Early Hearing Detection & Intervention Program

Guidelines for Infant Hearing Screening

Revised September 2014

TABLE OF CONTENTS

Preface iii
Background 1
Overview of the CT Early Hearing Detection & Intervention Program 1
Program Development 2
Service Delivery Flow Chart 3
Roles and Responsibilities 4
Policies and Procedures 4
Personnel and Training 5
Recommended Screening Technologies 6
Otoacoustic Emissions 6
Automated Auditory Brainstem Response 6
Risk Factors for Hearing Loss 6
Timing of the Hearing Screen 7
Testing Environment 8
Screening Protocols in the Well Baby Nursery 8
Screening Protocols in the Neonatal Intensive Care Unit 8
Screening Protocols for Readmission 8
Interpreting the Results 9
When the Screening Result is a “Pass” 9
When the Baby “Does Not Pass” the Screening 9
When a Complete Screening Cannot be Obtained 10
Conveying Screening Results 10
Documentation of the Results 11
Data Submission to DPH 11
Missed Screening 11
Right of Refusal 12
Adoption or Foster Care 13
Non Resident Births 13
Out of State Births 13
Transfers Out of State 13
In-State Transfers 13
Referral to Audiologist 13
Diagnostic Evaluation 14
Audiological Evaluation from Birth to 6 Months 14

Audiological Evaluation from 6 to 36 Months 15
Medical Evaluation 15
Primary Health Care Professional 16
Otolaryngologist.....16
Medical Geneticist 18
Other Medical Specialists 18
Early Intervention.....18

Appendices Index 19
A. CT Hearing Screening Legislation
B. Risk Indicators
C. Diagnostic Testing Center Locations
D. Refusal Waiver
E. Hearing Screening Reporting Form
F. CT EHDI Task Force Best Practice Recommendations
G. Diagnostic Testing Reporting Form
H. Birth to Three Hearing Specialty Programs
I. Scripts for conveying screening results to parents

PREFACE

The Connecticut Department of Public Health Early Hearing Detection and Intervention program developed the following guidelines in an effort to support a comprehensive and effective statewide mechanism to screen all newborns for hearing acuity, to provide prompt audiological follow-up testing for those infants who do not pass the newborn screen, and to facilitate timely and appropriate early intervention services for those infants who are diagnosed with a hearing loss.

This document should serve as a guide for birth facilities and pediatric healthcare providers in the development and implementation of each respective Universal Newborn Hearing Screening Program. These guidelines are not intended to supersede individual hospital policy or the independent clinical assessment and judgment of physicians and medical providers in any individual case.

> *The goal of early hearing detection and intervention (EHDI) is to maximize linguistic competence and literacy development for children who are deaf or hard of hearing. Without appropriate opportunities to learn language, these children will fall behind their hearing peers in communication, cognition, reading, and social-emotional development. Such delays may result in lower educational and employment levels in adulthood. To maximize the outcome for infants who are deaf or hard of hearing, the hearing of all infants should be screened at no later than **1 month** of age. Those who do not pass screening should have a comprehensive audiological evaluation at no later than **3 months** of age. Infants with confirmed hearing loss should receive appropriate intervention at no later than **6 months** of age from healthcare and education professionals with expertise in hearing loss and deafness in infants and young children. Regardless of previous hearing-screening outcomes, all infants with or without risk factors should receive ongoing surveillance of communicative development beginning at 2 months of age during well-child visits in the medical home. EHDI systems should guarantee seamless transitions for infants and their families through this process.*
>
> *~Joint Committee on Infant Hearing 2007 Position Statement*

The Connecticut Department of Public Health hopes that you find this document useful. Questions or comments related to these guidelines should be directed to:

Connecticut Department of Public Health
Early Hearing Detection & Intervention (EHDI) Program
410 Capitol Avenue, MS #11 MAT
Hartford, CT 06134-0308
Telephone: 860-509-8074
http://www.ct.gov/dph/ehdi

BACKGROUND

The Joint Committee on Infant Hearing (JCIH) was established in late 1969 and was composed of representatives from audiology, otolaryngology, pediatrics, and nursing. Throughout its over 30-year history, the Committee explored the complexities of hearing loss and its effect on a child's development, seeking to find newer and better methods to identify and serve the infants and their families. Today, the Joint Committee is comprised of representatives from the American Academy of Pediatrics, the American Academy of Otolaryngology and Head and Neck Surgery, the American Speech Language Hearing Association, the American Academy of Audiology, the Council on Education of the Deaf, and Directors of Speech and Hearing Programs in State Health and Welfare Agencies.

The Committee's primary activity has been publication of position statements summarizing the state of the science and art in infant hearing, and recommending the preferred practice in early identification and appropriate intervention of newborns and infants at risk for or with hearing loss.

In 1994, the JCIH endorsed universal detection of hearing loss in newborns and infants and stated that all infants with hearing loss be identified before 3 months of age and receive intervention by 6 months. With the support of the Health Resources Service Administration, Maternal and Child Health Bureau and Centers for Disease Control and Prevention, and other dedicated groups and individuals, universal screening gained momentum across the United States and all states now have newborn hearing screening programs in place.

The current JCIH Position Statement was released in 2007 after years of meetings, research, revision and hard work by many dedicated professionals and has been widely accepted as the “Gold Standard” of Universal Newborn Hearing Screening programs today.

The Connecticut Department of Public Health’s Early Hearing Detection and Intervention (EHDI) Program and the Connecticut EHDI Task Force fully embrace early hearing detection and intervention for infants with hearing loss through integrated, interdisciplinary community, state, and federal systems of universal newborn hearing screening, evaluation, and family-centered intervention.

This document is designed to serve as a guide to assist birth facilities and other healthcare providers in the provision of services for newborns from screening, through diagnostic testing and enrollment into early intervention and is the second revised version of the original *Connecticut Universal Hearing Screening Guidelines* that were distributed to Connecticut birth hospitals in 2000 and revised in 2008. The recommendations from the JCIH 2007 Position Statement were incorporated throughout this document.

OVERVIEW OF THE CT EARLY HEARING DETECTION AND INTERVENTION PROGRAM

Connecticut General Statutes, Section 19a-59 requires each institution that provides childbirth services, as defined in section 19a-490, to include a universal newborn hearing screening program as part of its standard of care and to establish a mechanism for compliance review. The provisions of this subsection do not apply to any infant whose parent/responsible party objects to hearing screening as being in conflict with their religious tenets and practice *(see Appendix A, CT Legislation).*

The Connecticut EHDI Program set benchmarks for birth facility newborn hearing screening programs, and monitors the programs and provides technical assistance when programs fall below the desired benchmarks. Birth facilities should strive to achieve the following:

- Screen 98% of all newborns prior to hospital discharge.
- Document the screening date, results and method on the nursery log and in the infant’s medical record.
- The percentage of infants who do not pass the hearing screening at birth and are referred for audiological follow-up should not exceed 4%.

- Hospitals should offer an outpatient rescreening if an infant is missed in one or both ears, prior to discharge.
- Physicians should be notified of all screening results, including those of infants who were missed or whose parents refused the screening.
- Newborn hearing screening should be offered to infants born at home or those born out-of-state and transferred to a CT birth facility.
- Physicians and hospitals are responsible for making the referral to a diagnostic audiologist following a referral from the screening at birth.
- It is important to obtain an appropriate diagnostic evaluation prior to 3 months of age in order to decrease the need for sedation of the infant, decrease parental anxiety, and identify the hearing loss within the recommended timeframe.
- Following confirmation of hearing loss the audiologist should notify the Connecticut Early Hearing Detection and Intervention program of the diagnostic results. The Connecticut Early Hearing Detection and Intervention program will provide support and care coordination services to assure the family receives early intervention services as early as possible or by 6 months of age.

