

## DIAGNOSTIC EVALUATION

Any audiologist accepting infants for initial audiological evaluations should have the equipment, training and skills to complete the procedures recommended by the CT EHDI Advisory Board (**see Appendix F, CT EHDI Advisory Board Best Practice Recommendations**). **It is important to note that both ears should be tested during the diagnostic evaluation, regardless of whether one ear passed in the initial screen.** The following recommendations outline the process for the diagnostic testing of infants, according to age.

### Audiological Evaluation from Birth to 6 Months of Age

For infants from birth to a developmental age of approximately 6 months, the test battery and audiological assessment should include:

- A child and family history.
- An evaluation of risk factors for congenital hearing loss.
- An evaluation of parental report of the infant's responses to sound.
- A frequency-specific assessment of the ABR using **air and bone**-conducted tone bursts, 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.
- Distortion product or transient evoked OAEs.
- Tympanometry using a probe tone greater than 1000-Hz.
- Clinician observation of the infant's auditory behavior as a crosscheck in conjunction with electrophysiologic measures.
- **Behavioral observation alone is not adequate** for determining whether hearing loss is present in this age group, and it is not adequate for the fitting of amplification devices.
- All NICU infants, or other infants at risk, should have a diagnostic evaluation conducted, not a rescreen.
- The initial diagnostic hearing evaluation with the pediatric audiologist should be scheduled within 2-4 weeks of parent/responsible party notification.
- The audiologist should have the ability to access ear mold impressions, fit, provide, dispense and repair hearing aids for the infants in a timely manner.
- Loaner hearing aids should be available, within a practical amount of time.
- If sedation is required, it should be administered in a medical facility where the child can be monitored safely.
- The audiologist will initiate the referral to the Connecticut Birth to Three System at the time of diagnosis by calling the Child Development Infoline at 1-800-505-7000.

- The diagnosing pediatric audiologist should work with the Birth to Three early intervention coordinator to provide the parent/responsible party with the information they need to make informed decisions regarding early intervention options. This transition period should include ongoing audiologic assessment as the families select intervention options.
- The audiologist should notify the infant's pediatric healthcare provider and the DPH EHDI Program of all testing results, including inconclusive results, failure to show for scheduled appointments and any subsequent referrals.
- The audiologist is responsible for faxing the initial diagnostic and any subsequent audiological evaluation results to the CT Early Hearing Detection & Intervention Program **within 2 days** following the appointment. Results should be faxed to the EHDI Program (860 509-8132) for **any child born after July 1, 2000 (see Appendix G, Diagnostic Testing Reporting Form)**.

### **Audiological Evaluation from 6 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.
- Parental report of auditory and visual behaviors and communication milestones.
- 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 and speech-detection and -recognition measures.
- OAE testing.
- Acoustic immittance measures (tympanometry and acoustic reflex thresholds).
- ABR testing if responses to behavioral audiometry are not reliable or if ABR testing has not been performed in the past.

### **MEDICAL EVALUATION**

Every infant with confirmed hearing loss and/or middle ear dysfunction should be referred for otologic and other medical evaluation. The purpose of these evaluations is to determine the etiology of hearing loss, to identify related physical conditions, and to provide recommendations for medical/surgical treatment as well as referral for other services. Essential components of the medical evaluation include:

- Clinical history,
- Family history of childhood-onset permanent hearing loss,
- Identification of syndromes associated with early or late-onset permanent hearing loss.
- A physical examination.
- Indicated radiologic and laboratory studies (including genetic testing).

Portions of the medical evaluation, such as urine culture for CMV, a leading cause of hearing loss, might even begin in the birth hospital, particularly for infants who spend time in the NICU.