ROLES AND RESPONSIBILITIES

The success of the Connecticut EHDI program depends on families working in partnership with healthcare professionals as a well-coordinated team. Each team member should clearly understand his or her role and responsibilities. Essential team members are the birth hospitals, families, medical home pediatricians or pediatric healthcare providers, audiologists, otolaryngologists, geneticists, speech-language pathologists, educators of children who are deaf or hard of hearing, and early intervention professionals.

The birth hospital is a key member of the EHDI team! The hospital staff has the primary responsibility of assuring that all infants are screened prior to discharge. Additionally, in collaboration with the state EHDI coordinator, the hospital staff should ensure that parents and pediatric healthcare professionals receive and understand the hearing-screening results, that parents are provided with appropriate follow-up and resource information, and that each infant is linked to a medical home. The hospital ensures that hearing-screening information is transmitted promptly to the medical home and appropriate data are submitted to the state EHDI coordinator.

POLICIES AND PROCEDURES

Each birth facility will develop written policies and procedures related to the newborn hearing screening program, including the necessary screener training, the screening process, data management, quality improvement and the testing of equipment. A copy of the policy and procedure manual shall be located in close proximity to the screening site, and be readily accessible to staff involved with newborn screening.
The policies and procedures must be reviewed at least annually, or as per facility policy, and should include, but not be limited to the following:

- Identify the title of the staff person(s) responsible for the training of personnel responsible for conducting the hearing screens.
- Include the contact information of the facility staff person who provides administrative oversight of the newborn hearing screening program.
- Document all job descriptions, qualifications, and roles and responsibilities for each newborn hearing screening position (e.g. audiologist, nurse, patient care assistant, rehabilitation aide, patient care technician, etc.), as well as orientation, minimum length of training, level of supervision and continuing education plans. Specific guidelines for periodic supervised performance appraisals should be included.
- Identify the name, model or type of hearing screening equipment used by the facility including the manufacturer's name, address and telephone number. Care, use, trouble-shooting, replacement of parts, maintenance and servicing of the screening equipment should be included.
- Identify the optimal testing environment as well as the desired condition or state of the newborn during testing.
- Identify the number of weeks of gestation at which infants will be screened.
- Include a mechanism to conduct a visual assessment of the newborn's skin for conditions that might necessitate the need for an alternate testing device.
- Identify ototoxic and other medications, which may interfere with testing. Include a plan to conduct the hearing screening after completion of the course of such medications.
- Identify safety measures and infection control practices.

- Identify risk factors associated with hearing loss that may necessitate the need for ongoing, periodic audiological evaluation and establish a mechanism to inform the parent of the risk. **(see Appendix B, Risk Indicators).**
- The policies must include a mechanism to identify the name, address and telephone number of the newborn's pediatric healthcare care provider who will follow the infant after discharge.
- Describe the method used to document and track all births, including the method, date, time and ear-specific results of all hearing screens conducted.
- Describe the method of communication to notify the infant's family and pediatric healthcare provider of all hearing screening results.
- Describe the screening method for the first screening. Otoacoustic emissions (OAE) or auditory brainstem response (ABR) are acceptable methods for the first screening for infants who are not at risk.
- Describe the screening method for the second screen. ABR should be the screening method for any NICU infant, any infant at risk, and or for any infant who did not pass the first screen and requires a second screen.
- Describe the mechanism to document all infants referred for further diagnostic testing, including the name, address and telephone number of the audiologist to whom the infant was referred.
- Describe the process to document a refused screening, including signing of the refusal waiver and DPH notification.
- Identify the title of the staff person responsible for notifying the parent/responsible party of a "refer" screening result (e.g. the primary care provider, audiologist, technician or nurse) and identify the method of such notification. Results should be relayed to the parent/responsible party face-to-face, in a private location, in a language that they understand and in a culturally sensitive manner with an appropriate level of concern.
- Each birthing facility should ensure that appropriate backup testing equipment be readily available in the event of equipment malfunction and that the equipment be readily accessible to the screening staff at all times.

PERSONNEL AND TRAINING

Staff training should include the purpose and scope of the birthing facility's newborn hearing screening program, as well as a review of all policies related to the newborn hearing screening program. The training should:

- Identify the roles, responsibilities, assigned tasks, and scope of practice and limitations of the duties of the screener.
- Be conducted by trainers who have had experience in newborn hearing screening and should be hands-on and competency based.

