



ARKANSAS DEPARTMENT OF HEALTH PEDIATRIC AUDIOLOGICAL ASSESSMENT

The following best practice recommendations were developed by the Arkansas Department of Health's Infant Hearing Program in collaboration with the Universal Newborn Hearing Screening, Tracking, and Intervention Advisory Board. The recommendations take into consideration information from national organizations, such as, the National Center for Hearing Assessment and Management (NCHAM), the Joint Committee of Infant Hearing (JCIH), the American Academy of Audiology (AAA) and the American Speech-Language-Hearing Association (ASHA).

This document serves to facilitate the diagnosis of hearing loss in infants. The Arkansas Department of Health recognizes individual cases may present unique challenges influencing the approach to evaluation and management. Therefore, these recommendations should be regarded as a guideline for practice, not as standards, and are not intended or designed to substitute for the reasonable exercise of independent clinical judgment. However, the protocol presented is consistent with generally accepted "standards of care" in pediatric audiology.

Our goals for babies born in Arkansas are:

- To screen all newborns' hearing before they leave the hospital;
- To re-test those who do not pass the first screen before one month of age;
- To confirm hearing loss by diagnostic audiologic testing before three months of age; and
- To enroll infants identified with hearing loss in appropriate early intervention services before six months of age.

I. INITIAL HEARING SCREEN/RESCREEN

- Otoacoustic emissions (OAEs) and automated auditory brainstem response (ABRs) are physiologic measures that can be used to screen newborns and infants for hearing loss. The following summarized protocols for the initial hearing screen/rescreen process were obtained from the 2007 JCIH Position Statement.
 - *Screening Protocols in the Well-Infant Nursery*
 - OAEs and automated ABRs are both acceptable screening measures for infants in the well-infant nursery.
 - Infants in the well-infant nursery who fail automated ABR testing should not be rescreened by OAE testing and "passed," because such infants are presumed to be at risk of having a subsequent diagnosis of auditory neuropathy/dyssynchrony.
 - *Screening Protocols in the NICU*
 - JCIH recommends ABR technology as the only appropriate screening technique for use in the NICU.
 - For infants who do not pass automated ABR testing in the NICU, referral should be made directly to an audiologist for rescreening and, when indicated, comprehensive evaluation, including diagnostic ABR testing, rather than general outpatient rescreening.
 - *Outpatient Rescreening for Infants Who Do Not Pass the Birth Admission Screening*
 - The outpatient rescreening should include the testing of both ears, even if only one ear failed the inpatient screening.
 - If the infant does not pass the rescreening, a referral for a diagnostic evaluation should be made to an audiologist.

II. PROFESSIONAL QUALIFICATIONS FOR PROVIDERS COMPLETING DIAGNOSTIC ASSESSMENTS

Any audiologist accepting infants for diagnostic audiological evaluations should be prepared with the equipment, training and skills to complete the procedures recommended by (JCIH). A licensed audiologist with experience in the pediatric population is the professional best qualified to perform diagnostic audiological assessments for infants. If the audiologist does not have the expertise and/or equipment necessary to evaluate and manage infants/young children, consideration for a referral should be made to professionals and facilities that provide pediatric diagnostic services.

III. RECOMMENDED PEDIATRIC AUDIOLOGIC ASSESSMENT

Birth to 6 Months of Age

When testing infants birth to 6 months of age, the confirmatory audiological test battery includes:

- Child and family history (including an evaluation of risk factors for congenital hearing loss).
- Assessment of parental report of the infant’s response to environmental sounds.
- Clinician observation of the infant’s auditory behavior as a crosscheck in conjunction with electrophysiological measures.
- Confirm middle ear status with one or more of the following techniques:
 - High frequency tympanometry.
 - Pneumatic otoscopy by a physician experienced in its application.
- Acoustic reflex threshold testing should be considered especially with concern of auditory neuropathy.
- Distortion product and/or transient evoked OAEs (TEOAE and/or DPOAE).
- Frequency specific assessment of hearing sensitivity using ABR air-conduction tonebursts at 500Hz, 1000Hz, 2000Hz, and 4000Hz with bone-conduction tonebursts utilized when indicated.
 - When permanent hearing loss is detected, frequency-specific ABR testing is needed to determine the degree and configuration of hearing loss in each ear for fitting of amplification devices.
- Click-evoked ABR testing using both condensation and rarefaction single-polarity stimulus, if there are risk indicators for neural hearing loss (auditory neuropathy/auditory dyssynchrony) such as hyperbilirubinemia or anoxia, to determine if a cochlear microphonic is present.
 - Because some infants with neural hearing loss have no risk indicators, any infant who demonstrates “no response” on ABR elicited by tone-burst stimuli must be evaluated by a click-evoked ABR.

