
Guidelines for

Identification and Management of Infants and Young Children with Auditory Neuropathy Spectrum Disorder



The Children's Hospital

Bill Daniels Center for Children's Hearing

**Guidelines Development Conference
at NHS 2008, Como, Italy**



DEBORAH HAYES, PHD

Preface

The “Guidelines Development Conference on the Identification and Management of Infants and Young Children with Auditory Neuropathy” evolved from an honorary *Advances in Children’s Hearing Lecture* delivered by Yvonne Sininger, PhD, at the *Bill Daniels Center for Children’s Hearing*, The Children’s Hospital – Colorado, on the topic of “auditory neuropathy.” As she updated that audience on the most current state-of-the-art in diagnosis and management of children with this disorder, Dr. Sininger also discussed the many questions and controversies about this perplexing and variable condition. After her thought-provoking lecture, Yvonne and I considered the possibility of developing an international conference with invited experts to share information and, hopefully, to arrive at some practical guidelines to help clinicians identify, diagnose, and manage infants and young children with this disorder.

As the idea of an international conference evolved, we concluded that a natural venue for such a conference would be the biennial NHS conference in Como Italy. Since 2000, Dr. Ferdinando Grandori and I have co-chaired the Newborn Hearing Systems (NHS) Conference to provide an international forum for scientists, clinicians, and parents to discuss issues relevant to the identification, diagnosis, and management of newborns and young infants with hearing loss. As this meeting grew over the years, it became apparent that an exceptional synergy emerged from the interactions of more than 500 participants from countries throughout Europe, Asia, Africa, Australia and New Zealand, and the Americas. After conferring with Dr. Grandori, Yvonne and I concluded that this venue was indeed the perfect place to host a guidelines development conference. We subsequently contacted a group of internationally recognized scientists and clinicians with expertise in the area of “auditory neuropathy” to invite their participation in this conference scheduled as a special component of the NHS 2008 Conference (19 – 21 June 2008). To our delight, each invited participant agreed to attend and to contribute not only an oral presentation but also a summary scientific paper in their area of expertise.

The panel of distinguished scientists and clinicians who assembled in Como Italy in June 2008 included Yvonne Sininger, PhD, Arnold Starr, MD, Christine Petit, MD, PhD, Gary Rance, PhD, Barbara Cone, PhD, Kai Uus, MD, PhD, Patricia Roush, AuD, Jon Shallop, PhD, and Charles Berlin, PhD. Given the expertise, experience, and stature of these individuals, it is not unexpected that the guidelines development conference exceeded our expectations for

quality presentations, lively discussions, and active panel and audience participation. The guidelines and summary scientific papers contained in this volume reflect the joint contributions of these eminent professionals. (The titles of some of the contributed papers in this monograph have been changed to reflect the terminology recommended by the expert panel. Terminology in the body of these papers has not been changed and is printed as originally submitted.)

In future years, we will undoubtedly learn more about how to identify, diagnose, and manage individuals with “auditory neuropathy.” In the interim, Dr. Sininger and I hope that clinicians will find these guidelines useful not only for identification and diagnosis of infants and young children with this disorder, but also for initiating a dialogue with parents and families about intervention options for their babies.

I am indebted to Yvonne Sininger for sharing her expertise and guidance in planning, developing, and implementing the conference. Neither the conference nor this publication would have been possible without her selfless contributions. Ferdinando Grandori offered unwavering support for inclusion of the untested concept of a “meeting within a meeting” at the NHS2008 conference. Through her organizational talents, careful attention-to-detail, and gracious kindness, Valerie Hernandez helped transform the concept of this conference from an exciting idea to a well-conducted reality. Jerry Northern added critical wisdom, editorial insight, and professional direction to the publication of this monograph. Lastly, the conference and publication were supported by substantial financial contributions by the Bill Daniels Center for Children’s Hearing and the Kelley Family/Schlessman Family Scottish Rite Masons Chair in Childhood Language Disorders at The Children’s Hospital – Colorado. To our fine panel, Yvonne Sininger, Ferdi Grandori, and my colleagues at The Children’s Hospital, I am forever grateful.

