

IX. DIAGNOSTIC TESTING GUIDELINES

It is the experience of other states that as many as 80% of the newborns referred from newborn hearing screening will pass a rescreen at a later date. Based on this information, it is recommended that all infants who are referred from a newborn hearing screening program because they failed two (2) hearing screenings in one or both ears should be seen for an outpatient ABR rescreen or an Evoked Otoacoustic Emissions rescreen. If the infant doesn't pass the outpatient rescreen, then a full diagnostic audiological assessment should be completed at the same appointment. It is suggested that the rescreen be conducted with a methodology different than that used in the hospital. For example, if the infant is screened with ABR equipment in the hospital, the outpatient rescreen should be done with Evoked Otoacoustic Emissions equipment.

Within the first two (2) months of life, the following procedures should be completed on all infants referred from the newborn hearing screening process.

A. Auditory Brainstem Response or Otoacoustic Emissions Rescreen Guidelines

Depending on the availability of equipment one of the following should be conducted:

1. Rescreen with an Auditory Brainstem Response Screen.
 - a. Obtain a 70 or 75 dBnHL response to click stimulus to assess the latency and morphology of waves I, III, V, I-III, and I-V.
 - b. Obtain a 30 or 35 dBnHL response to click stimulus to assess latency and morphology of wave V.
2. Rescreen with an Automated Auditory Brainstem Response Test (AABR).
3. Rescreen with Evoked Otoacoustic Emissions.
 - a. Obtain Transient Evoked Emissions and/or
 - b. Obtain Distortion Product Otoacoustic Emissions.
4. Infants who do not pass the rescreen in one or both ears should receive an audiologic diagnostic assessment at the same appointment.

B. Audiologic Diagnostic Assessment Guidelines

In 1999, the Illinois legislature passed the Hearing Screening for Newborns Act. By December 31, 2002, all hospitals delivering babies will be required to provide hearing screening to all babies born in their facility. The goals of the Illinois program are:

1. All infants born in Illinois will have their hearing screened.
2. All newborns referred from the Illinois Newborn Hearing Screening Program will have diagnostic testing completed by three (3) months of age.
3. All infants diagnosed with significant hearing loss will receive appropriate treatment, including hearing aids and Early Intervention Program services by six (6) months of age.

As of May, 2000, ninety-three (93) Illinois hospitals are providing universal newborn hearing screening. This is only the first step in the identification of babies with hearing loss, however.

Screening is not very useful if those who are referred for diagnostic testing are not able to receive appropriate diagnostic services.

It is essential that audiologists to whom babies are referred for diagnostic evaluations have experience using ageappropriate diagnostic techniques for such infants. Various organizations such as the American Speech-Language-Hearing Association have produced guidelines that can be helpful. In Illinois, the Department of Human Services Newborn Hearing Screening Advisory Committee has recommended diagnostic audiologic guidelines for the state. The purpose of this document is to inform audiologists testing Illinois infants of those guidelines.

It is also important for families of babies with suspected hearing loss have access to financial assistance so that no infant goes without diagnostic testing, medical treatment, or early intervention services due to financial reasons. All audiologists have been encouraged to become providers for the Illinois Department of Public Aid (IDPA), the University of Illinois Division of Specialized Care for Children (DSCC), and the Illinois Early Intervention Program (EI).

All audiologists working with infants referred from newborn hearing screening programs are encouraged to adopt the following guidelines in their practice.

Significant Hearing Loss means a dysfunction of the auditory system of any type or degree that is sufficient to interfere with the acquisition of speech and language skills. The methodology used to evaluate infant hearing should detect, at a minimum, unilateral or bilateral hearing loss > 35 dB HL or > 30 dBnHL.

The use of a test battery is strongly recommended in the evaluation of infants and young children in order to provide a cross-check system for accurate diagnosis. It is preferred that infants be tested without sedation to the extent possible. If sedation is required, the audiologist, the patient's physician and the patient's family should discuss whether or not to sedate and should ensure that the appropriate protocol and monitoring is employed.

The use of a test battery is strongly recommended in the evaluation of infant and young children in order to provide a crosscheck system for accurate diagnosis. It is preferred that infants be tested without sedation to the extent possible. For infants under 12 months of age, the use of sedation is absolutely not appropriate to determine unilateral loss. Sedation is appropriate for infants under 12 months of age only when bilateral hearing loss is suspected.

1. Perform an otoscopic evaluation.
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 2. Perform Auditory Brainstem Response/Auditory Evoked Potential (ABR/AEP):
 - a. Obtain threshold of each ear with air conductive click-evoked ABR utilizing <20 dB step threshold search.

Including:

 - 1) Evaluate response at a high level (i.e., 70 to 80 dBnHL)
 - 2) Evaluate absolute latencies for waves I, III, and V, compare to established norms.
 - 3) Evaluate interpeak latencies for waves I to III, III to V, and I to V.
 - 4) Evaluate waveform morphology relative to normal neonatal waveforms.
 - 5) Consider measuring wave V/I amplitude ratio.
 - 6) Consider evaluating click-rate functions.

