a foreign body or impacted cerumen. Conductive hearing loss may be temporary or permanent depending on the etiology.

Sensorineural hearing loss (SNHL) involves abnormalities in the cochlea, usually involving damage to the outer hair cells. Multiple risk factors are associated with this type of hearing loss in the NICU population. Among these factors are prematurity, low-birthweight, perinatal infection, meningitis, and exposure to aminoglycosides.<sup>4</sup> Sensorineural hearing loss also includes abnormalities of the auditory nerve that may result from space-occupying lesions or genetic disorders.

**Table 1. Glossary of Terms<sup>29,55-57</sup>**

Term	Definition
Auditory Brainstem Response (ABR)	An evoked response of the auditory system from a sound, generating electric potentials graphed as brain waveforms.
Air conduction testing	An audiogram using earphone transducers; evaluates the whole peripheral system from the outer ear to the inner ear.
Audiogram	Chart used to graph hearing sensitivity thresholds, with the x axis being frequency measured of a pure tone measured in Hertz and the y axis being the intensity of that particular tone heard measured in decibels.
Bone conduction testing	An audiogram using a bone conduction oscillator, which vibrates the skull and bypasses the outer and middle ear, testing the acuity of the cochlea alone.
Cochlear microphonic	Electrical signal from hair cells seen on ABR waveform when polarity of stimulus is inverted.
Conductive Hearing Loss (CHL)	Reduction in hearing sensitivity when tested by air conduction; normal hearing sensitivity by bone conduction.
Electrophysiologic testing	Umbrella of tests that do not require voluntary response from patient, including ABR and OAE testing.
Hearing thresholds	The softest sounds that someone perceives graphed on an audiogram.
Immittance testing	Includes tympanometry and acoustic reflex testing.
Middle ear acoustic reflex	A loud sound will cause a middle ear muscle contraction, measured by the resulting change of eardrum position.
Otoacoustic Emissions (OAE)	Pre-neural assessment of outer hair cells in response to a click or calibrated auditory stimuli.
Sensorineural Hearing Loss (SNHL)	Hearing loss in which air and bone conduction thresholds are essentially the same.
Mixed Hearing Loss	Hearing loss in which air and bone conduction thresholds are reduced in sensitivity and the bone conduction scores are better.
Tympanometry	Systematic displacement of the eardrum within a certain normal range as varied below and above ambient air pressure.

Mixed hearing loss includes both a conductive and sensorineural component.

In AN, the outer hair cells in the cochlea are intact

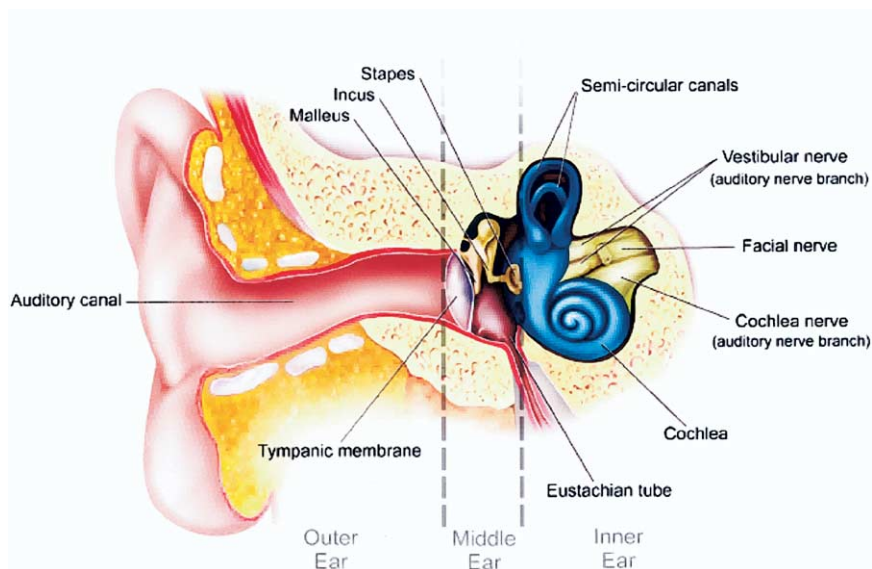


Figure 1. Anatomy of the auditory system. Sound passes through the auditory canal to the tympanic membrane. It is then transmitted as vibration of the middle ear ossicles to the fluid-filled cochlea in the inner ear.