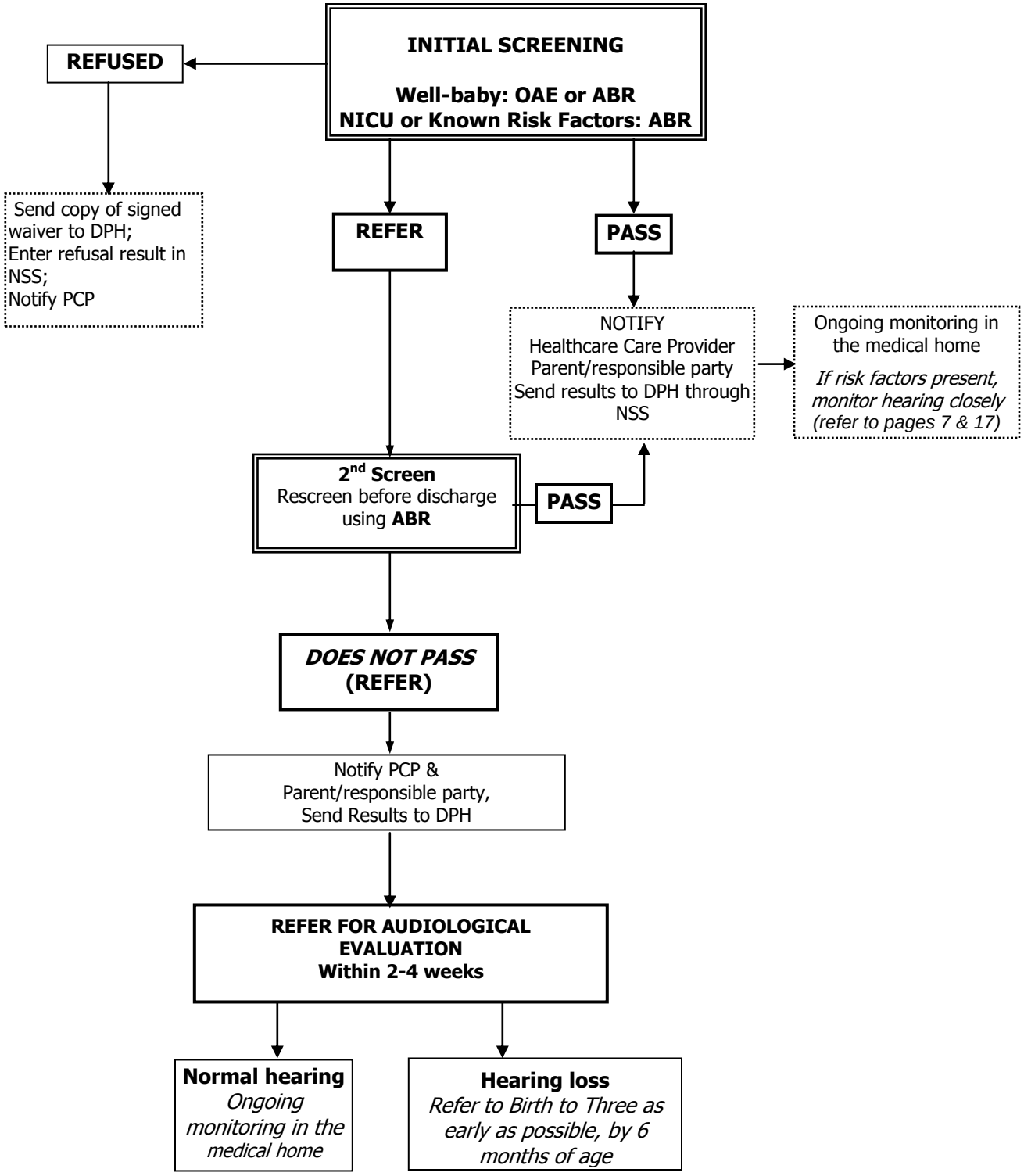
PROGRAM DEVELOPMENT

Since the Early Hearing Detection & Intervention program's inception in 2000, the CT Department of Public Health has assisted birth facilities in the development and implementation of Universal Hearing Screening Programs (UNHS) by drafting and disseminating program guidelines, through education and conferences, and by providing technical assistance, consultation, and support as needed.

Over the years, the DPH has assisted birth facilities to upgrade and/or standardize their screening equipment, identified infant diagnostic testing centers throughout the state, developed a web-based reporting system for the hospital's submission of screening data, conducted conferences and trainings for hospital staff and audiologists, developed a web-based training for pediatric healthcare providers titled, *"Newborn Screening in Connecticut"*, standardized equipment in the diagnostic testing centers, developed a multitude of printed educational materials for families and healthcare providers, and supported other provider and parent education and outreach initiatives.

Statewide hearing screening of all newborns began on July 1, 2000. Connecticut strives to meet the national goals of screening by 1 month, diagnosis by 3 months, and enrollment in early intervention by 6 months of age.

Early Hearing Detection & Intervention Program
Service Delivery Flow Chart



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 assurance and improvement mechanisms, 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 screenings.
- 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 <u>after completion of the course</u> 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 <u>must</u> include a mechanism to identify the name, address and telephone number of the newborn's pediatric healthcare care provider who will follow the infant <u>after discharge</u>.

- Describe the method used to document and track all births, including the method, date, time, and ear-specific results of all hearing screenings 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 method for the second screening. 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 screening and requires a second screening.
- 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 is 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 <u>all</u> 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: Automated Auditory Brainstem Response (AABR/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 two 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 percent.

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 response 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 screening 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 screening, 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* outlined those conditions that put infants at greater risk for late onset 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 anomalies
- **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 Task Force 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 Maven: Newborn Screening System, in the Hearing Screening question package. 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 Maven: Newborn Screening System (NSS).

The birth facility should notify the newborn's pediatric healthcare provider of any identified risk factors associated with the potential for late onset 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 or responsible party prior to discharge.

TIMING OF THE HEARING SCREENING

Infants should be screened prior to discharge, leaving ample time to conduct a second screening if the baby does not pass the first screening.

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 screening 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 screening 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 hearing screening is in progress. Newborns who have been discharged and return to the birthing facility for the initial or a repeat hearing screening, 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 "*Listen Up!"* brochure is distributed to birth facilities by the DPH and is available in both English and Spanish.

The equipment used for the initial screening 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 screening, regardless of the method, must have a repeat screening conducted <u>before</u> discharge using ABR equipment.

**Under no circumstances should an infant who does not pass the ABR screening, be rescreened with OAE equipment and passed. Until further audiological testing is conducted such infants are presumed to be at risk of having a subsequent diagnosis of auditory neuropathy/dys-synchrony.

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, a referral should be made directly to an audiologist for diagnostic testing and, when indicated, a comprehensive evaluation, including diagnostic ABR testing, rather than a rescreening.

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 (e.g. 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 (*Appendix E*).