*Deborah Hayes, PhD
Co-Chair, Bill Daniels Center for Children’s Hearing
Kelley Family/Schlessman Family Scottish Rite Masons
Chair in Childhood Language Disorders
The Children’s Hospital-Colorado
August 2008*

GUIDELINES: Identification and Management of Infants and Young Children with Auditory Neuropathy Spectrum Disorder

INTRODUCTION

“Auditory neuropathy” is a relatively recent clinical diagnosis used to describe individuals with auditory disorders due to dysfunction of the synapse of the inner hair cells and auditory nerve, and/or the auditory nerve itself. Unlike patients with sensory hearing loss who show clinical evidence of impaired outer hair cell function, patients with “auditory neuropathy” show clinical evidence of normally functioning outer hair cells. Individuals with “auditory neuropathy” typically demonstrate impaired speech understanding, and show normal to severely impaired speech detection and pure tone thresholds. It has been shown that “auditory neuropathy” affects an individual’s ability to process rapidly changing acoustic signals, known as auditory temporal processing.

The range of functional hearing abilities in individuals with “auditory neuropathy” is vast. Some individuals experience little or no difficulty hearing and understanding despite abnormal auditory test results. Others complain of “hearing but not understanding, especially in background noise.” Some individuals demonstrate fluctuant hearing abilities, reporting “good hearing days” and “bad hearing days.” Finally, some children and adults with “auditory neuropathy” are functionally deaf. For infants and young children, the deleterious effect of “auditory neuropathy” on language development and academic achievement can be significant.

Audiological management and speech and language intervention for infants and young children with this disorder is challenging. Because the range of functional hearing ability in “auditory neuropathy” is so great, each child with this diagnosis is unique. Furthermore, because the developmental consequences of “auditory neuropathy” cannot be predicted on the basis of auditory test results obtained in infants, guidelines that exist for identification and management of infants and young children with “typical” sensorineural hearing loss (SNHL) do not entirely fit the special needs of infants with “auditory neuropathy.”

To meet the need of the audiologists and other clinicians for guidance in identification and management of infants and young children with “auditory neuropathy,” these guidelines were formulated by an expert panel of audiologists, hearing scientists, and physicians to reflect contemporary practice. This document is not intended to duplicate or replace current guidelines for identification and management of children with “typical” SNHL, but rather seeks to supplement these existing documents with recommendations specific to infants and young children

with “auditory neuropathy.” As new information emerges, new techniques and strategies will undoubtedly evolve. In the interim, these guidelines for identification and management of children with “auditory neuropathy” offer practical guidance to audiologists and other clinicians, and families.

TERMINOLOGY

The term, auditory neuropathy, was originally proposed by Starr and colleagues (Starr et al., 1996) to describe the specific auditory disorder in a series of 10 patients, eight of whom demonstrated evidence of generalized peripheral neuropathy. The auditory disorder was characterized by evidence of normal cochlear outer hair cell function (preservation of otoacoustic emissions and cochlear microphonics) and abnormal auditory pathway function beginning with the VIII nerve (absent or severely abnormal auditory brainstem potentials).

Some investigators (Berlin et al., 2001a; 2001b; Rapin and Gravel, 2003; 2006) have expressed dissatisfaction with the term auditory neuropathy because the constellation of test results defining this disorder does not provide direct evidence of auditory nerve dysfunction or “neuropathy.” Indeed, only a subset of individuals with this disorder will be found to have abnormal auditory nerve function. Other lesions, for example, mutation of the otoferlin (OTOF) gene, which results in synaptic dysfunction at the junction of the inner hair cell/auditory nerve, will produce the same constellation of auditory test results in affected individuals (Yasunaga et al., 1999; Yasunaga et al., 2000). To address this, and other concerns, Berlin and colleagues (2001a; 2001b) proposed the term “auditory dys-synchrony.”

To address the potential confusion that arises from multiple designations for this disorder, the panel sought to identify simplified terminology that would unify the concept of an *auditory disorder* with a *range of presentations* secondary to a variety of etiologies. The panel considered multiple suggestions proposed by both panel and audience participants, and concurred that the most appropriate designation was “auditory neuropathy spectrum disorder” (ANSO). Three principle factors drove this consensus. First, despite potentially inexact usage, the term “auditory neuropathy” has gained wide-spread acceptance, both in the professional literature and among parent/consumer organizations. Renaming the disorder could lead to confusion for patients and professionals whereas retaining current terminology would provide continuity for the lay and scientific communities. Second, the expression of this disorder in everyday listening and communication behaviors encompasses a spectrum ranging

from limited or mild effects (complaints of difficulty “hearing” in noisy listening conditions) to profound effects (inability to “hear” in any listening condition, functionally “deaf”). Finally, the term “spectrum” was felt to expand the concept of this disorder to include sites of lesion other than the auditory nerve.

