Tennessee Department of Health

Newborn Hearing Program

Pediatric Audiology Assessment & Amplification Guidelines

February 2005

Table of Contents

Introduction 1

Pediatric Audiology Assessment Guidelines 2
- Primary Purpose Statement
- Professional Competency
- Equipment/Facilities
- American National Standards Institute
- Joint Commission on Accreditation of Healthcare Organizations
- Universal Precautions
- Moderate Sedation

Audiologic Assessment Procedures 5
- Case History
- Otoscopy
- Behaviorial Assessment
- Physiological Assessment
- Acoustic Immittance

Recommended Pediatric Audiologic Assessment Guidelines 11
- Infants 0-4 Months Developmental Age 12
- Infants 5-24 Months Developmental Age 15
- Toddlers and Preschoolers 25-60 Months Developmental Age 18

Audiologic Follow-Up Guidelines for the Pediatric Population 22

Pediatric Amplification Guidelines 27

Cochlear Implants in Children 45

Appendix 1. Tennessee Audiology Guideline Committee 49

Appendix 2. Joint Committee on Infant Hearing 2000 Position Statement 50
Risk Indicators for Progressive and
Delayed Onset or Acquired Hearing Loss

Appendix 3. National and Tennessee Hearing Resources and 51
Order Form for Tennessee Hearing Materials

Appendix 4. Tennessee Genetic Resources and Recommendations 54

Appendix 5. Audiology Reporting Form 60

Appendix 6. AAP UNHS Guidelines for Pediatric Medical Home Providers 61

Appendix 7. Audiology Acronyms 63

INTRODUCTION

The goal of the Tennessee Department of Health Universal Newborn Hearing Screening Program (NHS) 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.
- Assure all infants identified with a hearing loss receive appropriate and necessary intervention prior to 6 months of age.

The program is committed to assuring families have access to audiology providers that demonstrate the knowledge and skill necessary to provide current pediatric hearing assessment methods.

The following recommended guidelines were developed by the Tennessee Pediatric Audiology Guideline Committee convened by the Tennessee Newborn Hearing Screening Task Force. Members of the working group responsible for the development of the guidelines included the following pediatric audiologists: Barbara Nicodemus, Carol Runyan, Karen Clinton Brown, Helen Hallenback, Whitney Mauldin, Patricia Chase, Linda Gemayel, Laura Gifford, Cindy Brown Gore, Susan Lytle, Erin Plyler, Susie Robertson, Carol Thiele, Anne Marie Tharpe, and Kelly Yeager. Members representing other fields included: Jacque Cundall, Tennessee Department of Health Newborn Hearing, Teresa Blake, Genetic Counselor, U.T. Knoxville Developmental and Genetics Center, and Jamie Castle, Tennessee Department of Education, Early Intervention System. (Appendix 1-Tennessee Audiology Guideline Committee).

These guidelines were developed for the purpose of advancing an effective statewide system for assessing the hearing of infants and young children, birth to five years of age. In addition, these guidelines are meant to facilitate the diagnosis of hearing loss, obtain medical clearance for amplification, and implement amplification, prior to 3 months of age for infants, and for young children who are deaf and hard of hearing. 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 create an approach to care that is unique to the need of each individual patient.

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 were based largely upon 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. Any modifications to 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 failure of newborn hearing screening (NHS), risk indicators for hearing loss (per the Joint Committee on Infant Hearing 2000; JCIH), parental, caregiver or family concern, or from 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 2000 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 young children birth 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 these patients.

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 consortial 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).

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 over-sight 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. When air conduction thresholds obtained by behavioral methods are found to be abnormal, estimates of bone conduction sensitivity should be completed. Effective masking of the non-test ear should be utilized as necessary. Insert phones are recommended unless contra-indicated when testing infants and young children.

Acoustic stimuli used for behavioral assessment should provide frequency-specific information regarding auditory sensitivity. Therefore, responses to pure tones, FM tones, or narrow bands of noise should be obtained in behavioral testing of children regardless of the response levels obtained to broadband signals (e.g., speech). When using narrowband noise, the bandwidth must be sufficiently narrow to ensure accurate determination of frequency-specific thresholds. Because high-frequency spectral energy above 1000 Hz is critical to speech perception, audiologic assessment of children should always include test stimuli that allow the clinician to evaluate hearing sensitivity within the high-frequency range. At a minimum, thresholds should be obtained at 500 Hz and 2000 Hz for each ear to allow for the selection of appropriate amplification (The Pediatric Working Group, 1996).

It also 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 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).

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 dB 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 (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. Alternative electrode placements such as the earlobe or tragus or the use of tiptrodes should be considered.

Case History:
The case history is particularly important because it will often guide the selection of a strategy for the audiological 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).

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). 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 of age, 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). The basic paradigm used in the tangible reinforcement operant conditioning audiometry (TROCA) or visually reinforced operant conditioning audiometry (VROCA) procedure involves a bar press response coupled with either tangible or visual reinforcement. TROCA or VROCA has been shown to be most effective with children between 2 and 4 years of age developmentally, and also is effective with children with mental challenges (Wilson & Thompson, 1984; Diefendorf, 1988). In conditioned play audiometry (CPA), children learn to engage in an activity each time they hear the test signal. 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 techniques with infants and young children, the successful evaluation 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).

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).

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.

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).

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.

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.

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).

RECOMMENDED PEDIATRIC AUDIOLOGIC ASSESSMENT GUIDELINES FOR INFANTS, TODDLERS, PRESCHOOLERS BIRTH TO 60 MONTHS DEVELOPMENTAL AGE

Purpose Statement:

It is recommended that all infants who do not pass the newborn screen and any subsequent rescreening begin medical evaluation and receive a comprehensive pediatric audiologic assessment to confirm the presence of hearing loss and obtain medical clearance for amplification prior to 3 months of age. Comprehensive assessment should be completed on all young children referred for further hearing evaluation from other audiologic screening. Due to the complexity of the auditory mechanism and the fact that auditory dysfunction may result from pathology at one or more levels, a test battery approach is highly indicated. A test battery that includes physiologic, behavioral and developmental measures is recommended. 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 services prior to 6 months of age in interdisciplinary early intervention programs.

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, experienced pediatric audiologists. The cross check principle is essential to confirm behavioral assessment findings and evaluate individual ear function, particularly when behavioral assessment is limited to sound field measures.

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:

1. **Guidelines for Infants 0-4 Months Developmenal Age**
2. **Guidelines for Infants 5-24 Months Developmental Age**
3. **Guidelines for Toddlers and Preschoolers 25-60 Months Developmental Age**

Guidelines for Infants 0-4 Months Developmental Age

Tennessee Recommended Pediatric Audiologic Assessment

I. Case History

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

II. Otoscopy

- The purpose of otoscopy in this population 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.

III. 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]).

IV. 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]).

V. 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 nHL, bone conduction testing should be completed to assess the type of hearing loss).
- Protocol: Responses should be attempted down to 20 dB nHL. 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).

VI. 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

VII. 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.

VIII. Behavioral Audiologic Assessment

- **0-4 months**: Behavioral observation without reinforcement may be used to corroborate with parent/caregiver observation of child's auditory behavior, but is not recommended for threshold estimation.

IX. 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 (ELM; Coplan & Gleason, 1993)

X. 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 a sensorineural hearing loss should be referred immediately for consideration of 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.
- See Follow-Up section of the Pediatric Audiology Guidelines

XI. Parent Counseling and Resources

- The diagnosing audiologist needs to recognize the emotional impact the diagnosis of hearing loss can have on a family.
- The audiologist should provide the family with information about the type and degree of the hearing impairment, its potential impact on speech/language and cognitive development, the treatment and communication options available, and the positive impact of early intervention.
- Parent resources are available through the Tennessee Newborn Hearing Program. The "Information Packet for My Parents" was developed for families of children identified with hearing loss and may be obtained by calling 615-741-8530. The packet contains brochures and information regarding parent support, communication methods, hearing team members, helpful hints, web sites, and other local, state and national resources. (Appendix 3-Tennessee and National Hearing Resources).

Appendix 2 JCIH, (2000)-Risk Indicators for Progressive and Delayed Onset or Acquired Hearing Loss

Guidelines for Infants 5-24 Months Developmental Age

Tennessee Recommended Pediatric Audiologic Assessment

I. Case History

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

II. Otoscopy

- The purpose of otoscopy in this population 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.

III. 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 (Paridise, 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]).

IV. 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]).

V. 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.

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 nHL, bone conduction testing should be completed to assess the type of hearing loss).
- Protocol: Responses should be attempted down to 20 dB nHL. 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).

VI. 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.

VII. 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.

VIII. Behavioral Audiologic Assessment

- Visual Reinforcement Audiometry (VRA) preferred
- Ear specific testing. Alternate testing between ears to obtain some 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)
  - 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.

IX. 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).
  - Receptive-Expressive Emergent Language Test-3rd Edition (REEL-3; Broch, League, & Brown, 2003).