INTERPRETING THE RESULTS

When the Screening Result is a "Pass"

Infants who "p*ass"* the first hearing screening, 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 "p*ass"* result on the newborn hearing screening does not guarantee normal hearing for the rest of the child's life. Infants and children can "p*ass"* the hearing screening 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 screening at birth and has one or more risk factors present, the child should be referred for follow-up audiological monitoring. The CT EHDI Task Force 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 screening 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 Maven: Newborn Screening System **(reference separate document Maven: NSS user manual).**
- If the infant passed the hearing screening 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 Screening

Any infant who does not pass the first hearing screening in one or both ears, must have, at a minimum, a second hearing screening (rescreening) performed prior to discharge. The second, or repeat screening, should be conducted using ABR equipment.

- For rescreening, a complete screening on both ears is recommended, even if only one ear failed the initial screening. If the child does not pass the rescreening, it is recommended that an appointment for a diagnostic audiological evaluation be made prior to discharge, if possible.
- If the baby does not pass the second ABR screening, the child should be referred to one of Connecticut's Infant Diagnostic Testing Centers (*Appendix C*) for a rescreening, and, when indicated, comprehensive evaluation, including diagnostic ABR testing.
- The birth hospital should ensure that parents and primary healthcare providers 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 need for audiological follow-up should be documented on the discharge summary.
- The facility shall document that the infant has been screened and shall record the results on the nursery log and in the infant's medical record as well as report the results to DPH through the Maven: Newborn Screening System. The *Audiology Referral* should be reported in the appropriate data field, by selecting "Yes," as well as documenting the appointment date and audiology center, if known.

Note: Infants in the well baby nursery who do not pass an automated ABR screening 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/dys-synchrony.

When a Complete Screening Cannot be Obtained

If the hearing screening cannot be completed for a medical reason, including but not limited to structural abnormalities of the ear (i.e. congenital atresia), report the results electronically to DPH through the Maven: Newborn Screening System as "Refer" (which indicates an automatic need for audiological follow up testing) and document the explanation in the Comments section. Follow the same guidelines in the section above, entitled: "When the Baby 'Does Not Pass' the Screening,"

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:

- Sometimes babies don't pass the initial inpatient screen and need to be re-screened prior to discharge. If you're informing parents that their baby needs to be re-screened, you'll want to help them understand that it's common and that often the screening needs to be repeated once or twice before discharge.
- The parent of an infant who does not pass the final hearing screening 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. Remember to use simple and clear language when talking with parents and don't say more than needed because it may confuse the family. Reassure parents that this is a screening that tells us if further tests are needed. We can't know if a baby has hearing loss from the screening results.
- Parents should be told in a culturally sensitive and understandable manner that their infant did not pass the hearing screening 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)*.
- When advising a parent when a baby doesn't pass the screening - some common terms are "refer," "did not pass," and "fail." While they all mean essentially the same thing, your hospital has certain protocols on how to inform the parents. Check with your program manager to see which term they prefer to use. It is recommended that the birthing facility NOT use the word "fail" when an infant does not pass the hearing screening. *(See Appendix I, Scripts for conveying screening results to parents)*
- The birthing facility will notify the child's primary healthcare provider of the hearing screening results (pass, did not pass, refused, or missed), as documented in the hospital medical chart.
- The screening result should be entered into the web-based Maven: Newborn Screening System (NSS) reporting system.
- The need for audiological follow-up should be documented on the discharge summary. If the facility attaches the hearing screening sticker on the discharge summary, they should supplement it with language that explains that a "Refer" result indicates the need for follow-up audiological testing.
- All birthing facilities should be prepared to link the parent/responsible party of those infants who do not pass the screening to providers for community-based support and education, as needed.
- 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.

DOCUMENTATION OF THE RESULTS

In addition to submitting the newborn's hearing screening results to the DPH, the facility should document the screening results in the infant's medical record, according to facility policy. It is recommended that the documentation include:

- Screener's name
- Screening date(s)
- Type of equipment used
- Ear specific results
- Contact information for the audiology facility the infant was referred to
- Audiology appointment date and time, if known.

Referral information should also be documented in the discharge planning instructions. It is strongly suggested that the infant's Accession Number and hearing screening results be documented in the nursery log book, along with other pertinent birth information, so that it is readily accessible and can provide a quick view of infants who may be in need of a hearing screening before discharge. The Maven: Newborn Screening System also includes electronic workflow lists that identify newborn records with incomplete hearing screening information.

DATA SUBMISSION TO DPH

An electronic record should be created in the Maven: Newborn Screening System for each live birth. The Admin and Demographics data should be completed as soon as possible after birth, and prior to submission of the Newborn Screening laboratory filter paper form to the DPH. The Hearing Screening question package should be completed prior to infant discharge.

The screening result should be entered into the web-based Maven: Newborn Screening System (NSS) reporting system. When more than one screening is performed, provide the first and final screening results under Screening 1 and Screening 2. Remember, although there may be factors that require the screening to be repeated, it is not recommended that babies be screened more than three times. If a third screening takes place, overwrite the original second screening results with the final ear specific screening results. For more detailed reporting requirements, refer to the **Maven: NSS user manual**, a separate document provided to all birth facilities in November 2011 and available through the DPH EHDI program.

MISSED SCREENING

Connecticut birth facilities are required by State law to conduct a hearing screening on all newborns, as a "standard of care" *(see Appendix A, Hearing Screening Legislation)*. Staff should understand that when a newborn is discharged without a hearing screening there is an estimated 1 in 1,000 risk that the baby will have an undetected hearing loss.

- The birth facility should determine who will assume the initial responsibility for recall and screening a child who was discharged without the hearing screening.
- It is the responsibility of the primary care physician to know the hospital policies and procedures for recalling and rescreening infants who miss a screening.
- Screening should be performed by 1 month of age, as later ages require more time due to the infant's increased alertness and may even require sedation.
- The birth facility should notify the infant's pediatric healthcare provider that the baby was discharged without a hearing screening and inform the provider of any arrangements to recall the baby.
- The contact with the parent/responsible party and primary care provider should be documented in the infant's medical record.

- If the infant is recalled to the birth facility for the initial hearing screen, the screening should be conducted in an area that is separate from the newborn nursery, in accordance with hospital infection control policies.
- The hearing screening results should be submitted to the DPH through the Maven: NSS when conducted.
- If the infant does not return to the birth hospital for the hearing screening, the facility should report the result as "Missed" in the Maven: Newborn Screening System and provide an explanation in the notes section as a way of documenting it for DPH tracking purposes.

RIGHT OF REFUSAL

It is the responsibility of the birth facility to assure that the hearing screening is conducted. Parents should understand that if a newborn is discharged without a hearing screening there is an estimated 1 in 1,000 risk of the baby having an undetected hearing loss. Parents who refuse the hearing screening for their child should be counseled on the importance of early identification and encouraged to allow the hearing screening to be performed.

In accordance with the Connecticut General Statutes, Section 19a-59, a newborn's parent/responsible party has the right to oppose having the hearing screening conducted if it is in conflict with their religious tenets and practice. If a parent/responsible party refuses to allow the hearing screening to be conducted prior to discharge, they must sign the DPH Hearing Screening Refusal Waiver *(see Appendix D, Refusal Waiver)*. The Hearing Screening Refusal Waiver can be accessed online at www.ct.gov/dph/ehdi or on the home page of the Maven: Newborn Screening System:

- Log-in to the Maven: NSS.
- On the home page, select the following link: Click here for the Program Refusal Waivers and print out the document for signature.
- Access the infant's record.
- Click on the Hearing Screening question package.
- Enter the date refused as the screening date, enter the name of the hospital staff person providing the information, select "None" for the screening method, and then select "Refused" for right and left ear result.