and seem to work normally. However, functional abnormalities may exist with the inner hair cells, the synapse between the inner hair cells and their dendrites, the spiral ganglion, the auditory nerve fibers, or perhaps a combination of these areas.<sup>21</sup>

#### SCREENING FOR AUDITORY NEUROPATHY

The 2 hearing screening tests most commonly used for neonatal screening are otoacoustic emissions (OAE) and auditory brainstem response (ABR). Hospitals use either OAE or ABR screening equipment for infants, although ABR screening is the preferred method for high-risk infants.<sup>22</sup> The screening ABR presents sound to the outer ear that then travels to the cochlea. It continues beyond the cochlea to evaluate neural function of the auditory brainstem pathways.<sup>4</sup> Most screening ABR equipment is automated and the stimulus level for the hearing screening is at 35 dB.<sup>23</sup> A passed ABR screen suggests normal to near-normal hearing, but it does not rule out a mild hearing loss.

Otoacoustic emissions testing can be used as a screening or diagnostic test depending on the equipment and specific protocol that is used. Otoacoustic emissions testing specifically evaluates outer hair cell function within the cochlea. The presence of a response implies normal outer hair cell function, which suggests that hearing sensitivity is normal to near normal. A normal OAE does not rule out a mild hearing loss. See Tables 2 and 3 for implications of newborn hearing screening tests and newborn diagnostic hearing tests.

#### DIAGNOSING AUDITORY NEUROPATHY

An infant who does not pass a hearing screening should receive a full diagnostic evaluation by an audiologist, including ABR, OAE, immittance, and, if 6 months or older developmentally, an audiogram. A multitest battery for confirmation of hearing loss is recommended as a cross-check.<sup>24</sup> A diagnostic ABR allows the audiologist to pinpoint the decibel level at which the auditory system is responding. It also allows for bone-conduction testing, which uses a different transducer to determine if the hearing loss is sensorineural, conductive, or mixed.

Otoacoustic emission testing is usually repeated as part of the diagnostic evaluation to obtain more information about the auditory system. Otoacoustic emission testing is a preneural assessment and does not detect dysfunction along the auditory nerve or brainstem.<sup>25</sup> Both ABR and OAE are electrophysiological tests that provide information about the integrity of the auditory system by an evoked response.<sup>25</sup>

Immittance testing is also performed as part of the test battery and includes tympanometry and middle ear acoustic reflexes. Tympanometry yields information about middle ear status by looking at eardrum movement. Middle ear acoustic reflexes are measured by a change in eardrum status in response to a loud sound. Finally, an audiogram can be obtained from an infant as young as 6 months. It shows information about the hearing thresholds of 1 or both ears for different frequencies and requires a behavioral response from the infant, such as a head turn.

Children with a significant hearing loss, whether it is a conductive or sensorineural, will have a corresponding