- Include a review of nursery policies including, but not limited to, infection control, safety, and patient confidentiality.
- Not be limited to manufacturer representative's demonstrations. It should include supervised, return demonstrations of the screening process to evaluate the effectiveness of the training program and the competency of each individual screener. The length of training may be individualized.
- Include instruction on safe baby handling techniques.
- Include training on how to recognize typical versus atypical neonatal behaviors during the hearing screening process.
- Be readily accessible to all staff involved with newborn screening.
- Include education on the use, care, maintenance, routine function checks, and troubleshooting of the testing equipment used in performing the assigned tasks.
- Include the method of notifying the primary care provider and parent/responsible party of screening results.
- Include the mechanism for transmission of the necessary data elements to the DPH.
- Include the process for referring an infant to a pediatric audiologist, when indicated.

RECOMMENDED SCREENING TECHNOLOGIES

Each birthing facility will be responsible for selecting and securing appropriate hearing screening equipment according to standards. Currently there are two physiologic measures used to objectively screen hearing acuity in newborns: Auditory Brainstem Response (ABR) and Otoacoustic Emissions (OAE).

Both OAE and ABR technologies provide noninvasive recordings of physiologic activity underlying normal auditory function, both are easily performed in neonates and infants, and both have been successfully used in universal screening of newborns. It is important to note that there are important differences between the 2 methods.

Although both ABR and OAE screening tests have a high sensitivity and specificity, both tests can miss some mild hearing losses or unusual configurations. It is important to remember that all infants with risk factors, including but not limited to genetic factors, asymptomatic cytomegalovirus (CMV), or a family history, are at risk for late onset hearing loss.

Both OAE and ABR screening technologies can be used to detect sensory (cochlear) hearing loss; however, both technologies may be affected by outer or middle-ear dysfunction. Consequently, transient conditions of the outer and middle ear may result in a hearing screening referral in the presence of normal cochlear and/or neural function.

Otoacoustic Emissions (OAE) measures reflect the status of the peripheral auditory system extending to the cochlear outer hair cells. A soft click is presented through a small microphone placed in the baby's ear canal, and measures the echo that is returned from the baby's ear. Trained hospital personnel such as audiologists, nurses, or technicians can perform this procedure if automated OAE equipment is used.

There are two types of automated OAE technologies: Transient Evoked Otoacoustic Emissions (TEOAE) and Distortion Product Otoacoustic Emissions (a DPOAE).

Both TEOAE and DPOAE may miss a small percentage of hearing losses. The refer rates at discharge for newborns screened with OAE average 7-8%.

Automated Auditory Brainstem Response (AABR) measurements are obtained from electrodes placed on the infant's body that record neural activity generated in the cochlea, auditory nerve, and brainstem in response to acoustic stimuli delivered via an earpiece. AABR measurements reflect the status of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway.

AABR interpretation is fully automated and elicits a PASS/REFER respons and therefore, does not require interpretation on the part of the screener. Consequently the AABR allows for a variety of trained hospital personnel to perform the screen such as nurses, technicians, support staff, or volunteers.

AABR may miss a small percentage of hearing losses, such as a high frequency loss greater than 4000Hz. The refer rates at discharge for newborns screened with ABR are typically less than 4 percent.

Each birthing facility should establish baseline Pass/Refer rates for all screening equipment used and should establish policies and procedures that include the care, use and maintenance of the equipment.

Calibration, service and maintenance of the testing equipment should be followed as directed by the manufacturer. Maintenance and service records should be documented and maintained as per facility policy.

RISK FACTORS FOR HEARING LOSS

As many as 54% of infants who passed newborn hearing screening and were later identified with hearing loss, had one or more risk factors. Although an infant may "Pass" the initial hearing screen, the birth facility should assess the infant for other risk factors, which may precipitate the need for periodic audiological monitoring.

