



**Tennessee Department of Health**

# **Newborn Hearing Program**



## **Pediatric Audiology Assessment & Amplification Guidelines**

**EHDI**



**The Early Hearing Detection and  
Intervention Program**

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## INTRODUCTION

The goal of the Tennessee Department of Health Universal Newborn Hearing Screening Program (UNHS) for Early Hearing Detection and Intervention (EHDI) is to promote early screening, identification, and intervention of hearing loss utilizing existing Tennessee providers, agencies and organizations, and to:

- assure all newborns receive hearing screening using physiologic measures prior to discharge after birth or before 1 month of age;
- assure all infants referred for further hearing testing receive audiologic evaluation prior to 3 months of age and, upon diagnosis, are immediately referred to an otolaryngologist for evaluation and, if appropriate, medical clearance; and,
- assure all infants identified with a hearing loss receive appropriate and necessary intervention prior to 6 months of age.

As noted by Matkin (1998), these benchmarks must be balanced with the reality that issues related to medically-compromised/fragile infants, individual coping styles, family-systems, and cultural differences, may not always permit adherence to these guidelines.

The Tennessee UNHS program is committed to assuring families have access to audiology providers who demonstrate the knowledge and skills necessary to provide current pediatric hearing assessment methods as outlined in documents from the American Speech-Language-Hearing Association, ASHA, and the American Academy of Audiology, AAA (ASHA Guidelines for the Audiological Assessment of Children from Birth to 5 Years of Age, 2004; AAA Pediatric Amplification Protocol and the Exposition on Cochlear Implants in Children, 2003; AAA Cochlear Implant Guidelines, 1995).

The following recommended guidelines were most recently modified by the Tennessee Pediatric Audiology Guideline Committee convened by the Tennessee Newborn Hearing Screening Task Force. Members of the working group responsible for the updated guidelines included the following pediatric audiologists: Roxanne Jennemann Aaron, Julie Beeler, Aimee Biddle, Jan Dungan, Mary Edwards, Linda Gemayel, Beth Humphrey, Jennifer Pepper, Erin Plyler, Wendy Richardson, Anne Marie Tharpe, and Kelly Yeager. A student panel also participated in the review process. This panel included: Lynzee Alworth, Heather Porter, and Lindsey Rentmeester. Members representing other health care fields included: Jacque Cundall, John Phillips, and Carmen Lozzio (See Appendix 1).

These guidelines are in place for the purpose of advancing an effective statewide system for assessing the hearing of infants and young children, birth to five years of age. As members of the working groups updated each section of the guidelines, full consideration was given to the most recent position statement from the Joint Committee on Infant Hearing (JCIH), released in 2007 and published by the American Academy of Pediatrics (AAP). With that said, these guidelines are meant to facilitate the diagnosis of hearing loss prior to 3 months of age, obtain medical clearance for amplification from an otolaryngologist, and implement amplification and

early intervention as recommended no later than 6 months of age. While these guidelines strictly adhere to the “1-3-6” recommendations included in the JCIH ’07 statement, there are other areas of the JCIH statement that members of the working groups decided to modify, with the opinion that their recommendations were in the best interest of babies and young children in the state of Tennessee.

The guidelines are informational only and are not intended or designed as a substitute for the reasonable exercise of independent clinical judgment by audiologists, physicians and other medical providers. They can be used to form an approach to care that is unique to the needs of each individual child.

The Tennessee Department of Health Universal Newborn Hearing Screening Program (NHS) for Early Hearing Detection and Intervention (EHDI) is supported by funding through the Health Resources Services Administration (HRSA) grant for Universal Newborn Infant Hearing and Intervention (CFDA 93.251) and through the Centers for Disease Control and Prevention (CDC) grant for Early Hearing Detection and Intervention (EHDI) (CFDA93.283).

## **PEDIATRIC AUDIOLOGIC ASSESSMENT GUIDELINES**

The following Pediatric Audiologic Assessment Guidelines have been adopted from those developed by the American Speech-Language-Hearing Association (ASHA, 2004). A panel of nationally recognized experts in audiology developed the ASHA Guidelines for the Audiological Assessment of Children from Birth to 5 Years of Age. Some sections were adapted from the original guidelines have been made in acknowledgement of changes in the knowledge base in the field of audiology and needs specific to the state of Tennessee.

### **Primary Purpose Statement**

Infants and young children suspected of having a hearing loss should receive appropriate medical and audiologic evaluations as well as intervention services in a timely, efficient manner. Suspicion of hearing loss may occur as a result of any one of the following factors:

- 1) failure of the newborn hearing screening (NHS);
- 2) risk indicators for hearing loss (per the Joint Committee on Infant Hearing 2007 JCIH); or
- 3) expressed concern from parents, caregiver, family, or the child’s medical home provider.

Additionally, any infant or young child demonstrating a delay in speech/language development, regardless of prior hearing result, should also be evaluated. All infants who do not pass the NHS and any subsequent re-screening should receive appropriate audiologic evaluations to confirm the presence of hearing loss by three months of age. (Appendix 2 -Joint Commission on Infant Hearing 2007 Position Statement-Risk Indicators for Progressive and Delayed Onset or Acquired Hearing Loss).

When a hearing loss is diagnosed, family members should be notified and informed of intervention options. A family-centered and culturally-sensitive approach that advocates

involvement of the family to the fullest extent they desire should be maintained throughout the diagnostic and intervention process.

This document should be regarded as best practice guidelines, not standards. Each child presents unique individual characteristics, shaped by familial roles and culture that may influence an approach to the assessment and intervention process.

### **Professional Competency**

These best practice guidelines are intended for audiologists who serve infants and young children suspected of having a hearing loss. Therefore, it is assumed that clinicians considering these guidelines are familiar with specific audiologic tests. The guidelines are not intended to be a tutorial on test method or to provide specific protocols for individual test procedures. Other professional documents, literature, and web materials are available for such purposes. Rather, these guidelines are intended to delineate the specific technologies, skills, and knowledge that are considered fundamental to the provision of comprehensive audiologic services to infants, toddlers, and children to five years of age. Additionally, audiologists should be knowledgeable about federal and state laws and regulations impacting the identification, intervention and education of children who are deaf and hard of hearing.

Practitioners providing audiologic assessment and intervention services to this specialized pediatric population are expected to follow their professional code of ethics regarding their ability to provide such services. These audiologists must have the commensurate knowledge, skill and instrumentation necessary for use with current pediatric hearing assessment methods. Pediatric audiologists should also be knowledgeable about resources available within their region and be able to make appropriate referrals for the children they assess and their families.

Audiologists are the professionals singularly qualified to select and fit all forms of amplification for infants and young children. These include personal hearing aids, frequency-modulation (FM) systems, cochlear implants and other types of assistive listening devices.

### **Equipment/Facilities**

In order to obtain reliable and accurate measures of auditory function, the test facility should have all the proper equipment and personnel to provide comprehensive physiologic and behavioral audiologic evaluations, including sedated testing as needed. Facilities that lack appropriate equipment or personnel to perform the selected tests should establish collaborative arrangements with those that do. (Pediatric Working Group, 1996).

### **American National Standards Institute (ANSI) Standards**

All measurements of auditory function (behavioral and physiologic) must be completed in a test environment that meets current ANSI standards for background noise levels. Equipment must be maintained according to the manufacturer's specifications and recommendations and calibrated to comply with current ANSI standards. Daily listening checks are particularly important when working with the pediatric population. Documentation of listening checks and periodic electroacoustic calibration should be consistently maintained. When national

standards do not exist, as in the case with transient signals used in evoked potential testing or in sound field audiometry, calibration may be referenced to other published standards, to published data, or to values established by the clinic performing the audiologic tests. Appropriate sound field calibration is particularly critical in the behavioral audiologic assessment of children who cannot be tested under earphones or with insert phones (Morgan, Dirks, & Dower, 1979; Rochlin, 1990; Walker, Dillon, & Byrne, 1984).

### **Joint Commission on Accreditation of Healthcare Organizations**

Audiologists working in facilities accredited by the Joint Commission on Accreditation of Healthcare Organizations (JCAHO) must adhere to the standards encompassing patient contact. (JCAHO, 2002).

### **Universal Precautions**

All procedures must ensure the safety of the patient and clinician, and adhere to universal health precautions (e.g., prevention of bodily injury and transmission of infectious disease). Decontamination, cleaning, disinfection, and sterilization of multiple-use equipment before reuse must be carried out according to facility-specific infection control policies and procedures and according to manufacturer's instructions (ASHA, 1997; Centers for Disease Control, 1988). Handwashing prior to and at the completion of an evaluation is essential to deterring cross contamination between patients

### **Moderate Sedation**

To gain the cooperation of some infants and young children during physiologic assessments of auditory function, sedation may be required. Yet, sedation of pediatric patients has serious associated risks such as hypoventilation, apnea, airway obstruction, and cardiopulmonary impairment. As such, sedative medications should only be administered by or in the presence of individuals skilled in airway management and cardiopulmonary resuscitation. Additionally, the oversight by skilled medical personnel and the availability of age- and size-appropriate equipment, medications, and continuous monitoring are essential during procedures and in rescuing the child should an adverse sedation event occur.

The Joint Commission on Accreditation of Healthcare Organizations has adopted revisions to its anesthesia care standards (JCAHO, 2002), consistent with the American Society of Anesthesiologists (ASA) standards (2000). The most current terminology of the American Society of Anesthesiologists has replaced the term "conscious sedation" with the term "moderate sedation".

## AUDIOLOGIC ASSESSMENT PROCEDURES

### **Audiologic Assessment Procedures**

Audiologic assessment of infants and young children includes a thorough case history, otoscopy, behavioral, and physiologic measures. Because children undergo rapid sensory, motor, and cognitive development, and because some children will present with multiple developmental problems, it is vital that assessment tools are appropriate for the neurodevelopmental state of the child. In addition to the assessment of peripheral hearing status, it is essential for audiologists working with infants and young children to consider the functional implications of hearing loss. As is feasible within the time constraints of clinical practice, assessments of speech perception ability, and screening for communication skills, cognitive development, and social-emotional status should be included as part of the pediatric test battery. Such assessments and screenings are consistent with the objective of formulating recommendations and making additional referrals as needed.

A thorough assessment of hearing may require multiple sessions. As such, serial evaluations may be necessary to develop reliable profiles of hearing status and developmental abilities. Prolonged delays between assessments should be avoided. During the assessment process, the audiologist may be formulating a working diagnosis of the child's audiologic status while developing and perhaps, implementing initial management options.

Ear-specific assessment is the goal for both behavioral and physiologic procedures because a unilateral hearing loss, even in the presence of a normal-hearing ear, may place a child at significant developmental and/or educational risk (Bess, 1982; Bess, Klee, & Culbertson, 1988; Bovo et al., 1988; Oyler, Oyler, & Matkin, 1988). Therefore, determining hearing sensitivity for each ear is important for establishing supportive evidence for medical/surgical diagnosis and treatment, selecting amplification when appropriate, establishing baseline function, and monitoring auditory status when progressive, fluctuating, or late-onset hearing loss is suspected.

### **Case History**

The case history is particularly important because it will often guide the selection of a strategy for the audiologic evaluation. Moreover, accurate diagnosis of hearing loss relies on interpretation of a test battery within the context of the child's medical and/or developmental history. Case history information may suggest a need for modification of evaluation procedures. For example, the audiologist may want to include evaluation of the high-frequency region of the cochlea (above 4000 Hz) for a young child with a history of ototoxic drug exposure. Modification of routine assessment procedures also may be necessary when evaluating a child with multiple disabilities. The case history should be recorded using a standard form.

### **Otoscopy**

Several audiologic assessment procedures require the insertion of a probe into the external auditory canal. As such, a visual inspection of the outer ear canal should be conducted to verify

that there is no contraindication to placing a probe in the ear canal (e.g., drainage, foreign objects, occluding cerumen, atresia).

### **Acoustic Immittance**

Acoustic immittance measures are an integral part of the pediatric assessment battery. Clinical decisions should be made based on a quantitative assessment of the tympanogram, including consideration of equivalent ear canal volume, peak compensated static acoustic admittance, tympanometric width or gradient, and tympanometric peak pressure. The components of the immittance test battery, alone or in combination, have been used for many years to evaluate middle ear function and to screen for middle ear effusion (ASHA, 1997).

The acoustic reflex may provide supplemental information relevant to the functional status of the middle ear, cochlea, and brainstem pathway. Together, these measures are fundamental components of the pediatric audiology test battery. For neonates and young infants, however, optimal clinical procedures for application of tympanometric and acoustic reflex measurements are not well defined (ASHA, 1994; McMillan, Bennett, Marchant, & Shurin, 1985; Sprague, Wiley, & Goldstein, 1985). Under the age of approximately 4 months, interpretation of tympanograms and acoustic reflex findings may be compromised when a conventional low-frequency (220-Hz or 226-Hz) probe tone is used (Paradise, Smith, & Bluestone, 1976).

### **Behavioral Assessment**

Behavioral assessment of hearing sensitivity in children is complicated by developmental and maturational factors. It is now known that unconditioned behavioral observation techniques with infants are confounded by poor test re-test reliability, and high inter- and intra-subject variability (Bench, Collyer, Mentz, & Wilson, 1976; Weber, 1969; Wilson & Thompson, 1984). As such, Behavioral Observation Audiometry (BOA) is not recommended for estimating infant hearing thresholds.

Several studies have shown that once an infant reaches a developmental age of 5–6 months, it is possible to elicit reliable conditioned auditory responses using an operant, visually-reinforced behavioral response technique (Moore, Wilson & Thompson, 1977; Primus & Thompson, 1985; Thompson & Wilson, 1984; Thompson, Wilson, & Moore, 1979; Wilson, 1978; Widen, 1993). Typically-developing children as young as 5 months, may be conditioned to produce a motor response contingent upon the presence of an auditory stimulus (Wilson & Thompson, 1984). The behavior, usually a head turn, is reinforced by an appealing visual display. More recent studies confirm that frequency-specific thresholds may be obtained from infants at developmental levels of 5–6 months, enabling accurate evaluation of hearing sensitivity regardless of type, degree, or audiometric configuration (Bernstein & Gravel, 1990; Diefendorf, 1988; Gravel, 1989; Nozza & Wilson, 1984; Gravel & Wallace, 1999; Diefendorf, 2003; Widen et al., 2000). It has also been shown that for infants 5-6 months to 24 months of age (and children up to 4 years of age), tangible reinforcement operant conditioning audiometry (TROCA) or visually reinforced operant conditioning audiometry (VROCA) techniques can be effective. The basic paradigm used in the tangible reinforcement operant conditioning audiometry (TROCA) or visually reinforced operant conditioning audiometry



(VROCA) techniques involve a bar press response coupled with either tangible or visual reinforcement. (Wilson & Thompson, 1984; Diefendorf, 1988). From approximately 25 to 30 months, conditioned play audiometry (CPA) is recommended. When children are taught to perform play audiometry, it is usually not difficult to select a response behavior that they are capable of performing. The challenge in play audiometry is teaching the child to wait, listen, and respond with the play activity only when the auditory signal is audible. From 25 to 30 months, CPA is sometimes possible within the time constraints of clinical activity (Thompson, Thompson, & Vethivelu, 1989). After the developmental age of 30 months, CPA is the method of choice. Because overlap exists among VRA, TROCA/VROCA, and CPA as suitable evaluation techniques, successful testing of a child ultimately depends on the observational skills, interpersonal skills, and experience of the audiologist.

### **Physiologic Assessment**

Physiologic assessment procedures are of particular importance in the audiologic assessment of young children. Measurement of auditory evoked potentials, especially the ABR, can provide accurate estimates of threshold sensitivity. As such, ABR plays an important role in both identification and assessment, particularly with children too young or developmentally delayed for reliable assessment using conditioned behavioral techniques (Stein & Kraus, 1985). Tympanometry should be used along with other physiologic assessments to ensure accurate diagnosis and management of the patient.

Subject characteristics and recording parameters are known to influence the ABR. Under good recording conditions, visual detection levels of wave V are usually within 10 dB of behavioral audiometric thresholds for click stimuli. Data from several studies provide normative data for ABR latencies for infants and children to 3 years of age (Gorga, Reiland, Beauchaine, Worthington, & Jesteadt, 1987; Gorga, Kaminski, Beauchaine, Jesteadt, & Neely, 1989).

It is recommended that frequency-specific stimuli be used when comprehensive auditory brainstem response (ABR) testing is undertaken. At a minimum, responses to low- and high-frequency stimuli should be obtained for each ear to estimate audiometric configuration. High-frequency assessment should be completed using a 2000 and 4000 Hz tone burst (Pediatric Working Group, 1996) and low frequencies should be assessed using a 250 Hz or 500 Hz tone burst (Stapells, Gravel, & Martin, 1995; Stapells & Oates, 1997) The use of click stimuli alone is not sufficient for the estimation of audiometric configuration (Stapells, 1995; Stapells & Oates, 1997; Balfour, Pillion, & Gaskin, 1998). Appropriate contralateral masking should be used when indicated.

When air conduction thresholds obtained by physiologic methods are found to be abnormal, estimates of bone conduction sensitivity should be completed (Mauldin & Jerger, 1979; Stapells, 1989; Stapells & Ruben, 1989; Yang, Rupert, & Moushegian, 1987; Ysunza & Cone-Wesson, 1987). However, there are output limitations using bone conduction and transient stimuli (approximately 50 dBnHL maximum output for clicks). If bone conduction is not done and latency information only is used, precipitously sloping high-frequency losses can be confused with conductive losses. Generally, ABRs obtained by bone conduction have longer

latencies with normal or near normal interpeak intervals (Gorga et al., 1993). It is important when doing bone conduction ABRs that attention is paid to ensure adequate pressure of the bone vibrator (Yang & Stewart, 1990) on the mastoid. Care also must be taken to separate the bone vibrator from the electrode due to electromagnetic leakage. For example, padding such as 4x4s or foam cushioning between the top of the head and bone conduction headband may help to ensure adequate pressure and placement of the bone vibrator. Alternative electrode placements such as the earlobe or tragus or the use of tiptrodes should be considered.