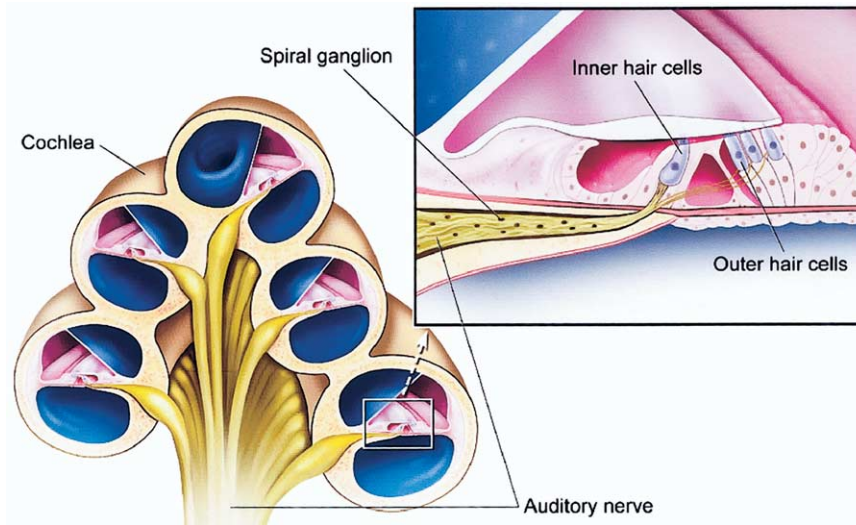


Figure 2. Cross-section of the cochlea with detailed view of the Organ of Corti. The hair cells in this structure create neural impulses that are transmitted by the spiral ganglia along the auditory nerve to the brain.

ABR response at the level of hearing loss and absent OAE responses. For example, an infant with a severe to profound hearing impairment would have no response on the ABR test, as well as no OAE responses. In contrast to CHL and SNHL, children with AN have absent or severely abnormal ABR responses, yet the OAE response is present. Table 3 reviews the implications of diagnostic hearing screening tests.

Auditory neuropathy cannot be diagnosed with an individual test. The definition of AN is based on a constellation of findings from audiological testing, including:

- Absent or severely abnormal ABR
- Present cochlear microphonics (a response from outer hair cells on ABR testing)
- Present OAE, normal tympanometry with absent middle ear acoustic reflexes
- Varying audiograms<sup>26</sup>

There is no specific test that evaluates inner hair cell function, which may be affected in AN.<sup>27</sup>

### PATHOPHYSIOLOGY OF AUDITORY NEUROPATHY

Much of the information about the pathophysiology of AN has been obtained through animal studies. In a study involving Gunn rats, the auditory nerve, spiral ganglia, and brainstem auditory nuclei were damaged at higher degrees of bilirubin toxicity. This would be consistent with the development of AN but has not been replicated in humans.<sup>28</sup> Animal studies using adult chinchillas suggested that chronic, mild hypoxia can produce changes compatible with AN.<sup>29</sup>

Two theories regarding the pathophysiology of AN

have been proposed. One theory is that there may be demyelination of auditory nerve fibers, which in turn slows conduction velocities. As demyelination may vary for each fiber, there may be a dys-synchronous neural response.<sup>30</sup> This has led some to refer to AN as auditory dys-synchrony.<sup>31,32</sup> Another theory is that AN may be the result of primary cochlear neuronal (inner hair cells or synaptic connection) degeneration.<sup>33</sup> More research is needed to determine the exact pathophysiology of AN and whether one or more sites of the auditory pathway are involved.

### REVIEW OF THE LITERATURE LINKING RISK FACTORS TO AUDITORY NEUROPATHY

The onset of AN can vary from the neonatal period to adulthood depending on the etiology of the disorder.<sup>34</sup> Factors that may place a child at increased risk of AN in the neonatal period include high-risk neonatal histories, family history of childhood hearing loss, or history of hyperbilirubinemia.<sup>4</sup> Additional factors that have been reported in children with AN include prematurity, exposure to aminoglycosides, hypoxia, and metabolic and mitochondrial disorders.<sup>7,9,10,32,35-38</sup> Reports describing these risk factors in the neonatal population are small in size with limited details about the neonatal histories.

One of the earliest studies to describe AN in high-risk neonates involved 3 children.<sup>7</sup> Two were premature with complicated neonatal courses, including respiratory distress syndrome, 1 of whom also had hyperbilirubinemia, and the third child had a metabolic disorder, Cytochrome C Oxydase Deficiency. In another study, 8 of 9 children with AN had either been born prematurely or had high-risk neonatal histories.<sup>35</sup>

In a study of 22 children with AN, 68% had a

**Table 2. Implications of Universal Newborn Screening Tests\*<sup>58</sup>**

Hearing Screen	Results
Present OAE	No CHL or SNHL greater than mild degree but cannot rule out AN.
Absent OAE	Possible CHL or SNHL that is mild or greater, or possible AN mixed with CHL.
Passed ABR screening	No CHL or SNHL greater than mild degree, no AN.
Did not pass ABR screening	Possible CHL, SNHL, or AN.