Starr and his colleagues (Starr et al., 2004) suggested segmenting the term auditory neuropathy into types, e.g. Type I (Pre-synaptic), Type II (Post-synaptic). In 2008, Starr and colleagues (Starr et al., 2008) proposed refining the terminology by site of disorder. For example, if the auditory nerve was involved but the inner hair cells and synapses were spared, the disorder would be classified as “auditory nerve disorder.” Similarly, if the inner hair cell synapses were disordered but the auditory nerve was normal, then the term “auditory synaptic disorder” would be appropriate. Currently, there are no clinical measures to distinguish site of disorder with this degree of precision. The panel concurred that subtypes or site-specific classification would be helpful to define the disorder more specifically, and that future research efforts should be directed to develop such a classification system.

DIAGNOSTIC CRITERIA

ANSD is characterized by evidence of normal or near normal cochlear hair cells (sensory) function and absent or abnormal auditory nerve function. Therefore, the (minimum) test battery needed to diagnose ANSD requires tests of cochlear hair cell (sensory) function and auditory nerve function.

Minimum Test Battery Required to Diagnose Individuals with ANSD:

1. Tests of cochlear hair cell (sensory) function:
 - a. Otoacoustic emissions (OAEs) for outer hair cell function: Standard screening or diagnostic protocol using Transient-Evoked OAEs (TEOAEs) or Distortion Product OAEs (DPOAEs), and/or
 - b. Cochlear microphonics: Click-evoked auditory brainstem response (ABR) to high-level click stimuli (80-90 dB nHL), tested with positive and negative polarity clicks in separate trials, through insert earphones (Starr et al, 2001; Berlin et al, 1998). A trial run with the sound-delivery tube clamped should be used to differentiate between the CM and stimulus artifact (Rance et al., 1999).
2. Test of auditory nerve function:
 - a. Auditory brainstem response (ABR) to high-level click stimuli (80-90 dB nHL). To avoid misinterpreting cochlear microphonics as components of the ABR, responses to positive and negative polarity clicks must be obtained in separate trials as described above. CMs

will show a characteristic reversal in polarity with reversal in polarity of the stimulating click; ABR will show a constant polarity regardless of polarity of the click (Berlin et al., 1998).

Additional Tests Useful for Diagnosing Individuals with ANSD:

Middle ear muscle reflexes (acoustic reflexes) are absent or elevated in individuals with ANSD (Berlin et al., 2005). Because normative data on acoustic reflex thresholds in very young infants using high probe-tone frequencies (1000 Hz) have not been established, this procedure is not required to diagnose ANSD. Nevertheless, a complete test battery for ANSD should include middle ear muscle reflex testing whenever possible.

Suppression of otoacoustic emissions by contralateral noise is abnormal in individuals with ANSD (Hood et al., 2003). Although this test has not gained widespread clinical usage, it is a potential candidate for further diagnostic studies in individuals with reliably recorded OAEs.

Special Considerations in Diagnosing Infants with ANSD:

Conventionally-recorded distortion product and transient OAEs are usually normal or near normal in individuals with ANSD. In newborns and very young infants, measurement of OAEs may be compromised by presence of residual fluid in the ear canal/middle ear (Doyle et al., 2000) or otitis media with effusion (OME). OAEs may be present initially and disappear over time in individuals with ANSD (Starr et al., 1996). Loss of OAEs, however, does not reflect change in auditory function or signal conversion of ANSD to typical SNHL.

Cochlear microphonics also provide a valid measure of hair cell function (see Cone in this volume for a discussion about the difference in generators of OAEs and CMs). CMs generally remain present in individuals with ANSD despite loss of OAEs (Starr et al., 1996). CMs are easily recorded from standard ABR recording protocols when insert earphones are used (Starr et al., 2001; Berlin et al., 2003). Stimulus artifact precludes effective recording of CMs when electromagnetic circumaural earphones are used (Stone et al. 1986; Berlin et al., 1998).

The auditory brainstem response (ABR) is markedly abnormal in individuals with ANSD. Recordings might appear as 1) a “flat” ABR with no evidence of any peaks or 2) presence of early peaks (waves up to III) with absence of later waves or 3) some poorly synchronized but evident later peaks (wave V) that appear only to stimuli at elevated stimulus levels.