X. 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 a sensorineural hearing loss should be referred immediately for amplification.
- ***Release of information forms*** should be signed by the parent/guardian to allow those evaluating the child to share information with the school and other service providers.
- See Follow-Up section of the Pediatric Audiology Guidelines

XI. Parent Counseling and Resources

- The diagnosing audiologist needs to recognize the emotional impact the diagnosis of hearing loss can have on a family.
- The audiologist should provide the family with information about the type and degree of the hearing impairment, its potential impact on speech/language and cognitive development, the treatment and communication options available, and the positive impact of early intervention.
- Parent resources are available through the Tennessee Newborn Hearing Program. The "Information Packet for My Parents" was developed for families of children identified with hearing loss and may be obtained by calling 615-741-8530. The packet contains brochures and information regarding parent support, communication methods, hearing team members, helpful hints, web sites, and other local, state and national resources. (Appendix 3-Tennessee and National Hearing Resources).

Appendix 2 JCIH, (2000)-Risk Indicators for Progressive and Delayed Onset or Acquired Hearing Loss

Guidelines for Toddlers and Preschoolers 25-60 Months Developmental Age

Tennessee Recommended Pediatric Audiologic Assessment

I. Case History

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

II. 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.

III. 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]).

IV. 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).

V. 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 nHL).
- Responses should be attempted down to 20 dB nHL. 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.

VI. 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.

VII. 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.

VIII. Behavioral Audiologic Assessment

- Assessments used should be determined based on the child's chronological age with respect to his or her developmental age, and adjusted for prematurity.
- Alternate between ears in order to obtain some ear specific information from each ear prior to the child's fatigue
- Utilize the following, as appropriate:
  - Air and bone conduction testing via insert phones
    - Fill in octave frequencies for 250-8000 Hz
    - Prioritize order of testing to obtain responses for low frequency and high frequency stimuli
- Conduct Conditioned Play Audiometry (CPA) or Visual Reinforcement Audiometry (VRA)
- Tangible (TROCA) or Visual Reinforcement Operant-Conditioning Audiometry (VROCA)
- 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, 1986])
- 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]).

IX. 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 Fluharty. (Fluarty, N.B. 1978)
- Referral for comprehensive speech/language evaluation may be necessary

X. Follow-up Schedule and Referral for Further Evaluation

- Children diagnosed with hearing loss should be monitored and referred for further evaluation and appropriate intervention services as deemed necessary by the intervention team and per TN Department of Health Guidelines.
- Children diagnosed with a sensorineural hearing loss should be referred immediately for amplification.
- ***Release of information forms*** should be signed by the parent/guardian to allow those evaluating the child to share information with the school and other service providers.
- See Follow-Up section of the Pediatric Audiology Guidelines

XI. Parent Counseling and Resources

- The diagnosing audiologist needs to recognize the emotional impact the diagnosis of hearing loss can have on a family.
- The audiologist should provide the family with information about the type and degree of the hearing impairment, its potential impact on speech/language and cognitive development, the treatment and communication options available, and the positive impact of early intervention.
- Parent resources are available through the Tennessee Newborn Hearing Program. The “Information Packet for My Parents” was developed for families of children identified with hearing loss and may be obtained by calling 615-741-8530. The packet contains brochures and information regarding parent support, communication methods, hearing team members, helpful hints, web sites, and other local, state and national resources. (Appendix 3-Tennessee and National Hearing Resources).

Appendix 2 JCIH, (2000)-Risk Indicators for Progressive and Delayed Onset or Acquired Hearing Loss

References

American Academy of Pediatrics, February 1999 Policy Statement, Newborn and Infant Hearing Loss: Detection and Intervention (RE9846). *Pediatrics*, 1999:527-530

Broch, League, Brown, (2003). Receptive-Expressive Emergent Language Test, 3rd Edition, Pro-Ed.

Coplan, J. & Gleason, J. R. (1993). Test-retest and inter-observer reliability of the Early Language Milestone Scale, Second Edition. *Journal of Pediatric Health Care*, 7 (5), 212-219

Fluharty, N.B., (1978). Fluharty Preschool Speech and Language Screening Test. DLM Teaching Sources, One DLM Park. Allen, TX.

Gorga, M.P., Worthington, D.W., Beauchaine, K.A, Goldgar, D.E. (1985). Some Comparisions Between Auditory Brainstem Response Thresholds, Latencies and Pure-Tone Audiogram. Ear and Hearing, 6 (2), 105-112.

Gorga M.P., Neely, S.T. (2001). Some Factors That May Influence the Accuracy of Auditory Brainstem Response Estimates of Hearing Loss. In R.C. Seewald, J.S Gravel (Eds.). A Sound Foundations Through Early Amplification. Proceeding of the Second International Conference (pp.149-161). Stafa, Switzerland: Phonak, A.G.

Gorga, M.P., Neely, S.T, Bergman, B., Beauchaine, K., Kaminski, J., Peters, J., Jesteadt. W. (1993). Otoacoustic emissions from normal-hearing and hearing-impaired subjects: Distortion product responses. *Journal of the Acoustical Society of America*, 93, 2050-2060.

Gorga, M. P., Neely, S. T., Olrich, B., Hoover, B., Redner, J., & Peters, J. (1997). From laboratory to clinic: A large scale study of distortion product otoacoustic emissions in ears with normal hearing and ears with hearing loss. *Ear and Hearing*, 18, 440-455.

Gorga, M .P., Kaminski, J. R., Beauchaine, K. L., Jesteadt, W., & Neely, S. T. (1989). Auditory brain stem responses from children three months to three years of age: II. Normal patterns of response. *Journal of Speech and Hearing Research*, 32, 281-288.

Hall, James W., III (1992). Handbook of Auditory Evoked Responses.

Joint Commission on Infant Hearing, 2000 Position Statement, Principles and Guidelines for Early Hearing Detection and Intervention.

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 identified with a risk indicator for hearing loss, the audiologist or other health care provider should provide the family 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), Tennessee Infant Parent Services (TIPS), Children's Special Services (CSS), Tennessee Genetics System and other medical specialists as outlined below:

Informed Consent for Referral:
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.

Medical/Genetic Evaluation:
Each child identified with hearing loss should be 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) 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 medical home or primary care provider in any decision-making processes that involve further referrals to otolaryngologists, geneticists, ophthalmologists or others. According to the ***ASHA Guidelines for Follow-Up Recommendations (2004).*** "In consultation with the infant's primary care provider, refer the infant/family to an otolaryngologist for medical assessment. As appropriate, discuss additional specialty evaluations, such as genetics, ophthalmology, child development with parents/caregivers and the infant's primary care provider."
- Genetic consultation is important in determining genetic hearing loss from non-genetic hearing loss. Hearing loss may be only one of a number of conditions associated with a genetic syndrome. Therefore, genetic evaluation may be significant in the identification of other medical and developmental diagnoses or conditions to be considered in the infant or child's plan of care. Genetic hearing loss is diagnosed by otologic, audiologic and physical examination, family history, ancillary testing (e.g.,

CT scan of temporal bone) and DNA-based testing. The genetic consultation provides the individual and family with information on the nature, inheritance and implications of a genetic condition and a review of available options to help families make informed decisions. Genetic counseling provides information in a culturally-sensitive manner. (Appendix 4-Tennessee Genetic Resources and Recommendations).

Vision Screening/Diagnostic:
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 Infant Parent's Services (TIPS; Referral to be made through TEIS)
- The Tennessee Technical Assistance and Resources for Enhancing Deaf/Blind Supports (TREDS) program for parent, provider and teacher education and support services. (Appendix 3-Tennessee and National Hearing Resources)

Early Intervention Services:
Infants and children age birth to three 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.

Tennessee Early Intervention System (TEIS):
The Tennessee Department of Education, Tennessee Early Intervention System (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 to age three) with developmental delays. TEIS provides service coordination to families of children with hearing loss age's birth to three years. There are no financial guidelines for eligibility. Families and providers can contact 1-800-852-7157.

Tennessee Infant Parent Services (TIPS):
TIPS is a statewide early intervention home visiting program for families of infants and toddlers with hearing loss, vision loss, hearing/vision loss, and/or developmental delays. TIPS provides parent/caregiver education and support using the SKI-HI Institute Model and Curricula. TIPS has a loaner hearing aid bank for audiologists to utilize until the child can be fitted with his/her own hearing aids.

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.

School System:
Children identified with hearing loss who 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.

Aural/Audiologic Rehabilitation Programs:
Auditory/Oral, Total Communication, sign languages, and Cued Speech are some of the communication approaches available to children and families. Parents should be counseled 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.

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.

Risk Indicators for Progressive and Delayed Onset or Acquired Hearing Loss and the Need for Re-screening and Re-evaluation:
The Joint Committee on Infant Hearing 2000 Position Statement (JCIH, 2000) outlines risk indicators for **birth through age 28 days** where universal hearing screening is not yet available and risk indicators for use with **neonates or infants (29 days through 2 years)**. The indicators that place an infant at risk for progressive or delayed-onset sensorineural hearing loss and/or conductive hearing loss can be found in Appendix 2. Any infant with these risk indicators for progressive or delayed-onset hearing loss who has passed the birth screen should, nonetheless, receive audiologic monitoring every 6 months until age 3 years. (Appendix 2-JCIH Risk Indicators for Progressive and Delayed Onset or Aquired hearing Loss)

Therefore, it is recommended that the audiologist, as well other hearing providers, report infants with a risk indicator to the State Newborn Hearing Screening (NHS) program on the appropriate form to enable NHS to assist the provider to monitor and track children to ensure receipt of timely and appropriate services.

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.

