Early Hearing Detection and Intervention

Arizona Pediatric Audiology Guidelines

GUIDELINES FOR SCREENING, HEARING ASSESSMENT, & TREATMENT

This document is the result of work completed by Arizona Audiologists who provide pediatric audiology services. Development meetings were held between February 1998 and December 1999.
Introduction

In 1998, 40% of all babies born in Arizona were screened for hearing loss before hospital discharge. In 1999 more than 85% will be given that opportunity and the expectation is that by the year 2000 screening will be universally available to newborns in Arizona.

This rapid change is going to mean a significant difference in the scope of practice of the Audiologists who are providing services to the pediatric population. It will mean that a growing and challenging percentage of caseloads will very newborns and infants. Working with children 0-3 years of age requires special skills, experience and equipment to assure that early identification efforts successfully result in optimal outcomes for children who are Deaf and hard of hearing.

Audiologists who are not able to provide these services, due to a lack of skill, experience or equipment are ethically obligated to refer families to facilities where the needed services can be obtained.

The goal of developing this document is to provide guidelines for care regarding providing pediatric audiology services while recognizing the diversity of individuals and communities. These guidelines have been developed specifically for services to children from birth to 36 months of age (0-3 years).

Definitions

Screening to detect, among apparently healthy persons, those individuals who demonstrate a greater probability for having a disease or condition, so they may be referred for further evaluation. 1

Hearing Assessment to determine frequency specific thresholds and other auditory behavioral or physiological information to identify and describe a hearing impairment as to degree, type and configuration of loss

Newborn Birth to 30 days
Infant 30 days to 1 year
Child birth to 36 months

I. Hearing Screening

Screening newborns before hospital discharge is the first step in helping children who are Deaf or hard of hearing reach their full potential. The goal should be:

**within the first month of age**
- identify those newborns who need further assessment,

**within the first three months of age**
- complete an appropriate audiological assessment

**before six months of age**
- fit amplification or other assistive devices based on the infant’s needs and to initiate appropriate early intervention services.

The purpose of screening is to identify those individuals having a defined disorder as early as possible, who would have otherwise not been identified, and to administer treatment at a time when it will either remediate the disorder or retard its development (Frankenberg W. K. Pediatrics. 54; 1971).

**Hearing screening for all age groups:**
If a recommended screening cannot be completed, a second attempt to screen should be conducted within 30 days. If the result of the rescreening is refer or if a rescreen cannot be completed a referral should be made for an assessment by an audiologist who is skilled, experienced and equipped to provide services to the pediatric population.
Screening-Newborn to 6 months of age:

At least one of the following screens must occur:
- Screening Auditory Brainstem Response (ABR)
  - Pass = presence of wave V; stimulus intensity of 35 dB n HL.
- Otoacoustic Emissions (OAE) including Distortion Product (DP) or Transient Evoked (TE)
  - DPOAE: Pass = Six (6) dB above the noise floor from 2000 through 5000 Hz with F1-F2 stimulus levels of 65 - 55
  - TEOAE: Pass = Reproducibility must be 70% or greater for three (3) or more individual frequency bands at 3 dB above the noise floor (one of which must be 3,600 - 4,000 Hz).

If a screening does not result in a pass in both ears for one of the above screens, a rescreen should occur within 30 days.

Newborns with high-risk conditions for auditory neuro dysfunction should have an OAE and ABR screening before hospital discharge. The high risk conditions may include (but are not limited to) the following:

- Hyperbilirubenemia
- Arnold-Chiari malformations
- Charcot-Marie Tooth Syndrome
- Velo Cardio Facial Syndrome
- De George Syndrome
- Hunter / Hurler Syndrome
- Other neurological disorders

Infants with conditions causing increased risk of progressive hearing loss due to auditory neuro dysfunction should have a screening ABR within one month of discharge if it is not completed before discharge. A list of these conditions is included as Appendix A. If the ABR is abnormal, an OAE should be completed and the child should be referred for periodic reassessment.