Six Months to 36 Months of Age

For subsequent testing of infants and toddlers at developmental ages of 6 to 36 months, the confirmatory audiological test battery includes:

- Child and family history including an evaluation of risk factors for congenital hearing loss and parental report of auditory and visual behaviors and communication milestones.
- Confirm middle ear status with one or more of the following techniques:
 - Tympanometry.
 - Pneumatic otoscopy by a physician experienced in its application.
- Acoustic reflex threshold testing should be considered especially when concern is present of auditory neuropathy.
- Distortion product and/or transient evoked OAEs (TEOAE and/or DPOAE), when indicated.
- Behavioral audiometry (either visual reinforcement or conditioned-play audiometry, depending on the child’s developmental level), including pure-tone audiometry across the frequency range for each ear.
- ABR testing if responses to behavioral audiometry are not reliable.

IV. ONCE ASSESSMENT IS COMPLETED

- It is important to discuss the results of the evaluation and follow-up recommendations with parents.
- The audiologist is responsible for entering the initial diagnostic and any subsequent audiological evaluation results into ERAVE within 2 days following the appointment.
- Notify the infant’s Medical Home Provider (MHP) of results.

Normal Hearing

- Provide information to the parents about hearing, speech and language milestones and information regarding risk indicators for progressive hearing loss. Encourage the parents to schedule the infant for a follow-up evaluation in the future should concerns arise.

Normal Hearing (Identified as “At Risk” for Acquired or Late Onset Hearing Loss)

- The JCIH Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs has outlined those conditions that put infants at greater risk for late onset/progressive or progressive hearing loss. These indicators include:
 - Caregiver concern regarding hearing, speech, language or developmental delay.*
 - Family history of permanent childhood hearing loss.*
 - Neonatal intensive care admission of more than 5 days or any of the following, regardless of length of stay: ECMO, assisted ventilation,* exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.*
 - In utero infections, such as CMV, herpes, rubella, syphilis, and toxoplasmosis.*
 - Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits and temporal bone anomalies.
 - Physical findings, such as white forelock, that are associated with a syndrome known to include sensorineural or permanent conductive hearing loss.
 - Syndromes associated with hearing loss or progressive or late-onset hearing loss such as neurofibromatosis,* osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
 - Neurodegenerative disorders, such as Hunter syndrome* or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
 - Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.*
 - Head trauma, such as basal skull/temporal bone fracture, * that requires hospitalization.
 - Chemotherapy*
- *Greater concern for delayed onset hearing loss
- The timing and number of hearing reevaluations for children with risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss. According to the JCIH 2007 Position Statement, all infants with a risk factor for hearing loss should be referred for an audiological assessment at least once by 24 to 30 months of age. Children with risk indicators that are highly associated with delayed-onset hearing loss, such as having received ECMO or CMV infection, should have more frequent audiological assessments.
- The Arkansas Infant Hearing Program collects risk factor data from the birth facilities in the ERAVE system. The birth facility has the responsibility of assessing all newborns for risk factors, and for reporting any recognized risk factors to the IHP through ERAVE. The birth facility should notify the newborn’s Medical Home Provider (MHP) of any identified risk factors associated with the potential for late onset/progressive or progressive hearing loss that warrants the need for ongoing audiological evaluations.

Confirmed Hearing Loss

- If a permanent hearing loss is present, the audiologist should make the necessary accommodations for the infant to be fit with amplification and be seen for follow up care, as needed. This includes a referral to Early Intervention (EI), medical clearance from an ENT, and all results/recommendations sent to the Medical Home Provider (MHP). The diagnosing audiologist should provide the parent/responsible party with the information they need to make informed decisions regarding early intervention options.

- If the presence of middle-ear fluid or pathology is noted with a hearing loss, the infant should be referred to the Medical Home Provider (MHP) for diagnosis, treatment, and possible referral to ENT. A follow-up appointment should be scheduled to determine middle ear status and complete testing to rule out sensorineural hearing loss within 4-6 weeks. If middle ear fluid persists, every effort should be made to assess hearing status within the three month time window.

V. ADDITIONAL RESOURCES

- Arkansas Department of Health – Infant Hearing Program: <http://www.healthy.arkansas.gov/programsServices/familyHealth/ChildAndAdolescentHealth/InfantHearing/Pages/default.aspx>
- JCIH 2007 Position Statement: Principals and Guidelines for Early Hearing Detection and Intervention Programs: <http://www.jcih.org/posstatemts.htm>.
- American Academy of Audiology (AAA): <http://www.audiology.org/publications-resources/document-library/pediatric-diagnostics>
- American Speech-Language Hearing Association: <http://www.asha.org/default.htm>

The Arkansas Department of Health provides resources/handouts to clinics and hospitals that are helpful in guiding parents through this process. The goal of the Arkansas Department of Health’s Infant Hearing Program is to provide resources and support to families and providers across the state of Arkansas. If you have any questions or are in need of additional information, please contact us.

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