At this time, elimination of the click evoked ABR is not recommended as it can provide useful information regarding neural integrity. Assessment of interwave latencies, ear asymmetries, and morphology relative to age-appropriate norms may be completed as part of the ABR evaluation and the information used in the context of other clinical and/or medical findings. Children who present with abnormal ABR findings regardless of otoacoustic emissions (OAEs) should undergo further evaluation to differentiate between cochlear and neural dysfunction. When the ABR is absent or abnormal, response to both rarefaction and condensation click stimuli should be obtained to evaluate the presence of the cochlear microphonic (CM; Berlin et al., 1998). In these instances, precautions must be taken to distinguish the CM from stimulus artifact. For example, performing repeat measurements with the stimulus tube open vs. pinched should cause the CM waveform to disappear because no signal is reaching the cochlea to generate a CM. If the alternating current (AC) waveform remains, then it is stimulus artifact, which results from the electrical signal at the back of the transducer being picked-up by the recording electrodes and amplified. (Durrant & Ferraro, 1999).

Caution should be used when testing children with neurological issues such as abnormal brain pathology, seizures, or confirmed auditory dys-synchrony when interpreting the ABR evaluation. The ABR is a test of neural integrity to the level of the brainstem. What a patient does cortically with this information cannot be interpreted by the ABR. Children should continue to have behavioral assessments until reliable ear specific thresholds can be obtained if possible. The preferred method of reporting ABR responses is dBeHL (estimated hearing level) which is the corrected level of the response.

The Auditory Steady State Response (ASSR) is an auditory evoked potential test with emerging clinical applications. It holds promise as a method of estimating frequency specific hearing sensitivity in patients who cannot or will not provide reliable or valid behavioral thresholds (Cone-Wesson, Dowell, Tomlin, Rance, & Ming, 2002; Dimitrijevic et al., 2002; Vander Werff, Brown, Gienapp, & Schmidt-Clay, 2002). The accuracy of ASSR predictions of hearing sensitivity in infants and young children is an area of active interest at this time (Sininger, 2002). Some concerns about recording artifact under certain stimulus conditions have been expressed (Gorga et al., 2004; Small & Stapells, 2003); research in this area is ongoing and improvements in methodology are expected. As with all developing clinical procedures, audiologists are expected to monitor the literature for methodological improvements in ASSR. ASSR has proven useful clinically when ABR responses are absent. As the ABR may only be obtained to the limits of the equipment, ASSR can often be obtained at higher thresholds. If the ASSR thresholds are

obtained successfully, they may provide valuable information for more accurate fitting of amplification.

Otoacoustic emissions (OAEs) also expand the pediatric audiology test battery by providing a physiologic means of assessing preneural auditory function (Kemp, Ryan, & Bray, 1990; Norton & Widen, 1990; Gorga et al., 1993). The presence of OAEs is with normal outer hair cell function which may be consistent with normal or near-normal hearing thresholds in a given frequency region. Although relations exist between OAEs and behavioral thresholds (Martin et al., 1990; Gorga et al., 1996; 2002) and there has been improvement in strategies for predicting thresholds using OAEs (Boege & Janssen, 2002; Gorga et al., 2003b), variability among individuals suggest that caution should be exercised when attempting to predict behavioral thresholds from OAEs. Because OAEs are generated in the cochlea, they provide information that further defines auditory system integrity and sensitivity. Used in conjunction with ABR, OAEs are not only useful in the differential diagnosis of cochlear hearing loss but also in the identification of children with neurological dysfunction.

Transient evoked OAEs (TEOAEs) are elicited either following a click/transient stimulus (TEOAE) while distortion product OAEs (DPOAEs) are elicited following stimulation with two tones. TEOAEs typically are measured in response to a click at approximately 80 dB pSPL (78-82 dB SPL). Although the click stimulus is a broad-band stimulus that is not frequency specific, the response is analyzed in the frequency domain, thus providing information across frequencies from 500 to 5000 Hz, although test performance is best for mid-to-high frequencies. Probe fit can affect the spectrum of the click stimulus in the ear canal. The stimulus spectrum, as measured in the ear canal, should have equal intensities across the frequency range. However, in neonates, this cannot be achieved and the stimulus typically has more high-frequency energy (Norton et al., 2000). In common clinical practice, TEOAEs need to be present above the noise floor by at least 6 dB, and/or have a reproducibility of greater than an established percentage at defined frequencies. For example, Kemp et al., (1990) recommended a minimum of 50% reproducibility for determining response presence while Prieve et al., (1993) found 70% to be a reasonable expectation when coupled with an overall minimum amplitude (wideband) of 6 dB SPL. For narrow frequency bands, levels of 3 dB above background noise may give reasonable assurance of a TEOAE response for that frequency region alone (Norton et al., 2000). Hussain et al., (1998) provided an approach in which data from normal and from impaired ears were used to develop diagnostic criteria, thus explicitly taking into account the fact that responses from normal and impaired ears are not completely separated for any criterion value. It should be noted that in the presence of very low noise levels, a low-level TEOAE response could result in an OAE-to-noise ratio (SNR) that exceeds passing criteria. A diagnostic approach in which SNR is used to establish the reliability of the measurement, followed by a clinical decision based on response level might avoid diagnostic errors associated with very low noise levels.

DPOAEs are measured in response to two tones (primaries) that interact to produce non-linear distortions in the cochlea. DPOAEs are measured at the frequencies of the distortion product  $2f_1 - f_2$  for each stimulus tone pair. The stimulus tones are designated by  $f_1$  for the lower frequency tone,  $f_2$  for the higher frequency tone, and  $L_1$  and  $L_2$  for the lower and higher

frequency intensity levels, respectively. The two tones typically are selected so that the frequency ratio between the tones ( $f_2/f_1$ ) is 1.22, which is known to produce the largest ( $2f_1 - 2f_2$ ) distortion product at most test frequencies in humans. Data from several studies suggest that the primaries should be unequal and of a moderate level (e.g.,  $L_1/L_2 = 65/55$  dB SPL) to most accurately classify auditory status (e.g., Stover et al., 1996). Response presence can be determined by examining response level or by examining the response level relative to the noise floor (SNR). SNR has generally good performance for identifying ears with normal cochlear function, but because it depends on the level of the noise as well as OAE level, the same potential problem mentioned above regarding use of SNR with TEOAEs also exists for the DPOAE. Gorga et al., (1997) provided an interpretative approach for DPOAEs that is similar to the one described by Hussain et al., (1998) for TEOAEs. It recognizes the fact that there is no criterion value that will separate normal or impaired function without error. However, their approach provides a means for determining the level of confidence with which any measured response indicates normal or impaired hearing. In their application, SNR is used first to determine that a response was reliably measured. If the SNR indicates that a reliable response was measured, DPOAE level is then used to determine auditory status. Varying protocols of DPOAEs may be used depending on the clinical relevance. For example, a screening protocol may be used for infants whereas a more comprehensive, diagnostic protocol may be used for those exposed to ototoxic drugs.

Schemes for trying to determine the degree of hearing loss and/or for predicting thresholds using DPOAEs have been investigated (Martin et al., 1990; Gorga et al., 1996; Dorn et al., 2001; Gorga et al., 2002; Boege & Janssen, 2002; Gorga et al., 2003a). Although some strategies have met with success, variability is such that threshold predictions should be viewed cautiously. In some approaches, predictions of behavioral thresholds from DPOAE thresholds require the measurement of DPOAE levels for several stimulus levels (i.e., DPOAE input/output functions). It may be difficult to obtain these data routinely under some clinical conditions.

# **RECOMMENDED PEDIATRIC AUDIOLOGIC ASSESSMENT GUIDELINES FOR CHILDREN *BIRTH TO 60 MONTHS DEVELOPMENTAL AGE***

## **Purpose**

It is recommended that all infants who do not pass the newborn hearing screen and any subsequent rescreening receive a medical evaluation and comprehensive pediatric audiologic assessment to quantify hearing levels and, for those for which hearing loss has been confirmed, obtain medical clearance for amplification prior to 3 months of age. Auditory dysfunction can result from pathology at one or more sites within the auditory system. Furthermore, a test battery approach is highly indicated. A test battery approach that includes physiologic, behavioral and developmental measures is recommended as a gold standard. The following guidelines include physiologic and behavioral assessment recommendations, by developmental age, supporting the use of a test battery approach. It is recommended that all infants confirmed with a hearing loss receive intervention services prior to 6 months of age.

## **Introduction**

This document provides guidelines for the purpose of choosing developmentally-appropriate test measures for infants and young children ages 0-5 years. The child's neurodevelopmental age should be considered in the test battery selection. These guidelines are intended for use by qualified, audiologists experienced in working with children. The cross check principle is essential for accurate hearing assessment and requires a test battery approach to assessment..

## **Equipment/Facilities:**

In order to obtain reliable, accurate results, the test facility should have proper equipment and personnel to provide comprehensive physiologic and behavioral audiologic evaluations, including sedated testing as needed. Those facilities that lack appropriate equipment or personnel to perform the selected tests should establish consortial arrangements with those that do have appropriate equipment (Pediatric Working Group, 1996).

**The following best practice pediatric audiologic assessment guidelines are divided into three age groups:**

- **Guidelines for Infants 0-4 Months Developmental Age**
- **Guidelines for Infants 5-24 Months Developmental Age**
- **Guidelines for Toddlers and Preschoolers 25-60 Months Developmental Age**

## GUIDELINES FOR INFANTS 0-4 MONTHS DEVELOPMENTAL AGE

### Case History

- Review newborn hearing screening results
- Identify risk indicators for progressive and delayed onset or acquired hearing loss

### Otoscopy

- The purpose of otoscopy is to ensure that there are no contraindications to placing an insert earphone or probe in the ear canal.
- Visual inspection for obvious structural abnormalities (i.e., ear pits, ear tags, atresia, low set ears) of the pinna and/or ear canal should be included.
- Because of the size and anatomy of the newborn ear, identifying the tympanic membrane or any landmarks may be difficult.

### Acoustic Immittance Measures

- Tympanograms should be obtained for both ears.
- Probe tones equal to or greater than 660 Hz should be used because of the poor validity of tympanometry when using a low-frequency probe tone with this population.
- Obtain ipsilateral acoustic reflexes at 1000, 500 and 2000 Hz.
- If ipsilateral reflexes are absent, obtain contralateral acoustic reflexes at 1000, 500 and 2000 Hz (no need for bone conduction [BC] auditory brainstem response [ABR] if acoustic reflexes within normal limits [WNL]).

### Evoked Otoacoustic Emissions (OAEs)

- Obtain distortion product otoacoustic emissions (DPOAE) or transient evoked otoacoustic emissions (TEOAE) or both to evaluate cochlear outer hair cell function.
- Attempt to get a good recording of evoked OAEs for each ear at 1000, 2000 and 4000Hz at a minimum (or per manufacturer's specifications in accordance with published norms [i.e., Gorga et al., 1993]).

### Auditory Brainstem Response (ABR) Testing for Threshold Estimation

Many children in this age group can be tested during natural sleep, without sedation, using sleep deprivation with nap and feeding times coordinated around the test session. For infants requiring sedation for testing, appropriate moderate sedation protocols should be followed.

- Stimuli: Frequency specific stimuli (tone bursts of low, mid and high frequency)
- Transducer: Insert earphones are recommended for air conduction testing; bone conduction transducer will be needed if air conduction is elevated (i.e. if air conduction thresholds are greater than 20 dB eHL, bone conduction testing should be completed to assess the type of hearing loss).

- Protocol: Responses should be attempted down to 20 dB eHL. Definition of threshold should be attempted in 10 dB steps. Twenty to 25 ms. recording epochs are necessary for adequate ABR threshold detection measures in infants, especially when tonal stimuli are used and hearing loss is present.
- Age appropriate normative values for wave latencies must be adhered to (i.e., Gorga et al., 1985; Hall, 1992; or own established norms).

### **Auditory Brainstem Response (ABR) Testing for Measuring VIIIth Nerve Integrity**

- Stimuli: Click stimuli at a high level (i.e., 70 dB nHL) will be adequate in most situations to identify waves I, III and V. If no response is obtained at the maximum output level, obtain one run of rarefaction clicks and one of condensation clicks to distinguish between cochlear and neural dysfunction. Use a catch trial (no signal) to rule out artifact that may be misinterpreted as the cochlear microphonic (CM).
- Transducer: Insert earphones
- Protocol: Compare interpeak latencies with corrected age norms
- Evaluate intra-aural latency differences and waveform morphology

### **Auditory Steady State Response (ASSR)**

- ASSR is an emerging auditory evoked potential test that holds promise as a method to estimate hearing sensitivity, however ASSR predictions of hearing sensitivity in infants and young children warrant further studies.
- When utilizing ASSR be aware that thresholds may be overestimated. Normative values for ASSR testing have not yet been specified. Caution should be exercised in interpretation.
- ASSR has proven useful clinically when ABR responses are absent. As the ABR may only be obtained to the limits of the equipment, ASSR can often be obtained at higher thresholds. If the ASSR thresholds are obtained successfully, they may provide valuable information for more accurate fitting of amplification.

### **Behavioral Audiologic Assessment**

Behavioral observation without reinforcement may be used to corroborate with parent/caregiver observation of child's auditory behavior, but should not be used for threshold estimation.

### **Speech/Language Screening**

- Parental report and behavioral observation
- Screening for communication skills using age appropriate normed assessment such as but not limited to:
- Early Language Milestone Scale-2 (ELM; Coplan & Gleason, 1993)

### **Developmental Screening**

- Parental report and behavioral observation

- Screening for developmental and social-emotional skills using age appropriate normed assessment tools such as, but not limited to:
- Ages & Stages Questionnaire – 3 (Squires, Potter, & Bricker 2009)
- Ages & Stages Questionnaires: Social-Emotional (Squires, Bricker, & Twombly, 2002)
- Parents' Evaluations of Developmental Status (Glascoe, 1997, 1998)
- Infant-Toddler Checklist for Language and Communication (Wetherby & Prizant, 1993, 1998)

#### **Follow-up Schedule and Referral for Further Evaluation**

- Infants diagnosed with hearing loss should receive ongoing hearing monitoring at least every three months, and should be referred for further evaluation and appropriate early intervention services as deemed appropriate by the intervention team and per Tennessee Department of Health Newborn Hearing Program Audiology Guidelines.
- Infants diagnosed with permanent hearing loss should be immediately referred to an otolaryngologist for medical clearance for amplification.
- Release of information forms should be signed by the parent/guardian to allow those evaluating the child to share information with other service providers.
- Please refer to the Follow-Up section of this document.

#### **Parent Counseling and Resources**

- The diagnosing audiologist needs to recognize the emotional impact the diagnosis of hearing loss can have on a family and provide emotional support, as needed (ASHA, 2008).
- Please refer to the Follow-Up section of this document.



## GUIDELINES FOR INFANTS 5-24 MONTHS DEVELOPMENTAL AGE

### Case History

- Review newborn hearing screening results
- Identify risk indicators for progressive and delayed onset or acquired hearing loss

### Otoscopy

- The purpose of otoscopy is to ensure that there are no contraindications to placing an earphone or probe in the ear canal.
- Visual inspection for obvious structural abnormalities (i.e., ear pits, ear tags, atresia, low set ears) of the pinna and/or ear canal should be included.

### Acoustic Immittance Measures

- Tympanograms should be obtained for both ears.
- Although a low-frequency (226 Hz) probe tone is appropriate for most of this age group, there is still a possibility of false negative tympanograms in ears with MEE according to some studies for infants in the 5-7 month age range (Paradise et al.; 1976; Purdy & Williams, 2000). Therefore, probe tones equal to or greater than 660 Hz should be used with this sub-set.
- Obtain ipsilateral acoustic reflexes at 1000, 500 and 2000 Hz.
- If ipsilateral reflexes are absent, obtain contralateral acoustic reflexes at 1000, 500 and 2000 Hz (no need for bone conduction [BC] auditory brainstem response [ABR] if acoustic reflexes within normal limits [WNL]).

### Evoked Otoacoustic Emissions (OAEs)

- Obtain distortion product otoacoustic emissions (DPOAE) or transient evoked otoacoustic emissions (TEOAE) or both to evaluate cochlear outer hair cell function.
- Attempt to get a good recording of evoked OAEs for each ear at 1000, 2000 and 4000Hz at a minimum (or per manufacturer's specifications in accordance with published norms [i.e., Gorga et al, 1993]).

### Auditory Brainstem Response (ABR) Testing for Threshold Estimation

In infants 5-24 months of age, ABR threshold testing will not be necessary in cases where acoustic immittance (including acoustic reflexes), OAE and behavioral audiologic assessments demonstrate consistent, replicable information with good reliability. The need for ABR threshold testing should be determined on an individual, case-by-case basis.

Some children in this age group can be tested during natural sleep, without sedation, using sleep deprivation with nap and feeding times coordinated around the test session. For infants requiring sedation for testing, appropriate moderate sedation protocols should be followed.

- Stimuli: Frequency specific stimuli (tone bursts of low, mid and high frequency)

- Transducer: Insert earphones are recommended for air conduction testing; bone conduction transducer will be needed if air conduction is elevated (i.e. if air conduction thresholds are greater than 20 dB eHL, bone conduction testing should be completed to assess the type of hearing loss).
- Protocol: Responses should be attempted down to 20 dB eHL. Definition of threshold should be attempted in 10 dB steps. Twenty to 25 ms. recording epochs are necessary for adequate ABR threshold detection measures in infants, especially when tonal stimuli are used and hearing loss is present.
- Age appropriate normative values for wave latencies must be adhered to (i.e., Gorga et al., 1985; Hall, 1992; or own established norms).

### **Auditory Brainstem Response (ABR) Testing for Measuring VIIIth Nerve Integrity**

- Stimuli: Click stimuli at a high level (i.e., 70 dB nHL) will be adequate in most situations to identify waves I, III and V. If no response is obtained at the maximum output level, obtain one run of rarefaction clicks and one of condensation clicks to distinguish between cochlear and neural dysfunction. Use a catch trial (no signal) to rule out stimulus artifact that may be misinterpreted as the cochlear microphonic (CM).
- Transducer: Insert earphones
- Protocol: Compare interpeak latencies with corrected age norms
- Evaluate intra-aural latency differences and waveform morphology.