The birth facility staff should complete the refusal waiver and have the infant's parent/responsible party sign and date it.

The original, signed waiver must be maintained as a permanent part of the infant's medical record. A copy of the signed waiver must be faxed to the state EHDI Program at **860 509-8132**.

The facility must notify the infant's pediatric healthcare provider of the refusal. The refusal and any parent counseling by staff should be documented in the infant's medical record.

ADOPTION OR FOSTER CARE

Newborns that will be placed for adoption will have the hearing screening and any repeat screenings conducted prior to discharge. If a referral for diagnostic follow-up is indicated, the follow-up information will be given to the representative of the adoption agency at the time the newborn is discharged, as well as to the birthing facility social worker and/or discharge planner, as per facility policy.

- The name of the infant's primary care provider who will care for the infant after discharge should be obtained and recorded in the Newborn Screening System, if known.
- If the name of the physician that will follow the infant after discharge is unknown, the name, address and telephone number of the Adoption Agency will be recorded in place of the physician's information. Provide this information by searching for and selecting "Other" from the provider list in the Maven: NSS, and then typing the agency information into the blank contact information fields.

NON RESIDENT BIRTHS

State legislation mandates that all birth hospitals implement a Newborn Hearing Screening Program as a "standard of care." This requires any infant born in a Connecticut birthing facility to have the hearing screening conducted, regardless of the state of residence and the child's screening information should be reported to DPH through the Maven: Newborn Screening System.

OUT OF STATE BIRTHS

Infants that are born out of state, who are transferred to a Connecticut hospital should have a hearing screening conducted before discharge. The results can be sent to the DPH on the back-up, paper reporting form and DPH will notify the Hearing Screening Coordinator in the infant's state of birth that the hearing screening was conducted *(see Appendix E, Hearing Screening Reporting Form).*

TRANSFERS OUT OF STATE

When a Connecticut-born infant is transferred to an out-of-state hospital, more than likely the hearing screening will not have been conducted prior to transfer due to the infant's medical condition. Since all states bordering CT have hearing screening programs, the hospital of transfer *should* conduct the hearing screening on the infant prior to discharge.

- The birth facility should document any screenings that need to be conducted on the inter-agency transfer report that is sent to the hospital of transfer.
- The newborn's electronic record cannot be accessed by an out of state hospital.
- However, the birth facility can inform the DPH that the infant was transferred out of state by selecting "Out of State/other" from the list of Transfer Locations in the Admin and Demographics question package and typing the name of the out-of-state hospital in the "Specify" field.
- DPH will contact the out of state hospital or EHDI program to obtain the hearing screening results when completed.
- Alternatively, if the appropriate authorization from the child's parent/responsible party has been obtained, the facility of birth can contact the out of state hospital of transfer and confirm that the newborn hearing screening was conducted and then enter the screening results into the Maven: NSS.

Remember, the NSS electronic record cannot be accessed by an out of state hospital. *Document all information related to the CT Newborn Screenings that need to be performed on the Inter-Agency Transfer Report and on the infant's Discharge Summary.*

IN-STATE TRANSFERS

If a newborn is transferred to a Connecticut hospital and was not screened prior to discharge because of an acute medical condition or other circumstances, it is the responsibility of the hospital of birth to assure that the hearing screening is conducted.

The facility of birth should transfer the infant's NSS record electronically to the hospital of transfer by selecting that facility from the list of Transfer Locations in the Admin and Demographics question package as well as documenting any screenings that are needed on the discharge summary and Inter-Agency Transfer Report.

REFERRAL TO AN AUDIOLOGIST

The key to a positive outcome of any newborn hearing screening program is to connect the family and infant to the appropriate services. Any infant who does not pass the final hearing screening should be referred to one of Connecticut's Infant Diagnostic Testing Centers for audiological testing (*see Appendix C, Diagnostic Testing Locations*). Infants who are in need of diagnostic testing should be referred to audiology centers that have the personnel, equipment, training, and skills to complete the procedures recommended by the CT EHDI Task Force *(see Appendix F, CT EHDI Task Force Best Practice Recommendations).* The timing and number of hearing re-evaluations for children with risk factors should

be individual and customized depending on the relative likelihood of a subsequent delayed onset hearing loss.

- The birth facility should explain to the parent/responsible party the importance of taking the child for follow-up audiological testing.
- The birthing facility staff should provide the parent/responsible party with the referral information necessary for diagnostic evaluation.
- The DPH strongly recommends that the parent/responsible party be given the appointment for follow-up with a pediatric audiologist, prior to discharge. Quality improvement studies demonstrate this practice significantly improves the likelihood a family will take their baby for follow-up testing.
- The follow-up appointment information should be explained to the parent/responsible party prior to discharge, documented in the infant's medical record, and is included on the discharge instructions (see **Conveying Results** section).
- The infant's pediatric healthcare provider who will follow the infant after discharge must be notified of all screening results including pass, refer, or missed screenings. If the primary care provider will be referring the family to the pediatric audiologist directly, the birthing facility should attempt to secure the name of the audiologist and appointment information, prior to discharge. This information should be explained to the parent/responsible party, documented in the infant's medical record, included in the discharge instructions, and entered into the DPH data system for tracking.
- Referrals to a pediatric audiologist may be scheduled and conducted prior to discharge. If an audiologist evaluates an infant prior to discharge, there shall be a written consultation for the services and such consultation will be properly entered into the infant's medical record. The audiologist will be responsible for submitting the diagnostic results to the DPH on the appropriate form *(see Appendix G, Diagnostic Audiology Reporting Form)*.

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 Task Force *(see Appendix F, CT EHDI Task Force Best Practice Recommendations).* **It is important to note that both ears should be tested during the diagnostic evaluation, regardless of whether one ear passed the initial screening.** 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 <u>and</u> 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 rescreening.
- 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 within the last three years and for any child under the age of 5 who is diagnosed with hearing loss** *(see Appendix G, Diagnostic Audiology 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.

PRIMARY HEALTH CARE PROFESSIONAL

The infant's pediatrician or other primary health care provider (PCP) is responsible for monitoring the general health, development, and well-being of the infant. The PCP should verify that the newborn's hearing screening was conducted during the child's first office visit. If the hearing screening was not done prior to discharge, the PCP should refer the family back to the birth facility or to a diagnostic center where the hearing screening can be conducted. The following outlines additional responsibilities of the infant/child's PCP:

- Must assume responsibility to ensure that the diagnostic testing/audiological assessment is conducted in a timely fashion on all infants who do not pass the hearing screening.
- Initiate referrals for any medical specialty evaluations necessary to determine the etiology of the hearing loss.
- Monitor the child's middle-ear status because the presence of middle-ear effusion can further compromise hearing.
- Partner with other specialists, including the audiologist, otolaryngologist, geneticist, and ophthalmologist, to facilitate coordinated care for the infant and family.
- Because 30% to 40% of children with confirmed hearing loss will demonstrate developmental delays or other disabilities, the primary care physician should closely monitor developmental milestones and initiate referrals related to suspected disabilities.
- Review every infant's medical and family history for the presence of risk indicators that require monitoring for delayed-onset or progressive hearing loss and ensure that an audiological evaluation is completed for children at risk of hearing loss, at least once by 24 to 30 months of age according to the Joint Committee on Infant Hearing, regardless of their newborn screening results. The CT EHDI Task Force recommends that infants with one or more risk factors have an audiological evaluation every six months, up until age three. This includes infants who passed the hearing screening at birth.
- Infants with specific risk factors, marked with an asterisk on page 7 – such as CMV infection or a family history, are at increased risk of delayed onset or progressive hearing loss and should be monitored closely.
- Conduct ongoing surveillance of parent concerns about language and hearing, auditory skills, and developmental milestones of <u>all infants and children</u> regardless of risk status, as outlined in the pediatric periodicity schedule published by the AAP.
- Children with cochlear implants may be at increased risk of acquiring bacterial meningitis compared with children in the general U.S. population. The PCP must assure that all children with, and all potential recipients of, cochlear implants receive the CDC recommended pneumococcal immunization that apply to cochlear implant users and that they receive age-appropriate *Haemophilus Influenzae* Type B vaccines.