When using these test procedures in newborns and very young infants, recording conditions must be optimum to obtain valid, artifact-free, unambiguous test results. Infants should be quietly sleeping in either

natural or sedated sleep to avoid movement artifact or “noisy” recordings. Caution should be used in interpretation of results when these tests are used in infants below 36 weeks gestational age. Repeated measures, over several weeks or months, are recommended to determine the reliability of test results. Because “transient” ANSD has been reported in some infants (Madden et al., 2002; Psarommatidis et al., 2006; Attias and Raveh, 2007), frequent monitoring by the ANSD test battery is recommended to establish the stability of test results, especially in the first two years of life.

Once the diagnosis of ANSD has been established, the infant should be referred for comprehensive medical, developmental, and communication assessments.

RECOMMENDED COMPREHENSIVE ASSESSMENTS

Many of the assessments recommended for infants with ANSD are similar to assessments recommended for infants with SNHL. (JCIH, 2007). The recommended assessments for infants with ANSD include:

1. Pediatric and developmental evaluation and history,
2. Otologic evaluation with imaging of the cochlea and auditory nerve (computed tomography, CT, and magnetic resonance imaging, MRI),
3. Medical genetics evaluation,
4. Ophthalmologic assessment,
5. Neurological evaluation to assess peripheral and cranial nerve function, and
6. Communication assessment.

Although not routinely recommended for infants and young children, vestibular assessment should be considered if developmental or otologic evaluation identifies potential vestibular disorder (e.g., nystagmus, delay in walking).

There are three principle reasons for infants with auditory disorders, including infants with ANSD, to receive comprehensive medical, developmental, and communication assessments. First, defining etiology of ANSD is important for predicting if the condition may be transient or is permanent (Madden et al., 2002; Psarommatidis et al. 2006 Attias and Raveh, 2007), determining if medical or surgical treatment is needed, and answering parent’s questions about cause of their infant’s hearing disorder. Second, because infants with ANSD, especially those who received care in the NICU, are at-risk for additional disabilities, early identification of developmental delays is important for optimum child development. Third, infants with ANSD may develop additional cranial or peripheral neuropathies secondary to a specific diagnosis (Starr et al., 1996).

ANSD may be unilateral or bilateral. The possibility of cochlear nerve

deficiency (absent or small cochlear nerves) should be considered for all children with ANSD, and especially for well-babies with unilateral ANSD and no medical history related to ANSD (Buchman et al., 2006) or infants with unilateral craniofacial anomalies (Carvalho et al., 1999). Contemporary imaging procedures (MRI and/or CT) are useful in these patients to assess integrity of the eighth nerve and internal auditory meatus.

Families of young infants benefit from early referral for communication assessment. Speech-language pathologists and deaf educators with expertise in early communication development can counsel families about the developmental sequence of pre-language, communicative behaviors, and support families in developing language-rich environments. Speech-language pathologists, deaf educators, and early intervention specialists can also help families monitor their infant’s language development and assist families in evaluating the effectiveness of their chosen language development strategy.

RECOMMENDED AUDIOLOGICAL TEST BATTERY

The audiological test battery recommended for assessing functional hearing and monitoring auditory development in infants and toddlers with SHNL (JCIH, 2007) is appropriate for infants and toddlers with ANSD. This test battery consists of measures of middle ear function, behavioral response to pure-tones, and speech reception and speech recognition. These measures include:

1. Otoscopic examination and acoustic immittance measures of middle ear function. As with any infant, infants with ANSD may develop middle ear dysfunction and otitis media with effusion resulting in mild conductive hearing loss. Because middle ear muscle (acoustic) reflexes are absent or elevated in individuals with ANSD, otoscopy and tympanometry will be most useful for identifying infants with middle ear dysfunction.
2. Behavioral assessment of pure-tone thresholds using developmentally-appropriate, conditioned test procedures such as visual reinforcement audiometry (VRA), or conditioned orientation reflex (COR) audiometry. For very young or developmentally-delayed infants, behavioral observation audiometry (BOA) may be used to observe the infant’s reflexive response to sound, however, results should not be interpreted as representing behavioral thresholds or minimal response levels.
3. Speech reception and speech recognition measures. For very young infants, response threshold to repetitive consonant-vowel combinations (e.g., ba-ba, ga-ga) is appropriate; for toddlers, pointing to body parts may yield acceptable speech threshold results. As children’s vocabulary develops, speech recognition measures using standardized picture-pointing (e.g., Word Intelligibility by Picture Identification, WIPI {Ross and Lerman, 1970}; Early Speech Perception Test {Moog and Geers, 1990}) or open-set tests should be