### **Auditory Steady State Response (ASSR)**

- ASSR is an emerging auditory evoked potential test that holds promise as a method to estimate hearing sensitivity, however ASSR predictions of hearing sensitivity in infants and young children warrant further study.
- When utilizing ASSR be aware that thresholds may be overestimated. Normative values for ASSR testing have not yet been specified. Caution should be exercised in interpretation.
- ASSR has proven useful clinically when ABR responses are absent. As the ABR may only be obtained to the limits of the equipment, ASSR can often be obtained at higher thresholds. If the ASSR thresholds are obtained successfully, they may provide valuable information for more accurate fitting of amplification.

### **Behavioral Audiologic Assessment**

- Visual Reinforcement Audiometry (VRA)
- Ear specific testing. Alternate testing between ears as appropriate to obtain ear specific information from each ear prior to child's fatigue.
- Prioritize order of testing to obtain responses for low and high frequency stimuli
- Minimum response levels should be obtained for the following stimuli:
  - Speech – (Speech Awareness Threshold [SAT] vs. Speech Recognition Threshold [SRT] when possible)

- Bone conduction testing should be obtained if any air conduction thresholds are elevated
- Frequency-specific stimuli at 2000, 500, 1000 and 4000 Hz (the order of presentation will vary according to the focus of the audiologic assessment)
- Numerous options for stimulus start-level, step-size and start-stop rules are available (Bernstein & Gravel, 1990; Tharpe & Ashmead, 1993; Widen et al., 2000; Widen et al., 2005).

### **Speech/Language Screening**

- Screening for communication skills using age-appropriate, normed tools such as, but not limited to:
- Early Language Milestone Scale-2 (ELM-2; Coplan & Gleason, 1993).  
Receptive-Expressive Emergent Language Test-3rd Edition (REEL-3; Broch, League, & Brown, 2003).

### **Developmental Screening**

- Parental report and behavioral observation
- Screening for developmental and social-emotional skills using age appropriate normed assessment tools such as, but not limited to:
- Ages & Stages Questionnaire – 3 (Squires, Potter, & Bricker 2009)
- Ages & Stages Questionnaires: Social-Emotional (Squires, Bricker, & Twombly, 2002)
- Parents' Evaluations of Developmental Status (Glascoe, 1997, 1998)
- Infant-Toddler Checklist for Language and Communication (Wetherby & Prizant, 1993, 1998)

### **Follow-up Schedule and Referral for Further Evaluation**

- Infants diagnosed with hearing loss or auditory deficit should receive ongoing hearing monitoring at least every three months, and should be referred for further evaluation and appropriate early intervention services as deemed appropriate by the intervention team and per Tennessee Department of Health Newborn Hearing Program Audiology Guidelines.
- Infants diagnosed with permanent hearing loss should be immediately referred to an otolaryngologist for medical clearance for amplification.
- Release of information forms should be signed by the parent/guardian to allow those evaluating the child to share information with other service providers.
- Please refer to the Follow-Up section of this document.

### **Parent Counseling and Resources**

- The diagnosing audiologist needs to recognize the emotional impact the diagnosis of hearing loss can have on a family and provide emotional support, as needed (ASHA, 2008).
- Please refer to the Follow-Up section of this document.

## **GUIDELINES FOR TODDLERS AND PRESCHOOLERS 25-60 MONTHS DEVELOPMENTAL AGE**

### **Case History**

- Review newborn hearing screening results
- Identify risk indicators for progressive and delayed onset or acquired hearing loss

### **Otoscopy**

- The purpose of otoscopic examination is to ensure there are no contraindications for placing an earphone or probe in the ear canal.
- Visual inspection for obvious structural abnormalities (i.e., ear pits, ear tags, atresia, low set ears) of the pinna and/or ear canal should be included.

### **Acoustic Immittance Measures**

- Obtain 226 Hz probe tone tympanometry
- Obtain ipsilateral acoustic reflexes at 1000, 500, 2000 and 4000 Hz.
- Obtain contralateral acoustic reflexes at 1000, 500, 2000 and 4000 Hz (no need for bone conduction [BC] auditory brainstem response [ABR] if acoustic reflexes are within normal limits [WNL]).

### **Evoked Otoacoustic Emissions (OAEs)**

- Obtain Distortion Product Otoacoustic Emissions (DPOAE), Transient Evoked Otoacoustic Emissions (TEOAE), or both to evaluate cochlear outer hair cell function.
- Attempt to get a good, repeatable recording of evoked OAE's for each ear at 1000, 1500, 2000, 3000, 4000 and 6000 Hz (or the standard protocol with norms per manufacturer's specifications).

### **Auditory Brainstem Response (ABR) Testing for Threshold Estimation**

If audiologic results are unreliable or unobtainable, ABR testing should be completed. For children requiring sedation for testing, appropriate moderate sedation protocols should be followed.

- Stimulus: Frequency specific tonebursts of low, mid and high frequency
- Transducer: Insert earphones for air conduction testing. Bone vibrator for bone conduction testing (needed if air conduction thresholds greater than 20 dB eHL).
- Responses should be attempted down to 20 dB eHL. Definition of threshold should be attempted in 10 dB steps.
- Age appropriate normative values for wave latencies must be adhered to (i.e., Gorga et al., 1985; Hall, 1992; or own established norms).
- Follow-up testing should occur for all infants with risk factors per Tennessee Newborn Hearing Program Audiology Guidelines for follow-up.

## **Auditory Brainstem Response (ABR) Testing for Measuring VIIIth Nerve Integrity**

Conduct assessment **if**:

- ABR is abnormal with present OAEs
- ABR is abnormal regardless of OAE results
  - Stimulus: a click stimulus at a high intensity level (i.e., 70 dB – 80 dB nHL) will be adequate in most cases to identify waves I, III and V.
  - If no response is obtained at the maximum output level, obtain one run of rarefaction clicks and one of condensation clicks to distinguish between cochlear and neural dysfunction. Use a catch trial (no signal) to rule out artifact that may be misinterpreted as the cochlear microphonic (CM).
- Transducer: Insert earphones
- Protocol: Compare interpeak latencies with age appropriate norms
- Evaluate intra-aural latency differences and waveform morphology.

## **Auditory Steady State Response (ASSR)**

- ASSR is an emerging auditory evoked potential test that holds promise as a method to estimate hearing sensitivity, however ASSR predictions of hearing sensitivity in infants and young children warrant further study.
- When utilizing ASSR be aware that thresholds may be overestimated. Normative values for ASSR testing have not yet been specified. Caution should be exercised in interpretation.
- ASSR has proven useful clinically when ABR responses are absent. As the ABR may only be obtained to the limits of the equipment, ASSR can often be obtained at higher thresholds. If the ASSR thresholds are obtained successfully, they may provide valuable information for more accurate fitting of amplification.

## **Behavioral Audiologic Assessment**

- Pure tone assessments should be selected based on the child's developmental age. Options include:
  - Visual Reinforcement Audiometry (VRA)
  - Conditioned Play Audiometry (CPA)
  - Tangible (TROCA) or Visual Reinforcement Operant-Conditioning Audiometry (VROCA)
- Alternate between ears (as appropriate) to obtain some ear specific information from each ear prior to the child's fatigue
- Obtain the following, as appropriate:
  - Air and bone conduction testing
    - Frequency-specific thresholds for 250-8000 Hz
    - Prioritize order of testing to obtain, at a minimum, thresholds for low and high frequency stimuli
- Speech Reception Threshold (SRT)
  - spondee pictures if needed
  - point to body parts

- Although word recognition testing may not be possible with some young children because of their age, degree of hearing loss, or language skills, it is possible to assess speech perception skills in very young children.
- Speech Perception Skills: The ability of audiologists to determine if a child's auditory development is at the detection, discrimination, or comprehension stage is important for management purposes.
  - Detection (e.g., Early Speech Perception Test [ESP; Moog & Geers, 1990]; Ling 6-Sound Test [Ling, 1989])
  - Discrimination (e.g., Screening Inventory of Perception Skills [SCIPS; Osberger et al., 1991]; Low-Verbal ESP [Moog & Geers, 1990])
  - Comprehension (e.g., SPICE Curriculum [Moog, Biedenstein, & Davidson, 1995]; Mr. Potato Head [Robbins, 1994]; or, following simple commands [Makins, 1979; Olsen & Matkin, 1979]).

### **Speech/Language Screening**

- Screening for communication skills using age appropriate normed assessment such as but not limited to:
  - Early Language Milestone Scale-2 (ELM-2; Coplan & Gleason, 1993)
  - The Fluarty-2. (Fluarty, N.B. 2001)
- Referral for comprehensive speech/language evaluation may be necessary

### **Developmental Screening**

- Parental report and behavioral observation
- Screening for developmental and social-emotional skills using age appropriate normed assessment tools such as, but not limited to:
  - Ages & Stages Questionnaire – 3 (Squires, Potter, & Bricker 2009)
  - Ages & Stages Questionnaires: Social-Emotional (Squires, Bricker, & Twombly, 2002)
  - Parents' Evaluations of Developmental Status (Glascoe, 1997, 1998)
  - Infant-Toddler Checklist for Language and Communication (Wetherby & Prizant, 1993, 1998)

### **Follow-up Schedule and Referral for Further Evaluation**

- Infants diagnosed with hearing loss or auditory deficit should receive ongoing hearing monitoring at least every three months, and should be referred for further evaluation and appropriate early intervention services as deemed appropriate by the intervention team and per Tennessee Department of Health Newborn Hearing Program Audiology Guidelines.
- Infants diagnosed with permanent hearing loss should be immediately referred to an otolaryngologist for medical clearance for amplification.
- Release of information forms should be signed by the parent/guardian to allow those evaluating the child to share information with other service providers.
- Please refer to the Follow-Up section of this document.

**XI. Parent Counseling and Resources**

- The diagnosing audiologist needs to recognize the emotional impact the diagnosis of hearing loss can have on a family and provide emotional support, as needed (ASHA, 2008).
- Please refer to the Follow-Up section of this document.



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## **AUDIOLOGIC FOLLOW-UP GUIDELINES FOR THE PEDIATRIC POPULATION**

To assure best practice in the follow-up of pediatric patients identified with a hearing loss or a risk indicator for hearing loss, the audiologist or other health care provider should provide the family with information regarding the child's diagnosis and need for ongoing care. In addition, the family should be informed of the services provided by agencies and organizations such as the Tennessee Early Intervention System (TEIS) and Children's Special Services (CSS) as well as several different medical specialists that are outlined below.

### **Informed Consent for Referral**

In all cases of audiologic care, the individual's privacy must be protected. The referring practitioner is responsible for obtaining/confirming informed consent or informed parental/legal guardian permission.

Written and electronic records, documentation, and communication must follow recommended laws and standards such as:

- Health Insurance Portability and Accountability Act (HIPAA)
- Joint Commission on Accreditation of Healthcare Organizations (JCAHO)
- Family Educational Rights and Privacy Act (FERPA)
- State statutes, regulations, or institutional policies may supersede some recommendations.

## **Audiologic Management and Follow-Up for Children Identified with Hearing Loss**

### **Referral for Medical/Genetic Evaluation**

Each child identified with hearing loss should be immediately referred to an otolaryngologist or otologist for medical evaluation to determine if medical intervention or genetic counseling is appropriate and to obtain medical clearance for amplification.

The medical team serving infants and children who are deaf and hard of hearing may consist of many professionals. The physician (primary care provider/medical home provider) has the primary responsibility for medical care, including referrals for the infant or child. The audiologist who identifies the infant or child with a hearing loss maintains an obligation to include the primary care/medical home provider in any decision-making processes that involve further referrals to otolaryngologists, geneticists, ophthalmologists or others per the ASHA Guidelines for Follow-Up Recommendations (2004).

Genetic consultation is important to differentiate between genetic hearing loss and non-genetic hearing loss. Hearing loss may be only one of a number of conditions associated with a genetic

syndrome. Therefore, a genetic evaluation may be significant in the identification of other medical and developmental diagnoses or conditions to be considered in the child's plan of care. Genetic hearing loss is diagnosed by physical examination, otologic evaluation, audiologic assessment, family history, ancillary testing (e.g., CT scan of temporal bone) and DNA-based testing. The genetic consultation provides the family with information on the nature, inheritance, and implications of a genetic condition and offers the family a review of available options so that informed decisions can be made. Please refer to Appendix 4-Tennessee Genetic Resources for additional information.

### **Referral for Vision Assessment**

Children with sensorineural (SNHL) hearing loss should be referred to an ophthalmologist for assessing any ocular deficits or vision problems.

Children diagnosed with hearing loss and vision loss should be referred to:

- The Tennessee Early Intervention System (TEIS).
- The Tennessee Technical Assistance and Resources for Enhancing Deaf/Blind Supports (TREDS) program for parent, provider, and teacher education/support services. (Appendix 3-Tennessee and National Hearing Resources)

### **Referral for Early Intervention Services**

Infants and children age birth through age two years identified with a condition that has a high probability of resulting in developmental delay need to be referred to the Tennessee Early Intervention System (TEIS) within two working days of the diagnosis. Parents should be advised of the availability of intervention services through TEIS. TEIS is responsible for the Federal, Individual with Disabilities Education Act (IDEA), Part C, Child Find and for planning, implementation, supervision, monitoring, and technical assistance for the statewide early intervention system for infants and toddlers (birth through age two) with developmental delays. TEIS provides service coordination to families of children with hearing loss. There are no financial guidelines for eligibility. Families and providers can contact 1-800-852-7157.

### **Referral to Children's Special Services (CSS)**

CSS provides medical and care coordination services for children birth to 21 years. The program is available for children with disabilities who meet medical and financial guidelines. The provider should refer parent(s) to their local County Health Department to schedule an appointment with the CSS coordinator to be evaluated for eligibility for enrollment.

### **Referral to Family Voices of Tennessee**

Family Voices has regional Newborn Hearing Parent Consultants who provide advice, guidance, and support to families affected by hearing impairments in children. The consultants, who are parents of children with hearing loss, can help families identify possible resources and offer referral assistance as requested or appropriate.

### **Information on Programs for Communication Development**

Auditory/Oral, Total Communication, sign languages, and Cued Speech are some of the



communication approaches available to children and families. Parents should be counseled in an unbiased manner on the different communication approaches and be informed of the programs available in their community to allow them to make the best decision based on the needs of their child and family. Parent and caregiver education should be integrated into all aspects of the child's audiologic and early intervention services.

### **Referral for Speech and Languages Services**

All children with hearing loss should be seen by a speech-language pathologist who is designated to provide assessment and management of infants and children with hearing loss and has the commensurate knowledge and skills to do so.

### **Referral for Developmental Screening**

Pediatric patients identified with hearing loss should be monitored to ensure that developmental milestones are being met in order to rule out any other possible developmental delays or deficits.

### **Schedule for Audiologic Monitoring**

All children with identified hearing loss (i.e. unilateral or bilateral, permanent or fluctuating) should receive periodic audiologic monitoring. An immediate audiologic evaluation should be scheduled when there is concern related to change in hearing or hearing aid function.

- Bilateral sensorineural hearing loss and permanent conductive hearing loss
  - Age 0-3 years: At least every 3 months, after hearing loss is confirmed;
  - Age 4-6 years: At least every 6 months, if intervention progress is satisfactory;
- Transient conductive hearing loss (i.e., otitis media with effusion), unilateral or bilateral:
  - Monitor after medical treatment (completion of antibiotic treatment, PE tubes, etc.), and/or at least every 3 months until resolved and normal hearing is confirmed;
- Unilateral hearing loss (sensorineural or permanent conductive):
  - Infants with unilateral hearing loss should be monitored at least every 3 months during the first year and at least every 6 months after the first year to rule out changes in the normal hearing ear or progression of hearing loss in the poorer ear.

## **Audiologic Follow-Up for Children Identified with Risk Indicators**

### **Risk Indicators for Permanent Congenital, Delayed-Onset, or Progressive Hearing Loss**

The Joint Committee on Infant Hearing (JCIH) 2007 Position Statement has outlined indicators that place infants and children at risk for permanent, progressive or delayed-onset hearing loss. In contrast to previous position statements, JCIH '07 differentiated risk indicators that are of greater concern for delayed-onset hearing loss versus those indicators that are considered to be at a lower risk. For a complete list of the risk indicators, please refer to Appendix 2. The risk indicators that JCIH has judged to be of greater concern for permanent congenital, delayed-

onset, or progressive hearing loss have been marked with a section sign ( § ).

Tennessee’s Newborn Hearing Screening Hospital Guidelines require hospital hearing screening personnel to record risk indicators on the bloodspot form when the hearing screening is performed. Each infant with a reported risk indicator is then entered into the Tennessee Department of Health Newborn Hearing Screening (TDH NHS) database which generates reminder letters to both parents and family physicians that the child needs continued surveillance even if the initial hearing screen was passed. TDH NHS reminder letters will be periodically sent until the child turns three years of age, or until an audiologist determines that a child can be released from audiologic care and this is reported to the TDH NHS Program. Audiologists should expect phone calls from families and physicians to schedule hearing assessments for babies and young children with risk indicators.

#### **Schedule for Audiologic Monitoring:**

The JCIH ’07 Statement recommends that, “all infants with a risk indicator for hearing loss, regardless of surveillance findings, should be referred for an audiological assessment at least once by 24 to 30 months of age. Children with risk indicators that are highly associated with delayed-onset hearing loss...should have more frequent audiological assessments.” Considering the recommendations of JCIH and also the success of Tennessee’s previous audiologic monitoring schedule for babies and children with risk indicators, the following schedule is advised for audiologic assessment of the pediatric population with risk indicators in Tennessee:

- All infants identified with any risk indicator at birth should receive an audiologic assessment between 6 and 9 months of age.
- All infants with a risk indicator that is of greater concern for delayed-onset hearing loss, should continue to receive audiologic assessments every 6 months until age 3.
- All infants with a risk indicator that is of less concern for delayed-onset hearing loss, should receive annual audiologic assessments until age 3.