OTOLARYNGOLOGIST

Otolaryngologists are medical doctors who diagnose, treat, and manage a wide range of diseases of the head and neck and specialize in treating hearing and vestibular disorders. They perform a full medical diagnostic evaluation of the head and neck, ears, and related structures, including a comprehensive

history and physical examination, leading to a medical diagnosis and appropriate medical and surgical management. Often, a hearing or balance disorder is an indicator of, or related to, a medically treatable condition or an underlying systemic disease. The otolaryngologists work closely with other healthcare professionals, including other physicians, audiologists, speech-language pathologists, educators, and others, in caring for patients with hearing, balance, voice, speech, developmental, and related disorders. It is important for a family to select an otolaryngologist that specializes in working with infants and children.

- The otolaryngologist's evaluation includes a comprehensive history to identify the presence of risk factors for early-onset childhood permanent hearing loss, such as family history of hearing loss, having been admitted to the NICU for more than 5 days, and/or having received ECMO.
- A complete head and neck examination for craniofacial anomalies should document defects of the auricles, patency of the external ear canals, and status of the eardrum and middle-ear structures. Atypical findings on eye examination, including irises of 2 different colors or abnormal positioning of the eyes, may signal a syndrome that includes hearing loss. Congenital permanent conductive hearing loss may be associated with craniofacial anomalies that are seen in disorders such as Crouzon disease, Klippel-Feil syndrome, and Goldenhar syndrome. The assessment of infants with these congenital anomalies should be coordinated with a clinical geneticist.
- In large population studies, at least 50% of congenital hearing loss has been designated as hereditary and nearly 600 syndromes and 125 genes associated with hearing loss have already been identified. The evaluation, therefore, should include a review of family history of specific genetic disorders or syndromes, including genetic testing for gene mutations such as *GJB2* (connexin-26), and syndromes commonly associated with early-onset childhood sensorineural hearing loss. As the widespread use of newly developed conjugate vaccines decreases the prevalence of infectious etiologies such as measles, mumps, rubella, *Haemophilus Influenzae* Type B, and childhood meningitis, the percentage of each successive cohort of early-onset hearing loss attributable to genetic etiologies can be expected to increase, prompting recommendations for early genetic evaluations.
- Approximately 30% to 40% of children with hearing loss have associated disabilities, which can be of importance in patient management. The decision to obtain genetic testing depends on informed family choice, in conjunction with standard confidentiality guidelines.
- In the absence of a genetic or established medical cause, a computed tomography scan of the temporal bones may be performed to identify cochlear abnormalities, such as Mondini deformity with an enlarged vestibular aqueduct, which have been associated with progressive hearing loss.
- Temporal bone imaging studies may also be used to assess potential candidacy for surgical intervention, including reconstruction, bone-anchored hearing aid, and cochlear implantation. According to the JCIH, recent data have shown that some children with electrophysiologic evidence suggesting auditory neuropathy/dyssynchrony may have an absent or abnormal cochlear nerve that may be detected with MRI.

Historically, an extensive battery of laboratory and radiographic studies was routinely recommended for newborn infants and children with newly diagnosed sensorineural hearing loss. However, emerging technologies for the diagnosis of genetic and infectious disorders have simplified the search for a definitive diagnosis, which obviates the need for costly diagnostic evaluations in some instances. If, after an initial evaluation, the etiology remains uncertain, an expanded multidisciplinary evaluation protocol including electrocardiography, urinalysis, testing for CMV, and further radiographic studies is indicated. The etiology of neonatal hearing loss may remain uncertain in as many as 30% to 40% of children.

- Once hearing loss is confirmed, medical clearance for hearing aids and enrollment into Birth to Three should be initiated as soon as possible. Amplification and early intervention <u>should not be delayed pending the outcome of the diagnostic process</u>.
- The otolaryngologist should conduct careful longitudinal monitoring to detect and promptly treat any coexisting middle-ear effusions.

MEDICAL GENETICIST

A referral to the Medical Geneticist can provide families with information on the etiology of the hearing loss, prognosis for progression, associated disorders (e.g., renal, vision, cardiac), and the likelihood of recurrence in future offspring. This information may influence the parents' decision-making regarding intervention options for their child. All families of children with confirmed hearing loss should be offered a genetics evaluation and counseling. The medical geneticist is responsible for the following:

- Interpreting family history data.
- Clinically evaluating and diagnosing inherited disorders, if present.
- Performing and assessing genetic tests.
- Providing genetic counseling to the family.

OTHER MEDICAL SPECIALISTS

Every infant with a confirmed hearing loss should have an evaluation by an ophthalmologist to document visual acuity and rule out concomitant or late-onset vision disorders such as Usher syndrome. Indicated referrals to other medical subspecialists, including developmental pediatricians, neurologists, cardiologists, and nephrologists, should be facilitated and coordinated by the primary health care professional.

EARLY INTERVENTION

Before newborn hearing screening was instituted universally, children with severe-to-profound hearing loss, on average, completed the 12th grade with a 3rd- to 4th-grade reading level and language levels of a 9- to 10-year-old hearing child. In contrast, infants and children with mild-to-profound hearing loss who are identified in the first 6 months of life and provided with immediate and appropriate early intervention (EI) services have significantly better outcomes than later-identified infants and children in vocabulary development, receptive and expressive language, syntax, speech production, and social-emotional development.

According to federal guidelines and Connecticut State law (C.G.S. 19a-59), once <u>any degree</u> of hearing loss is confirmed in a child, a referral should be initiated to the CT Birth to Three System, Connecticut's early intervention program, within two days of confirmation of hearing loss. Referrals are made to Birth to Three by calling the Child Development Infoline at 1-800-505-7000. More information about the CT Birth to Three System can be obtained by visiting their website at: http://www.birth23.org.

- The family should be referred to Birth to Three by the diagnosing audiologist at the time of diagnosis and EI services should be initiated as soon as possible, and no later than 6 months of age.
- Connecticut has three Birth to Three programs that specialize in working with infants and children who are deaf or hard of hearing: American School for the Deaf, CREC/Soundbridge, and New England Center for Hearing Rehabilitation (NECHEAR). (*See Appendix H, Birth to Three Hearing Specialty Programs).* Upon referral to Birth to Three the family will receive information about each of the three programs. Although the audiologist and PCP should be available to answer any questions that the family may have about the three programs, the choice as to which program is selected should be made by the family.
- Children with other medical conditions, in which hearing loss is not the primary disability, should have access to intervention with a provider who is knowledgeable about hearing loss.