If a newborn passes the hearing screening, rescreening should occur if there are parental concerns or if the child has risk factors of progressive hearing loss such as family history, kidney problems, etc. See appendix A for more information about risk factors.
Screening 6 to 36 months of age:

A multi-dimensional process should occur.

The following screenings may be performed:
- ABR, if it can be done unsedated: Pass = presence of wave V; intensity of 35 dBn HL (a sedated ABR is considered an assessment)
- Otoacoustic Emissions (OAE): Distortion Product or Transient Evoked
  - DPOAE: Pass = Six (6) dB above the noise floor from 2000 through 5000 Hz with a stimulus level of 65-55.
  - TEOAE: Pass = Reproducibility must be 70% or greater for three (3) or more individual frequency bands one of which must be 3,600-4,000 Hz
- In children between 24 and 36 months of age, behavior hearing screening using pure tone play audiometry may be used if the child is developmentally ready

Middle ear function tests such as tympanometry, reflectometry or reflex thresholds can not be used as a hearing screening but may be used along with another screening method.

Parental concerns or behavioral history should indicate the need for assessment (not a screening) by an audiologist

If a flat tympanogram is obtained or reflectometry results suggesting middle ear involvement, with otherwise normal hearing screening, referral should be made for medical assessment prior to the rescreen.

Screening may be provided by:

1. Nurses, technicians or aides/volunteers who have been specifically trained and have demonstrated competence in providing the screening.
2. An audiologist should provide oversight and direction to any individual providing screening.

Communicating with Families at the time of the screening.

- Provide information that a pass does not mean hearing will always be normal; referral for re-screening does not mean there is definitely a problem.
- Inform parents that their child has passed or is being referred for rescreening.
- Provide brochures listing the normal development stages of language and auditory behavior.
- Tell parents what to do or who to contact if they have concerns.
- Instruct parents on where should go for follow-up screening or tests.
- Provide information in writing in their native language if possible.

All personnel involved in the screening process need to understand and reinforce with the parent that this is a screen not a test.
II. Audiolcic Assessment Following Refer from Screening

The following principles underlie these guidelines:

Audiologic assessment should occur within 30 days and no later than 90 days of the screening.

Frequency specific and ear specific information must be obtained to confirm presence or absence of hearing loss.

Assessment should be initiated after a two stage screening process unless there are other concerns such as speech, language, or parental concerns. If these concerns exist, the child should go directly to the audiolcic assessment.

A battery of tests is important (some behavioral as well as more objective physiological assessment).

Confirmation of hearing loss should be provided by an Audiologist who is experienced and skilled in pediatric assessment and has access to the equipment required.

The order of tests may vary based on the individual child, the case history, the results of the screening, and the age of the child.

A child being assessed for diagnostic purposes usually must pass the test that was failed during the screening process before a determination can be made that hearing is normal.
**Assessment 0 to 6 months:**

Any of the following assessment tools are acceptable; however, at a minimum an ABR should be completed.

**ABR**

For newborns and infants 0 to six months of age an ABR should be completed.

- Frequency Specific ABR- 500 and 4,000 Hz
- If tone burst stimuli is not available, click ABR without tone burst and a follow-up recommendation for a behavioral assessment in 3 – 6 months if the click ABR is passed.
- If elevated ABR, a bone conduction ABR, High frequency tympanometry, and OAE (TE and/or DP) should be assessed.

**OAE/TEOAE or DPOAE in conjunction with ABR**

- DPOAE: Pass = Six (6) dB above the noise floor from 2000 through 5000 Hz with $F_1$-$F_2$ stimulus level of 65 – 55.
- TEOAE: Pass = Reproducibility must be 70% or greater for three (3) or more individual frequency bands one of which must be 3600 – 4,000 Hz.