Of course, if families develop a significant concern regarding their child’s hearing at any time between assessments, a hearing evaluation should be scheduled immediately. Audiologists may use their professional discretion to determine the need for continued follow-up. Audiologists should contact the TDH NHS Program to report newly identified cases of babies or children with risk indicators or to report the discontinuation of audiologic monitoring based on documented stability of hearing levels and the level of associated risk with given indicators. In both cases, the TDH NHS Program will begin or cease mailing reminder letters accordingly to parents and physicians.

## **Other Audiologic Responsibilities for the Pediatric Population**

#### **Documentation/Reporting**

Documentation must be completed for each visit or interaction to provide a full archive of the child’s audiological history. Documentation of assessment must address interpretation of test

results, the type and severity of the hearing loss, and other relevant background information (e.g., birth history, etiology, medical diagnosis, co-existing conditions) as well as a complete summary of recommendations. Recommendations may address the need for further assessment, follow-up, and/or referral. When treatment is recommended, information must be provided concerning the frequency, estimated duration and type of service (e.g., individual, group, home program) required (ASHA, 1997). Patient records should follow HIPPA standards. Requests for a child's records must respect parental rights to confidentiality and protected health information mandates and require necessary and appropriate informed consent (Pediatric Work Group,1996).

A complete report should be sent to the child's parent/legal guardian, primary care physician/pediatrician and any referral sources upon parental consent. Providers should report all cases of confirmed hearing loss to the TDH NHS Program on infants, toddlers, and children birth to five years old. This includes any baby or child that was born outside of Tennessee, but now resides within the state. The Audiology Reporting Form is the means to communicate these results via fax (615-262-6159). This form can be found in Appendix 5 of this document. Please complete the Audiology Reporting Form in its entirety so accurate documentation can be reported to state and federal agencies for our state.

### **Provision of Counseling**

Parents, primary caregivers, grandparents, and immediate family members of children with diagnosed hearing loss should be counseled on topics that include but are not limited to: 1) type and degree of hearing loss; 2) expectations of having a child with hearing loss; 3) current amplification technology; 4) communication options and cultural considerations; 4) local/state/national resources for children with hearing loss; and 5) family support options. Counseling should also be provided to families of children who exhibit risk indicators for delayed-onset or progressive hearing loss. It is imperative that these families understand the typical development of listening and speech-language skills and signs that may indicate a change in their child's hearing. A family-centered and culturally-sensitive approach needs to be maintained during all aspects of counseling. Family members who require counseling services that are beyond the scope of the managing audiologist should be referred for psychology services in the local community that will meet the needs of the individual.

### **Sharing of Relevant Literature**

To support families who have a baby or young child with hearing loss, Family Voices of Tennessee and TDH NHS Program have made available the "Family Voices Parent Notebook". This free resource can serve as a beginning point for families to learn more about hearing loss in children while also serving as a spring board for discussions between the audiologist and the family on many related topics. The Parent Notebook was also designed to serve as a care notebook for families to use for filing paperwork (audiograms, medical reports, treatment plans, etc.) related to their child's hearing care and maintaining important records (provider contacts, amplification serial numbers, warranty information, etc.). Audiologists can request copies of the Parent Notebook via email at [familyvoices@tndiability.org](mailto:familyvoices@tndiability.org) or by calling 1-888-643-7811.

TDH NHS brochures and posters can be obtained at no cost by contacting TDH Newborn Hearing Screening Program at 615-262-6160 or by faxing the Newborn Hearing Screening Order Form for Materials to 615-262-6159 (See Appendix 3).

**School System Referrals**

Children who are identified with hearing loss and are three years and older should be referred to the Local Education Agency (LEA) in compliance with Tennessee Department of Education, Federal Individual with Disabilities Education Act (IDEA), Part B, Child Find and Special Education recommendations.

# PEDIATRIC AMPLIFICATION GUIDELINES

## HEARING AIDS

The following pediatric amplification guidelines were based upon those developed by the American Academy of Audiology (AAA, 2003). The AAA Pediatric Amplification Protocol was developed by panels of nationally recognized experts in their respective fields. Any modifications to the original guidelines have been made in acknowledgement of advances in technology and intervening growth of knowledge in the field of audiology. These guidelines have been adopted by the Tennessee Newborn Hearing Screening Program, with permission from AAA, and with the clear understanding that a child's family has the final choice as to whether or not the infant should use hearing aids, assistive technology, or other methods of communication.

### **Purpose**

The purpose of this section is to provide a detailed guideline regarding to which children should be considered for amplification, what data are necessary to start and continue the amplification process, how essential features of the amplification system should be chosen, what testing should constitute verification and validation of the amplification system, and suggestions for appropriate orientation, training, and follow-up. These guidelines are intended for application to newborns, infants, and children. These guidelines are not meant to suggest specific communication modes or academic settings for these children. In addition, children may have a variety of other co-existing conditions with hearing loss and these guidelines must be considered within the context of each child's individual characteristics. The general goal of any amplification is to provide a signal that makes soft, moderate, and loud sounds audible but not uncomfortable and to provide excellent sound quality in a variety of listening environments.

### **Candidacy**

Amplification with hearing instruments should be considered for a child who demonstrates a significant hearing loss, including sensorineural, conductive, central, or mixed hearing losses of any degree. The duration and configuration (bilateral or unilateral) will assist the audiologist in the decision to fit a child with personal hearing aids. Additional factors such as the child's health, cognitive status, and functional needs also will influence the time-line of fitting hearing aids. The presence of chronic or recurrent middle ear conditions that can affect hearing threshold results or the ability to wear an occluding ear mold should be considered. When determining hearing aid candidacy for infants or children with borderline or minimal hearing losses, middle ear status is of particular concern in determining the likelihood of a transient condition.

### **Special Considerations**

Special consideration should be given to the fitting of amplification on children with unilateral hearing loss, minimal or mild hearing loss, profound hearing loss, and auditory neuropathy.

#### *A. Unilateral hearing loss*

Use of hearing aid amplification is indicated for some children with unilateral hearing losses. The decision to fit a child with a unilateral hearing loss should be made on an individual basis, taking into consideration the child's or family's preference as well as audiologic, developmental, communication, and educational factors. Amplification options such as personal FM systems also should be considered. Use of communication strategies (noise reduction, positioning, etc.) may prove to be beneficial and easily accomplished for the infant or toddler with unilateral hearing impairment. The use of contralateral-routing-of-signal (CROS) amplification requires particular care. Its design is to overcome the problem caused by the head shadow effect. This could be especially helpful in a quiet environment and when the signal of interest originates from the direction of the nonfunctioning ear. However, one study (Kenworthy, Klee, & Tharpe, 1990) indicated that CROS amplification may not be beneficial for children in a classroom setting, because of the introduction of additional noise to the normal-hearing ear.

#### *B. Minimal-mild hearing loss*

Current evidence suggests that children with minimal and mild hearing losses are at high risk for experiencing academic difficulty (Yoshinaga-Itano, 1996; Bess, Dodd-Murphy, & Parker, 1998; Bess & Tharpe, 1984). As such, children with minimal and mild hearing loss should be considered candidates for amplification and/or personal FM system or soundfield systems for use in school.

#### *C. Profound hearing loss*

A finding of no response by ABR should not exclude a child from hearing aid candidacy, as residual hearing may exist at intensity levels greater than those capable of eliciting a standard ABR response. Children with confirmed profound hearing loss still may experience benefit from hearing aid amplification. An infant or child with severe to profound hearing loss or auditory neuropathy should be considered as a candidate for a cochlear implant.

#### *D. Normal peripheral hearing sensitivity*

In some cases, children with normal peripheral hearing sensitivity may benefit from amplification (Matkin, 1996). These cases may include children with auditory processing disorders (APD), auditory neuropathy or dysynchrony (AN/AD), and attention deficit disorder/attention deficit hyperactivity disorder (ADD/ADHD). In such cases, close audiologic monitoring of hearing sensitivity, and careful control of the output of the amplification is required.

### **Pre-Selection Issues and Procedures**

Many decisions must be made prior to selecting amplification for a child. These decisions may be based on individual needs and abilities, diagnostic information (e.g., degree of hearing loss, physical characteristics, etc.), environment in which the individual functions, empirical evidence, and/or clinician experience. Many of these decisions must be revisited on an ongoing basis as the child matures.

### *A. Air vs. Bone Conduction*

Air conduction hearing aids are considered the more conventional hearing aid type and provide amplified sound into the ear canal of the user. A bone conduction hearing aid typically is considered for children who are unable to wear air conduction devices as a result of malformation of the outer ear or recurrent middle ear drainage. A bone conduction hearing aid may be considered for children with unilateral conductive hearing loss to insure that the intact cochlea on the side with the conductive hearing loss is stimulated during development while waiting for possible corrective surgery. The bone anchored hearing aid is a device that is surgically implanted into the skull behind the ear and produces a bone-conducted signal that is transmitted through the skull to the inner ear. This type of device is useful for an individual who must use a bone-conducted rather than an air-conducted signal on a permanent basis. At this time, bone anchored hearing aids do not have the approval of the U.S. Food and Drug Administration (FDA) for use in children less than five years of age. A bone anchored hearing aid may be considered as an option for an older child. A bone conduction hearing aid may be used on younger children; however it is not surgically implanted, but couples to the head with a specially designed headband.

### *B. Style: behind-the-ear (BTE) vs. in-the-ear (ITE) vs. in-the-canal (ITC) vs. completely-in-the-canal (CIC)*

Style will be dictated by the child's hearing loss and potential for growth of the outer ear and individual needs. The outer ear may continue to grow well into puberty, thus dictating the BTE style. When growth occurs, only the earmold has to be replaced. The BTE is more durable (with no circuitry directly exposed to cerumen) than in-the-ear styles, is less likely to produce feedback when fitted with an appropriate earmold, and allows for a variety of features that may be essential for the child (i.e., telecoil circuitry, direct audio input (DAI) connection, built-in FM circuitry). An in-the-ear or even completely-in-the-canal hearing aid may be an option for older children as long as the audiologist, child, and parents recognize the pros and cons of each style.

### *C. Routing of the Signal*

#### 1) Bilateral vs. unilateral listening

It is well documented that bilateral hearing is necessary for localization and for best performance in noise (Hawkins & Yacullo, 1984; Valente, 1982a, 1982b). In addition, investigations have reported auditory deprivation in children fitted with unilateral amplification (Boothroyd, 1993; Hattori, 1993). Therefore, it is recommended that, unless contraindicated, children be fitted with bilateral amplification.

#### 2) CROS, BICROS, transcranial fitting

For children with severe to profound unilateral hearing loss (or very poor word recognition unilaterally), contralateral routing of signal (CROS) system may be considered. A CROS system can be achieved by putting a microphone at the location of the impaired ear and transmitting the signal to the normal ear through:

- a.) a wire or FM signal (conventional CROS),
- b.) through bone conduction

For the child with severe to profound hearing loss (or very poor word recognition) in one ear and an aidable hearing loss in the other ear, a BICROS system may be considered.

### 3) Implantable devices

No middle ear implantable devices for children are available at this time.

#### *D. Bandwidth*

Research in adults supports the use of a wide bandwidth for individuals with mild to moderate hearing losses (Skinner, 1983). A number of investigators have studied bandwidth effects in adults with moderate-to-severe hearing loss (Ching, Dillon, & Byrne, 1998; Hogan & Turner, 1998; Turner & Cummings, 1999). These studies suggest that the provision of high-frequency amplification may not always be beneficial and can even degrade speech perception for some individuals. In these studies, there is considerable variability in performance across individuals and no consensus on the degree of hearing loss at which benefit from high-frequency amplification no longer occurs (Moore, 2001). Kortekaas & Stelmachowicz (2000) and Stelmachowicz, Pittman, Hoover, & Lewis (2001) found that children with hearing loss require a wider bandwidth than adults with similar hearing losses to perceive high-frequency speech sounds, particularly when listening to female and child talkers. Ching, Dillon, & Katsch (2001) indicate that there is no conclusive evidence in this area at this point and time. Therefore, the clinician must consider each child as an individual as we wait for more evidence in this area. In addition, the clinician should not confuse a lack of increased performance with high frequency amplification with an actual decrease in performance.

#### *E. Memories*

Memories allow more than one amplification characteristic for use by the wearer in different listening situations. The user (or parent) can choose among memories based on the listening situation. In the pediatric population, multiple memories may be very useful if there is a predictable fluctuating hearing loss so that the hearing aid output can be easily adjusted accordingly. In addition, a programmable telecoil memory may also be useful.

#### *F. Ear Mold*

The audiologist should consider the style, material, color, length, and frequency of remakes for the ear mold. The need for well-fitted ear molds has increased with the advent of wide dynamic range, wideband hearing aids. The audiologist is able to make a wide range of sounds audible in an automatic way by using compression circuitry with no volume control. Without a volume control, the child (or parent) cannot turn down the hearing aid if it starts to feed back as a result of poor ear mold fit (after growth of the outer ear). The use of automatic technology forces the audiologist to be more proactive about regular ear mold changes. The recent advent of automatic feedback control through various digital signal processing techniques may alleviate this problem temporarily while the new ear mold is ordered. For infants, ear mold replacement may be as frequent as monthly.



Venting in the ear mold may be appropriate for some children depending on the configuration and degree of hearing loss as well as the status of their outer and middle ear. The audiologist should approach venting ear molds in children cautiously. Diagonal venting may cause the hearing aid to lose some of its high frequency response and certain placements of venting may create problems in sound channel tubing retention.

#### *G. Sound Channel*

The sound channel consists of the earhook and tube that leads through the ear mold and sends sound into the ear canal. Just as a horn (increased diameter at the end of a sound channel) increases the high frequency response, a reverse horn will roll off the high frequencies. These are often the frequencies where the child needs the most amplification. A reverse horn is a common concern in an infant or young child because the ear mold is so small. It is essential that the end of the sound channel be checked visually for any crimping. An electroacoustic measure that includes the ear mold will reveal any roll off in high frequency response as will probe microphone measurements that include the individual's ear mold connected to the hearing aid.

Manufacturers generally send adult size earhooks unless otherwise instructed. A pediatric earhook can be the difference between a well situated BTE and a BTE that falls off of the ear. Earhooks add resonant peaks to the hearing aid response. These peaks can increase the chance of acoustic feedback and may dictate the maximum output setting of the hearing aid thereby unnecessarily decreasing the headroom (the difference between the level of speech and the saturation level of the hearing aid) of the instrument. A filtered (damped) earhook will smooth the response (Scollie & Seewald, 2002).

#### *H. Microphone*

Microphone location impacts the response of the signal that is presented to the ear. For most pediatric users, the microphone will be at the top of the ear because they will use the BTE style. The BTE and ITE styles can be equipped with omni-directional microphones (microphones that respond to signals equally around the head) or directional microphones (microphones that reduce signals from the sides and back). Directional microphones can enhance hearing in noise in adults (Hawkins & Yacullo, 1984). The user may switch between microphone types by using a toggle switch, button, or remote control device. This is not a realistic choice for infants and young children. The use of a traditional directional microphone also implies that the signal of interest is in front of the listener. Young children learn by listening to the adults around them and may not be looking at them directly. In such situations, there may not be a primary talker. In some of the newest digital hearing aids, this switching occurs automatically based on a sampling of the incoming signal. Type of microphone technology will be dictated by the age and abilities of the child as well as listening environment. Through the selection and de-selection of memories, some hearing aids allow the audiologist to choose when to introduce the use of directional microphone technology (activating the programmable memory), thereby equipping hearing aids with potential that may not be used right away with a young child. When directional microphones are used with children, the audiologist should ensure that the microphone response in the directional setting is equalized to the microphone response in the omni-directional setting or audibility for low frequency sounds is lost (Ricketts & Henry, 2002).

### *I. Controls for Fine-Tuning*

With children, it is frequently necessary to conduct fine-tuning of the hearing aids' gain and output characteristics. As more and more infants are fitted with hearing aids as a result of universal newborn screening, the use of flexible technology becomes even more critical. The hearing abilities of these babies continue to be defined as they mature and flexible hearing aids can be changed to reflect the new information obtained from the diagnostic procedures. In addition, children may have progressive hearing losses. A flexible hearing aid is a cost-effective solution for these children because the response of the hearing aid can be changed to meet the child's needs as the hearing loss changes or as more complete information is obtained.

### *J. Previous Experience*

The audiologist's decisions for all of the features described in this section may be impacted by the child's previous experience. Only the older child will have previous experience, but the impact of previous experience should be considered when working with the infant. There are data to suggest that hearing aid users will become accustomed to whatever signal processing they experience and will come to prefer it (Palmer, 2001). This puts a great deal of burden on the audiologist to provide the very best audibility and sound quality to the first-time user as this is the signal to which he/she will adapt. This is not to say that a current user of one technology (e.g., linear processing) cannot adapt and benefit from another technology that the audiologist may deem appropriate at the time of a replacement hearing aid fitting (e.g., wide dynamic range compression). Children may require an adjustment period before they tolerate and benefit from the newer technology, just as we expect adjustment to frequency transposition, cochlear implant signal processing, etc.

### *K. Telephone Access*

The Developmental Index of Audition and Listening (Palmer & Mormer, 1999) illustrates that the telephone is an integral part of a child's life from the time when they know that someone is calling, extending through their attempts to participate in telephone communication with a parent's help, to the time when they are using the telephone to make plans with their friends. It is essential that the audiologist provide telephone access for even the youngest hearing aid wearers and take the time to educate the parents on how the solution works (this may take a variety of training sessions until the parents or guardians are comfortable).

### *L. Ability to Couple to Assistive Listening Technology*

The child's hearing aids may be coupled to assistive technology through the telecoil, direct audio input, built-in FM receiver, or FM receiver attachment. The assistive listening device will be the best solution for listening in noise and/or listening at a distance. Selection of instruments that are compatible with FM systems, particularly the specific FM system provided at school may be warranted. It is critical to know the coupling requirements of the school system.

### *M. Battery Doors*

The audiologist should recommend tamper-resistant battery doors for younger children.

#### *N. Volume Control*

The need for a volume control is dictated by the signal processing scheme that is used in the hearing aid and the user's previous experience (if any). If the audiologist does not expect the child to make these adjustments, wide dynamic range compression signal processing will be advantageous.