Audiological Monitoring:
All children with identified hearing loss (i.e., hearing loss less than or equal to 25dB HL, unilateral or bilateral, permanent or fluctuating) should receive periodic audiological 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:

    - Should be monitored after medical treatment (completion of antibiotic treatment, PE tubes, etc.), and/or at least every 3-4 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.

Amplification:
Refer to the amplification section of these guidelines.

Counseling:
Parents/primary caregivers, including grandparents and immediate family members, should be counseled regarding the child's diagnosed hearing loss (type, degree/severity, and developmental or educational impact). Recommendations should include communication methods, amplification, referrals to early intervention services or to the local educational agency, and the discussion regarding future services for the child. A family centered and culturally-sensitive approach needs to be maintained during all aspects of counseling.

Documentation/Reporting:
- Documentation must be contemporaneous with each visit or interaction to provide a complete and cogent 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 associated conditions (e.g., medical diagnosis, disability, home program). In addition, documentation must contain pertinent background information, assessment procedures employed, assessment results, interpretation, and specific recommendations. Recommendations may address the need of further assessment, follow-up 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 a documentation standard of HIPPA. Requests for a child's records must respect a parent's 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 are encouraged to report follow-up and confirmatory hearing testing to the Tennessee Department of Health (TDH) Newborn Hearing Program on infants, toddlers, and children birth to five years old.
- The Confirmation of Evaluation for Hearing Loss form can be acquired by contacting the Tennessee Department of Health Newborn Hearing Program by phone, 615-741-0310 or 615-262-6160. The form is available in PDF format on the Newborn Screening web site. (www2.state.tn.us/health/MCH/NBS; Appendix 5-Audiology Reporting Form).

Hearing Loss Support Programs/Organizations/Resources:
For State and National Resources (Appendix 3-Tennessee and National Hearing Resources)

Tennessee Newborn Hearing Program's "Informational Packet for My Parents": Parent resources are available through the Tennessee Newborn Hearing Program. An excellent resource for parents and professionals is the "Information Packet for My Parents". The packet was developed for families of children identified with hearing loss. The packet contains brochures and information regarding parent support, communication methods, hearing team members, helpful hints, web sites, and other local, state and national resources. Tennessee hearing brochures and posters are available on the state web site. Documents can be obtained at no cost by contacting, the TDH Newborn Hearing Program, Cordell Hull Building, 5th Floor, 425 Fifth Avenue North, Nashville, TN 37247-4750. Phone 615-741-8530 or 615-262-6160. (Appendix 3-Tennessee and National Resources).

PEDIATRIC AMPLIFICATION GUIDELINES

The following pediatric amplification guidelines were based upon those developed by the American Academy of Audiology (AAA, 2003). The AAA Pediatric Amplification Protocol and the Exposition on Cochlear Implants in Children were 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, cochlear implants, other assistive technology or other methods of communication.

Purpose

The purpose of this document 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.

Outline:

1. Personnel Qualifications
2. Candidacy
3. Pre-selection issues and procedures
4. Circuitry—Signal Processing
5. Hearing Instrument Selection/Fitting Considerations
6. Verification
7. Hearing Instrument Orientation and Training
8. Validation
9. Follow-up and Referral

1. Personnel Qualifications

A. Audiologists are the professionals singularly qualified to select and fit all forms of amplification for children, including personal hearing aids, frequency-modulated (FM) systems, cochlear implants and other assistive listening devices (The Pediatric Working Group, 1996). Audiologists have a master's and/or doctoral degree in audiology from a regionally-accredited university.

B. Audiologists must meet all state licensure and/or regulatory requirements.

C. Pediatric audiologists are qualified by unique experience or formal training to fit hearing aids on infants and young children and should have the expertise and the test equipment necessary to complete all tests for hearing aid selection, evaluation, and verification procedures described herein.

D. Audiologists should adhere to procedures consistent with current standards of practice to assess auditory function in infants and children (ASHA, 2004).

E. 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.

2. 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.

A. Methods for the Assessment of Hearing

For newborns and infants under the developmental age of 6 months, estimates of hearing sensitivity must be supported by electrophysiological measures including auditory brainstem response (ABR) threshold assessment. Frequency-specific air-conduction and bone-conduction ABR thresholds should be obtained. Frequency-specific ABR is necessary for accurate estimation of the degree and configuration of hearing loss. A click-ABR threshold alone is not sufficient for accurate hearing aid fitting. Acoustic immittance measures, including tympanometry and middle ear muscle reflexes, and otoacoustic emissions (OAE) are necessary to determine the type of hearing loss present.

Differential diagnosis continues to be refined and these measures should be applied to the assessment of hearing in children as they become available and interpretable. Currently researchers are suggesting that the summating potential may have value in diagnosis and that a lack of response in this measure may relate to inner hair cell function. These and other electrophysiologic measures may become a valued part of the assessment of hearing in the pediatric population. At a minimum, low and high frequency, ear specific information should be obtained in order to prescribe appropriate amplification. These data are developed over the course of evaluating the infant or child and the hearing aid fitting may begin before all data are obtained.

For older infants and young children, behavioral thresholds should be obtained using visual reinforcement audiometry (VRA), or conditioned play audiometry (CPA) test techniques appropriate for the child's developmental level. Ear-specific and frequency-specific air and bone conduction thresholds are essential for providing information needed for accurate hearing aid fitting (The Pediatric Working Group, 1996).

Additional Factors

1) Middle Ear Conditions

The presence of chronic or recurrent middle ear conditions that can affect hearing threshold results or the ability to wear an occluding earmold 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.

2) Other Health Concerns

Other health concerns or conditions that may affect the ability to obtain reliable threshold information must be considered. The use of physiologic test methods (ABR, OAE) may be necessary even with older children who have additional disabilities.

B. 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.

1) 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.

2) 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.

3) 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.

4) 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 children with unilateral hearing impairment when an FM system is coupled to the normal hearing ear. In such cases, close audiologic monitoring of hearing sensitivity, and careful control of the output of the amplification is required.

3. Pre-Selection Issues and Procedures

A. Introduction

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.

B. 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.

C. Style: body aid vs. 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 (e.g., increased cost, lack of DAI coupling to assistive technology, susceptibility to damage, etc.).

D. 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.

E. 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.

F. 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.

G. Earmold

The audiologist should consider the style, material, color, length, and frequency of remakes for the earmold. The need for well-fitted earmolds 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 earmold fit (after growth of the outer ear). The use of automatic technology forces the audiologist to be more proactive about regular earmold changes. The recent advent of automatic feedback control through various digital signal processing techniques may alleviate this problem temporarily while the new earmold is ordered. For infants, earmold replacement may be as frequent as monthly.

Venting in the earmold 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 earmolds 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.

H. Sound Channel

The sound channel consists of the earhook and tube that leads through the earmold 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 earmold 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 earmold will reveal any roll off in high frequency response as will probe microphone measurements that include the individual's earmold 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).

I. 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 & Yaccullo, 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. Benefits and limitations of directional microphone technology with children are currently unknown. Through the selection and deselection 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 older children, the audiologist should ensure that the microphone response in the directional setting is equalized to the microphone response in the omnidirectional setting or audibility for low frequency sounds is lost (Ricketts & Henry, 2002).

J. 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.

K. 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.

L. 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).

M. 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.

N. Battery Doors

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

O. 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 earmolds. 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.

4. 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. It is likely that all hearing aids will be digital within the next five years and the analog vs. digital decision will be irrelevant. 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. 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.

5. 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 n 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.

6. Verification

A. The electroacoustic performance of the instrument should be matched to the prescribed 2 cm3 coupler target values for gain and output limiting where the 2 cm3 coupler values have been derived using an individualized real ear to 2 cm3 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).

7. Hearing Instrument Orientation and Training

Orientation and training should include family members, caregivers, and 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

8. Validation

A. 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.

B. 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.

C. 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.

The tools listed above should be helpful in planning for the individual child. The majority of these tools, however, do not have published psychometric data at this time. With these data, it would not be appropriate to use these tools to document significant change in performance.

9. 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.

References

American National Standards Institute. Specification of hearing aid characteristics. ANSI S3.22-1996, New York.

Anderson, K.L. (1989). Screening Instrument for Targeting Educational Risk (SIFTER).Tampa, FL, Educational Audiology Association.

Anderson, K.L., Matkin, N.D. (1996). Screening Instrument for Targeting Educational Risk in Preschool Children (age 3-kindergarten) (Preschool SIFTER). Tampa, FL, Educational Audiology Association.

Anderson, K.L., Smaldino, J. (2000). Children's Home Inventory of Listening Difficulties (CHILD). Educational Audiology Review, 17(3), Suppl.

Anderson, K.L., Smaldino, J. (1996). Listening Inventory for Education (LIFE), an efficacy tool. Educational Audiology Association, [on-line]. Available: www.edaud.org.

Anderson, K.L. (2002). Early Listening Function (ELF). Educational Audiology Association, [on-line]. Available: www.edaud.org.

Bess, F.H., Dodd-Murphy, J., Parker, R.A. (1998). Children with minimal sensorineural hearing loss: Prevalence, educational performance, and functional status. Ear and Hearing, 19(5): 339-354.

Bess, F.H, Tharpe, A.M. (1984). Unilateral hearing impairment in children. Pediatrics, 74: 206-216.