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 Congenital Hearing Loss (PCHL)
- **Neonatal intensive care >5 days,** including any of the following:
  Extracorporeal membrane oxygenation (ECMO)*, Assisted ventilation, Exposure to ototoxic medications (Gentamycin and Tobramycin) in combination with loop diuretics (Furosemide/Lasix)
- **Hyperbilirubinemia requiring exchange transfusion**
- **In-utero infections** such as cytomegalovirus*, herpes, rubella, syphilis, and toxoplasmosis
- **Craniofacial anomalies** including those involving the pinna, ear canal, ear tags, ear pits, and temporal bone anomalie
- **Physical findings** such as white forelock, associated with a syndrome known to include a sensorineural or permanent conductive hearing loss
- **Syndromes** associated with hearing loss or progressive or late onset hearing loss* such as neurofibromatosis, osteopetrosis, and Usher's syndrome, Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson
- **Neurodegenerative disorders*** such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich's ataxia and Charcot-Marie-Tooth syndrome
- **Culture positive** postnatal infections associated with sensorineural hearing loss* Confirmed bacterial and viral (especially herpes viruses and varicella) meningitis
- **Head trauma,** especially basal skull/temporal bone fracture* requiring hospitalization
- **Chemotherapy***

* Risk indicators marked with an asterisk are of greater concern for delayed onset hearing loss.

Source: JCIH Year 2007 *Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs*

The CT EHDI Advisory Board has recommended that infants who Pass the hearing screening at birth, and have one or more risk factors as outlined by the JCIH, receive audiological monitoring every six months up until age three.

The Connecticut EHDI Program collects risk factor data from the birth facilities in the Newborn Screening System, on the Hearing panel. The birth facility has the responsibility of **assessing all newborns for risk factors**, and for reporting any recognized risk factors to the DPH through the Newborn Screening System.

The birth facility should notify the newborn's pediatric healthcare provider 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.

Any recommendations for risk factor monitoring and audiological follow-up should be documented on the discharge summary and be explained to the parent/responsible party prior to discharge.

TIMING OF THE HEARING SCREEN

Infants should be screened prior to discharge, leaving ample time to conduct a 2nd screening if the baby does not pass the 1st screen.

For premature infants, it is recommended that the infant be screened at **34 weeks corrected gestational age** or greater. If a newborn is receiving ototoxic and other medications that may interfere with testing, the hearing screen should be conducted after the completion of the course of the medications.

Birth facilities should take caution to **avoid over-screening** newborns! Although there may be factors that require the screen to be repeated, it is not recommended that babies be screened more than three times. The goal of the screening is NOT to get every baby to Pass, but rather to identify those that require further audiological evaluation. Over-screening increases the likelihood that you will get a false-negative result and that a child with a hearing loss may not be identified.

TESTING ENVIRONMENT

The facility should provide an area conducive to hearing testing that is free from excessive light ambient noise and/or other distractions that may impair the testing. It is recommended that signage be posted to indicate that a hearing screen is in progress. Newborns who have been discharged and return to the birthing facility for the initial or a repeat hearing screen, shall be screened in an area that is separate from the newborn nursery, in accordance with hospital infection control policies.

SCREENING PROTOCOLS IN THE WELL-BABY NURSERY

The parent/responsible party should be given the hearing screening brochure titled, *"Listen Up!"* prior to the screening and staff should be available to answer any questions that they may have about the procedure. The parent should be permitted to observe the screening, if they so request.

The equipment used for the initial screen in the well-baby nursery varies from hospital to hospital. Some facilities utilize OAE equipment and others only use ABR. Use of either technology in the well-baby nursery will detect peripheral (conductive and sensory) hearing loss of 40 dB or greater. When automated ABR is used as the single screening technology, neural auditory disorders can also be detected.

Any infant that does not pass the first hearing screen, regardless of the method, must have a repeat screen conducted before discharge using ABR equipment.

Under no circumstances should an infant who does not pass the ABR screen, be rescreened by OAE testing and "Passed." Until further audiological testing is conducted such infants are presumed to be at risk of having a subsequent diagnosis of auditory neuropathy/dyssynchrony.

SCREENING PROTOCOLS IN THE NICU

A neonatal intensive care unit (NICU) is defined as a unit in a facility in which a neonatologist provides primary care for the infant. Newborn units are divided into 3 categories:

- Level I: basic care, well-baby nurseries
- Level II: specialty care by a neonatologist for infants at moderate risk of serious complications
- Level III: a unit that provides both specialty and subspecialty care including the provision of life support (mechanical ventilation)

The DPH 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 diagnostic testing and, when indicated, a comprehensive evaluation, including diagnostic ABR testing, rather than a rescreen.