\*Note that all abnormal screening tests need additional confirmatory testing before diagnosis.  
Abbreviations: OAE = otoacoustic emissions; ABR = auditory brainstem response; CHL = conductive hearing loss; SNHL = sensorineural hearing loss; AN = auditory neuropathy.

complicated perinatal course, including prematurity, use of ototoxic medications, and mechanical ventilation.<sup>9</sup> Hyperbilirubinemia was reported in 50% of the patients. Total bilirubin levels ranged from 12.3 to 40.0 mg/dL (210.33 to 684  $\mu\text{mol/L}$ ) with a mean level of 19.4 mg/dL (331.74  $\mu\text{mol/L}$ ). Onset was noted day 2 to 3 of life and mean duration was 6.8 days. Thirty-six percent of patients had family members with hearing loss. Similarly, in a study of 20 patients with AN, 50% had a history of hyperbilirubinemia, 20% had a history of hypoxia; 30% had no neonatal risk factors.<sup>10</sup>

In another study which involved 12 children with kernicterus, 10 of the children met the clinical criteria for AN.<sup>28</sup> Findings compatible with AN were reported in a small study of 4 prematurely born children, who all had histories of hyperbilirubinemia; 2 required exchange transfusion.<sup>36</sup> Peak serum bilirubin levels for the 2 requiring exchange transfusion were 15.5 mg/dL (265.05  $\mu\text{mol/L}$ ) at 42 hours with birthweight of 2370 g, and 12.4 mg/dL (212.04  $\mu\text{mol/L}$ ) at 4 days with birth weight of 2005 g. One had Rhesus hemolytic disease and the other Rhesus sensitization. The remaining 2 children included 1 with a peak bilirubin level of 4.9 mg/dL (83.79  $\mu\text{mol/L}$ ) and birthweight of 885 g, and another with peak bilirubin level of 14.9 mg/dL (254.79  $\mu\text{mol/L}$ ) with birthweight of 2615 grams. Both were treated with phototherapy.

In a study of subjects with AN who presented before 2 years of age, 44% had a family history of hearing loss. The study included several sibling pairs.<sup>11</sup> Other small case reports involving siblings can be found in the literature.<sup>20,37</sup> In a study of familial AN, 3 family pedigrees suggested an autosomal recessive inheritance,

**Table 3. Implications of Diagnostic Audiologic Tests<sup>58</sup>**

Test Result	Implication
Abnormal ABR	SNHL, AN, or CHL
Abnormal ABR + Absent OAE	SNHL, CHL, or AN with CHL
Abnormal ABR + Present OAE	AN

Abbreviations: ABR = auditory brainstem response; OAE = otoacoustic emissions; SNHL = sensorineural hearing loss; CHL = conductive hearing loss; AN = auditory neuropathy.

and in a fourth family an X-linked recessive hereditary pattern was suggested.<sup>38</sup>

In addition to family history, other hereditary conditions may be associated with AN; these include Charcot-Marie-Tooth (also known as hereditary motor and sensory neuropathy) and Friedreich's ataxia.<sup>14,21,39-41</sup> Charcot-Marie-Tooth has a 17p11.2 locus and Friedreich's ataxia has a 9q13-q21.1 locus. Both involve progressive neurological degeneration and typically are not diagnosed until after the neonatal period.<sup>42</sup> One child with a mitochondrial disorder has also been reported to have AN.<sup>43</sup>

A history of meningitis has been reported in 2 patients with AN.<sup>44,45</sup> One child developed meningitis at 1 year of age; the age of onset was not reported for the second child. Because the information about risk factors is based on small case studies, more research is needed to determine the significance and role of each these factors in the development of AN.

## INCIDENCE OF AUDITORY NEUROPATHY

The prevalence of AN in the general population has not been clearly determined.<sup>4</sup> A few studies have reported the incidence of AN in children who have been diagnosed with a hearing loss.<sup>9,10,17,46</sup> The incidence ranged from 5% to 15% with an average of 10%. This range was in a subset of children with an identified hearing loss and ABR abnormality, inferring that it is an uncommon disorder in the general population. There is limited research in this area and more is needed to determine the actual incidence of AN in the general population.