APPENDICES INDEX

A. Connecticut Newborn Hearing Screening Legislation

B. Risk Indicators for Hearing Loss

C. Infant Diagnostic Testing Center Locations

D. Refusal Waiver

E. Newborn Hearing Screening Reporting Form

F. CT EHDI Task Force Best Practice Recommendations

G. Diagnostic Audiology Reporting Form

H. Birth to Three Hearing Specialty Programs

I. Scripts for conveying screening results to parents

Appendix A
Connecticut Hearing Screening Regulations

Connecticut General Statutes, Section 19a-59: Program to identify newborn infants at high risk for hearing impairments.

a) Each institution, as defined in section 19a-490, that provides childbirth service shall, no later than July 1, 2000, include a universal newborn hearing screening program as part of its standard of care and shall establish a mechanism for compliance review. The provisions of this subsection shall not apply to any infant whose responsible party objects to hearing screening as being in conflict with their religious tenets and practice.

b) The Department of Public Health shall establish a plan to implement and operate a program of early identification of infant hearing impairment. The purpose of such plan shall be to: (1) Identify infants at high risk of having hearing impairments; (2) notify responsible party of such infants of the risk; (3) inform responsible party of resources available to them for further testing and treatment, including rehabilitation services for such infants, and (4) inform responsible party of financial assistance available through the Department of Public Health, including, but not limited to, parental eligibility criteria, which may result in reduced cost or no cost to responsible party for testing, evaluation or treatment, including rehabilitation of such infants. The department shall develop such plan in consultation with persons including, but not limited to, pediatricians, otolaryngologists, audiologists, educators and responsible party of deaf and hearing impaired children.

c) The Commissioner of Public Health shall adopt regulations, in accordance with Chapter 54, to implement the provisions of subsection (a) of this section.

Newborn Hearing Screening Program

19a-59-1. Newborn hearing screening program

(a) Not later than July 1, 2000, each institution as defined in section 19a-490 of the Connecticut General Statutes that provides childbirth services shall develop and implement a universal newborn hearing screening program which shall at a minimum include the following:

(1) A physiologic technologies testing mechanism which employs automated or diagnostic auditory brainstem response (ABR) or otoacustic emissions (OAE), or subsequently developed or improved physiologic technologies that substantially enhance newborn hearing assessment that are recognized by the American Academy of Audiology or American Speech Language Association; and

(2) a mechanism for monitoring the institution's compliance with the newborn hearing screening program which shall include, but not necessarily be limited to, the following information:

(A) name of each newborn infant;

(B) date of birth;

(C) date infant received hearing screening or documentation of parent refusal for newborn hearing screening;

(D) method of screening;

(E) results of screening;

(F) person performing screening; and

(G) to whom referral for further evaluation was made, if applicable.

(b) A parent who refuses to allow his or her infant to be screened for a hearing impairment based upon religious tenets and practice shall sign a statement attesting to said refusal which shall include a statement by a licensed health care provider that the parent was informed of the medical consequences of such refusal. The document shall identify the specific reasons for the refusal and shall be placed in the infant's medical record. If a parent declines to sign the refusal statement, the institution shall document in the infant's medical record the reason for the refusal by the parent to permit newborn hearing screening and a statement that the parent refused to sign the document and was informed of the medical consequences of such refusal.

(a) Documentation of the screening shall be maintained in the infant's medical record and shall be retained for a minimum of twenty-five (25) years after discharge of the infant except that original medical records may be destroyed sooner if they are microfilmed by a process approved by the department.

(b) The institution shall develop and implement policies and procedures for the requirements of this section which shall be reviewed and approved by the institution's medical staff and governing body.

(c) The department may review implementation of Section 19a-59-1 of the Regulations of Connecticut State Agencies at the time of licensure inspections.

(Effective March 4, 1999; Amended effective April 10, 2000.)

Appendix B
Risk Factors for Hearing Loss

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) or 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 anomalies

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*

***** Are of greater concern for delayed onset hearing loss

CONNECTICUT EARLY HEARING DETECTION AND INTERVENTION (EHDI) INFANT DIAGNOSTIC TESTING LOCATIONS

The following is a list of audiologists who conduct the test battery as recommended by the Connecticut Early Hearing Detection and Intervention Task Force, for the diagnostic hearing testing of infants who do not pass the hearing screening conducted at birth.

FARMINGTON
Connecticut Children's Medical Center
Audiology Department
505 Farmington Ave.
1st Floor
Farmington, CT 06032
(860) 545-9642
www.connecticutchildrens.org
Medical facility affiliation for sedation: Connecticut Children's Medical Center - Hartford

HARTFORD
Connecticut Children's Medical Center
282 Washington Street
Hartford, CT 06106
(860) 545-9642
www.connecticutchildrens.org
Medical facility affiliation for sedation: Connecticut Children's Medical Center - Hartford

STORRS
University of Connecticut
Speech & Hearing Clinic
850 Bolton Road
Unit 1085
Storrs, CT 06269-1085
(860) 486-2629
http://slhs.uconn.edu/
Note: Does not perform sedation.

GLASTONBURY
Connecticut Children's Medical Center
Glastonbury Satellite Office
310 Western Blvd.
Glastonbury, CT 06033
(860) 545-9642
www.connecticutchildrens.org
Medical facility affiliation for sedation: Connecticut Children's Medical Center - Hartford

NEW HAVEN
ENT Medical & Surgical Group
46 Prince Street
New Haven, CT 06519
(203) 752-1726
www.entmedicalsurgical.com
Note: Does not perform sedation.

WATERFORD
Lawrence & Memorial at Waterford
Outpatient Rehabilitation Services
40 Boston Post Road
Waterford, CT 06385
(860) 271-4900
www.lmhospital.org
Note: Does not perform sedation.

HAMDEN
Hearing, Balance & Speech Center
2661 Dixwell Avenue
Hamden, CT 06518
(203) 287-9915
www.hearingbalance.com
Note: Does not perform sedation.

NEW HAVEN
Yale New Haven Hospital
Yale Hearing & Balance Center 800
Howard Ave, 4th Floor
New Haven, CT 06519
(203) 785-2467
http://hearing.yale.edu
Medical facility affiliation for sedation: Yale New Haven Hospital

For more information, contact the
Connecticut Department of Public Health
Early Hearing Detection and Intervention (EHDI) Program at:
(860) 509-8074
or visit our web site:
http://www.ct.gov/dph/ehdi

Rev. 9/2014

Appendix D
Hearing Screening Refusal Waiver

Connecticut Department of Public Health

Family Health Section
Early Hearing Detection & Intervention Program
410 Capitol Avenue, MS #11 MAT
Hartford, Connecticut 06134-0308

Newborn Hearing Screening Refusal Waiver

As defined in Section 19a-59 of the Connecticut General Statutes, I, _____ (the responsible party), of _____(infant's name), a baby born on _____(date of birth), in _____ (birthing facility/hospital), refuse permission for the Newborn Hearing Screening test to be performed on my baby, because such a test is in conflict with my religious tenets and practice. The risks and benefits of the Newborn Hearing Screening have been fully explained to me and I understand and accept responsibility for choosing not to have the screening performed.