Adjustment of a volume control wheel can provide a short-term solution to feedback caused by poorly fitting ear molds. If a volume control is present, the clinician must decide if the child should have access to manipulating the control or if a locking volume control is preferred (access is then limited to the clinician and perhaps parent/caregiver). Linear signal processing implies that a volume control is not only included, but is manipulated since the gain for a linear system is targeted to moderate level input signals. One assumes that the user would need to turn down more intense inputs and turn up quiet inputs to maintain audibility and comfort.

The unique combination of the above decisions will lead to the selection of particular hearing aids for a particular child. Some decisions exclude other choices and a compromise may have to be reached by prioritizing these choices.

## **Circuitry - Signal Processing**

Although certain signal processing schemes require digital processing, the discussion here is only relevant to the strategies, not digital versus analog processing to implement those strategies. That is, the appropriate signal processing question is not, in our opinion, whether we should select digital or analog hearing aids, but rather, what signal processing schemes are appropriate. In some cases the desired signal-processing scheme may require digital signal processing, in other cases it may not. Most of the hearing aids are digital; however, analog is still available for the unique cases where it is necessary. The choice of appropriate features for each individual will be paramount.

### *A. Basic Requirements*

- 1) The system should avoid distortion.
- 2) The system should allow frequency/output shaping to provide audibility base on an appropriate prescriptive method.
- 3) The system should allow frequency/output shaping to avoid tolerance issues based on an appropriate prescriptive method.
- 4) The system should employ amplitude processing that ensures appropriate audibility over a range of typical speech sounds from soft to loud. It is likely that some form of amplitude compression may be necessary to achieve this goal for the common cases of reduced residual dynamic range of hearing. Wide-dynamic range amplitude processing may routinely be necessary to allow for optimal audibility of soft to loud inputs (Jenstad et al., 1999, 2000).
- 5) Output limiting is independent of the signal processing that is provided in the dynamic range. Compression output limiting has been shown to provide superior sound quality as compared with peak clipping output limiting (Hawkins & Naidoo, 1993; Preves & Newton, 1989).
- 6) The system should include sufficient electroacoustic flexibility to allow for changes in required frequency/output characteristics related to growth of the child (e.g., a larger ear canal will result in a smaller real-ear-to-coupler difference, etc).

### *B. Current and Future Processing Schemes*

Until sufficient data become available to exclude the following schemes, each should be considered viable for pediatric fitting of hearing aids.

- 1) Automatic feedback control, to allow for use of amplification while the child or infant is held or placed in close proximity to other objects. Caution is advised in cases in which the hearing aid requires a gain reduction in order to prevent feedback. In such cases, the potential loss of audibility of important sounds must be considered.

- 2) Multiple channels to allow for finer tuning of the response for fitting unusual or fluctuating audiograms, application of wide dynamic range compression, increasing the specificity of noise reduction, allowing specialized feedback and occlusion management.
- 3) Expansion to reduce low-level noise (e.g., microphone noise and over-amplification of soft sounds associated with very low-threshold compression).
- 4) Compression to allow fitting of the large variation of input levels found in speech and environmental sounds into the dynamic range of the child with hearing loss. Compression also is used as a limiter, providing comfort and good sound quality for the output of intense signals.
- 5) Frequency transposition and frequency compression have yet to be sufficiently validated. This type of signal processing might be recommended only when the frequencies to be transposed cannot be made audible with non-transposing aids.

### C. Future Developments

Many schemes under development to reduce background noise (e.g., envelope modulation counters [digital noise reduction]) and/or enhance speech perception (e.g., spectral enhancement, temporally or spectrally based selective speech enhancement) cannot be recommended until data relative to their effectiveness become available.

### **Hearing Instrument Selection/Fitting Considerations in Children**

During the selection process, a determination of appropriate circuitry and processing schemes should be based on the degree, configuration, and type of hearing impairment as well as consideration of familial and economic factors. Selection and verification protocols are predicated on the availability of frequency-specific threshold data.

- A. Individual or age appropriate ear acoustics should be accounted for in the hearing instrument selection fitting process. Measurement and application of the real-ear-to-coupler-difference (RECD) accomplishes this goal (Moodie, Seewald, & Sinclair, 1994). Real-ear-coupler-differences are used to individualize the HL to SPL transform. This is important in a population whose earcanals and eardrum impedance generally are different from the adult averages that typically are used to conduct these transforms (Scollie et al., 1998; Seewald & Scollie, 1999). In addition, the RECD is used to adjust the electroacoustic fitting so the final output in the real-ear will be correct for an individual child (Seewald et al., 1999). This use of the measurement is especially important when real-ear aided response measures are not possible.
- B. Minimally, the fitting method employed to determine hearing instrument electroacoustic characteristics should be audibility based (i.e., the goal would be to provide audibility of an appropriate amplified long-term amplified speech spectrum). When nonlinear circuitry is considered, the prescriptive formula should take into account speech

audibility at different input levels (eg., NAL-NL1 or DSL [i/o; Byrne et al., 2001; Cornelisse, et al., 1995). That is, the primary goal is the audibility of speech regardless of input level or vocal effort.

- C. Target values for gain and output are determined through the use of a prescriptive formula (evidence-based independent or evidence-based device-related) by using hearing sensitivity data and the RECD.
- D. Although none of the threshold-based selection procedures are guaranteed to ensure that a child will not experience loudness discomfort or that output levels are safe, the use of a systematic objective approach that incorporates age-dependent variables into the computations is preferred. Frequency-specific loudness discomfort levels should be obtained when children are old enough to provide reliable responses (Gagné, Seewald, Zelisko, & Hudson 1991a, 1991b).
- E. The audiologist may consider the need to reduce gain recommended by a particular fitting strategy if binaural summation is not considered in the fitting strategy and the fitting is binaural. Currently, there are not data that clearly illustrate binaural summation experienced through hearing aids in the soundfield. Scollie et al. (2000) reported no binaural summation as measured through preferred listening levels in children who were using hearing aids. In addition, the desired frequency/gain response and output limiting may need to be modified from the prescription if the hearing loss is primarily conductive or if there is a conductive component.
- F. The electroacoustic parameters of the hearing instrument are pre-set so as to achieve the targeted response. Coupler measurement allows for pre-setting the hearing aids prior to fitting them to the child. Pre-setting in the pediatric population is especially important because the child may not provide reliable feedback for fine-tuning.
- G. Further electroacoustic measurement after the desired output (gain) has been set should include verification of low distortion at varying inputs at user prescribed settings.

### **Verification**

- A. The electroacoustic performance of the instrument should be matched to the prescribed 2 cm<sup>3</sup> coupler target values for gain and output limiting where the 2 cm<sup>3</sup> coupler values have been derived using an individualized real ear to 2 cm<sup>3</sup> coupler transform (e.g., the RECD).
- B. Aided soundfield threshold measurements may be useful for the evaluation of audibility of soft sounds but they are not recommended and should not be used for verifying electroacoustic characteristics of hearing instruments in infants and children for several reasons:
  - 1) prolonged cooperation from the child is required
  - 2) frequency resolution is poor

- 3) test-retest reliability is frequently poor (Seewald, Moodie, Sinclair, & Cornelisse, 1996)
  - 4) misleading information may be obtained in cases of severe to profound hearing loss, minimal or mild loss, or when non-linear signal processing, digital noise reduction, or automatic feedback reduction circuitry is used
- C. Probe microphone measurements employing an insertion gain protocol are not the preferred procedure for verifying electroacoustic characteristics of hearing instruments in infants and children for several reasons:
- 1) targets are provided outside of any relevant context (i.e., threshold) and consequently are not directly audibility based
  - 2) targets assume an average adult REUG
- D. Output characteristics should be verified using a probe microphone approach that is referenced to ear canal SPL. Determination of audibility at several input levels is the ideal method of verification. This requires the placement of a probe microphone and hearing aid in the child's ear while sound is presented through a loudspeaker at several intensity levels (e.g., soft, moderate, loud). The resulting real ear aided response (REAR) can be compared to thresholds and UCLs (measured or age-appropriate estimation) converted to ear canal SPL. This provides a direct measurement of the predicted levels of amplified speech. The clinician must select signals for this type of testing that ensure accurate electroacoustic verification. As hearing aid technology changes (processing various input signals in different ways), the clinician must update his/her knowledge as to the appropriate signal to use for testing and may need to update his/her equipment with newly developed signals (Scollie & Seewald, 2001). All air conduction hearing aid technology can be measured electroacoustically in some appropriate manner.
- E. If probe-microphone measures of real-ear hearing aid performance are not possible, hearing aid performance can be predicted accurately in the real ear by applying age appropriate average RECD values to the measured 2-cc coupler electroacoustic results (Seewald et al., 1999).
- F. As audibility is one of the main goals of the pediatric fitting, the Situational Hearing-Aid Response Profile (SHARP; Stelmachowicz, Lewis, Kalberer, & Creutz, 1994) may be used to verify predicted audibility in a variety of settings that cannot easily be measured in a clinical setting. Measured hearing aid characteristics (test chamber or probe-microphone data) are entered into this software program and the audibility for twelve different listening situations (e.g., cradle position, hip position, 1 meter, 4 meters, child's own voice, etc.) is evaluated. Estimated performance displayed on a hearing aid manufacturer screen during programming without the direct measurement of a probe microphone is an estimate of performance based on a variety of estimations associated with the individual's ear and hearing aid. These data cannot be relied on for verification purposes.

Note: In the various procedures described under Verification, a signal must be presented to the hearing aid whether it is being tested with a microphone in the test chamber or with a probe microphone in the real ear. The test signal should adequately represent the frequency, intensity, and temporal aspects of speech. Recent investigations have illustrated that various advanced signal processing interacts with the test signal and that the most accurate representation of the hearing aid's response will be through the use of a speech-like signal or by turning off signal processing during test that attempts to reduce output that it considers noise (Scollie & Seewald, 2002; Scollie, Steinberg, & Seewald, 2002).

### **Hearing Instrument Orientation and Training**

Orientation and training should include family members, caregivers, and, when appropriate, the child. This information also must be communicated to the child's educators through interactions with the educational audiologist, deaf and hard-of-hearing specialist, or other qualified personnel. Orientation and training should be discussed, demonstrated, and sent home in a written or video format. Orientation and training may take place over several appointments based on the family and child's ability to perform tasks.

Orientation and training should include:

- A. care of the hearing aids, including cleaning and moisture concerns
- B. suggested wearing schedule and retention
- C. insertion
- D. removal
- E. overnight storage (including the mechanism for turning off the hearing aids)
- F. insertion and removal of the batteries
- G. battery life, storage, disposal, toxicity
- H. basic troubleshooting (batteries, feedback, plugged earmold and/or receiver)
- I. telephone coupling and use
- J. assistive device coupling and use
- K. moisture solutions (e.g., dehumidifying systems and covers)
- L. tools for maintenance and care (e.g., battery tester, listening stethoscope, earmold air blower)
- M. issues of retention/compliance/loss (including spare hearing aids and any loaner program)
- N. recommended follow-up appointments to monitor use and effectiveness

### **Validation**

Validation of aided auditory function is a demonstration of the benefits and limitations of aided hearing abilities and begins immediately after the fitting and verification of amplification. Validation is an ongoing process designed to ensure that the child is receiving optimal speech input from others and that his or her own speech is adequately perceived (Pediatric Working Group, 1996). In addition to ongoing monitoring of the amplification device, objective measures of aided performance in controlled clinical environments and in real world settings may be included in the validation process. Functional assessment tools assist in the monitoring process by evaluating behaviors as they occur in real-world settings. These tools are typically



questionnaires designed for administration to parents and teachers or assessments that can be conducted in the child's school environment.

*A. Aided speech perception measures*

Aided speech perception tasks including, but not limited to, the Low-Verbal Early Speech Perception Task and the Early Speech Perception Task (ESP; Moog & Geers, 1990), Phonetically Balanced Kindergarten List (PBK; Haskin, 1949), Northwestern University's Children's Perception of Speech Test (NUCHIPS; Katz & Elliott, 1978), Pediatric Speech Intelligibility Test (PSI; Jerger, Lewis, Hawkins, & Jerger, 1980) may be used in the validation process.

## B. Functional Assessment Tools

1) Tasks conducted in the classroom setting or questionnaires completed by educators such as the Functional Listening Evaluation (FLE; Johnson & Von Almen, 1997), the Screening Instrument for Targeting Educational Risk (SIFTER; Anderson, 1989), the Screening Instrument for Targeting Educational Risk in Pre-School Children (pre-school SIFTER; Anderson & Matkin, 1996) may be used for functional assessment, and the Listening Inventory for Education questionnaire (LIFE; Anderson & Smaldino, 1996).

2) Questionnaires completed by parents or caregivers such as the Children's Home Inventory of Listening Difficulties (CHILD; Anderson & Smaldino, 2000), the Family Expectation Worksheet (FPW; Palmer & Mormer, 1999), the Early Listening Function (ELF; Anderson, 2002), the Meaningful Auditory Integration Scale (MAIS; Robbins, Renshaw, & Berry, 1991), the Infant-Toddler MAIS (IT-MAIS; Zimmerman, Osberger, Robbins, 1998), the Meaningful Use of Speech Scale (MUSS; Robbins, Svirsky, Osberger & Pisoni, 1998), and the Functional Auditory Performance Indicators (FAPI; Stredler-Brown & Johnson, 2001) also may provide useful validation mechanisms.

### **Follow-up and Referral**

Parents and other family members or individuals who will assist in caring for the amplification system should receive orientation, training, and ongoing support and appropriate referral as needed from the audiologist. The audiologist is a key professional who can provide education or refer families to those who can educate them about hearing loss.

Fitting of personal amplification in an infant or young child is an on-going process.

- Minimally, an audiologist should see the child every three months during the first two years of using amplification and every 4-6 months after that time (The Pediatric Working Group, 1996).

Follow-up appointments should include:

- A. Behavioral audiometric evaluations
- B. Current assessment of communication abilities, needs, and demands
- C. Adjustment of the amplification system based on updated audiometric information and communication demands
- D. Periodic electroacoustic evaluations
- E. Listening checks
- F. Earmold fit check
- G. Periodic probe-microphone measurements (at a minimum, following replacement of earmolds)
- H. Periodic functional measures to document development of auditory skills (see previous section number 8: Validation)
- I. Long-term follow-up including academic progress (tools may include the Meadow-Kendall Social-Emotional Scales (Meadow-Orlans, 1983).

On-going auditory habilitation should be provided as part of a team of professionals including, but not limited to, audiologists, early interventionists, deaf and hard-of-

hearing specialists, speech-language pathologists, classroom teachers, pediatricians, or pediatric otologists with the primary focus to support families in the development of the communication abilities of their children.

- J. The prudent audiologist will want to help the parent or guardian make sure that the hearing aids are covered for loss, damage, and repair at all times. For a variety of reasons, the pediatric population has a fairly high rate of loss, damage, and repair. Coverage may be available through the hearing instrument company, a hearing aid insurance company, or a homeowner's policy.

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# PEDIATRIC AMPLIFICATION GUIDELINES

## COCHLEAR IMPLANTS

This particular section of the Tennessee Newborn Hearing Screening Pediatric Audiology Assessment and Amplification Guidelines document is a culmination of the contributing authors' expertise and adapted information from national consensus and technical statements on cochlear implants, from organizations including the American Academy of Audiology (AAA – 1995, 2003) and the American Speech-Language-Hearing Association (ASHA - 2004).

### **Introduction**

Some children with profound deafness develop oral communication skills with conventional hearing aids; however, many do not. Failure to develop adequate communication skills may have a detrimental impact on educational and eventual employment opportunities. It is widely recognized that cochlear implants are an appropriate option for children with prelingual or postlingual severe to profound hearing impairments who demonstrate limited or no functional benefit from conventional hearing aid amplification. Cochlear implants may offer improved sound and speech detection and improved auditory perception of speech. It is further recognized that parents (or legal guardians) have the right to choose cochlear implants if they decide that they are the most appropriate option for their child.

### **Background & History**

A cochlear implant is an electronic prosthetic device which includes both internal and external components that is surgically placed in the inner ear and under the skin behind the ear for the purpose of providing useful sound perception via electrical stimulation of the auditory nerve. The internal components consist of the receiver/stimulator and electrode array while the external components consist of the sound processor, microphone, and transmitter. Internal and external components communicate via a radio frequency signal that is transmitted across the skin. Cochlear implant research and development is ongoing to improve the devices, refine the sound processing strategies, and increase the ability of the user to perceive auditory information.

The law requires the safety and efficacy of a cochlear implant to be demonstrated through clinical investigations before the device can be commercially marketed as accepted clinical practice. Following years of extensive testing, the U.S. Food and Drug Administration approved the first multichannel cochlear implant as medically safe for use in adults with profound hearing loss (1984) and children over age two (1990). Currently, the FDA has broadened the candidacy criteria to include adults with severe hearing loss, older children with severe-to-profound hearing loss and reduced speech discrimination ability and children as young as 12 months of age. Cochlear implants also have been found to be medically safe by the American Academy of Otolaryngology-Head and Neck Surgery, the American Medical Association, and are supported by many health insurance companies.

## **Cochlear Implant Benefits**

Studies on the efficacy of cochlear implants in the pediatric population have reported postoperative speech perception and speech production results in postlingually deafened children and in children with congenital or acquired prelingual deafness. All children, especially those implanted at a young age, demonstrated improvement in sound detection and in their auditory perception skills following implantation. In addition, research has shown that children with cochlear implants achieved performance levels that exceeded those of their non-implanted peers who used other sensory aids, including conventional hearing aids and vibrotactile aids.

Further, studies also have shown improvement in speech production skills and overall speech intelligibility in children with prelingual deafness. Although much sound awareness is observed following implantation, improvements in auditory speech recognition and speech production occur over an extended time in prelingually deafened individuals who receive cochlear implants. There are large individual differences in the benefit that children derive from cochlear implants due to factors such as age at onset of deafness, age at implantation, amount of cochlear implant experience, educational training, and familial support. Cochlear implant outcomes can be characterized with wide variability across individuals for both adults and children (Eisenberg, 2009). However, measurable performance benefits can be observed and may include : the adjustment to use of the device, growth in auditory skills, and growth in speech-language skills.