Bess, F., Tharpe, A.M. (1988). Performance and management of children with unilateral sensorineural hearing loss. Barajas, J.J., Borg, E. (Eds), Overview and Evaluation of the Hearing-Impaired Child – Imaging and Audiology. Scandinavian Audiology Supplement, 30, 75-79.

Boothroyd, A. (1993). Recovery of speech perception performance after prolonged auditory deprivation: Case Study. Journal of the American Academy of Audiology, 4, 331-337.

Byrne, D., Dillon, H., Ching, T., Katsch, R., Keidser, G. (2001). NAL-NL1 procedure for fitting nonlinear hearing aids: Characteristics and comparisons with other procedures. J Amer Acad Audiol. 12, 37-51.

Ching, T., Dillon, H., Byrne, D. (1998). Speech recognition of hearing-impaired listeners: Predictions from audibility and the limited role of high-frequency amplification. Journal of the Acoustical Society of America, 103, 1128-1140.

Ching, T., Dillon, H., Katsch, R. (2001). Do children require more high-frequency audibility than adults with similar hearing losses? A sound foundation through early amplification: Proceedings of the Second International Conference. Phonak, Inc.: Chicago, IL. 141-152.

Cornelisse, L., Seewald, R., Jamieson, D. (1995). The input/output formula: A theoretical approach to the fitting of personal amplification devices. Journal of the Acoustical Society of America, 97(3), 1854-1864.

Gagné, J.P., Seewald, R.C., Zelisko, D.L., Hudson, S.P. (1991a). Procedure for defining the auditory area of hearing impaired adolescents with severe/profound hearing loss I: Detection thresholds. Journal of Speech Language Pathology and Audiology, 15(3):13-20.

Gagné, J.P., Seewald, R.C., Zelisko, D.L., Hudson, S.P. (1991b). Procedure for defining the auditory area of hearing impaired adolescents with severe/profound hearing loss II: Loudness discomfort levels. Journal of Speech Language Pathology and Audiology, 15(4): 27-32.

Gravel, J., Fausel, N., Liskow, C., Chobot, J. (1999). Children's speech recognition in noise using omnidirectional and dual-microphone hearing aid technology. Ear and Hearing, 20(1), 1-11.

Haskin, H.A. (1949). A phonetically balanced test of speech discrimination for children. Master's thesis, Northwestern University: Evanston, Illinois.

Hattori, H. (1993). Ear dominance for nonsense-syllable recognition ability in sensorineural hearing-impaired children: Monaural versus Binaural amplification. Journal of the Academy of Audiology, 4, 319-330.

Hawkins, D., Naidoo, S. (1993). Comparison of sound quality with asymmetrical peak clipping and output limiting compression. American Journal of Audiology, 4, 221-228.

Hawkins, D., Yacullo, W. (1984). Signal-to-noise ratio advantage of binaural hearing aids and directional microphones under different levels of reverberation. Journal of Speech and Hearing Disorders, 49(3), 278-286.

Hogan, C., Turner, C. (1998). High frequency audibility: Benefits for hearing-impaired listeners. Journal of the Acoustical Society of America, 104(1), 432-441.

Jenstad, L., Seewald, R., Cornelisse, L., Shantz, J. (1999). Comparison of linear gain and WDRC hearing aid circuits: Aided speech perception measures. Ear and Hearing, 20, 117-126. Jenstad, L., Pumford, J., Seewald, R., Cornelisse, L. (2000). Comparison of linear gain and WDRC hearing aid circuits II: Aided loudness measures. Ear and Hearing, 21, 32-44.

Jerger, S., Lewis, S., Hawkins, J., and Jerger, J. (1980). Pediatric speech intelligibility test: I. Generation of Test materials. International Journal of Pediatric Otorhinolaryngology 2: 217-230.

Johnson, C.D., Von Almen, P. (1997). Functional listening evaluation. In Johnson, C.D., Benson, P.V., Seaton, J.B. Educational audiology handbook. San Diego: Singular Publishing Group, Inc., pp. 336-339.

Johnson, W., Killion, M. (1994). Amplification: Is class D better than class B? American Journal of Audiology, 3, 11-13.

Joint Audiology Committee. (2000). Audiology Clinical Practice Algorithms and Statements, Audiology Today, Special Issue, August 2000.

Katz, D.R., Elliott, L.L. (1978). Development of a new children's speech discrimination test. Paper presented at the convention of the American speech and Hearing Association, November 18-21, Chicago.

Kenworthy, O.T. Klee, T., Tharpe, A.M. (1990). Speech recognition ability of children with unilateral sensorineural hearing loss as a function of amplification, speech stimuli, and listening condition. Ear and Hearing, 11, 4.

Killion, M., Wilber, L., Gudmundsen, G. (1988). Zwislocki was right… a potential solution to the "hollow voice" problem. Hearing Instruments, 39(1), 14-18.

Kortekaas, R., Stelmachowicz, P. (2000). Bandwidth effects on children's perception of the inflectional morpheme (s): Acoustical measurements, auditory detection, and clarity rating. Journal of Speech, Language, and Hearing Research, 42, 645-660.

Kuk, F., Kollofski, C., Brown, S., Melim, A., Rosenthal, A. (1999). Use of a digital hearing aid with directional microphones in school-aged children. Journal of the American Academy of Audiology, 10(10): 535-548.

Matkin, N.D. (1996). The potential benefits of amplification for young children with normal hearing, in: Bess, F.H., Gravel, J.S., Tharpe, A.M. (eds) Amplification for Children with Auditory Deficits. Nashville: Bill Wilkerson Center Press.

Meadow-Orlans, K.P. (1983). Meadow-Kendall Social/Emotional assessment Inventory (SEAI) for Deaf and Hearing-Impaired Students. Gallaudet University.

Moog, J.S., Geers, A.E. (1990). Early speech perception test for profoundly hearing-impaired children. St. Louis: Central Institute for the Deaf.

Moore, B. (2001). Dead regions in the cochlea: implications for the choice of high-frequency amplification. A sound foundation through early amplification: Proceedings of the Second International Conference. Phonak, Inc.: Chicago, IL., 153-166.

Palmer, C. (2001). The impact of hearing loss and hearing aid experience on sound quality judgments. Seminars in Hearing, 22, 125-138.

Palmer, C., Killion, M., Wilber, L., Ballad, W. (1995). Comparison of two hearing aid receiver-amplifier combinations using sound quality judgments. Ear and Hearing, 16(6), 587-598.

Palmer, C., Mormer, E. (1999). Goals and expectations of the hearing aid fitting. Trends in Amplification, 4 (2), 61-71.

Preves, D., Newton, J. (1989). The headroom problem and hearing aid performance. The Hearing Journal, 42, 19-26.

Ricketts, T., Henry, P. (2002). Low-frequency gain compression in directional hearing aids. American Journal of Audiology, 11 (1), 1-13.

Ricketts, T., Mueller, G. (2000). Predicting directional hearing aid benefit for individual listeners. Journal of the American Academy of Audiology, 11(10), 561-569.

Robbins, A.M., Renshaw, J.J., Berry, S.W. (1991) Evaluating meaningful auditory integration in profoundly hearing impaired children. American Journal of Otolarygology 12:144-150.

Robbins, A.M., Svirsky, Osberger, Pisoni (1998). Beyond the audiogram: The role of functional assessments. In: Bess, F.H., ed. Children with Hearing Impairment: Contemporary Trends. Nashville: Bill Wilkerson Center Press, 1998, pp 105-124.

Scollie, S., Seewald, R., Cornelisse, L., Jenstad, L. (1998). Validity and repeatability of level dependent HL to SPL transforms. Ear and Hearing, 19, 407-413.

Scollie, S., Seewald, R. (2001). Electroacoustic verification measures with modern hearing instrument technology. A sound foundation through early amplification: Proceedings of the Second International Conferences. Phonak, Inc.: Chicago, IL. 121-137.

Scollie, S., Seewald, R. (2002). Hearing aid fitting and verification procedures for children. In Katz, J. (5th Edition). Handbook of Clinical Audiology, Lippincott/Williams and Wilkins: New York. 695-696.

Scollie, S., Seewald, R. (2002). Evaluation of electroacoustic test signals. 1 – Comparison with amplified speech. Ear and Hearing, 23(5), 477-487.

Scollie, S., Steinberg, M.J., Seewald, R. (2002).. Evaluation of electroacoustic test signals. 2 – Development and cross validation of correction factors. Ear and Hearing, 23(5), 488-498.

Seewald, R., Scollie, S. (1999). Infants are not average adults: Implications for Audiometric Testing. Hearing Journal, 52(10), 64-72.

Seewald, R.C., Moodie, K.S., Sinclair, S.T., Cornelisse, L.E. (1996). Traditional and theoretical approaches to selecting amplification for infants and children. In Bess, F.H., Gravel, J.A., Tharpe, A.M. (Eds). Amplification for Children with Auditory Deficits. Vanderbilt University Press.

Seewald, R., Moodie, S., Sinclair, R., Scollie, S. (2000). Predictive validity of a procedure for pediatric hearing instrument fitting. American Journal of Audiology, 8(2), 143-152.

Skinner, M., Miller, J. (1983). Amplification bandwidth and intelligibility of speech in quiet and noise for listeners with sensorineural hearing loss. Audiology, 22, 253-279.