employed. Standardized taped materials are preferable to live-voice presentation to obtain consistency of stimuli across test sessions and should be employed once children are old enough to repeat recorded materials. Because ANSD can significantly affect speech understanding in background noise, tests of speech recognition in noise or competing messages should be conducted as soon as developmentally appropriate.

5. Otoacoustic emissions utilizing either TEOAEs and/or DPOAEs. Although initially present, OAEs may disappear in individuals with ANSD (Starr et al., 2001; Deltenre et al., 1999).

Obligatory cortical auditory evoked potentials to speech or speech-like signals are not yet a standard clinical measure for infants or toddlers. These measures show promise, however, as objective clinical tools for predicting speech recognition performance in young children with ANSD (Rance et al., 2002; Cone-Wesson et al., 2003; Pearce et al., 2007).

Infants and young children with ANSD should receive frequent audiological evaluation to assess their behavioral response to sound and auditory development. Some youngsters with ANSD will experience fluctuations in detection thresholds for pure-tones (Starr et al., 1996; Rance et al., 1999; Rance et al., 2002). For children who demonstrate consistently elevated pure-tone thresholds, amplification should be considered to improve audibility of speech.

RECOMMENDED AMPLIFICATION STRATEGIES

For infants with typical SNHL, hearing aid fitting can proceed in the earliest months of life based on electrophysiological estimates (e.g., click ABR, ABR to tone bursts, and/or auditory steady state response) of hearing sensitivity. For infants with ANSD, however, electrophysiological methods do not predict auditory detection thresholds. Clinicians and parents must rely upon the infant's or young child's behavioral response to sound to guide the hearing aid fitting decision. If an infant or young child with ANSD demonstrates elevated pure-tone and speech detection thresholds with consistent test-retest reliability, hearing aid fitting should be considered and a trial use of hearing aids should be offered to families.

Hearing aid fitting strategies for children with ANSD should follow established guidelines for the fitting of amplification in infants and toddlers (The Pediatric Working Group of the Conference on Amplification for Children with Auditory Deficits, 2001; American Academy of Audiology Pediatric Amplification Protocol, 2003). Special considerations for infants and young children with ANSD include:

1. Infants and young children with ANSD should be fitted with amplification as soon as ear-specific elevated pure-tone and speech detection thresholds are demonstrated by conditioned test

procedures (VRA or COR, see above). "Thresholds" or minimum response levels obtained by these techniques should be used to set amplification targets.

2. Significant improvement in auditory function, including "recovery" from ANSD, has been reported in some infants with this diagnosis ((Madden et al., 2002; Psarommatis et al., 2006; Attias and Raveh, 2007)). Careful monitoring of infant's auditory function by ABR and behavioral response by conditioned test procedures is required to adjust and modify amplification as needed. Although some risk factors for "transient" ANSD have been identified ((Madden et al., 2002; Psarommatis et al., 2006; Attias and Raveh, 2007)), at the present time, all infants and young children with ANSD, regardless of presumed etiology, should be carefully monitored for changes in auditory function and behavioral response to sound.
3. For infants with developmental delay where conditioned test procedures are unsuccessful, amplification fitting may proceed using behavioral observation of auditory behaviors and/or cortical evoked potentials when a) indications of auditory sensitivity are clearly outside developmental norms until more reliable measures can be obtained, and b) generally not before 6 months of age.

Temporal processing, or encoding the temporal characteristics of speech, is affected in individuals with ANSD (Zeng et al., 1999; Rance et al., 2004) resulting in a disproportionate loss in speech understanding ability relative to the individual's pure-tone thresholds (Starr et al., 1996; Rance et al., 1999; Rance et al., 2002). Although conventional hearing aids improve sound audibility, they do not resolve temporal processing deficits. Therefore, children with ANSD may not experience the same benefits from hearing aids expected from children with typical SNHL in whom temporal processing is relatively unaffected. Parental observation by formal questionnaire or survey (e.g., Infant-Toddler Meaningful Auditory Integration Scale, IT-MAIS {Zimmerman-Phillips et al., 2001}) may be helpful for assessing amplification benefit. In addition, speech recognition testing, including speech-in-noise or competing messages, should be incorporated into the hearing aid monitoring protocol as soon as developmentally appropriate for the child.