## **Guidelines for Candidacy**

The criteria for cochlear implant candidacy have significantly changed since the first multichannel device was approved for children by the Food and Drug Administration (FDA) in 1990. These criteria continue to evolve and are updated with FDA approval of each new cochlear implant system. Thus, specific criteria for candidacy published by cochlear implant manufacturers may vary slightly from one manufacturer to another.

Current general pediatric candidacy criteria for cochlear implants are as follows:

- Age 12 months and older (Children under the age of 12 months may be considered for cochlear implantation if medically indicated.)
- Profound sensorineural hearing loss bilaterally for infants ages 12 to 24 months
- Severe to profound sensorineural hearing loss bilaterally for children older than 24 months
- Limited benefit from the use of appropriately fitted hearing aids after a significant trial period
- Poor aided speech perception scores
- Lack of progress in the development of auditory skills
- High motivation of the child and family to receive the device and participate in follow-up and habilitation
- Realistic expectations about device use



The audiologic evaluation for candidacy defines the type and extent of hearing loss as well as the ability to hear and understand speech when using hearing aids. The techniques used for this assessment depend on the age and developmental status of the child.

For the hearing evaluation, the audiologist chooses a battery of age-appropriate physiologic and behavioral tests to determine hearing status. Physiologic measures of hearing may include auditory brainstem response thresholds for clicks and tonal stimuli, otoacoustic emissions, tympanometry using an appropriate probe tone frequency, and acoustic reflex thresholds. Behavioral hearing testing via air and bone conduction may include behavioral observation audiometry, visual reinforcement audiometry, tangible reinforcement operant conditioning audiometry, conditioned play audiometry or standard methods. Speech audiometry may be conducted using age-appropriate word lists and response format.

The assessment of amplification and aided performance includes objective hearing aid measures, parent/child questionnaires, and aided speech perception testing. The appropriateness of the child's hearing aid fitting should be evaluated using real ear measures. When actual real ear measures cannot be obtained, simulated real ear measures with real-ear to coupler-difference corrections are a suitable alternative. Parent/child questionnaires are used to determine how the child functions with the current amplification devices in everyday environments. Examples of such questionnaires may include but are not limited to:

- **Early Listening Function - ELF** (ages 0 to 3 years),
- **Infant Toddler Meaningful Auditory Integration Scale - IT-MAIS** (ages 0 to 2 years)
- **Meaningful Auditory Integration Scale - MAIS** (ages 3 to 5 years)
- **Children's Home Inventory for Listening Difficulties - CHILD** (school-age child)
- **The Listening Inventory for Education - LIFE** (school-age child)
- **Screening Identification for Targeting Educational Risk – SIFTER** (preschool, school-age, secondary) also can be used when appropriate.

Aided speech perception scores satisfy an important criterion in the determination of candidacy. This testing may be conducted using closed-set and/or open-set materials. Recorded presentation is recommended, however, monitored live voice may be used as needed. The audiologist chooses at least one developmentally appropriate test to be administered in right aided, left aided and binaurally aided conditions. This will establish the child's best aided condition which can be used for the remainder of the aided speech perception test battery. When possible, it is recommended that scores for both word and sentence materials be obtained. It also is ideal to obtain at least one measure in the presence of background noise. Children ages 25 months to 17 years are candidates to receive cochlear implants if they have poor performance (20 to 30 % correct) on open-set word recognition tasks.

An otologist performs a medical evaluation to determine if a child is a candidate to receive a cochlear implant. This evaluation may include history, physical examination and imaging studies of the temporal bone. The child should be free of active ear disease and be an

acceptable candidate for general anesthesia. High resolution computed tomography (CT) scan, magnetic resonance imaging (MRI), or both are necessary to identify the cochlear landmarks and patent internal auditory canal. Electrical promontory stimulation may be indicated in suspected cases of auditory nerve insufficiency.

While the audiologist and the cochlear implant surgeon form the core of the cochlear implant team, comprehensive candidacy evaluation may include speech-language evaluation, assessments of cognitive skills, educational performance, and general behavior, as well as assessment of family and child expectations for device use. Additional evaluations by professionals from other disciplines such as social work, developmental pediatrics, neurology, ophthalmology, or occupational therapy may be sought by the cochlear implant team as needed.

The implant components and function, the risks, limitations, and potential benefits of implantation, the surgical procedure, and the postoperative follow-up schedule should be discussed with parents or guardians and the child (if appropriate). Children should be enrolled in educational programs that support the use of cochlear implants and the development of auditory and speech skills, regardless of the communication method employed. It is further recommended that the family be fully informed about alternatives to implantation, and Deaf Culture.

In 2002, the CDC and FDA determined that children with cochlear implants are more likely to get bacterial meningitis than children without cochlear implants. Therefore, the CDC recommended in 2003 that people with cochlear implants follow these recommended guidelines for pneumococcal vaccinations:

- Children with cochlear implants less than 2 years of age should receive pneumococcal conjugate vaccine (PCV-7) (Prevnar®) as is recommended for all children
- Children with cochlear implants 2 years of age and older who have completed the PCV-7 series should receive one dose of the pneumococcal polysaccharide vaccine (PPV-23) (Pneumovax®). If they have just received PCV-7, they should wait at least two months before receiving PPV-23.
- Children with cochlear implants between 24 and 59 months of age who have never received either PCV-7 or PPV-23 should receive two doses of PCV-7 two to more months apart and then received one dose of PPV-23 at least two months later.
- Persons 5 years of age and older with cochlear implants should receive one dose of PPV-23. Revaccination is not indicated.

Despite the efforts of the CDC to recommend these vaccinations, many children have still not been vaccinated. Therefore, in October, 2009 the American Academy of Otolaryngology- Head and Neck Surgery (AAO-HNS) supported all three cochlear implant manufacturers in an effort to raise awareness of the importance of every cochlear implant patient receiving the pneumococcal vaccination. The audiologist should provide each candidate with information regarding these vaccinations prior to cochlear implantation. Those who plan to receive a

cochlear implant should be up-to-date on age appropriate pneumococcal vaccination at least 2 weeks before surgery, if possible.

Additional information may be found at [www.cdc.gov/mmwr/preview/mmhtml/m2e731a1.htm](http://www.cdc.gov/mmwr/preview/mmhtml/m2e731a1.htm).

### **Guidelines for Managements: Initial Stimulation**

Children who receive cochlear implants require ongoing audiological management and medical follow-up. Ongoing management by an audiologist includes programming the implant processor and monitoring device and user performance.

Typically, the initial stimulation is scheduled (4) four weeks post implantation; however, schedules for initial activation can vary widely by physician preference. This time is set to allow for healing of the incision and for reduction of swelling. At the initial stimulation appointment, the audiologist should check the incision to be sure it is healing. If any concerns are noted, the surgeon should be contacted immediately. A copy of the surgical report and a post-operative x-ray, if obtained, will reveal any variations in the patient's individual ear anatomy and if the surgeon was able to complete a full insertion of the electrode array. The audiologist should communicate with the surgeon about which electrodes have acceptable impedance values.

The cochlear implant is programmed based on each patient's individual auditory perception in response to auditory stimuli. The goal of initial stimulation is to obtain audibility for speech and environmental sounds while maintaining comfort for all sounds. The initial map or program should be created by measuring Threshold (T) and Comfort (C)/Most Comfortable (M)/Most Comfortable Loudness (MCL) levels. The T and C/M/MCL levels will change while the child is beginning to listen with their cochlear implant and eventually stabilize. Behavioral measures may be difficult to obtain for younger children and children with little or no auditory skills. Depending on the child's age and ability to participate in the mapping session, objective measures such as Electrically Evoked Compound Action Potentials (ECAP) such as Neural Response Telemetry (NRT), Neural Response Imaging (NRI), Auditory nerve Response Telemetry (ART) and Electrically evoked Stapedial Reflex Thresholds (ESRT) may be used to help determine the map settings. However, every effort to obtain actual behavioral responses should be pursued. Objective measures are useful to support behavioral responses or as a last resort to set program levels. There may also be cases when objective measures cannot be obtained. The audiologist should remember that these behavioral and objective measures may change over time and should, therefore, be monitored and re-measured on a consistent basis or as needed. The audiologist should have age appropriate toys, reinforcement and rewards that are easily accessible. The clinic area should have space for family and video equipment. The family should have a clear understanding of what to expect during an initial mapping session. Some audiologists recommend that the child continue to use a hearing aid even if the child is not receiving any benefit or utilize an unmapped speech processor prior to the initial stimulation. Use of the hearing aid or processor may allow the child some time to adjust to the physical comfort of the device *and* allow the child to be initially more accepting of their own processor.

The audiologist should refer to each manufacturer's individual programming guidelines and training manual to specific details regarding initial stimulation and re-programming. The

following are suggested guidelines and are not intended to replace the training provided by each manufacturer.

#### *Prior to Initial Stimulation Appointment*

- Check all equipment to ensure all equipment and parts were included in order.
- Charge the rechargeable batteries.
- Visually inspect all cables and parts and be certain that the speech processor is recognized by the programming software.
- Optimally, the clinic should have extra parts, supplies and accessories on hand.

#### *Initial Stimulation Appointment*

- Review Expectations with family and child (if age appropriate).
- Check incision to be certain it is healing well and notify surgeon immediately if there is any concern.
- Complete otoscopy and tympanometry.
- Connect device to computer and coil to child's head. Verify serial number of connected device.
- Verify the strength of the magnet. The magnet should show appropriate attraction but not be so tight that it leaves an impression in the skin.
- Complete Impedance Telemetry and view results: Allows for a quick check of individual electrode function, actual impedance levels and any flags for short or open electrodes, manually flag any known extra cochlear electrodes (if not completed in the OR). Electrodes that are either open or short as well as electrodes that elicit non-auditory side effects may need to be deactivated.
- Complete Objective Measures: ECAP or ESRT
- Measure Behavioral Responses: Attempt to measure at least 5 channels and interpolate between channels to create an initial map. If measuring T levels, VRA, CPA or counting may be used. If measuring M or C or MCL levels, visual supports for loudness scaling such as picture cards with images of graduated size may be used. Since children may not indicate first hearing until the sound has become sufficiently loud, set M or C or MCL levels at measured T levels and activate the live map. T levels may be autoset within the software or can be manually set at a fixed range below the upper levels.
- Activate map and gradually increase or decrease C/M/MCL levels based on patient response
- Confirm equal loudness and pitch changes by loudness balancing and sweeping channels
- Be sure indicator lights, volume and sensitivity, etc are set according to preference
- Create progressive maps
- Save to primary processor and backup processor
- Use Ling 6 sound test to verify perception of speech sounds for children who are old enough to detect and/or discriminate

#### *Bilateral Programming*

Bilateral programming may be completed sequentially or simultaneously. The audiologist should be certain that both implants are entered into the same record in the programming software. When programming sequentially, follow the above recommendations for device #1 and then for device #2. Once each device has an appropriate map, then balance the two

devices to ensure bilateral equalization. This step may not be possible for very young children who cannot report equalization. If balancing cannot be completed, set the volume slightly lower than the manufacture's recommended settings and gradually increase to recommended volume. The steps for bilateral programming may also be completed simultaneously. The audiologist should use caution and carefully note which serial number is connected to which programming interface to ensure correct programming of each individual device.

### *Counseling and Orientation*

- Parts of Processor
- Coil/Headpiece
- Magnet: strength, placement, watch incision site
- Batteries: disposable, rechargeable (charger), precautions
- Listening Check: daily listening check with monitor earphones
- Care and Troubleshooting: storage, cleaning, Dry and Store
- Accessories
- Wearing schedule: begin listening in P1 and gradually progress through programs
- Parents should always have extra rechargeable and/or disposable batteries on hand.
- Parent may want to use a "wake up" program one level softer than where child is using device for 30 minutes to an hour each morning.
- Complete product registration and warranty forms. Discuss extended warranty and supplemental insurance options.
- Schedule appointments for follow-up.

### *Programming Schedule*

- Initial Stimulation (IS)
  - 4-6 weeks post-op (or sooner at physician's discretion)
- Follow-up Programming
  - 1-2 weeks post IS
  - 1 month post IS
  - 2 months post IS
  - 3 months post IS
  - 6, 9, 12 months post IS
- Under age 7, continue reprogramming every three months for at least the first year, then every six months for the second and third years.
- Over age 7, continue reprogramming every six months for the first three years of implant use.

Follow-up programming may be requested at any time if there is a change in a child's responsiveness to sound, a change in the quality or intelligibility of a child's speech, an unusual change in a child's general behavior that is unexplained by an external equipment problem or if any other concerns arise.

## ***Guidelines for Management: Ongoing Monitoring***

### *Medical Follow-Up*

It is imperative that CI users consult their CI surgeon and programming audiologist on a regular basis. The audiologist supports the child and works in conjunction with the surgical office to care for the patient. The CI user should return to the surgical office as recommended by the surgeon; however, the audiologist may also refer back to the surgeon if the following conditions occur:

- Middle ear dysfunction
- Pain or irritation at the surgical site or at the magnet site
- Facial nerve stimulation or irritation upon activation of the cochlear implant
- Poor patient follow-up
- Consultation regarding bilateral implantation
- Any other extenuating circumstance/concerns

The audiologist should also be aware of “red flags” which could indicate the map may need adjusting: facial nerve stimulation, increased channel interaction, low battery life, and voltage compliance issues.

### *Audiologic Follow-Up*

The goal of on-going mapping is to maintain audibility of speech and environmental sounds as well as comfort for loud sounds. The cochlear implant user may be scheduled as indicated in the Follow-Up Programming Schedule noted above. Follow-up appointments should include objective, subjective, and verification measures of cochlear implant benefit, but may be completed at various intervals of care. The following are recommended tools to assess CI function and benefit. Post-activation subjective and verification measures may be compared to pre-implant evaluation results to assess overall benefit from cochlear implant use and to monitor audiologic progress.

### *Objective Measures*

- Tympanometry to confirm middle ear function. CI user should be referred back to surgeon for treatment if middle ear effusion is present.
- Visual inspection of cochlear implant system and listening check of processor microphone, if possible. Audiologist should inspect all parts of cochlear implant to ensure proper function.
- Telemetry (i.e. Impedance, ECAP, Voltage Compliance levels). Impedances should be considered at each visit; however, ECAP and check of compliance may be evaluated as needed to adjust map settings.
- Electrically evoked stapedial reflex thresholds (ESRT) may be measured using stimulation through the cochlear implant programming software and recorded via standard impedance test equipment. These measures, if available, may provide

information for the setting of C, M or MCL levels within a child's cochlear implant program or map.

### *Behavioral Measures*

Psychophysical measures (T, C, M, MCL) are reassessed at follow-up visits in an effort to maintain audibility and comfort for each listening program. As mentioned above, T, C, M, and MCL levels are expected to change significantly in the first few months of cochlear implant use and then eventually to stabilize. While initial programs may be created using levels collected for a small group of electrodes, further programming sessions will allow for the assessment of all electrodes.

As children mature and develop auditory skills, they also can participate in more advanced listening tasks designed to make the setting of their map levels more precise. Program levels may initially be assessed in large step sizes which can be refined to smaller units during subsequent programming sessions. If thresholds are assessed, children may move from simple detection to counting tasks. Upper loudness levels may initially be set using clinician judgment about what seems comfortable or uncomfortable based on behavioral observation. Later, children may be engaged in making their own judgments about loudness using a "big sound vs. little sound" task. As they become more sophisticated listeners, children may discern finer gradations of loudness using a three to seven step loudness scaling procedure.

### *Verification and Validation Measures (Sound Awareness/Speech Perception)*

Individual variability amongst cochlear implant users presents a challenge in selecting the appropriate outcome measure. Factors that affect the selection process range from age of the user, amount of residual hearing, age of onset of hearing loss, speech perception, speech production, language, and cognitive skills. Because the goal of cochlear implantation in children in relation to the development of speech and language is extremely broad, so should be the assessment battery. A hierarchy of skills that range from sound discrimination to comprehension of connected speech is necessary for a comprehensive evaluation. However, it is at the audiologist's discretion when each skill will be assessed as the child progresses. The following are examples of assessment tools:

- **Sound Field Verification:** Assessment of detection skills for frequencies ranging from 250 to 4000Hz with either a warble tone (preferred) or narrowband noise while the processor is set to a typical use setting. It is common for implant recipients to detect sounds in the range of 20 to 30 dB HL. If thresholds exceed those levels, the audiologist should check the settings of the processor, troubleshoot the equipment, and consider re-mapping the processor to increase audibility.
- **Ling Six Sound Test:** Traditionally performed live voice to assess detection or identification of sounds that lie within the speech spectrum of hearing (/m/, /oo/, /ah/, /ee/, /sh/, and /s/). The level of difficulty of the task may be varied to fit the user's abilities.
- **Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS):** For children younger than 2 years of age. This is a structured interview schedule designed to assess the child's spontaneous responses to sound in his/her everyday environment. Based upon

parental report that assesses vocalization behavior, alerting to sounds and deriving meaning from sound.