Behavioral assessment with insert earphones or in a calibrated sound field with either warble tone or narrow band noise as well as speech awareness. The Behavioral assessment results should be confirmed by an ABR.
Assessment at developmental age of 6 months to 36 months of age:
The goal of the assessment is to obtain separate ear, discreet frequency, hearing threshold information. The following assessment(s) should be completed:

1. History on auditory behavior, health and family, birth, developmental status
2. Determine threshold hearing levels; frequency specific and ear specific. The assessment may include:
   - VRA audiogram using insert earphones for both ears at 500, 1000, 2000 and 4000 Hz.
   - OAE same on each side. If OAEs are not available, ear specific behavioral information with symmetrical acoustic reflexes is adequate.
   - Frequency specific ABR
   Hearing levels must be normal in both ears for hearing to be considered normal.
3. Tympanometry. If abnormal results are obtained a medical referral should be made.
4. With normal sound field responses, follow up in 3 months with another attempt at ear specific assessment, if a sedated ABR / OAE cannot be completed within that 3 months.
5. Equipment must be calibrated to current ANSI standards annually.

Communicating with Families when a hearing loss is identified:
Families need information when a baby is identified as having a hearing loss. Some parents will be ready for more information sooner than others. The following topics should be discussed with parents within the first few months after a hearing loss is diagnosed:

- Results of the assessment
- Information about the functioning of the ear and the implications of the test results.
- Recommendations for referrals the audiologist will be making to the primary care physician.
- Information about the need for medical follow-up, as applicable.
- Communication options
- Recommendations for intervention
- Funding / payment options
- The importance of early intervention
- Counseling and social work support
- A resource guide
- Educational options - settings from mainstream to ASDB
- Auditory development skills list
- Parent support groups/deaf adult groups
III. Treatment

If a hearing loss is diagnosed, the following steps should be taken:
- Referral to Otologist/Otolaryngologist
- Referral to ASDB Parent Outreach
- Referral for hearing aid fitting consultation
- Pediatric Audiologist
- Referral for pediatric speech/language assessment

The following guidelines apply to fitting of hearing aids:
A child needs hearing aids when there is a significant permanent, bilateral peripheral hearing loss. Some children with variable and/or unilateral losses may also need hearing aids. There are no empirical studies that delineate the specific degree of hearing loss at which need for amplification begins. However, if one considers the acoustic spectrum of speech at normal conversational levels in the 1000–4000 Hz range, hearing thresholds of 25dB HL or greater can be assumed to impede a child’s ability to perceive the acoustic features of speech necessary for optimum aural/oral language development. Hence, thresholds equal to or poorer than 25 dB HL would indicate candidacy for amplification in some form. For children with unilateral hearing loss, rising or high-frequency hearing loss above 2000Hz and/or milder degrees of hearing loss (<25dB HL), need should be based on the audiogram plus additional information including cognitive function, the existence of other disabilities, and the child’s performance within the home and classroom environment.²

The following information should be obtained:
- Threshold estimates for better ear based on the assessment tools identified above
- Parental consent
- Medical clearance from Physician (ENT preferred)
- Binaural fitting if there is a bilateral loss
- RECD-real ear to coupler difference measures

The following factors must be taken into consideration when fitting amplification devices:

**Preselection**  
**Physical Characteristics**

Even at a very young age, consideration should be given to the availability of appropriate coupling options on hearing aids so that the child will have maximum flexibility for accessing the various forms of current assistive device technology. Consequently, hearing aids for most children should include the following features: Direct Audio Input (DAI), telecoil (T), and microphone-telecoil (m-T) switching options. Hearing aids used with young children also require more flexibility in electro-acoustic parameters (e.g. tone, gain, output limiting) than for adults, as well as more safety-related features such as battery and volume controls that are tamper resistant.

The physical fit of the hearing aids (in most cases worn behind the pinnae) and ear molds is important for both comfort and retention. Color of the hearing aids and earmolds needs to be considered across ages, and size of hearing aids is an especially important cosmetic concern for older children. Earmolds should be constructed of a soft material.

While consideration needs to be given to the aforementioned physical factors, the ultimate goal is the consistency and integrity of the amplified signal that the child receives. Providing the best possible amplified speech signal should not be compromised for cosmetic purposes, particularly in the early years of life when speech-language learning is occurring at a rapid pace.