Strategies to improve the signal-to-noise ratio for children with ANSD should, theoretically, improve speech recognition and language learning (Hood et al., 2003). Trial use of an FM system, especially in structured and spontaneous language-learning activities, should also be considered for children with ANSD.

SPECIAL CONSIDERATIONS FOR COCHLEAR IMPLANTATION

Despite an adequate trial with appropriately-fitted amplification, some children with ANSD may demonstrate poor progress in speech

understanding ability and aural/auditory language development. For these children, cochlear implantation should be considered, regardless of behavioral audiometric thresholds.

In addition to standard cochlear implantation criteria for children, special considerations for cochlear implantation in children with ANSD include:

1. As noted above, significant improvement in auditory function, including “recovery” from ANSD has been reported in a subset of infants with this diagnosis. Families should be informed that spontaneous improvement in auditory function has been reported up to two years of age. Cochlear implantation, therefore, should not be considered until auditory test results (ABR and estimates of behavioral sensitivity) are stable and demonstrate unequivocal evidence of permanent ANSD (no change in or recovery of ABR). Deferring the decision for cochlear implantation until age two years may be appropriate. All infants with ANSD, including those being monitored for possible recovery, should be enrolled in early intervention and language stimulation programs to prevent delay in language acquisition.
2. Evidence of auditory nerve sufficiency should be obtained prior to surgery using appropriate imaging technology (Buchman et al., 2006).
3. Children with ANSD who do not demonstrate good progress in speech recognition ability and language development should be considered candidates for cochlear implantation *regardless of audiometric thresholds*. Children in this category with elevated pure-tone and speech detection thresholds should receive a trial of amplification fitted by pediatric amplification guidelines prior to consideration for implantation.

Emerging data suggest that pre-implantation electrical stimulation testing may be useful in determining CI candidacy in some cases (Gibson et al., 2007). At the present time, pre-implantation electrical stimulation is not a requirement for implantation.

Cochlear implants offer the possibility of improving auditory temporal processing by stimulating synchronous discharge of the auditory nerve. For example, ABR, which requires neural synchrony, can be electrically-evoked in many individuals with cochlear implants (Peterson et al., 2003; Shallop et al., 2003). Furthermore, speech recognition ability, which is strongly dependent on temporal processing ability, is similar in many cochlear-implant users with ANSD to speech recognition ability measured in cochlear implant users with typical SNHL (Madden et al., 2002, Mason et al., 2003; Rance and Barker, 2008). For families who wish to consider cochlear implantation for their child with ANSD, referral to a center with experience with managing children with this diagnosis is strongly encouraged.

RECOMMENDED HABILITATION FOR COMMUNICATION DEVELOPMENT

Families of infants with ANSD should be informed that their baby’s auditory capacity or speech, language, and communication development cannot be predicted on the basis of the initial evaluation. Ongoing monitoring of their infant’s auditory, speech, language, communication, and general development is essential. As with all infants and children with hearing loss (JCIH, 2007), families should be made aware of all communication options presented in an unbiased manner. Informed family choice and desired outcome guide the decision-making process. For most children with ANSD, use of any combination of communication systems that incorporates visual support is appropriate (e.g., auditory/aural with lipreading and natural gesture, cued speech, total communication, sign language). Decisions regarding mode of communication must ultimately be made by the family and respected by all professionals involved.

Infants with this diagnosis should receive referral to early intervention programs that assess the language, cognitive skills, auditory skills, speech, vocabulary, and social-emotional development of children at six month intervals during the first three years of life. Appropriate assessment tools include those that have been standardized on children with normal hearing and norm-referenced assessment tools that are appropriate to measure progress in verbal and visual language (JCIH, 2007).

SCREENING NEWBORNS FOR AUDITORY NEUROPATHY SPECTRUM DISORDER

The panel concurred with the Joint Committee on Infant Hearing 2007 Position Statement in which the definition of targeted hearing loss was expanded to include “neural hearing loss” in infants admitted to the NICU. Because screening by OAEs will fail to detect infants with “neural hearing loss” or ANSD, the panel further concurred with the JCIH recommendation that infants who receive care in the NICU for five days or more receive hearing screening by ABR.