- 7) Each clinic will need to establish its own normative values for infants. As a general guideline, normal sensitivity for clicks may be defined as a repeatable wave V or 30 dBnHL or less in each ear.
 - b. If the air conduction click ABR is abnormal, then perform a toneburst ABR at 500 Hz or 4000 Hz. Also obtain a bone conduction click ABR. Air bone gaps of 20dB or more or absent bone conduction response should be considered significant.
2. Perform Auditory Brainstem Response/Auditory Evoked Potential (ABR/AEP): Obtain ABR threshold in each ear using a click or high frequency (e.g. 4000 Hz) tone-burst stimuli. Actual thresholds should be determined in steps no greater than 10 dB, although larger step sizes may be appropriate early in the evaluation to initially bracket threshold region.

If ABR thresholds in response to click or high frequency tone-bursts are abnormal, ABR thresholds in response to tone-burst stimuli centered at 1000 Hz or lower should be assessed. An abnormal ABR threshold in response to click stimuli is defined as > 30 dBnHL. In some cases, the use of bone conducted stimuli may also be appropriate.

When appropriate, analysis of ABR morphology and waveform latencies in response to high intensity stimuli may be performed. Latencies may be compared to established norms. Additional measures may also be utilized as a part of an assessment for auditory neuropathy or other neurologic dysfunction of the auditory pathway.

3. Evaluate evoked otoacoustic emissions in each ear (TEAOE and/or DPOAE) to further evaluate cochlear function. OAEs may be evaluated even when click ABR thresholds are normal since it is possible to obtain frequency specific information for the low frequencies.
4. Behavioral testing should be conducted, as well, for normally developing infants over 6 months of age. Behavioral procedures should include separate-ear visual reinforcement procedures.
 - 4a. In addition, behavioral testing and audiometry should be considered, particularly in infants with suspected hearing loss.
5. If the click-evoked ABR thresholds are normal and OAEs are normal, the parents should receive written information about hearing, speech, and language developmental milestones and information regarding risk indicators for progressive hearing loss. If questions arise about the infant's hearing, speech, and/or language development, the infant should be referred for an age appropriate audiologic assessment.
 - 5a. All parents should receive written information about hearing, speech, and language developmental milestones and information regarding risk indicators for progressive or late onset hearing loss. If questions arise about the infant's hearing, speech, and/or language development, the infant should be referred for an age appropriate audiologic assessment.
6. Incorporate an immittance battery with caution for infants less than four months of age. Multi-component/multifrequency testing is the only appropriate immittance test for this age group.
7. Complete the "Neonatal Hearing Screening Follow-up Services Report" and mail it to the Illinois Department of Public Health - Newborn Screening Program (attached).

8. If family does not have Medicaid or KidCare coverage for the infant and the family needs financial assistance to pay for the diagnostic evaluation, complete DSCC form 3.44 (Diagnostic Evaluation Authorization for Genetic Diseases Program/Newborn Hearing Screening Program) to obtain reimbursement from DSCC for diagnostic evaluation. If hearing loss has been confirmed, DSCC staff will contact the family to invite them to apply for assistance from DSCC.
9. If hearing loss is confirmed, refer the family to the appropriate Child and Family Connections office for early intervention services within 2 days of diagnosis.

C. Confirmed Hearing Loss Follow-Up Guidelines

The following should be completed by three (3) months of age:

1. If a bilateral/unilateral hearing loss of > 35 dB HL or 30 dBnHL in either ear is detected, refer the infant to an otolaryngologist for an examination and medical clearance (See Recommended Guidelines for Medical Protocol for Infants With Confirmed Hearing Loss). Begin the process of fitting amplification, if appropriate.

* Hearing loss is defined as an average of the frequencies 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz or if the hearing level of any two (2) of these frequencies is > 35 dB HL or 30 dBnHL.
2. If significant air-borne gaps are present or if other evidence of middle ear disease is seen, i.e., click-evoked ABR is normal, but OAE results are abnormal, refer the infant to an otolaryngologist for evaluation and treatment.
Repeat the diagnostic evaluation following medical treatment.
- 2a. If evidence of middle ear disease is present, refer the infant to an otolaryngologist for evaluation and treatment.
Repeat the diagnostic evaluation following medical treatment.
3. If results indicate a mixed hearing loss, refer the infant to an otolaryngologist for evaluation, treatment, and medical clearance if appropriate. Begin the process of fitting amplification if appropriate and repeat the diagnostic evaluation following medical treatment.
- 3a. If the ABR threshold is significantly elevated or the morphology is abnormal (i.e., prolonged interpeak latencies) and OAE results are normal, refer the infant to an otolaryngologist for evaluation of possible retrocochlear dysfunction (auditory neuropathy). Refer to local Child and Family Connections and initiate appropriate intervention for auditory neuropathy.
4. If a unilateral hearing loss is detected with normal sensitivity in the “good” ear, provide the family with information regarding the effects of unilateral hearing loss on auditory, speech, and language skills, as well as the importance of hearing conservation for the “good” ear. Continued audiologic monitoring of the child’s hearing and speech and language development is recommended every six (6) months until the child is three (3) years of age. Also, consider amplification options and refer the infant to an olaryngologist for evaluation, treatment, and medical clearance, if appropriate.
- 4a. Defer the fitting of hearing aids, but consider referral to the local CFC office for early intervention services and consider the use of assistive listening devices (e.g., an FM system). Repeat the diagnostic evaluation at three to six month intervals to monitor neuromaturation.