Binaural amplification should always be provided to young children unless there is a clear contraindication. Even if there is audiometric asymmetry between ears as evidenced by pure tones or speech perception, hearing aids should be fitted binaurally until it is apparent from behavioral evidence that a hearing aid fitted to the poorer ear is detrimental to performance.

In general, behind-the-ear (BTE) hearing aides are the style of choice for most children. However, for children with profound hearing loss, body aids or FM systems may be more appropriate because of acoustic feedback problems limiting sufficient gain to provide full audibility of the speech signal in BTE arrangements. Other circumstances that may indicate the need for body-worn amplification include children with restricted motor capacities and those confined by a head restraint. In-the-ear (ITE) hearing aids may be considered appropriate when ear growth has stabilized at about 8-10 years of age as long as the flexibility and options available are not markedly restricted by concha and ear-canal size. 3

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3 Ibid., p. 54
Other considerations:

Primary amplification vs. supplemental
Soft earmolds
Programmable hearing aids
Safety features like:
  - Volume control covers
  - Battery door devices
Small behind-the-ear hearing aids
Loss and damage insurance for the hearing aid
Cochlear implants (some information to everyone even if not a candidate)
  (See Appendix for specific information about cochlear implants)
  - who qualifies
  - who to refer to
  - how much they cost
  - what realistic expectations are
  - what the cochlear implant is and does
Tactile aid (information only if indicated)
FM systems
  - what they do
  - who is a candidate
  - use with unilateral, mild or central auditory nervous system deficits
Alerting devices/dogs

The following equipment is needed to fit amplification on this population:
Test equipment that meets current ANSI standards to verify that the electroacoustical characteristics meet the manufacturer’s specifications.
In situ or simulated measurements to verify the frequency gain characteristics and maximum power output.

Communicating with Families when fitting an amplification device:
Families need information about the hearing aid or other amplification device. Communication should be in written as well as 1:1 to allow questions to be answered.
  - Information from the manufacturer about the device
  - use/care/function
  - disposal of batteries
  - Information about follow up
  - Information on repeat testing
  - Information on replacing the hearing aid and/or earmolds
  - Information on realistic expectations
  - Instruction on how to do listening checks
  - Instruction on taking care of the hearing aids
IV. Ongoing Assessment & Monitoring

Monitoring for children who are hard of hearing or Deaf should occur every three months during the first two years of using amplification and every six months thereafter.

Monitoring exams may include:

- An audiometric evaluation
- Electroacoustic evaluation and analysis
- Listening checks of the hearing aid
- Re-evaluation of the RECD
- Other probe microphone measures as appropriate
- Validation of expected amplification

Monitoring for children who are at risk for progressive hearing loss should occur at least every 6 months or more frequently as determined by the audiologist and the parent/caregiver.

The parent/caregiver should be provided a log/checklist for documenting certain behaviors in order to monitor auditory function in their natural environment.

An evaluation of the child’s needs should include:
- Home and/or child care environment
- Family dynamics
- Any other factors unique to the child’s natural environment

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4 Ibid. p. 56.
V. Referrals

The following recommendations for referrals should be made to the primary care physician for children with newly identified hearing loss:

- Medical referral to an Otologist/Otolaryngologist with pediatric experience
- Referral to ASDB Parent Outreach Program
- Referral for speech/language assessment with a Pediatric Speech/Language Pathologist
- Referral for hearing aid consultation with a Pediatric Audiologist

The Pediatrician or Primary Care Provider should be informed at each step.

The audiologist should consider additional recommendations to the Primary Care Physician (PCP) for referrals to the following specialists depending upon the specific needs of the child:

- Vision screening
- Genetic counseling
- Kidney
- Cardiology
- Neurology
- Developmental assessment

The recommendation for referral should state why the specific referral is being recommended. The Pediatric Audiologist should be aware of other related conditions and should inform the PCP and the parents of the likelihood of these conditions.

Children referred to a physician for conductive hearing loss should be re-assessed following completion of the medical intervention. The audiologist should inform the parents of this need for re-assessment and should include the documentation in the referral to the primary care physician.