SCREENING PROTOCOLS FOR READMISSION

The JCIH Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs recommends that readmissions in the first month of life when there are conditions associated with potential hearing loss (eg, hyperbilirubinemia that requires exchange transfusion or culture-positive sepsis), an ABR screening should be performed before discharge. The results should be submitted to the DPH on the "back-up" paper reporting form.

INTERPRETING THE RESULTS

When The Screen Result Is A "Pass"

Infants who "*Pass"* the first hearing screen, or any subsequent screenings can be assumed to have adequate hearing function for speech/language development, at that point in time. It is important to note that a "*Pass"* result on the newborn screen does not guarantee normal hearing for the rest of the child's life. Infants and children can "*Pass"* the hearing screen at birth and develop a delayed onset or progressive hearing loss at a later time.

Therefore, primary care providers have the responsibility for surveillance of all infants to monitor for delayed onset or progressive hearing loss and certainly, parental concern at any age, should prompt a referral for an audiological evaluation.

- If the infant passes the hearing screen at birth and has one or more risk factors present, the child should be referred for follow-up audiological monitoring. The CT EHDI Advisory Board recommends that infants with one or more risk factors have an audiological evaluation **every six months**, up until age three.
- The infant's parent/responsible party will be notified of the hearing screen result and of the recommendation for follow-up audiological monitoring, both verbally and in writing, according to facility policy.
- The newborn's pediatric healthcare provider will be notified of the screening results and any identified risk factors associated with the potential for hearing impairment, which may warrant the need for audiological follow-up.
- Any recommendations for audiological follow-up should be documented on the discharge summary and be explained to the parent/responsible party, prior to discharge.

- The facility will document that the infant has been screened and will record the results on the nursery log, and in the infant's medical record as well as report the results electronically to DPH through the Newborn Screening System **(see Appendix X, Electronic Reporting Guidelines).**
- If the infant passed the hearing screen at birth and has NO risk factors, the child should receive ongoing surveillance of communicative development beginning at 2 months of age, during well-child visits in the medical home.

When The Baby "Does Not Pass" The Screen
Any infant who does not pass the first hearing screening in one or both ears, must have, at a minimum, a second hearing screening performed prior to discharge. The second, or repeat screening, should be conducted using ABR equipment.

- If the baby does not pass the 2nd ABR screen, the child should be referred to one of CT's Diagnostic Testing Centers for a rescreening, and, when indicated, comprehensive evaluation, including diagnostic ABR testing. For rescreening, a complete screening on both ears is recommended, even if only one ear failed the initial screening.
- The birth hospital should ensure that parents and primary healthcare professionals receive and understand the hearing screening results, that parents are provided with appropriate follow-up and resource information, and that each infant is linked to a medical home.
- The birth facility should ensure that hearing screening information is transmitted promptly to the medical home and appropriate data are submitted to the state EHDI coordinator.

Note: Infants in the well-baby nursery who do not pass an automated ABR screen should NOT be rescreened by OAE testing and "passed", because they are presumed to be at risk of having a subsequent diagnosis of auditory neuropathy/dyssynchrony.

CONVEYING SCREENING RESULTS
Screening results should be conveyed immediately to families so that they understand the outcome and the importance of follow-up when indicated and are allowed ample time to ask any questions. Communication with parents should be face-to-face, confidential and presented in a caring and sensitive manner. The person conveying the results should consider the following:

- The parent of an infant who does not pass the hearing screen should be given the brochure titled, *"A Parent's Guide to Diagnostic Hearing Testing of Infants".* The brochure is distributed to birth facilities by the DPH and is available in both English and Spanish.
- Information conveyed to families should be accurate, at an appropriate reading level and in a language they are able to understand.
- Parents should be told in a culturally sensitive and understandable manner that their infant did not pass the hearing screen and informed of the importance of prompt audiological follow-up testing.
- An appointment should be made for follow-up testing before discharge, if possible (**see Appendix C, Diagnostic Testing Locations).**