A study of children at risk of hearing impairment provided some insight into the incidence of AN in the high-risk neonatal population.<sup>10</sup> The children had their hearing evaluated because their high-risk neonatal or family histories placed them at risk of hearing loss. Of the 5199 children who were assessed, 12 children had AN (1:433). Again, there is not enough information in the literature to determine the actual incidence of AN in the high-risk neonatal population. This is an important area of future research.

## COURSE AND OUTCOME OF AUDITORY NEUROPATHY

Currently, the literature on AN is limited and consists primarily of small case reports that suggest that the course of AN varies widely.<sup>9</sup> Auditory neuropathy can occur unilaterally or bilaterally; bilateral presentation is the most common.<sup>9,21,47,48</sup> Some patients have normal hearing, whereas others are deaf.<sup>9,32,35,45,49</sup> Some children have shown progressive or fluctuating hearing loss; others have shown improvement over time; and still others have remained unchanged.<sup>9,25,35,50</sup>

Even more intriguing are reports that the course of AN can be variable within the same patient. Fluctuations in hearing acuity have been reported in 3 children associated with changes in body temperature. When febrile, these children exhibited transient profound hearing loss and, when afebrile, were able to comprehend speech in a quiet environment.<sup>37</sup>

One of the most common difficulties exhibited by those with AN is difficulty understanding speech, especially when in noisy environments. This can be a problem for some patients even when there is only a low level of background noise.

Risk factors associated with AN may impact the course of the disorder. In 1 study of children with AN, those with a history of hyperbilirubinemia were more likely to show spontaneous audiological improvement than those without hyperbilirubinemia.<sup>9</sup> Of the 22 children included in the study, 10 were born premature. Some patients with AN may also have coexisting neurological morbidity, such as cerebral palsy, apraxia, feeding problems, motor delays, or evidence of subtle peripheral neuropathy noted only on detailed neurological examination.<sup>14</sup>

## MANAGEMENT OF AUDITORY NEUROPATHY

The variability of AN makes it a challenge to manage. There is no consensus on a particular approach. A child needs to be viewed as an individual, and a plan should be developed in conjunction with the child's family, as is done with any type of hearing disorder.

A critical first step is to confirm the diagnosis with a thorough audiological assessment as described above. Once the diagnosis has been confirmed, a multidisciplinary evaluation is recommended to assist in identifying the etiology, if possible, and to rule out comorbidities.<sup>35</sup> In addition to the child's primary care provider, specialists involved in assessing a child may include an audiologist, speech-language pathologist, neurologist, geneticist, otorhinolaryngologist, ophthalmologist, and developmental specialist. Periodic assessments are recommended to monitor the child's auditory development and also to provide support to the family. These supports may include individual counseling or support groups.<sup>35</sup> In addition, periodic develop-

mental assessments are necessary to screen for any language delays, neurological sequelae, or other developmental delays and to help provide resources to families to meet their child's developmental needs.

It is important that all children with AN have careful periodic monitoring of their auditory development and speech and language development. This can be achieved through various methods including early intervention services, periodic assessment by a speech and language pathologist, and/or through involvement with a developmental follow-up program.<sup>35</sup> The benefits of early intervention for a hearing-impaired child have been clearly demonstrated.<sup>51,52</sup> In their Year 2000 Position Statement, the Joint Committee on Infant Hearing called for consensus regarding appropriate early intervention strategies for infants with AN.<sup>4</sup>

If delays are noted or if a child appears to be having difficulty processing auditory information, increasing a child's exposure to language is an important approach.<sup>25</sup> Exposing the child to opportunities to listen in on conversations can increase opportunities for language exposure. In addition, a bimodal approach to language, that is, the use of visual augmentation to spoken language, may be a useful tool.<sup>25,32,50</sup> For infants, this generally involves the use of basic gesturing for common words, such as mommy, daddy, more, etc. There are various formal visual communication methods available for more complex communication. The most common include Cued Speech, American Sign Language, and Signed Exact English (Table 4). In postlinguistic patients who are diagnosed with AN, training in speech reading skills may be helpful for situations when the patient is in a noisy environment.<sup>25</sup>