Stedler-Brown, A., Johnson, C., DeConde, (2001). Functional Auditory Performance Indicators: an integrated approach to auditory development. [on line] , Colorado Department of Education, Special Education Services Unit. Available: www.cde.state.co.us/cdesped/specificdisability-hearing.htm

Stelmachowicz, P. (2001). The importance of high-frequency amplification for young children. A sound foundation through early amplification: Proceedings of the Second International Conference. Phonak, Inc.: Chicago, IL., 167-175.

Stelmachowicz, P., Lewis, D., Kalberer, A., Creutz, T. (1994). Situational Hearing-Aid Response Profile (SHARP, Version 2.0). User's Manual. Boys Town National Research Hospital, 555 North 30th Street, Omaha, Nebraska 68131.

Stelmachowicz, P., Pittman, A., Hoover, B., Lewis, D. (2001). Effect of stimulus bandwidth on the perception of /s/ in normal – and hearing-impaired children and adults. Journal of the Acoustical Society of America, 110, 2183-2190.

Teder, H. (1993). Compression in the time domain. American Journal of Audiology, 2(2), 41-46.

The Pediatric Working Group. (1996). Amplification for infants and children with hearing loss, Nashville, TN: Bill Wilkerson Press.

Turner, C., Cummings, K. (1999). Speech audibility for listeners with high-frequency hearing loss. American Journal of Audiology, 8, 47-56.

Valente, M. (1982a). Binaural amplification: Part 1. Audiol: J for Continuing Education 7(6).

Valente, M. (1982b). Binaural amplification: Part 2. Audiol: J for Continuing Education 7(6).

Zimmerman, S., Osberger, M.J., Robbins, A.M. (1998). Infant-Toddler: Meaningful Auditory Integration Scale (IT-MAIS). In: W. Estabrooks (ed). Cochlear Implants for Kids. Washington: AG Bell Association for the Deaf.

COCHLEAR IMPLANTS IN CHILDREN

It is well established that profound deafness in childhood affects the development of auditory speech perception, speech production, and spoken language skills. Some children with profound deafness develop viable oral communication skills with conventional hearing aids but most do not. Failure to develop adequate communication skills can have a significant negative effect on educational and employment opportunities for individuals. It is recognized that multichannel cochlear implants are options for children with profound hearing impairments who demonstrate limited or no functional benefit from conventional hearing aid amplification. Multichannel cochlear implants are appropriate for children with prelingual or postlingual deafness. It is further recognized that parents (or legal guardian) have the right to choose a cochlear implant if they decide that it is the most appropriate option for their child.

Background

A cochlear implant is an electronic prosthetic device 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. Cochlear implants are intended to provide prelingually or postlingually deafened children, who obtain limited functional benefit from conventional amplification, improved sound and speech detection and improved auditory perception or speech. Because research in adults and children has shown significantly greater benefit with multichannel than single-channel cochlear implants, only multichannel devices should be used in the pediatric population. Multichannel cochlear implants attempt to mimic the place representation of frequencies along the cochlea by tonotopic arrangement and stimulation of electrodes.

The law requires that 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 (1984) and children (1990). 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 virtually all health insurance companies.

Cochlear Implant Benefits

Studies on the efficacy of multichannel 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 multichannel 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. Studies also have shown improvement in speech production skills and overall speech intelligibility in children with prelingual deafness. Improvements in auditory speech recognition and speech production occur over a long time-course in prelingually deafened children who receive multichannel cochlear implants. There are large individual differences in the benefit that children derive from multichannel cochlear implants due to factors such as age at onset of deafness, age at

implantation, amount of cochlear implant experience, and educational training. However, the reliable predictors of cochlear implant performance have not been identified.

Guidelines for Determining Candidacy for Cochlear Implants

Accurate assessment of hearing impairment by an audiologist is a critical factor in the determination of implant candidacy. The audiologist should use an age-appropriate combination of behavioral and physiological measures to determined hearing status. A pure tone audiogram demonstrating severe-to-profound, bilateral sensorineural hearing loss should be confirmed by acoustic reflex data and, when appropriate, auditory brainstem responses to both clicks and tonal stimuli. Behavioral audiological tests should be repeated following the provision of appropriate electroacoustic amplification and training. A cochlear implant is indicated only after the child has had a sufficient trial with hearing aid amplification.

At the time of this writing, the audiological criteria for implantation are a congenital or acquired profound sensorineural hearing loss and limited or no functional benefit from electroacoustic hearing aid amplification. Generally, a pure tone average (500, 1000, 2000 Hz) of 90dB HL or greater in both ears is indicated. The criteria for limited functional hearing aid benefit continue to evolve and are influenced by the performance results reported for pediatric multichannel cochlear implant users. Hearing aid benefit is examined in terms of: (1) aided thresholds with conventional hearing aids relative to aided results in the high frequencies where important consonant cues occur, and (2) performance on word recognition tasks, administered with auditory cues only in a closed- or open-response set. Transtympanic promontory stimulation immediately prior to surgery may aid in the selection of the ear to be implanted.

Candidates for cochlear implantation require medical evaluation by an otolaryngologist, including history, physical examination and imaging studies of the temporal bone. The patient should be free of active ear disease, have an intact tympanic membrane, and be acceptable candidate for general anesthesia. High resolution computed tomography (CT) scan, magnetic resonance imaging (MRI), or both, are necessary to identify the implantable cochlea and patent internal auditory canal. Electrical promontory stimulation is indicated when auditory nerve integrity is in doubt.

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 age appropriate. Ideally, children should be enrolled in educational programs that support the use of auditory prostheses and the development of auditory and speech skills, regardless of the particular communication method employed. It is further recommended that parents (or guardians), and the child, if age appropriate, be fully informed about alternatives to implantation, horizontal acculturation, and Deaf Culture.

Guidelines for Management of Children with Cochlear Implants

Children who receive cochlear implants require ongoing audiological management and otolaryngological follow-up. Ongoing management by an audiologist includes programming the implant parameters and monitoring device performance from electrical threshold and dynamic range data. Electrically evoked auditory brainstem responses (EABR), middle latency responses (MLR), or acoustic reflexes (EART) may be used intraoperatively with

stimuli delivered to the cochlear implant prior to leaving the operating room or postoperatively on a outpatient basis to facilitate the fitting process. These objective measures can be particularly useful in children who are either difficult to condition or otherwise unable to respond consistently to the electrical stimuli used to program the speech processor. Follow-up audiological evaluations are required to assess improvement in sound and speech detection and auditory reception of speech following implantation. Medical evaluation by an otolaryngologist should be performed as needed to monitor the postoperative course and medical status of the child.

Pediatric cochlear implant users require training to maximize the benefits that they receive from their devices. Rehabilitation should focus on the development of a wide range of listening behaviors within meaningful communicative contexts. Ideally, there should be close interaction between the audiologist at the implant center, the clinician who provides rehabilitative services, and educators working on a day-to-day basis with the child. For a child to realize optimal benefit from a multichannel cochlear implant, educators should have an understanding of device function and maintenance, as well as an appropriate level of expectation regarding the child's progress with the implant.

Future Needs

The field of cochlear implants is still in its infancy. Technological advances will lead to the development of more sophisticated and improved devices. It appears inevitable that as technology for cochlear prostheses advances, candidacy criteria for implantation will continue to expand to include a wider range of the population with severe and profound hearing impairments. Audiological training programs must provide course work and clinical experience with cochlear prostheses. Audiologists with expertise in the diagnosis (including the use of electrophysiological techniques), management, and habilitation of children with hearing impairments are necessary to ensure competent provision of professional services by pediatric cochlear implant programs.

References

Beiter, A.L., Brimacombe, J. (1993): Cochlear implants. In Alpiner, J.G., McCarthy, P.A. (Eds.) Rehabilitative Audiology: Children and Adults. Baltimore, Williams & Wilkins.

Brown, C.J., Abbas, P.J., Fryauf-Bertschy, H., Kelsay, D., Gantz, B.J. (1994). Intraoperative and postopoerative electrically evoked auditory brain stem responses in Nucleus cochlear implant users: Implications for the fitting process. *Ear Hear*, 15:168-176.

Cohen, N.L., Waltzman, S.B., Fisher, S.G. (1993). A prospective, randomized study of cochlear implants. *New Eng J Med*, 328:233-2372.

Gantz, B.J. et al. (1994). Results of multichannel cochlear implants in congenital and acquired prelingual deafness in children: Five year follow up. *Am J Otol*, 15(Suppl 2):1-7

Geers, A.E., Moog, J. (Eds.) (In press) Effectiveness of cochlear implants and tactile aids for deaf children: A report of the CID sensory aids study. *Volta Review*.

Kileny, P.R., Zwolan, T.A., Zimmerman-Phillips, S., Teilan, S.A. (1994). Electrically evoked auditory brain-stem response in pediatric patients with cochlear implants. *Arch Otolaryngol Head Neck Surg*, 120:1083-1090.

Miyamoto, R.T., Osberger, M.J., Robbins, A.M., Myres, W.A., Kessler, K (1993). Prelingually deafened children's performance with the Nucleus multichannel cochlear implant. *Am J Otol*, 14:437-445.

Miyamoto, R.T., Osberger, M.J., Todd, S.L., Robbins, A.M. (1993). Speech perception skills of children with multichannel cochlear implants. *Advances in cochlear implants*. Manz, Vienna: Datenkonvertierung, Reproduktion und Druck.