Medical assessment of possible middle ear disorders should not preclude assessment for permanent hearing loss. It is ideal if transient middle ear fluid can be cleared before audiological assessments are complete (within 30 days of screening) but, if middle ear fluid persists, every effort should be made to assess hearing status within the three month window. Amplification and early intervention should not be delayed beyond six months of age.
VI. When is an Interdisciplinary Team needed and who should be involved?

Any child with an identified hearing loss must have multiple disciplines involved in their care and treatment. It is important that the disciplines communicate and share information, however, although desired, it is often not practical for the multiple disciplines to actually meet as a team. Involvement of other disciplines may include but is not limited to the following:

- Audiologist
- Speech Pathologist
- Parent Outreach Services (In Arizona, the Parent Outreach Program may be the coordination point for the multiple disciplines)
- Primary Care Physician
- Neurologist if the child has auditory neuropathy
- The parent and/or primary caregiver should always be involved
VIII. Pediatric Audiologist Qualifications

Pediatric Audiologists must meet the following qualifications prior to providing hearing assessment and amplification for newborns and infants:

- Hold a current licence to practice Audiology in Arizona
- Hold a current license to dispense hearing aids in Arizona (if required)
- Have experience working, as an audiologist, with newborns and infants.
- Access to adequate equipment and environment to allow accurate assessment of degree and type of hearing loss
- Have the training and experience necessary to assure adequate skills in assessing the hearing of newborns and infants.
IX. Recommendations

There remain several unmet needs that should be addressed to improve outcomes for newborns and infants who are Deaf and hard of hearing. Those needs include:

- Assuring that families are not lost during the process
- Educating pediatricians and other primary care providers about where they can obtain a rescreening and what the screening means.
- Addressing the inherent delays and cost involved in obtaining preauthorization.
- Educating health care providers on appropriate referrals
- Standardizing the eligibility criteria for CRS
- Educating AHCCCS and remove barriers to providing timely, cost effective and appropriate services including:
  - Simplifying authorization for follow up services
  - Assuring reimbursement for first and second screening
  - Clarifying EPSDT requirements to include a physiological screen
- Developing license, certification or specialty recognition for pediatric audiologists.
- Developing and publishing a list of where infants can be screened
- Developing and using functional assessment tools to validate hearing aid benefit
Appendix A  Risk Factors

Example 1

Infants at risk for delayed onset or progressive hearing loss include:

1.71.1 Meningitis
1.71.2 Long-term ototoxic medications
1.71.3 Intrauterine TORCH infections
1.71.4 Infrequently found syndromes associated with progressive hearing loss including:

1.71.4.1 Albers-Schonber Disease of Osteopetrosis
1.71.4.2 Alport’s Syndrome
1.71.4.3 Muckel-Wells Syndrome
1.71.4.4 Cleindocranial Dysostosis
1.71.4.5 Ectodermal Dysplasia
1.71.4.6 Fanconi Anemia Syndrome
1.71.4.7 Harboyan Syndrome
1.71.4.8 Griedrich’s Ataxia
1.71.4.9 Norrie’s Syndrome
1.71.4.10 Optic Atrophy
1.71.4.11 Polyneuropathy
1.71.4.12 Juvenile Paget’s Disease
1.71.4.13 Pyle’s Disease
1.71.4.14 Tichards-Rundle Syndrome
1.71.4.15 Sensory Tadicular Neuropathy
1.71.4.16 Von Tecklinghausen’s Neurofibromatosis
1.71.4.17 Wardenburg’s Syndrome
Example 2.5

The following risk factors are identified in the literature to cause hearing impairment. They are recommended as a guideline to obtain an Automated Auditory Brainstem Evoked Response (AABR or BAER).