Accession Number: _____

Parent/Responsible Party Name (Please print):_____

Relationship (if other than parent):_____

Street Address:_____

Town/State/Zip Code:_____

Infant's Primary Care Physician:_____

Physician's Address:_____

Physician's Telephone:_____

Parent/Guardian Signature: _____

Witness:_____

Date:_____

To be filed with the Hospital/Birthing Facility Medical Record of this infant
Send a copy of this signed waiver to:
Connecticut Department of Public Health
Early Hearing Detection & Intervention Program
410 Capitol Avenue, MS #11 MAT
Hartford, Connecticut 06134-0308

PHONE: 860-509-8074 FAX: 860 509-8132

Rev. 7/2014

CONNECTICUT
DEPARTMENT OF PUBLIC HEALTH

NEWBORN HEARING SCREENING REPORTING FORM

Birth Place

Mother's Last Name

First

Accession # / / / / / / /

Baby's Medical Record Number

Baby's Last Name

First

Address

Telephone ( ) -

Date of Birth / /

Weight (grams)

EGA (weeks)

Birth Sequence

Sex: Male Female

Race

Hospital Transferred to

HEARING SCREENING

Date / / Method Right Left

Date / / Method Right Left

Primary Care Provider Name

PCP Address

Telephone ( ) -

Please return this form to:

Connecticut Department of Public Health
Early Hearing Detection and Intervention Program
410 Capitol Avenue, MS# 11 MAT, P.O. Box 340308
Hartford, CT 06134-0308

or Fax to: (860) 509-8132

Contact the CT EHDI Program at (860) 509-8074 with any questions.

INFANT DIAGNOSTIC HEARING TESTING
CONNECTICUT EARLY HEARING DETECTION AND INTERVENTION TASK FORCE

BEST PRACTICE RECOMMENDATIONS

Early infant hearing detection and intervention are keys to the development of speech and language. Universal newborn hearing screening has been mandated in Connecticut since July 2000. Approximately 1-2% of all infants screened will be referred for diagnostic hearing testing. The initial diagnostic hearing evaluation should be scheduled promptly upon discharge and be completed by the time the child is three months of age.

The pediatric audiologist to whom the child is referred must decide on the **battery of tests** that is appropriate for each child based on: screening results, the medical history of the child, risk factors if present, and the type, number, and timing of screenings carried out prior to the referral.

The Connecticut Early Hearing Detection and Intervention Task Force strongly recommends that any audiologist accepting infants for initial diagnostic audiological evaluations have the ability to complete the following procedures (*):

Auditory Brainstem Response [ABR (a.k.a BAERS, BAER)]:

- Threshold measurement with frequency specific tone bursts
- Threshold measurement with bone conduction ABR
- Sedation in a medical facility where the child can be appropriately monitored

Immittance Testing:

- Tympanometry with high frequency probe tone **greater than 1000 Hz**
- Acoustic reflex testing

Otoacoustic Emissions (OAE):

- Transient evoked or distortion product

Behavioral Audiometry:

- May be useful in addition to the above

(*) An otological evaluation must be included as part of the diagnostic process, although it may occur at a different facility and time.

The audiologist must report the results of the initial diagnostic audiological evaluation to the referring physician and to the Connecticut Department of Public Health, in the requested format. For reporting instructions, please call (860) 509-8057.

The audiologist must refer any child with a hearing loss to the CT Birth to Three System at the time of diagnosis. The Birth to Three System provides a range of services including ongoing audiological testing, acquisition of assistive technology, family training and communication intervention. For referrals, more information, or to receive the *Service Guidelines for Children Who are Hard of Hearing or Deaf*, contact CT Birth to Three at 1-800-505-7000 or visit their website: http://www.birth23.org/.

Compiled by the Connecticut Early Hearing Detection and Intervention Task Force. Endorsed by: Connecticut Department of Public Health, Connecticut Birth to Three System, Connecticut Speech, Language and Hearing Association, and the Commission on the Deaf and Hearing-Impaired.

Connecticut Department of Public Health
Early Hearing Detection and Intervention
DIAGNOSTIC AUDIOLOGY REPORTING FORM

Child's Last Name **First Name** **DOB** **Birth Hospital** **Accession Number**

Parent / Responsible Party Name **Parent/ Responsible Party Address** **Parent/ Responsible Party Telephone** ( )

Pediatrician Name **Address** **Telephone**

Date of Evaluation: _____ Did not Keep Appointment: _____

Purpose of Appointment: **INITIAL SCREEN** ☐ **RESCREEN** ☐ **Screening Method: OAE / ABR** **Results RIGHT: PASS / REFER** **Results LEFT: PASS / REFER**

DIAGNOSTIC TESTING ☐ **FOLLOW UP TESTING** ☐ **RISK FACTOR MONITORING** ☐

DIAGNOSTIC ABR RESULTS:

	Right Ear	Left Ear
Hearing within Normal Limits (-10 to 15 dB HL)	☐	☐
TYPE OF HEARING LOSS:		
Sensorineural Hearing Loss	☐	☐
Conductive Hearing Loss	☐	☐
Mixed Hearing Loss	☐	☐
Undetermined Type Hearing Loss	☐	☐
DEGREE OF HEARING LOSS: (Degree of AC Thresholds at 500, 1000, 2000 Hz)		
Slight (16 to 25 dB HL)	☐	☐
Mild (26 to 40 dB HL)	☐	☐
Moderate (41 to 55 dB HL)	☐	☐
Moderately severe (56 to 70 dB HL)	☐	☐
Severe (71-90 dB HL)	☐	☐
Profound (91+ dB HL)	☐	☐

Other Tests Conducted (Please Specify):_____

Was this a Progressive or Late Onset Hearing Loss? **Yes** ☐ **No** ☐ **Unknown** ☐

Hearing Aid Candidate? Yes ☐ **No** ☐ **Not Determined** ☐ **Date of Amplification:**_____

Cochlear Implant Candidate? Yes ☐ **No** ☐ **Not Determined** ☐ **Date of Implant if Known (mm/yy):** _____

Referred for Genetic Testing? Yes ☐ **No** ☐ **Unknown** ☐

Referred to Birth to Three? **Yes** ☐ Date:_____

(Referral Line: 800-505-7000) **No** ☐ Reason:_____

RISK FACTORS: (Check all that Apply)

- ☐ **None Known**
- ☐ **Culture Positive Postnatal Infections**
- ☐ **Hyperbilirubinemia**
- ☐ **NICU Care >5 days**
- ☐ **Syndromes – Specify:**_____
- ☐ **Caregiver Concern**
- ☐ **Family History**
- ☐ **In-utero Infections**
- ☐ **Ototoxic medications**
- ☐ **Craniofacial Anomalies**
- ☐ **Head Trauma**
- ☐ **Neurogenerative Disorder**
- ☐ **Physical Findings**

AUDIOLOGIST'S RECOMMENDATIONS:

ENT REFERRAL? Yes ☐ **No** ☐ Referred to: _____ Telephone:_____

_____

_____

TESTING CONDUCTED BY: _____

NAME OF CENTER: _____ **TELEPHONE:**_____

FAX RESULTS TO: Connecticut Dept. of Public Health, Early Hearing Detection & Intervention Program: (860) 509-8132

Appendix H
Birth to Three Hearing Specialty Programs
http://www.birth23.org/programs/hsp/

Capitol Region Education Council (CREC)/Soundbridge
123 Progress Drive
Wethersfield, CT 06109
Telephone: (860) 529-4260
http://www.crec.org/

New England Center for Hearing and Rehabilitation (NECHEAR)
354 Hartford Turnpike
Hampton, CT 06247-1320
Telephone: (860) 455-1404
http://www.nechear.com

American School for the Deaf
139 North Main Street
West Hartford, CT 06107
Telephone: (860) 570-2300 (Voice)
(860) 570-2222 (TTY)
http://www.asd-1817.org/

Appendix I
Scripts for conveying screening results to parents

Scripts developed by the National Center for Hearing Assessment and Management (NCHAM) and provided via the:

Interactive Web Based Newborn Hearing Screening Training Curriculum
http://www.infanthearing.org/nhstc/

NCHAM
National Center for Hearing Assessment and Management
Utah State University™

Communicating with Parents and Medical Providers

Passing Script for Babies

"Congratulations on the birth of your baby. We just completed the hearing screen; the results are a pass. Here is a brochure that talks about development of speech and language. It is always important to monitor the progress of your baby's development, especially their speech and language because your baby's hearing can change any time. If you are ever worried that your baby can't hear, talk to your baby's doctor right away and ask for a referral to an audiologist that is skilled at testing infants and young children."