- Meaningful Auditory Integration Scale (MAIS): Minimum age for this test is 2 years of age. The questionnaire, from which the IT-MAIS was modified, assesses a child's meaningful listening skills in everyday situations. Parental response to client administered questions is sought to determine the child's history with their hearing device.
- Early Speech Perception (ESP) Test, Low Verbal Version: The minimum age for this test is 2 years or when vocabulary has been acquired. Estimates speech perception abilities in very young children with limited verbal abilities with a closed-set object identification procedure.
- Multi Syllabic Lexical Neighborhood Test (MLNT): For use with children ages 2 to 5 years. This test was designed to assess a child's open-set speech recognition abilities using multi-syllabic words. Twenty-four words, either lexically "easy" or "hard" are presented.
- Northwestern University-Children's Perception of Speech (NU-CHIPS) Test: Minimum age requirement is an age equivalency of 2.5 years on the Peabody Picture Vocabulary Test. The NU-CHIPS assesses the word recognition abilities in children in a closed-set format containing four possible pictures.
- Word Intelligibility by Picture Identification (WIPI) Test: Minimum suggested age for this test is 5 years, but it may be attempted on younger children. The WIPI is designed to evaluate a child's ability to perceive words using a closed-set containing six possible picture stimuli.
- Lexical Neighborhood Test (LNT): (ages 5 to 17 years) Designed to assess a child's open-set speech recognition abilities using monosyllabic words. Fifty words, either lexically "easy" or "hard" are presented.
- Glendonald Auditory Screening Procedure (GASP): Minimum age is 5 years. Designed to assess a child's open-set speech recognition abilities using both words and sentences that are familiar. Three lists of 12 words and 2 lists of 10 sentences are presented.
- Early Speech Perception (ESP) Test, Standard Version: Minimum age for this test is 6 years of age. This is a closed-set, picture identification test designed to assess the progression of speech discrimination skills in children. The child is asked to point to a picture of the word that is presented live voice or via recording.
- PBK-50 Word List: Minimum age is 6 years. Open-set format with 50 monosyllabic words that are scored for accuracy.
- Bamford-Kowal-Bench Sentences (BKB): Minimum age is 6 years. This test assesses speech recognition at a sentence level using key words in sentences. A percent correct score is derived from an open-set.
- Hearing-In-Noise Test for Children (HINT-C): Designed for children ages 6 to 12 years. Assesses a child's open-set speech recognition abilities with a signal to noise ratio. Stimuli may be initially presented in quiet and progress to noise as deemed appropriate.



Using these tools as well as patient, parent, teacher and aural habilitation therapist report regarding the use, benefit, and progress with the implant, the audiologist may decide to re-program the device to optimize speech perception abilities.

### ***Other Considerations***

#### ***FM Systems***

Children with cochlear implants may greatly benefit from the use of an FM system, just as a child with hearing aids. Post-implantation verification and validation measures may indicate significant improvements in sound awareness and discrimination, but the implementation of an assistive listening device, such as an FM system, may support the child in making listening easier and less stressful.

All current sound processors are FM compatible via telecoil or direct audio input; however, unlike with hearing aids, it is not possible to monitor the FM signal through the cochlear implant processor. For this reason, it is advisable to postpone the acquisition of a personal FM system until the child acquires the ability to report on the sound quality of the FM system. Until that time, a sound field system or portable desktop system may be considered.

School systems can provide support to the programming audiologist in many ways. Schools can assist in device acquisition and maintenance. In addition, school personnel can communicate with the programming audiologist issues related to the child's functional performance while wearing the FM in the classroom (ie. changes in speech perception abilities). In order for the child to consistently reap the benefits of FM technology, school personnel must be trained in the use, care, and troubleshooting of the FM system. In many cases, the school system has a supply of FM system components that may be compatible with the child's implant processor. However, once a child is ready for a personal FM system, the audiologist should consider all current technology as well as the option for future upgrades in processor equipment to determine the most suitable FM device for the child.

#### ***Bimodal Stimulation***

Children who use one cochlear implant may benefit from the use of a hearing aid for the non-implanted ear. Evidence has not yet suggested a preferred hearing aid to be used in conjunction with a cochlear implant, so decisions about devices need to be made on an individual basis. When such fitting is conducted, attention should be devoted to loudness balancing of the cochlear implant and hearing aid. Speech perception measures should be conducted to determine the ability of the child to understand speech with stimulation to the cochlear implant only, hearing aid only, and bimodally (CI+HA). This will ensure that the hearing aid does not interfere with the overall perception of speech.

#### ***Clinical Preparedness***

In recent years, cochlear implant sound processors have become smaller, device programs more sophisticated, and surgical techniques more refined. With the rapid rate of advancing technology in the cochlear implant industry and the introduction of new applications of cochlear implant devices, it is imperative that audiologists strive to remain current in their knowledge and skills through professional development opportunities. Examples of these

opportunities would include, but not be limited to: manufacturer-sponsored workshops and updates, web-based training, attendance at national or regional cochlear implant conferences, and independent study of current research and relevant topics. In addition to having up-to-date skills and knowledge related to implant technology, audiologists who will be working with children who have cochlear implants must also be prepared to appropriately counsel their families both prior to and following the implant surgery and provide continued guidance during the early intervention process. With this said, the concept of clinical preparedness must include professional development opportunities that address issues of family support.

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- <http://www.cdc.gov/mmwr/preview/mmwrhtml/m2e731a1.htm>

## Appendix 1

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#### Section Key:

Behavioral Assessment (B), Cochlear Implant (CI), Follow-Up (FU),  
Genetics (G), Hearing Aids (HA), Physiologic Assessment (P)

## Appendix 2

### **Risk Indicators for Progressive and Delayed Onset or Acquired Hearing Loss**

*per Joint Committee on Infant Hearing (JCIH) Position Statement '07  
and American Academy of Pediatrics*

Caregiver concern § regarding hearing, speech, language, or developmental delay.

Family history § of permanent childhood hearing loss.

Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO §, assisted ventilation, exposure to ototoxic medications (Gentamycin and Tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.

In utero infections, such as CMV§, herpes, rubella, syphilis, and toxoplasmosis.

Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.

Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.

Syndromes associated with hearing loss or progressive or late-onset hearing loss §, such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.

Neurodegenerative disorders§, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.

Culture-positive postnatal infections associated with sensorineural hearing loss §, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.

Head trauma, especially basal skull/temporal bone fracture § that requires hospitalization.

Chemotherapy §.

*Indicators marked with the section symbol (§) are of greater concern  
for the development of delayed-onset hearing loss.*

## Appendix 3

### National and Tennessee Hearing Resources

#### National Hearing Resources

- American Academy of Pediatrics (AAP) Universal Newborn Hearing Screening Diagnosis and Intervention, Guidelines for Pediatric Medical Home Providers (chart) [www.aap.org](http://www.aap.org)
- American Association for Deaf Children 800-942-ASDC [www.deafchildren.org](http://www.deafchildren.org)
- Alexander Graham Bell Association for the Deaf and Hard of Hearing (AG Bell) 866-337-5220 [www.agbell.org](http://www.agbell.org)
- Auditory Verbal International 703-739-1049 [www.auditory-verbal.org](http://www.auditory-verbal.org)
- BEGINNINGS 800-541-HEAR <http://www.ncbegin.org/index.php>
- Boys Town National Research Hospital [www.babyhearing.org](http://www.babyhearing.org)
- Centers for Disease Control and Prevention Early Hearing Detection and Intervention (CDC-EHDI) <http://www.cdc.gov/ncbddd/hearingloss/index.html>
- Cochlear Implant Association, Inc. (CIAI) [www.cici.org](http://www.cici.org)
- Families for Hands and Voices [www.handsandvoices.org](http://www.handsandvoices.org)
- Hearing Loss Association of America <http://www.hearingloss.org/>
- John Tracy Clinic 800-522-4582 [www.johntracyclinic.org](http://www.johntracyclinic.org)
- Laurent Clerc National Deaf Education Center [www.clercenter.gallaudet.edu](http://www.clercenter.gallaudet.edu)
- The Listen-Up! [www.listen-up.org](http://www.listen-up.org)
- National Association of the Deaf (NAD) 301-587-1788 [www.nad.org](http://www.nad.org)
- National Center for Hearing Assessment and Management (NCHAM) [www.infanthearing.org](http://www.infanthearing.org)
- National Cued Speech Association [www.cuedspeech.org](http://www.cuedspeech.org)
- National Institute of Deafness and Other Communication Disorders [www.nidcd.nih.gov](http://www.nidcd.nih.gov)
- Oberkotter Foundation - Oral Deaf Education [www.oraldeafed.org](http://www.oraldeafed.org)
- The S.E.E. (Signing Exact English) Center [www.seecenter.org](http://www.seecenter.org)

#### Tennessee Directories

- Tennessee Directory of Services for People who are Deaf and Hard of Hearing 800-342-3262 [www.tndeaflibrary.nashville.gov](http://www.tndeaflibrary.nashville.gov)
- Tennessee Department of Health Newborn Hearing Program Pediatric Audiology and Hearing Providers List (Full directory or region-by-region one page list of providers) 615-262-6160
- Tennessee Department of Health Newborn Hearing Program Hospital Guidelines 615-262-6160
- TEIS District Directories of Services for Infants and Toddlers with Disabilities 800-852-7157

#### Tennessee Departments, Agencies and Organizations

- Newborn Hearing Screening (NHS) Tennessee Department of Health program that promotes and coordinates statewide newborn hearing screening, assessment, intervention, and follow-up. 615-741-8530 or 615-262-6160. [www.state.tn.us/health/MCH/NBS/index.html](http://www.state.tn.us/health/MCH/NBS/index.html)
- Tennessee Early Intervention System (TEIS) 800-852-7157 [www.state.tn.us/education/teishome.htm](http://www.state.tn.us/education/teishome.htm)

- Children’s Special Services (CSS) Tennessee Department of Health program that provides medical services, care coordination and the Parents Encouraging Parents (PEP) support services to qualifying children under 21 yrs. with a chronic illness or medical condition. 615-741-8530 [www.state.tn.us/health/MCH](http://www.state.tn.us/health/MCH)
- Family Voices: Family support network and advocacy group for all children and youth with special health care needs, 888-643-7811 [http://www.tndisability.org/coalition\\_programs/family\\_voices/newborn\\_hearing\\_program](http://www.tndisability.org/coalition_programs/family_voices/newborn_hearing_program)
- Library Services for the Deaf and Hard of Hearing 800-342-3262 [www.tndeaflibrary.nashville.gov](http://www.tndeaflibrary.nashville.gov)
- TREDIS (Tennessee Project for Individuals with Combined Vision and Hearing Loss) 800-288-2266 or 615-936-2862 <http://www.treds-deafblindproject.com/>

## Appendix 3 (cont.)



# NEWBORN HEARING SCREENING ORDER FORM FOR MATERIALS

Tennessee Newborn Hearing Screening (NHS) Program materials are available for hospitals, audiology providers, and others and at no charge. You are encouraged to share materials with families of child bearing age, pregnant women and parents of newborns. Materials provide helpful information about NHS and contain contact information for parents to obtain services.

### Please complete the information below:

Indicate number of materials requested in appropriate boxes. Brochures and forms come in packs of 100.

Hospital or Provider Name: \_\_\_\_\_

Send Attention: \_\_\_\_\_

Address: \_\_\_\_\_

Phone: \_\_\_\_\_

#### BROCHURES:

- Newborn Hearing Screening (for expectant or new parents) (English)
- Your New Baby's Hearing (for expectant or new parents) (Spanish)
- Newborn Hearing Screening: What Next? (for families with baby who referred on initial screen) (English)
- Your Baby's Hearing Screening Suggests a Referral (for families with baby who referred on initial screen) (Spanish)
- Talking with Parents about Hearing Loss (for physicians, hearing screeners, nurses, audiologists) English

#### POSTERS:

- No Child Is Too Young To Test (8"x17") (Picture of Infant) English
- No Child Should Miss Out On Life Because of Hearing Loss (8"x17") (Picture of Adolescent) English

#### AUDIOLOGIST, MEDICAL PROVIDER AND INTERVENTION RESOURCES and FOLLOW-UP REPORTS:

- Report of Infant Hearing Rescreen or Diagnostic Evaluation (to be used by physicians and audiology providers to report results to State)
- Family Voices Newborn Hearing Parent Notebook (to be given to families of child with newly-identified hearing loss)
- Communicate with Your Child (brochure published by National Centers for Hearing Assessment and Management – NCHAM; specifically designed for parents of children with hearing loss)

#### REPORTING FORMS for HOSPITALS:

- **Hearing Screening Only** form and instructions for use  
These forms are to be used by hospitals and birthing facilities to submit hearing screening results on newborns who received a hearing screening **after the newborn screening blood collection** specimen had been submitted to the TN State Lab. Please document the specimen control number (SCN), if available, from the previous blood specimen form to assure a link to the initial screening.

#### GUIDELINES:

- TN Hospital and Birthing Center, Newborn Hearing Screening Guidelines
- TN Early Intervention, Newborn Hearing Follow-Up Guidelines
- TN Pediatric Audiologic Assessment and Amplification Guidelines
- TN Directory of Pediatric Hearing Screening & Audiologic Diagnostic and Early Intervention Providers (full directory)
- Region-by-Region Directory of Pediatric Hearing Screening & Audiologic Diagnostic Providers (a one page list for each of the three state regions: east, middle, west)

**Fax Completed Form to: 615-262-6159**

Questions: 615-262-6160 (Jacque Cundall)



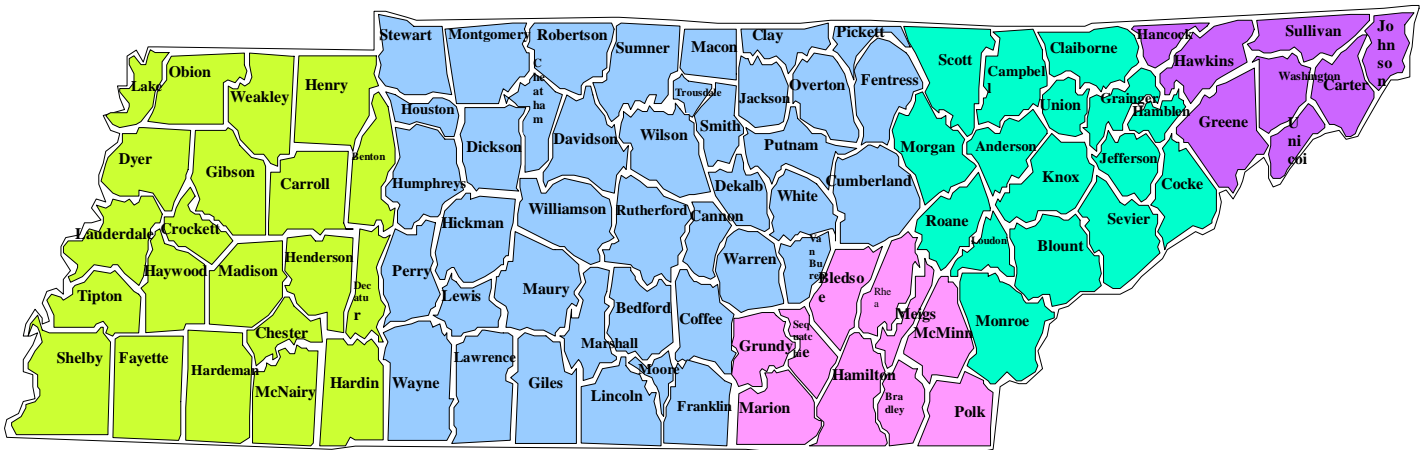
# Appendix 4

## Tennessee Genetic Resources

The Tennessee Department of Health Newborn Metabolic and Hearing Screening programs collaborate with genetic centers located in five regions of the state. Centers provide consultation and evaluation to healthcare providers and families of individuals at risk for or found to have hearing loss.

### Tennessee Genetic Centers

Tennessee Department of Health



**U T Memphis**  
901-528-6594

- |          |            |         |
|----------|------------|---------|
| Benton   | Hardeman   | Madison |
| Carroll  | Hardin     | Obion   |
| Chester  | Haywood    | Shelby  |
| Crockett | Henderson  | Tipton  |
| Decatur  | Henry      | Weakley |
| Dyer     | Lake       |         |
| Fayette  | Lauderdale |         |
| Gibson   | McNairy    |         |

**Vanderbilt Univ.**  
615-322-7601

- |            |           |            |            |
|------------|-----------|------------|------------|
| Bedford    | Giles     | Montgomery | Trousdale  |
| Canon      | Hickman   | Moore      | Van Buren  |
| Cheatham   | Houston   | Overton    | Warren     |
| Clay       | Humphreys | Perry      | Wayne      |
| Coffee     | Jackson   | Pickett    | White      |
| Cumberland | Lawrence  | Putnam     | Williamson |
| Davidson   | Lewis     | Robertson  | Wilson     |
| Dekalb     | Lincoln   | Rutherford |            |
| Dickson    | Macon     | Smith      |            |
| Fentress   | Marshall  | Stewart    |            |
| Franklin   | Maury     | Sumner     |            |

**U T Chatt  
TC Thompson**  
423-778-6112

- Bledsoe
- Bradley
- Grundy
- Hamilton
- Polk
- Marion
- McMinn
- Meigs
- Rhea
- Sequatchie

**U T Knox**  
865-544-9030

- Anderson
- Blount
- Campbell
- Claiborne
- Cocke
- Grainger
- Hamblen
- Jefferson
- Knox
- Loudon
- Monroe
- Morgan
- Roane
- Sevier
- Union

**ETSU**  
423-439-8541

- Carter
- Greene
- Hancock
- Hawkins
- Johnson
- Sullivan
- Unicoi
- Washington

Jan. 2004



## Appendix 4 (cont.)

### Genetic Consultation and Evaluation Related to Hearing Loss

Genetic information provided below has been respectfully taken from the following resources: Smith R., Hildebrand M., and Van Camp G., *Deafness and Hereditary Hearing Loss Overview*, initial posting February 14, 1999; last update October 14, 2010 (<http://www.ncbi.nlm.nih.gov/bookshelf/br.fcgi?book=gene&part=deafness-overview>) and the American College of Medical Genetics Policy Statement (2002) Genetic Evaluation Guidelines for Etiologic Diagnosis of Congenital Hearing Loss. *Genet Med* 4(3):162-171 (<http://www.geneticsinmedicine.org>).

**Diagnosis/testing:** Genetic forms of hearing loss must be carefully distinguished from acquired (non-genetic) causes of hearing loss. The genetic forms of hearing loss are diagnosed by otologic, audiologic, and physical examination, family history, ancillary testing (such as CT examination of the temporal bone), and DNA-based testing. DNA-based genetic tests are available for many types of syndromic and nonsyndromic deafness, although usually only on a research basis. On a clinical basis, DNA-based testing is available for the diagnosis of branchio-oto-renal (BOR) syndrome (EYA1 gene), Mohr-Tranebjaerg syndrome (deafness-dystonia-optic atrophy syndrome; TIMM8A gene), Pendred syndrome (SLC26A4 gene), Usher syndrome type IIA (USH2A gene), one mutation in USH3A, DFNB1 (GJB2 gene), DFNB3 (POU3F4 gene), DFNB4 (SLC26A4 gene), and DFNA6/14 (WFS1 gene). Testing for deafness-causing mutations in the GJB2 gene (which encodes the protein connexin 26) and GJB6 (which encodes the protein connexin 30) plays a prominent role in diagnosis and genetic counseling.