1. Severe asphyxia  Apgar score of 0-3 at 5 minutes who fails to institute spontaneous respiration with ten (10) minutes or with hypotonia that persists during the first two (2) hours of age.
2. Birth weight less than 1,500 grams.
3. Hyperbilirubinemia that requires exchange transfusion
4. Meningitis
5. Defects of head/neck/face such as ear deformities, cleft palate, dysmorphic appearance.
6. Persistent pulmonary hypertension of the newborn (PPHN/PFC).
7. Extracorporeal membrane oxygenation (ECMO)
8. Family history of a congenital childhood hearing impairment.
10. Congenital perinatal infections:
   - Rubella
   - Herpes Simplex
   - Toxoplasmosis
   - Syphilis
   - Cytomegalovirus (CMV)
11. Length of stay greater than 72 hours.
12. Maternal drug or alcohol use.

5 Chandler Regional Hospital, Hearing Screening, Newborn Nursery Guidelines
Appendix B  Cochlear Implants  
(Submitted by Karla Balko, M.S., CCC-A; November 20, 1998)

Who Qualifies for Cochlear Implants?

Severe-Profound (>90 dB) sensorineural hearing loss in both ears (if all other candidacy criteria are met, hearing threshold levels are reviewed on an individual basis)  
Age 18 months or older (Studies (Cohen NL, et al. 1996) have shown that the earlier a child is implanted the potential benefits can be enhanced.)  
Use of appropriate amplification and participation in intensive aural habilitation for 3-6 months (If the child has etiology of meningitis and serial CT Scans show evidence of ossification development, this is waived.)  
Receive little or no useful benefit from hearing aids (candidacy cannot be determined by hearing sensitivity thresholds and/or aided thresholds alone)  
Younger children (18 months-5 years) defined as failure to develop basic auditory skills as quantified by either a Meaningful Auditory Integration Scale (MAIS) or Early Speech Perception (ESP) Test.  
Older children (5 years and up) defined as minimal open-set word recognition, i.e. <20% correct on the Multisyllabic Lexical Neighborhood Test (MLNT) or the Lexical Neighborhood Test (LNT)  
No medical contraindications  
Hearing loss of neural or central origin  
Absence of cochlear or VIII nerve development  
Active middle ear infections  
Tympanic membrane perforation in the presence of active middle ear disease  
Other contraindications for surgery  
Psychological contraindications  
High motivation and appropriate expectations by the family, and the child as well (if age appropriate).

Who to Refer To?

A physician who is trained in the delivery of medical care that is specific to implantation in pediatric patients, i.e. assessment of potential implant candidates, surgical procedures, and post-operative management.

An audiologist who is trained in the delivery of audiological service that is specific to implantation in pediatric patients, i.e., assessment of potential implant candidates, device fitting, application of electrical stimulation, and post-operative management.
**How much does an implant cost?**

Costs for the pre-implant assessment, the implant system, surgery, post-surgical fitting and initial follow-up visits are generally $40,000 to $50,000. If this is not a covered benefit with the child’s health plan, the implant team will assist in the appeal process or identify other resources.

**What are realistic expectations?**

The cochlear implant does not restore normal hearing function. In addition, not all individuals using a cochlear implant will receive the same benefit; nor can the amount of benefit be predicted prior to implantation. The Cochlear Implant Team will analyze the pre-implant test results to determine if a cochlear implant will provide significant improvement over what can be achieved via current hearing aid technology for that particular child.

**What is a cochlear implant?**

It is an electronic device that is placed in only one ear. It is designed to bypass the damaged or absent hair cells by providing electrical stimulation to the nerve fibers, which in turn allows the recipient to receive sound. The American Academy of Otolaryngology - Head and Neck Surgery recognizes the cochlear implant as a standard treatment for bilateral profound hearing loss. The cochlear implant has been an FDA approved device for adults since 1985 and for children since 1990. There are three implant systems that are FDA approved for use in the United States for Children:

- Cochlear Corporation
  - Nucleus 22 Channel Cochlear Implant System
  - Nucleus 24 Channel Cochlear Implant System
- Advanced Bionics Corporation
  - Clarion Multi-Strategy Cochlear Implant

Appendix C - Participants

The following individuals attended one or more of the planning sessions.

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