"Felicitaciones por el nacimiento de su bebé. Acabamos de finalizar el tamizaje auditiva de su bebé y él/ella la pasó. Este es un folleto que trata sobre el desarrollo del habla y del lenguaje. Es importante observar el desarrollo de su bebé especialmente de su habla y lenguaje ya que la audición de su bebé puede cambiar en cualquier momento. Si usted está preocupado de que su bebé no pueda oír, hable con el médico pediatra inmediatamente y pídale que lo envíe a donde un audiólogo especializado en hacer pruebas a bebés y niños pequeños."

NCHAM
National Center for Hearing Assessment and Management
Utah State University™

Communicating with Parents and Medical Providers

Passing Script for Babies at High Risk for Hearing Loss

"Congratulations on the birth of your baby. We just finished screening your baby's hearing and your baby passed the screen today. However, because your baby's had some medical problems at birth, there is a chance that your baby can develop hearing loss after you leave the hospital. Your baby's hearing is critical in order for "on time" development to occur. Your doctor can help you to monitor your babies hearing development and tell you when your baby should have further tests with an audiologist that's skilled at testing infants and young children."

"Felicitaciones por el nacimiento de su bebé. Acabamos de terminar de hacerle el tamizaje auditivo a su bebé el cual pasó. Sin embargo, debido a que su bebé tuvo algunos problemas médicos al nacer, existe la posibilidad pueda desarrollar una pérdida auditiva después de salir del hospital. La audición de su bebé es muy importante para que ocurra un desarrollo normal. Un médico le puede ayudar a hacer un seguimiento del desarrollo auditivo de su bebé y le puede indicar cuando un audiólogo, con experiencia con infantes y niños pequeños, necesite realizarle más pruebas."

NCHAM
National Center for Hearing Assessment and Management
Utah State University™

Communicating with Parents and Medical Providers

Not Passing Script for Babies

"Congratulations on the birth of your baby. We just finished screening your baby's hearing. Your baby did not pass the screen today. This does not necessarily mean that your baby has a permanent hearing loss, but without additional testing we can't be sure. The screening results will be provided to your baby's doctor. Please be sure you make or keep (depending on your hospital's protocol) the appointment for further hearing testing. "

Felicitaciones por el nacimiento de su bebé. Los resultados del tamizaje auditivo que le hicimos hoy a su bebé indican que él/ella no lo pasó. Esto no necesariamente significa que su bebé tenga una pérdida auditiva permanente, pero sin hacer pruebas adicionales no podemos estar seguros. Los resultados del tamizaje le serán enviados al médico de su bebé. Asegúrese de hacer una cita para hacer más exámenes auditivos o acudir a esta (dependiendo del protocolo de su hospital)."

Clarification for Year 2007 JCIH Position Statement

2007 JCIH Position Statement Update

Separate hearing screening protocols are recommended for NICU and well-infant nurseries. High risk NICU infants (admitted for more than 5 days) are to have automated auditory brainstem response (ABR) included as part of their hearing screening so that neural hearing loss will not be missed.

Rationale for different protocols:

The JCIH 2007 recommendation is to identify infants with congenital permanent sensory, conductive and neural (auditory neuropathy/auditory dyssynchrony) hearing loss. ABR technology is needed to identify neural hearing loss. Consequently, the JCIH recommends ABR screening to identify neural hearing loss in the infants most at risk of a neural loss. NICU infants represent 10% of the newborn population or approximately 400,000 infants per year. Infants cared for in the NICU are at increased risk of neural hearing loss. Data from the National Perinatal Research Center (NPIC) (Quality Analytic Services)(QAS) was utilized to make the recommendation of ABR for the subgroup of NICU infants that require NICU care for >5 days. NPIC data indicated that approximately 25% of NICU infants are considered "low" risk (includes infants with diagnoses such as transient respiratory distress, observation for temperature instability, and negative sepsis workup) and are discharged by 5 days of age. High risk NICU or Level 2 infants hospitalized for > 5 days for medical reasons (not social) were then identified as the target population for ABR to rule out neural hearing loss. Since specific risk factors are often difficult for screeners to identify in the medical record, establishing a time criterion (>5 days) was felt to be easier to implement. Implementation of this recommendation for all NICU and well baby infants was not felt to be indicated based on current evidence. Therefore, a NICU baby who meets this >5 day criterion cannot be screened and passed by OAE alone.

Risk Factor Clarifications:

1. **Recommendations for audiology follow-up of infants with risk factors**. The prior recommendation for follow-up audiology assessments every 6 months for all screen fails was felt to place a great burden on audiologists and could not be accomplished in most parts of the country. In addition, there are infants with "unknown risk factors" who develop late onset hearing loss. Therefore, the responsibility for surveillance of all infants was shifted to the primary care provider, who will refer to audiologists, as needed, any patient for whom there are concerns or findings consistent with hearing loss. The document lists accepted risk factors for hearing loss, identifies risk factors which are known to be associated with late onset or progressive hearing loss, and makes recommendations for standard or more frequent follow-up by the audiologist. Therefore, for infants with a risk factor which may be considered low risk, at least one audiology

assessment by 24-30 months is the recommendation. This management would change if there is a referral from the primary care provider because of a new concern regarding hearing. In contrast, for an infant with risk factors known to be associated with late onset or progressive hearing loss, such as, CMV or family history, early and more frequent assessment is appropriate. Early and more frequent can be interpreted as every 6 months, or more, depending on the clinical findings and concerns. See following excerpt from document.

> The timing and number of hearing re-evaluations for children with risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss. Infants who pass the neonatal screening but have a risk factor should have at least 1 diagnostic audiology assessment by 24 to 30 months of age. Early and more frequent assessment may be indicated for children with cytomegalovirus (CMV) infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, or culture-positive postnatal infections associated with sensorineural hearing loss; for children who have received ECMO or chemotherapy; and when there is caregiver concern or a family history of hearing loss.

Appendix 1 in JCIH 2007 lists the risk factors and identifies the risk factors associated with late onset or progressive loss with an asterisk.

2. **Recommendations regarding ototoxic medications.**

The following are listed as risk factors in Appendix 1 of the JCIH 2007 Statement:

> Neonatal intensive care of >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 requiring exchange transfusion.

To be consistent with the intent of simplifying the referral process to NICU >5 days and for clarification the recommendation has been reworded.

All infants with or without risk factors requiring neonatal intensive care for greater than 5 days, including any of the following: ECMO,* assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/lasix). In addition, regardless of length of stay: hyperbilirubinemia requiring exchange transfusion.

JCIH
February 13, 2008