Some audiologists recommend a trial with hearing aids. Although controversy exists, there are AN patients who have benefited from amplification.<sup>45</sup> In 1 study, the use of hearing aids improved speech perception in half of the subjects in the study.<sup>45</sup> Others express concern that hearing aids increase the volume of sound but may not clarify sounds for children with AN.<sup>50</sup> As hearing aids compensate for abnormal outer hair cells and the outer hair cells are normal in children with AN, the use of hearing aids may potentially damage these normal outer hair cells. To avoid this, some audiologists have recommended a trial of low maximum-power-output hearing aids.<sup>25</sup> Others recommend using only 1 hearing aid during trials.<sup>34</sup> Auditory neuropathy patients with hearing aids need to be carefully monitored by an audiologist to assess tolerance and responsiveness.<sup>50</sup>

## ADVANCED TECHNOLOGY: COCHLEAR IMPLANTS

Cochlear implantation has been used in some children with AN who do not seem to be benefiting from standard interventions. Children who have AN

**Table 4. Most Common Methods of Visual Communication and Web Resources<sup>59-61</sup>**

Type of Speech	Definition	Resource
<b>Cued Speech</b>	Involves the use of 8 hand shapes placed in 4 locations near the mouth. These are combined with natural speech so that each spoken sound looks different.	National Cued Speech Association <a href="http://cuedspeech.mit.edu/">http://cuedspeech.mit.edu/</a> Information for parents and professionals. Offers legal and research resources and information about cue camps. Links to Cued Speech Discovery Web site, which offers telephone resource for information, information about cued speech, workshops and books, videos, DVDs, games and flash cards for purchase.
<b>American Sign Language (ASL)</b>	A complete language that is not based on English. Involves the use of hand shapes, body movements, gestures, and facial expressions to form words.	National Institute on Deafness and Other Communication Disorders <a href="http://www.nidcd.nih.gov/health/hearing/asl.asp">http://www.nidcd.nih.gov/health/hearing/asl.asp</a> General information as well as links to Web sites of national deafness organizations.
<b>Signing Exact English (S.E.E.)</b>	A code system that uses ASL signs modified to create English-like utterances including prefixes and suffixes, English word order, etc. Not a complete language.	The S.E.E. Center <a href="http://www.seecenter.org">http://www.seecenter.org</a> Bulletin board, parent information packets, telephone resource. Offers books, videos, and CD-ROMs for purchase.

with severe to profound audiograms are potential candidates for cochlear implantation, as are children with severe to profound SNHL.<sup>53</sup> All centers that perform cochlear implantation have teams that evaluate candidates.

If a child has been using a visual communication method before cochlear implantation, it is recommended that it be continued after implantation to facilitate the acquisition of speech and language.<sup>50</sup> In a review of recent reports of 23 children with AN who received cochlear implants, the age of cochlear implantation ranged from 15 months to 5.8 years. Outcome data were reported on 18 of these children and all showed improvement in auditory and communication skills after implantation.<sup>9,53,54</sup>

#### IMPLICATIONS FOR NICU NURSES

Because AN is a newly named disorder, many questions remain regarding the incidence, etiology, risk factors, course, and management. Parents of newly diagnosed infants may ask questions for which there are not yet answers. The technology now exists to identify more cases, and knowledge about this complicated disorder is rapidly expanding. Until more is known about this disorder, it is important to remember to approach each child as an individual, as there is wide variation among children with AN.

Because long-term prospective studies of children with AN are lacking, it is difficult to predict an infant's auditory and speech and language potential. The audiologists involved in a child's care are the frontline

resources for families in regard to the latest information on this topic.