Screening well-babies for ANSD is more problematic. In many well-baby nurseries, the hearing screening protocol is screening by OAEs. Although this technology will detect infants with sensory hearing loss, it will “pass” infants with ANSD. Even if the nursery uses a “two-stage” protocol, e.g., OAEs followed by automated ABR for those infants who “fail” OAE screening, infants with ANSD will not receive the second, automated ABR screening because they “passed” OAE screening. In those well-baby nurseries where automated ABR is the first screening technology, infants who fail this test should not be rescreened by OAEs and “passed” because these infants may have ANSD.

Because the probable cause of ANSD in well-babies is genetic, infants with a family history of childhood hearing loss or sensory motor neuropathy should receive hearing screening by ABR.

As more information becomes available on the prevalence of ANSD in the well-baby population, stronger recommendations for screening all infants for ANSD, regardless of nursery care level, may emerge.

For infants who “pass” newborn hearing screening, subsequent parent or caregiver concern about the child’s auditory, speech, or language development should trigger a referral for audiological assessment including behavioral pure-tone and speech threshold measures, speech recognition testing (as developmentally appropriate), and tympanometry and middle ear muscle reflexes. Re-screening these infant’s or young children’s hearing with OAEs is not sufficient because such re-screening will “pass” infants and young children with ANSD.

MONITORING INFANTS WITH “TRANSIENT” ANSD

Some infants with an initial diagnosis of ANSD may demonstrate improved auditory function and even “recovery” on ABR testing (Madden et al., 2002; Psarommatas et al., 2006; Attias and Raveh, 2007). For those infants who “recover” from ANSD, the panel recommends regular surveillance of developmental milestones, auditory skills, parental concerns, and middle ear status consistent with the Joint Committee on Infant Hearing 2007 Position Statement (JCIH, 2007). Because the residual effects of transient ANSD are unknown, ongoing monitoring of the infant’s auditory, speech, and language development as well as global (e.g., motor, cognitive, and social) development is critical. Those infants and young children whose speech and language development is not commensurate with their general development should be referred for speech and language evaluation and audiological assessment.

The Joint Committee on Infant Hearing recognizes sensory motor neuropathies such as Friedreich ataxia and Charcot-Marie-Tooth syndrome as risk indicators for delayed onset hearing loss (JCIH, 2007). Per the Joint Committee’s recommendation, infants with a risk indicator should be referred for an audiological assessment at least once by 24 to 30 months of

age. Given the possibility of late onset ANSD in infants with family history of sensory motor neuropathies, audiological assessment including ABR, OAEs, tympanometry and middle ear muscle reflexes is warranted.

COUNSELING FAMILIES OF INFANTS WITH ANSD

Counseling families of infants and young children with ANSD is one of the greatest challenges associated with this disorder. Because the developmental effects of ANSD cannot be predicted from test results obtained in the earliest months or even years of life, families struggle with the uncertainty of what the diagnosis means relative to their infant’s growth and development. Many infants with ANSD have had difficult perinatal or neonatal courses with complications including prematurity, birth asphyxia, infections, or other conditions requiring neonatal intensive care. The significance of the ANSD diagnosis may be difficult for families to appreciate as they struggle to understand their infant’s complex medical and developmental needs. Strong support systems, including parents of children with similar diagnoses and professionals with expertise in clinical social work and family counseling, should be available to meet the ongoing and changing needs of families.

Clinicians working with infants and young children with ANSD and their families must remain flexible in approaching rehabilitative options. All members of the team, including the family, should be encouraged to question specific methodologies and strategies if the child’s language and communication development is not commensurate with his or her developmental potential.

Children with ANSD can develop into healthy and dynamic citizens with happy personal lives, successful academic experiences, and satisfying careers. Clinicians should help families realize this goal by identifying and supporting the unique strengths and abilities of the child and family.



Participants in the “Guidelines Development Conference on the Identification and Management of Infants and Young Children with Auditory Neuropathy” included (from left): Kai Uus, Barbara Cone, Yvonne Sininger, Patricia Roush, Deborah Hayes, Charles Berlin, Ferdinando Grandori, and Jon Shallop. Not pictured: Gary Rance, Arnold Starr, and Christine Petit.

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