Miyamoto, R.T., Robbins, A.M., Osberger, M.J., Todd, S.L. (1995). Comparison of tactile aids and cochlear implants in children with profound hearing impairments. *Am J Otol*, 16:8-13.

Osberger, M.J., Robbins, A.M., Miyamoto, R.T., Berry, S.W., Myres, W.A., Kessler, K.S., Pope, M.L. (1991). Speech perception abilities of children with cochlear implants, tactile aids, or hearing aids. *Am J Otol*, 12 (Suppl.): 105-115.

Osberger, M.J., Robbins, A.M., Todd, S.L., Miyamoto, R.T. (1993). Speech production skills of children with multichannel cochlear implants. Advances in cochlear implants (pp. 503-508). Manz, Vienna: Datenkonvertierung, Reproduktion und Druck.

Staller, S.J., Dowell, R.C., Beiter, A.L., Brimacombe, J.A. (1991). Perceptual abilities of children with the Nucleus 22-channel cochlear implant. *Ear Hear*, 12 (Suppl.): 34S-47S.

Tobey, E.A., Angelette, S., Murchinson, C., Nicosia, J., Sprague, S., Staller, S.J., Brimbacombe, J.A., Bieter, A.L. (1991). Speech production performance in children with multichannel cochlear implants. *Am J Otol*, 12 (Suppl.) 165-173.

Tobey, E.A., Hasenstab, M.S.: (1991). Effects of a Nucleus multichannel cochlear implant upon speech production in children. *Ear Hear*, 12 (Suppl.), 48S-54S.

Waltzman, S.B., Cohen, N.L., Gomolin, R.H., Shapiro, W.H., Ozdamar, S., Hoffman, R. (1994). Long-term results of early cochlear implantation in congenitally and prelingually deafened children. *Am J Otol*, 14 (Suppl.):9-13.

Waltzman, S.B., Cohen, N.L., Gomolin, R.H., Ozdamar, S., Shapiro, W., Hoffman, R. (1995). Effects of short-term deafness in young children implanted with the Nucleus cochlear prosthesis. Proceedings of the International Cochlear Implant, Speech and Hearing Symposium.

Appendix 1

Tennessee Pediatric Audiology Guideline Committee

Committee Chair:
Barbara Nicodemus (Knoxville)
UT Consulting Audiologist
Newborn Hearing Program
Tennessee Department of Health
Ph. 865-688-4694
E-mail: bnicodem@utk.edu

Physiologic Sub-Committee Chair:
Carol Runyan (Kingsport)
Dr. Daniel R. Schumaier and Associates
Ph. 423-928-5771
E-mail: LooperC1961@aol.com
crunyon@eartec.com

Behavioral Sub-Committee Chair:
Karen Clinton Brown (Memphis)
University of Memphis
Ph. 901-678-5800
E-mail: kcbrown@memphis.edu

Amplification Sub-Committee Chair:
Helen Hallenbeck (Cookeville)
Cookeville Hearing Center
Ph. 931-526-8863
E-mail: hellenhallenbeck@hotmail.com

Follow-up Sub-Committee Chair:
Whitney Mauldin (Chattanooga)
The Speech and Hearing Center
Ph. 423-622-6900
E-mail: wroop@speechhearing.com

Jacque Cundall (Nashville)
Nursing Consultant/Project Coordinator
Newborn Hearing Program
Tennessee Department of Health
Ph. 615-741-0310 or 262-6160
E-mail: Jacque.Cundall@state.tn.us

Teresa Blake (Knoxville)
UTK Developmental & Genetics Center
Ph. 865-544-9030
E-mail: tblake@mc.utmck.edu

Jamie Castle (Nashville)
Tennessee Department of Education
Early Childhood
Ph. 615-523-2809
E-mail: jamie.castle@state.tn.us

Patricia Chase (Johnson City)
East Tennessee State University
Ph. 423-439-5252
E-mail: chase@mail.etsu.edu

Linda Gemayel (Kingsport)
Wellmont Medical Center
Ph. 423-224-5540
E-mail: Linda_C_Gemayel@wellmont.org

Laura Gifford (Columbia)
Middle TN Audiology
Ph. 931-388-3646
Email: legifford@comcast.net

Cindy Brown Gore (Memphis)
Baptist Memorial Health Care
Ph. 901-226-5682
E-mail: Cynthia.gore@bmhcc.org

Susan Lytle (Knoxville)
UTK Medical Center
Ph. 865-544-9151
E-mail: slytle@mc.utmck.edu

Erin Plyler (Knoxville)
University of Tennessee Audiology Services
Ph. 865-974-5453
E-mail: erinp@utk.edu

Susie Robertson (Maryville)
Blount Hearing and Speech
Ph. 423-982-8557
E-mail: srobertson@bhssinc.com

Carol Thiele (Memphis)
University of Memphis
Ph. 901-678-5800
E-mail: cthiele@memphis.edu

Anne Marie Tharpe (Nashville)
Vanderbilt Bill Wilkerson Center
Vanderbilt University
E-mail: anne.m.tharpe@vanderbilt.edu

Kelly Yeager (Knoxville)
University of Tennessee Audiology Services
Ph. 865-974-5453
E-mail: kyeager@utk.edu

Appendix 2

Joint Committee on Infant Hearing (JCIH) 2000 Position Statement

Risk Indicators for Progressive and Delayed Onset or Acquired Hearing Loss

1. The JCIH risk indicators for **birth through age 28 days** where universal hearing screening is not yet available. These indicators are as follows:
   a. An illness or condition requiring admission of 48 hours or greater to a neonatal intensive care unit.
   b. Stigmata or other findings associated with a syndrome known to include a sensorineural and or conductive hearing loss.
   c. Family history of permanent childhood sensorineural hearing loss.
   d. Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal.
   e. In utero infection such as cytomegalovirus, herpes, toxoplasmosis, or rubella.

2. The JCHI recommends the following indicators for use with **neonates or infants (29 days through 2 years)**. These indicators place an infant at risk for progressive or delayed-onset sensorineural hearing loss and/or conductive hearing loss. Any infant with these risk indicators for progressive or delayed-onset hearing loss who has passed the birth screen should, nonetheless, receive audiologic monitoring every 6 months until age 3 years. These indicators are as follows:
   a. Parental or caregiver concern regarding hearing, speech, language, and/or developmental delay.
   b. Family history of permanent childhood hearing loss.
   c. Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or eustachian tube dysfunction.
   d. Postnatal infections associated with sensorineural hearing loss including bacterial meningitis.
   e. In utero infections such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis.
   f. Neonatal indicators–specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation (ECMO).
   g. Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher's syndrome.
   h. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich's ataxia and Charcot-Marie-Tooth syndrome.
   i. Head trauma.
   j. Recurrent or persistent otitis media with effusion for at least 3 months.

Appendix 3

National and Tennessee Hearing Resources and Order Form for Tennessee Hearing Materials

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**
- American Association for Deaf Children 800-942-ASDC **www.deafchildren.org**
- Alexander Graham Bell Association for the Deaf and Hard of Hearing (AG Bell) 866-337-5220 **www.agbell.org**
- Auditory Verbal International 703-739-1049 **www.auditory-verbal.org**
- BEGINNINGS 800-541-HEAR **www.beginningssvcs.com.**
- Boys Town National Research Hospital **www.babyhearing.org**
- Cochlear Implant Association, Inc. (CIAI) **www.cici.org**
- Families for Hands and Voices **www.handsandvoices.org**
- John Tracy Clinic 800-522-4582 **www.johntracyclinic.org**
- Laurent Clerc National Deaf Education Center **www.clerccenter.gallaudet.edu**
- The Listen-Up! **www.listen-up.org**
- National Association of the Deaf (NAD) 301-587-1788 **www.nad.org**
- National Center for Hearing Assessment and Management (NCHAM**)** **www.infanthearing.org**
- National Cued Speech Association **www.cuedspeech.mt.edu**
- National Institute of Deafness and Other Communication Disorders **www.nidcd.nih.gov**
- Oberkotter Foundation - Oral Deaf Education **www.oraldeafed.org**
- The S.E.E. (Signing Exact English) Center **www.seecenter.org**
- Self Help for Hard of Hearing People (SHHH**) www.shhh.org**

Tennessee Directories

- Tennessee Directory of Services for People who are Deaf and Hard of Hearing 800-342-3262 **www.tndeaflibrary.nashville.gov**
- Tennessee Department of Health Newborn Hearing Program Pediatric Audiologogy and Hearing Providers List 615-741-8530 or 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. **www2.state.tn.us/health/MCH/NBS/index.html**
- Tennessee Early Intervention System (TEIS) 800-852-7157 **www.state.tn.us/education/teishome.htm**
- Tennessee Infant Parent Services (TIPS): Knoxville 865-579-3099; Nashville 615-741-5002; Cookeville 931-372-6247; Chattanooga 423-634-3010; Jackson 731-423-6592; Memphis 901-678-3501; Johnson City 423-926-4388
- 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 **www2.state.tn.us/health/MCH**
- Family Voices: Family support network and advocacy group for all children and youth with special health care needs, familyvoices@tndisabilities 888-643-7811 **www.tndisabilities.org/familyvoices**.
- Library Services for the Deaf and Hard of Hearing 800-342-3262 or **www.tndeaflibrary.nashville.gov**
- Tennessee Services for the Blind and Visually Impaired, cathy.steger@state.tn.us, 800-270-1349 (TTY); 800-628-7818 (voice)
- TREDS (Technical Assistance & Resources for Enhancing Deaf/Blind Support) 800-288-2266 **kc.Vanderbilt.edu/TREDS**