Some major audiology centers also have support groups for families of children with AN. In addition, there is an AN Listserv and informational Web sites available for families, and a textbook directed toward professionals working with AN patients (Table 5). Emphasize that information found on the Internet should be verified with the professionals. Close professional monitoring of a child's auditory and developmental progress is important, especially during the period of early speech and language development. In particular, those children with AN who have normal or near-normal hearing thresholds may be at risk of inadequate surveillance. In 1 study, families of children with normal or near-normal hearing thresholds were the most likely to express doubt or denial about their child's test results.<sup>35</sup> Disbelief of the diagnosis led to discontinuation of audiologic follow-up by some families. Close monitoring is recommended well beyond the period of early speech and language because of the variability of AN and the unknown long-term outcome of this disorder.<sup>35</sup>

Regardless of whether AN is diagnosed before or after discharge, NICU nurses can help facilitate the communication of hearing screening results to primary care providers to assure follow-up. Educate parents about the meaning of hearing screening tests. Passing a hearing screening does not rule out a mild hearing loss, nor does it preclude the development of a hearing loss. Teach parents about normal auditory milestones. Brochures about newborn hearing screening and auditory

**Table 5. Auditory Neuropathy Informational Resources**

Resource Name	Description
<b>Auditory Neuropathy: A New Perspective on Hearing Disorders.</b> <sup>30</sup>	Textbook written for professionals. Albany, NY: Delmar; 2001.
<b>Yahoo! Group: Auditory Neuropathy</b> <a href="http://groups.yahoo.com/group/AuditoryNeuropathy/">http://groups.yahoo.com/group/AuditoryNeuropathy/</a>	Provides information and support to families of children with AN.
<b>National Institute on Deafness and Other Communication Disorders</b> <sup>60</sup> <a href="http://www.nidcd.nih.gov/health/hearing/neuropathy.asp">http://www.nidcd.nih.gov/health/hearing/neuropathy.asp</a>	General information on AN and links to Web sites of national deafness organizations.
<b>Auditory Neuropathy Information</b> <a href="http://auditoryneuropathy.tripod.com/ANindex.html">http://auditoryneuropathy.tripod.com/ANindex.html</a>	Parent-developed Web site that offers a chat room, information about AN, personal stories, articles and abstracts, books, and tips for professionals.
<b>Computer Simulation of AN</b> <a href="http://www.bsos.umd.edu/hesp/zeng/simulations.html">http://www.bsos.umd.edu/hesp/zeng/simulations.html</a>	Developed by Drs. Fan-Gang Zeng and Arnold Starr to help families understand AN. <sup>33</sup>
<b>American Academy of Audiology</b> <a href="http://www.audiology.org/store/metools/">http://www.audiology.org/store/metools/</a>	<i>Your Baby's Hearing.</i> Brochure for parents, containing information on infant hearing milestones and hearing loss. Available in English and Spanish.  <i>Newborn Hearing Screening.</i> Brochure for parents and professionals, explaining hearing testing and infant hearing milestones. Available in English.

milestones are available through the American Academy of Audiology (Table 5). Encourage parents to alert their child's primary care provider regarding any concerns they have about their child's development. Additional information about parent reaction to hearing screening results, as well as additional recommendations for parent education have been published previously in this journal.<sup>6</sup>

#### IMPLICATIONS FOR FUTURE RESEARCH

Research about AN is in the early stages. More research is needed to determine the prevalence in both the general newborn and high-risk newborn populations. In addition, large prospective outcome studies to determine the natural course of the disorder are lacking. The exact pathophysiology still needs to be determined. A systematic study of risk factors and their role in AN is another important area of research. Once clarified, ways in which to prevent the disorder may become apparent. The most beneficial management and intervention approaches, including NICU interventions, remain to be identified. Finally, when more information is available, current newborn hearing

screening protocols need to be reviewed to determine if changes in protocol are necessary.

#### CONCLUSION

High-risk infants are thought to be at increased risk of AN. Auditory neuropathy is potentially under-recognized in NICUs using only otoacoustic screening. Because this is a relatively newly named disorder, much research is needed to determine prevalence, risk factors, ways to prevent AN, natural course, management, and intervention approaches for this disorder. Revision of current screening procedures may be recommended after further study.

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