**Prevalence:** Hearing loss is the most common birth defect and the most prevalent sensorineural disorder in developed countries [Hilgert et al 2009]. One of every 500 newborns has bilateral permanent sensorineural hearing loss  $\geq 40$  dB; by adolescence, prevalence increases to 3.5 per 1000 [Morton & Nance 2006].

A small percentage of prelingual deafness is syndromic or autosomal dominant nonsyndromic. More than 50% of prelingual deafness is genetic, most often autosomal recessive and nonsyndromic. Approximately 50% of autosomal recessive nonsyndromic hearing loss can be attributed to the disorder DFNB1, caused by mutations in the *GJB2* gene (which encodes the protein connexin 26) and the *GJB6* gene (which encodes the protein connexin 30). The carrier rate in the general population for a recessive deafness-causing *GJB2* mutation is approximately one in 33.

In the general population, the prevalence of hearing loss increases with age. This change reflects the impact of genetics and environment, and also interactions between environmental triggers and an individual's genetic predisposition, as illustrated by aminoglycoside-induced ototoxicity, middle ear effusion, and possibly otosclerosis.

**Evaluation Strategy:** Correctly diagnosing the specific cause of hearing loss in an individual can provide information on prognosis and is essential for accurate genetic counseling. The following is usually required:

- **Family history:** A three-generation family history with attention to other relatives with hearing loss and associated findings should be obtained. Documentation of relevant findings in relatives can be accomplished either through direct examination of those individuals or through review of their medical records, including audiograms, otologic examinations, and DNA-based testing.
- **Clinical examination:** All persons with hearing loss of unknown cause should be evaluated for features associated with syndromic deafness. Important features include branchial cleft pits, cysts or fistulae; pre-auricular pits; telecanthus; heterochromia iridis; white forelock; pigmentary anomalies; high myopia; pigmentary retinopathy; goiter; and cranio-facial anomalies. Because the autosomal dominant forms of syndromic deafness tend to have variable expressivity, correct diagnosis may depend on careful physical examination of the proband as well as other family members.
- **Audiologic findings:** Hearing status can be determined at any age. Individuals with progressive hearing loss should be evaluated for Alport syndrome, Pendred syndrome, and Stickler syndrome and have temporal bone-computed tomography. Sudden or rapidly progressive hearing loss can be seen with temporal bone anomalies (as in Pendred syndrome and BOR syndrome), neoplasms (associated with NF2), and immunologic-related deafness, as well as trauma, infections (syphilis, Lyme disease), and metabolic, neurologic, or circulatory disturbances.
- **Temporal bone CT:** Computed tomography of the temporal bones is useful for detecting malformations of the inner ear (i.e., Mondini deformity, Michel aplasia, enlarged/dilated vestibular aqueduct), which should be considered in persons with progressive hearing loss. Because inner ear defects (enlarged/dilated vestibular aqueduct and Mondini dysplasia) are associated with mutations in SLC26A4 (see Pendred syndrome), detection of temporal bone anomalies by CT examination can help direct molecular genetic testing (see below).
- **Testing:** Cytomegalovirus (CMV) testing needs to be considered in infants with sensorineural hearing loss. The diagnosis of in utero CMV exposure requires detection of elevated CMV antibody titers or a positive urine culture in the neonatal period. Although these tests can be obtained at a later time, their interpretation is confounded by the possibility of postnatally acquired CMV infection, which is common and is not associated with hearing loss.
- **Molecular genetic testing:** Molecular genetic testing of the GJB2 gene (which encodes the protein connexin 26) and the GJB6 gene (which encodes the protein connexin 30) (see DFNB1), molecular genetic testing should be considered in the evaluation of individuals with congenital nonsyndromic sensorineural hearing loss. Strong consideration also should be given to "pseudo-dominant" inheritance of DFNB1. Pseudo-dominant inheritance refers to occurrence of an autosomal recessive disorder in two or more generations of a family; such inheritance tends to occur when the carrier rate in the general population is high. GJB2 and GJB6 molecular genetic testing should be performed in families with nonsyndromic hearing loss in which two generations are involved.

- Inner ear defects: (enlarged/dilated vestibular aqueduct and Mondini dysplasia) are associated with mutations in SLC26A4 (see Pendred syndrome), and the detection of these temporal bone anomalies by CT examination should prompt consideration of molecular genetic testing.
- Although molecular genetic testing is available for a number of these genes, the large size of many (MYO7A, MYO15) and their low relative contribution to deafness (DFNB9, HDIA1, TECTA, COCH, POU4F3) makes it impractical to offer such testing on a clinical basis at this time.

**Genetic Counseling:** Genetic counseling is the process of providing individuals and families with information on the nature, inheritance, and implications of genetic disorders to help them make informed medical and personal decisions. The following section deals with genetic risk assessment and the use of family history and genetic testing to clarify genetic status for family members. This section is not meant to address all personal or cultural issues that individuals may face or to substitute for consultation with a genetic professional.

Genetic counseling and risk assessment depend on accurate determination of the specific genetic diagnosis. In the absence of a specific diagnosis, empiric recurrence risk figures, coupled with GJB2 and GJB6 molecular genetic testing results, can be used for genetic counseling.

**Mode of Inheritance:** Hereditary hearing loss may be inherited in an autosomal dominant manner, an autosomal recessive manner, or an X-linked recessive manner. Mitochondrial disorders with hearing loss also occur.

#### 1. Risk to Family Members - Autosomal **Dominant** Hereditary Hearing Loss

##### Parents of a Proband

- Most individuals diagnosed as having autosomal dominant hereditary hearing loss have an affected parent; the family history is rarely negative.
- A proband with autosomal dominant hereditary hearing loss may have the disorder as the result of a de novo gene mutation. The proportion of cases caused by de novo mutations is unknown but thought to be small. Recommendations for the evaluation of parents of a proband with an apparent de novo mutation include audiometry and genetic testing. Although most individuals diagnosed with autosomal dominant hereditary hearing loss have an affected parent, the family history may appear to be negative because of alternate paternity, adoption, early death of a parent, failure to recognize hereditary hearing loss in family members, late onset in a parent, reduced penetrance of the mutant allele in an asymptomatic parent, or a de novo mutation for hereditary hearing loss.

##### Sibs of a proband

- The risk to sibs depends upon the genetic status of a proband's parents. If one of the proband's parents has a mutant allele, the risk to the sibs of inheriting the mutant allele is 50%. Depending upon the specific syndrome, clinical severity and

disease phenotype may differ between individuals with the same mutation; thus, age of onset and/or disease progression may not be predictable.

#### Offspring of a Proband

- Individuals with autosomal dominant hereditary hearing loss have a 50% chance of transmitting the mutant allele to each child.

Depending upon the specific syndrome, clinical severity and disease phenotype may differ between individuals with the same mutation; thus, age of onset and/or disease progression may not be predictable.

### 2. Risk to Family Members - Autosomal **Recessive** Hereditary Hearing Loss

#### Parents of a Proband

- The parents are obligate heterozygotes and, therefore, carry a single copy of a disease-causing mutation.
- Heterozygotes are asymptomatic.

#### Sibs of a Proband

- At conception, the sibs have a 25% chance of being affected, a 50% chance of being unaffected and carriers, and a 25% chance of being unaffected and not carriers. Once an at-risk sib is known to be unaffected, the risk of his/her being a carrier is 2/3. Heterozygotes are asymptomatic.

#### Offspring of a Proband

- All of the offspring are obligate carriers.

Depending upon the specific syndrome, clinical severity and disease phenotype may differ between individuals with the same mutations; thus, age of onset and/or disease progression may not be predictable. For probands with GJB2-related deafness and severe-to-profound deafness, siblings with the identical GJB2 genotype have a 91% chance of having severe-to-profound deafness and a 9% chance of having mild-to-moderate deafness. For probands with GJB2-related deafness and mild-to-moderate deafness, siblings with the identical GJB2 genotype have a 66% chance of having mild-to-moderate deafness and a 34% chance of having severe-to-profound deafness.

#### Other Family Members of a Proband

- The sibs of obligate heterozygotes have a 50% chance of being heterozygotes.

### 3. Risk to Family Members - **X-Linked Recessive** Hereditary Hearing Loss

#### Parents of a Proband

Women who have an affected son and another affected male relative are obligate heterozygotes. If pedigree analysis reveals that an affected male is the only affected individual in the family, several possibilities regarding his mother's carrier status need to be considered:

- He has a de novo disease-causing mutation and his mother is not a carrier;
- His mother has a de novo disease-causing mutation, as either: a “germline mutation” (i.e., at the time of her conception and thus present in every cell of her body); or “germline mosaicism” (i.e., in her germ cells only);
- His maternal grandmother has a de novo disease-causing mutation.
- No data are available, however, on the frequency of de novo gene mutations nor on the possibility or frequency of germline mosaicism in the mother.

#### Sibs of a Proband

- The risk to sibs depends upon the genetic status of the proband's mother. A female who is a carrier has a 50% chance of transmitting the disease-causing mutation with each pregnancy. Sons who inherit the mutation will be affected; daughters who inherit the mutation are carriers and are likely to be unaffected.
- If the mother is not a carrier, the risk to sibs is low but greater than that of the general population because the possibility of germline mosaicism exists. Depending upon the specific syndrome, clinical severity and disease phenotype may differ between individuals with the same mutation; thus, age of onset and/or disease progression may not be predictable.

#### Offspring of a Proband

- Males with X-linked hereditary hearing loss will pass the disease-causing mutation to all of their daughters and none of their sons.

#### Other Family Members of a Proband

- The proband's maternal aunts may be at risk of being carriers and the aunt's offspring, depending upon their gender, may be at risk of being carriers or of being affected.

### 4. Risk to Family Members - **Mitochondrial** Disorders with Hearing Loss as a Possible Feature Parents of a Proband

- The mother of a proband (usually) has the mitochondrial mutation and may or may not have symptoms. The father of a proband is not at risk of having the disease-causing mtDNA mutation. Alternatively, the proband may have a de novo mitochondrial mutation.

#### Sibs of a Proband

- The risk to the sibs depends upon the genetic status of the mother. If the mother has the mitochondrial mutation, all sibs are at risk for inheriting it.

#### Offspring of a Proband

- All offspring of females with an mtDNA mutation are at risk of inheriting the mutation. Offspring of males with an mtDNA mutation are not at risk.

#### Other Family Members of a Proband

- The risk to other family members depends upon the genetic status of the proband's mother. If she has a mitochondrial mutation, her siblings and mother are also at risk.

#### 5. Risk to Family Members - **Empiric Risks**

If a specific diagnosis cannot be established (and/or the mode of inheritance cannot be established), the following empiric figures can be used:

- The subsequent offspring of a hearing couple with one deaf child and an otherwise negative family history of deafness have an 18% empiric probability of deafness in future children. If the deaf child does not have DFNB1 based on molecular genetic testing of GJB2 (which codes for the protein connexin 26), the recurrence risk is 14% for deafness unrelated to connexin 26. If the hearing couple is consanguineous, the subsequent offspring have close to a 25% probability of deafness due to the high likelihood of an autosomal recessive disorder.
- The offspring of a deaf person and a hearing person have a 10% empiric risk of deafness. Most of the risk is attributed to autosomal dominant syndromic deafness. If both syndromic deafness and a family history of autosomal recessive inheritance can be excluded, the risk of deafness is chiefly related to pseudo-dominant occurrence of recessive deafness. GJB2 (which codes for the protein connexin 26) testing can identify much of this risk.
- The child of a non-consanguineous deaf couple in whom autosomal dominant deafness has been excluded has an approximately 15% empiric risk for deafness. However, if both parents have connexin 26-related deafness, the risk to their offspring is 100%. Conversely, if the couple has autosomal recessive deafness known to be caused by mutations at two different loci, the chance of deafness in their offspring is below that of the general population.
- The child of a hearing sib of a deaf proband (presumed to have autosomal recessive nonsyndromic deafness) and a deaf person has a 1/200 (0.5%) empiric risk for deafness, or five times the general population risk. GJB2 and GJB6 molecular genetic testing can clarify if the risks are higher. If the hearing sib is a carrier of a GJB2 mutation or a GJB6 mutation and marries a person with DFNB1 deafness, the chance of having a deaf child is 50%.

#### **Related Genetic Counseling Issues:**

- Communication with individuals who are deaf requires the services of a skilled interpreter.
- Deaf persons may view deafness as a distinguishing characteristic and not as a handicap, impairment, or medical condition requiring a "treatment" or "cure," or to be "prevented."
- Many deaf people are interested in obtaining information about the cause of their own deafness, including information on medical, educational, and social services rather than information about prevention, reproduction, or family planning. As in all genetic counseling, it is important for the counselor to identify, acknowledge, and respect the individual's/family's questions, concerns, and fears [Middleton et al 1998, Arnos 2003].
- The use of certain terms is preferred: probability or chance versus risk; deaf and hard of hearing versus hearing impaired. Terms such as "affected," "abnormal," and "disease-causing" should be avoided.

**DNA Banking:**

- DNA banking is the storage of DNA (typically extracted from white blood cells) for possible future use. Because it is likely that testing methodology and our understanding of genes, mutations, and diseases will improve in the future, consideration should be given to banking DNA of affected individuals. DNA banking is particularly relevant in situations in which molecular genetic testing is available on a research basis only.

**References**

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- Morton CC, Nance WE. Newborn hearing screening – a silent revolution. *N Engl J Med.* 2006; 354: 2151–64.
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# Appendix 5



Tennessee Department of Health  
**Newborn Hearing Screening Program**  
 Women's Health and Genetics, Newborn Hearing Screening  
 Laboratory Services, 630 Hart Lane, Nashville, Tennessee 37243  
 615-262-6160 Fax 615-262-6159

## Report of Infant Hearing Re-Screen or Diagnostic Evaluation

Child's Last Name \_\_\_\_\_ First Name \_\_\_\_\_ Middle Name \_\_\_\_\_ Sex \_\_\_\_\_ Birth Date \_\_\_\_\_

Mother's Last Name \_\_\_\_\_ First Name \_\_\_\_\_ Mother's Maiden Name \_\_\_\_\_ State Lab TDH# (if available) \_\_\_\_\_

Address \_\_\_\_\_ City \_\_\_\_\_ State/Zip \_\_\_\_\_ Phone \_\_\_\_\_

Referred by:     Hospital Screening     pass     refer    Name of Hospital \_\_\_\_\_  
                    Other Specify \_\_\_\_\_  
                    Out of State \_\_\_\_\_

**Date of Evaluation:** \_\_\_\_\_     Initial Screen     Re-screen     Diagnostic     3 mo. F/U     6 mo. F/U

**Risk Indicators for Hearing Loss:** \_\_\_\_\_

**Type(s) of Evaluation:**     ABR     OAE     TEOAE     DPOAE     ASSR     Tympanometry     Behavioral Testing

<b>Degree of Hearing Loss:</b>	<b>Ear</b>
Hearing Within Normal Limits	<input type="checkbox"/> R <input type="checkbox"/> L
Mild (21-40 dB HL)	<input type="checkbox"/> R <input type="checkbox"/> L
Moderate (41-70 dB HL)	<input type="checkbox"/> R <input type="checkbox"/> L
Severe (71-90 dB HL)	<input type="checkbox"/> R <input type="checkbox"/> L
Profound (>90 dB HL)	<input type="checkbox"/> R <input type="checkbox"/> L
Sloping Hearing Loss	<input type="checkbox"/> R <input type="checkbox"/> L
Unspecified Hearing Loss	<input type="checkbox"/> R <input type="checkbox"/> L
Inconclusive due to: _____	
<b>Type of Hearing Loss:</b>	<b>Ear</b>
Hearing Within Normal Limits	<input type="checkbox"/> R <input type="checkbox"/> L
Fluctuating Conductive HL	<input type="checkbox"/> R <input type="checkbox"/> L
Permanent Conductive HL	<input type="checkbox"/> R <input type="checkbox"/> L
Sensorineural Hearing Loss	<input type="checkbox"/> R <input type="checkbox"/> L
Auditory Neuropathy/Dyssynch	<input type="checkbox"/> R <input type="checkbox"/> L
Mixed Hearing Loss	<input type="checkbox"/> R <input type="checkbox"/> L
Unspecified Hearing Loss	<input type="checkbox"/> R <input type="checkbox"/> L
Inconclusive due to: _____	

<b>Referrals:</b>	<b>Date</b>
<input type="checkbox"/> No Referral	_____
<input type="checkbox"/> Repeat Hearing Testing	_____
<input type="checkbox"/> Primary Care Provider (PCP)	_____
<input type="checkbox"/> Medical Specialist (ENT/OTO)	_____
<input type="checkbox"/> Early Intervention Program	_____
<input type="checkbox"/> TEIS <input type="checkbox"/> Other _____	_____
<input type="checkbox"/> Children's Special Services (CSS)	_____
<input type="checkbox"/> Speech/Language Services	_____
<input type="checkbox"/> Hearing Aid Fitting	_____
<input type="checkbox"/> Genetic Referral	_____
<input type="checkbox"/> Family Support/Family Voices	_____
<input type="checkbox"/> Vision Referral	_____
<input type="checkbox"/> Other _____	_____
Type and Location	

**Follow-up date:** \_\_\_\_\_

**Comments:** \_\_\_\_\_  
 \_\_\_\_\_

**Provider:** \_\_\_\_\_  
 Audiologist, Medical Provider, Hospital, Early Intervention Provider, Other

**Address:** \_\_\_\_\_

**City:** \_\_\_\_\_ **State/Zip:** \_\_\_\_\_

Mail to above address or **Fax to 615-262-6159**    Attn: Newborn Hearing Coordinator  
 PH-3684