Tennessee Centers for Speech, Hearing and Aural Rehabilitation

- Baptist Memorial Healthcare Pediatric Audiology Center, Memphis 901-226-5682
- Blount Hearing and Speech Services, Maryville 865-982-8557
- ENT Associates of Middle TN, Shelbyville 931-684-3504
- ETSU Audiology Clinic, Johnson City 423-439-5252
- Hearing Services of TN, Franklin 615-591-6410
- Memphis Oral School for the Deaf 901-448-8490 **www.oraldeafed.org/schools/memphis**
- Mid-East TN Speech and Hearing Center, Dayton 423-775-0303
- Mountain Region Speech and Hearing Center Kingsport 423-246-4600
- Speech and Hearing Center, Chattanooga 423-622-6900
- Tennessee School for the Deaf, Knoxville 865-594-6022 **www.tsdeaf.org**
- University of Memphis Speech and Hearing Center 901-678-5800
- UT Child Hearing Services, Knoxville 865-974-5453 **www.hearingandspeech.org**
- Vanderbilt Bill Wilkerson Center, Nashville 615-936-5000 **www.mc.vanderbilt.edu/VanderbiltBillWilkersonCenter/dhss.html**
- West TN School for the Deaf, Jackson 731-423-5705 **www.wtsd.tn.org**

NEWBORN HEARING SCREENING ORDER FORM FOR MATERIALS

The Tennessee Universal Newborn Hearing Screening Program materials are available for hospital, providers and others and are free of charge. You are encouraged to share materials with families of child bearing age, pregnant women and parents newborns. The brochures explain why and how newborn hearing screening is performed, and what the results mean. Materials 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:

- ☐ Your New Baby's Hearing (for expectant moms and their obstetricians) English & Spanish
- ☐ Your Baby's Hearing Screening Suggests a Referral (for parents of an infant that did not pass the screen), also in English & Spanish.
- ☐ Talking with Parents about Hearing Loss (for physicians, hearing screeners, nurses, audiologists) English
- ☐ Why Bother with Hearing Screens and Hearing Tests? (for pediatricians, family practitioners, and other professionals and parents) 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).
- ☐ Tennessee Newborn Hearing Parent Resource Packet (for parents of children who have been confirmed with a 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) from the previous blood specimen form to assure a link to the initial screening.

PROTOCOLS:

- ☐ TN Hospital and Birthing Center, Newborn Hearing Screening Protocols
- ☐ TN Early Intervention, Newborn Hearing Follow-Up Protocols
- ☐ TN Pediatric Audiologic Assessment and Amplification Guidelines
- ☐ TN Directory of Pediatric Hearing Screening Audiological Diagnostic and Early Intervention Providers

Call: 615-741-8530 or 615-262-6160 (Kathy Miller/Jacque Cundall)
Mail: Women's Health and Genetics, Newborn Hearing Screening Program, Cordell Hull Bldg., 5th Floor
425 5th Ave North, Nashville, TN 37247-4701

Fax: Newborn Hearing Screening 615-262-6159

Appendix 4

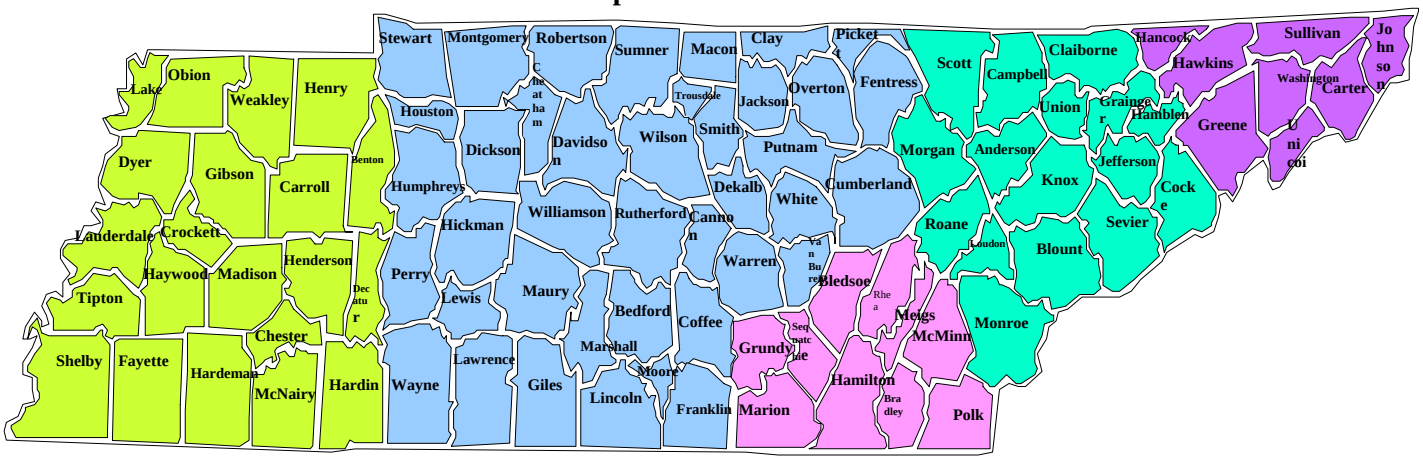
Tennessee Genetic Resources and Recommendations

The Tennessee Department of Health Newborn Metabolic and Hearing Screening programs collaborates 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

Referral Pattern for Hearing Loss

Tennessee Department of Health



U T Memphis
901-528-6594

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

Vanderbilt Univ.
615-322-7601

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

U T Chattanooga
TC Thompson
423-778-6112

Bledsoe
Bradley
Grundy
Hamilton
Polk
Marion
McMinn
Meigs
Rhea
Sequatchie

U T Knoxville
865-544-9030

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

ETSU
423-439-8541

Carter
Greene
Hancock
Hawkins
Johnson
Sullivan
Unicoi
Washington

Jan. 2004

Genetic Consultation and Evaluation Related to Hearing Loss

Genetic information provided below, as well as other information on other genetic conditions, may be found at http://www.genetests.org 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), DFN3 (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.

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 (see Definition). 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”. In fact, having a child with deafness may be preferred over having a child with normal hearing [Arnos et al 1992].
- Many deaf people are interested in obtaining information 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].
- The use of certain terms is preferred: probability or chance vs. risk; deaf and hard of hearing vs. 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.

Appendix 5

Audiology Reporting Form

Tennessee Department of Health
Newborn Hearing Screening, Identification and Monitoring Program
Women's Health and Genetics, Newborn Hearing Screening
Laboratory Services, 310 Hart Lane,Nashville, Tennessee 37247-0801
615-262-6160

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	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 ☐ TPOAE ☐ DPOAE ☐ ASSR ☐ Tympanometry ☐ Behavioral Testing

Test Results Suggest:

Degree of Hearing Loss:		**Ear**
Hearing Within Normal Limits		☐ R ☐ L
Mild	(<=40 dB HL)	☐ R ☐ L
Moderate	(41-60 dB HL)	☐ R ☐ L
Severe	(61-80 dB HL)	☐ R ☐ L
Profound	(>80 dB HL)	☐ R ☐ L
Sloping Hearing Loss		☐ R ☐ L
Unspecified Hearing Loss		☐ R ☐ L

Inconclusive, due to: _____

Type of Hearing Loss:	**Ear**
Hearing Within Normal Limits	☐ R ☐ L
Fluctuating Conductive HL	☐ R ☐ L
Permanent Conductive HL	☐ R ☐ L
Sensorineural Hearing Loss	☐ R ☐ L
Mixed Hearing Loss	☐ R ☐ L
Unspecified Hearing Loss	☐ R ☐ L

Inconclusive, due to: _____

Referrals:	**Date**
☐ No Referral	_____
☐ Repeat Hearing Testing	_____
☐ Medical Referral	_____
☐ Early Intervention Program	
☐ TEIS ☐ TIPS ☐ Other_____	_____
☐ Children's Special Services (CSS)	_____
☐ Speech/Language Services	_____
☐ Hearing Aid Fitting	_____
☐ Genetic Referral	_____
☐ Other_____	_____
Type and Location	

Follow-up date: _____

Comments: _____

_____

Provider: _____**Phone: (____)**_____
Audiologist, Medical Provider, Early Intervention Provider, Other

Address: _____

City:_____ **State/Zip:**_____

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

Appendix 6

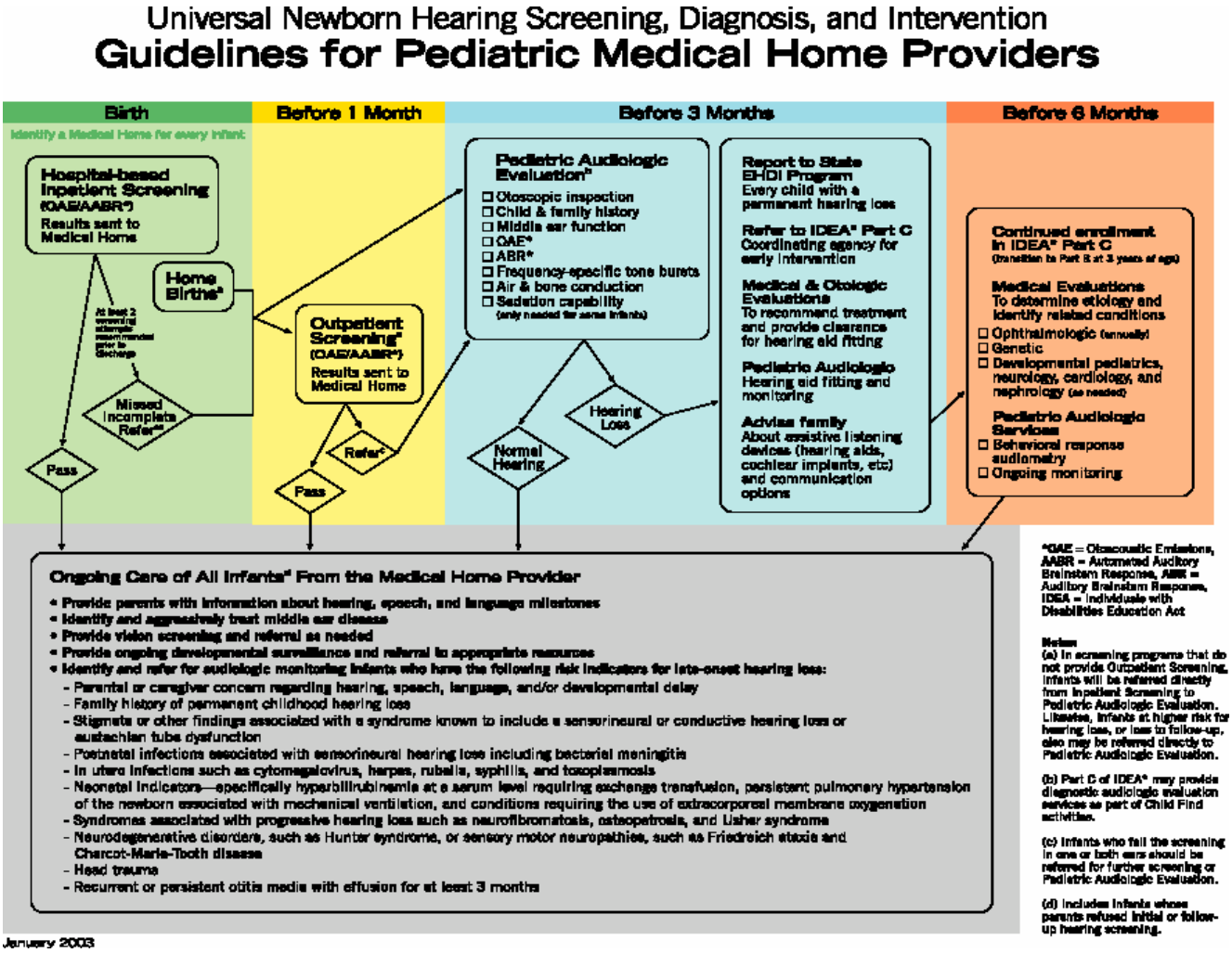
American Academy of Pediatrics (AAP) Guidelines for Medical Home Providers

Universal Newborn Hearing Screening Diagnosis and Intervention

This chart may be obtained through the AAP web site **www.aap.org**

The chart outlines the role of the Pediatric Medical Home Provider and the Audiologist. The back of the form has sections for physicians to record the name and number of local hearing team providers.

Side 1



American Academy of Pediatrics

Universal Newborn Hearing Screening, Diagnosis and Intervention
Guidelines for Pediatric Medical Home Providers

Side 2

Appropriate Referrals

1. Audiologist knowledgeable in pediatric screening and amplification

Name:	
Telephone number:	
Fax:	
Date of referral:	

2. Otolaryngologist knowledgeable in pediatric hearing loss

Name:	
Telephone number:	
Fax:	
Date of referral:	

3. Local early intervention system

Name:	
Telephone number:	
Fax:	
Date of referral:	

4. Family support resources, financial resources

Name:	
Telephone number:	
Fax:	
Date of referral:	

5. Speech/language therapy and/or aural rehabilitation therapy

Name:	
Telephone number:	
Fax:	
Date of referral:	

6. Sign language classes if parents choose manual approach

Name:	
Telephone number:	
Fax:	
Date of referral:	

7. Ophthalmologist knowledgeable in co-morbid conditions in children with hearing loss

Name:	
Telephone number:	
Fax:	
Date of referral:	

8. Clinical geneticist knowledgeable in hearing impairment

Name:	
Telephone number:	
Fax:	
Date of referral:	

9. Equipment vendor(s)

Name:	
Telephone number:	
Fax:	
Date of referral:	

10. State EHDI coordinator
http://www.infanthearing.org/statelists/ehdi.html

Name:	
Telephone number:	
Fax:	
Date of referral:	

11. AAP Chapter champion
http://www.medicalhomeinfo.org/screening/Champions%20Roster.pdf

Name:	
Telephone number:	
Fax:	
Date of referral:	

12. Family physician(s)

Name:	
Telephone number:	
Fax:	
Date of referral:	

National Resources

Alexander Graham Bell Association for the Deaf and Hard of Hearing (AG Bell)
202/337-5220
www.agbell.org

American Academy of Audiology (AAA)
800/AAA-2336
www.audiology.org

American Academy of Pediatrics
www.aap.org

American Society for Deaf Children
717/334-7922
www.deafchildren.org

American Speech-Language-Hearing Association (ASHA)
800/498-2071
www.asha.org

Boys Town Center for Childhood Deafness
www.babyhearing.org

Centers for Disease Control and Prevention
www.cdc.gov/ncbddd/ehdi

Cochlear Implant Association, Inc.
202/835-2781
www.cicia.org

Families for Hands and Voices
303/830-6753
www.handsandvoices.org

Laurent Clerc National Deaf Education Center and Gallaudet University
www.gallaudet.edu/clerc_center.html

National Association of the Deaf (NAD)
301/587-1788
www.nad.org

National Center on Hearing Assessment and Management (NCHAM)
www.infanthearing.org

National Institute on Deafness and Other Communication Disorders
www.nidcd.nih.gov

Oberkotter Foundation
www.pediatrics.org

The recommendations in this document do not indicate an exclusive course of treatment, or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

Copyright © 2002 American Academy of Pediatrics. No part of this document may be reproduced in any form or by any means without prior written permission from the American Academy of Pediatrics except for 1 copy for personal use.

This project is funded by an educational grant from the Maternal and Child Health Bureau, Health Resources and Services Administration, US Department of Health and Human Services.

American Academy of Pediatrics
DEDICATED TO THE HEALTH OF ALL CHILDREN™

NCHAM
National Center on Hearing Assessment and Management
Utah State University

Appendix 7

Audiology Acronyms

AAA	American Academy Audiology
ABR	Auditory Brainstem Response
AN/AD	Auditory Neuropathy/Dysynchrony
ANSI	American National Standards Institute
APD	Auditory Processing Disorder
ASA	American Society of Anesthesiology
ASHA	American Speech and Hearing Association
ASL	American Sign Language
ASSR	Auditory Steady State Response
BC	Bone Conduction
BTE	Behind-the-Ear
CIC	Completely-in-the-Ear
CM	Cochlear Microphonic
CPA	Conditioned Play Audiometry
CROS	Contralateral Routing of Signal
CSS	Children's Special Services
CT	Computed Tomography Scan
DAI	Direct Audio Input
dB	Decibels
DNA	Dioxyribonucleic Acid
DPOAE	Distortion Product Otoacoustic Emissions
FDA	Food and Drug Administration
FERPA	Federal Education Rights and Privacy Act
FM	Fequency Modulation
HIPAA	Health Insurance Portability and Accountability Act
HL	Hearing Loss
Hz	Hertz
IDEA	Individuals with Disabilities Education Act
ITC	In-the-Canal
ITE	In-the-Ear
JCAHO	Joint Commission on Accreditation of Healthcare Organizations
JCIH	Joint Commission on Infant Hearing
LEA	Local Education Agency
MRI	Magnetic Resonance Imaging
nHL	Normed Hearing Loss
NHS	Newborn Hearing Screening Program
NU-CHIPS	Northwestern University Children's Perception of Speech
OAE	Otoacoustic Emissions
PSI	Pediatric Speech Intelligibility Test
REAR	Real Ear Aided Response
RECD	Real Ear to Coupler Difference
SAT	Speech Awareness Threshold
SHARP	Situational Hearing Aid Response Profile
SNHL	Sensorineural Hearing Loss
SRT	Speech Reception Threshold
TDH	Tennessee Department of Health
TEIS	Tennessee Early Intervention System
TEOAE	Transient Evoked Otoacoustic Emissions
TIPS	Tennessee Infant Parents Services
TREDS	Tennessee Technology Assistance and Resources for Enhanced Deaf/Blind Supports
TROCA	Tangible Reinforcement Operant Conditioning Audiometry
VRA	Visual Reinforcement Audiometry
VROCA	Visual Reinforcement Operant Conditioning Audiometry
WIPI	Pediatric Speech Intelligibility Test
WNL	Within Normal Limits
WRS	Word Recognition Scores