Have You Heard?

Policies and Procedures Manual

Georgia Universal Newborn Hearing Screening and Intervention Program

Georgia's Universal Newborn Hearing Screening and Intervention (UNHSI) program policy and procedural manual was developed in an effort to provide guidance as to the implementation of newborn hearing screening and follow up in Georgia. This guide is intended to be used as a resource to Georgia UNHSI stakeholders so that all babies and parents in Georgia receive the best care and information from initial hospital screening to any follow up and intervention services necessary.

Georgia Department of Public Health

Georgia Department of Public Health,
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Newborn Hearing Screening History and Background

Newborn hearing screening is standard practice throughout the United States and territories and has been successfully implemented in other countries according to the World Health Organization (2009). The first newborn hearing screening programs in the United States in the 1990's, were limited to screening babies on the high risk registrar, which excluded and missed a lot of infants with congenital hearing loss. The evolution and spread of newborn hearing screening in the U.S. began in 1993, when a panel from the National Institutes of Health (NIH) reviewed evidence on early identification of hearing loss and made recommendations (NIH Consensus Statement, 1993). The NIH panel concluded that the best practice to ensure an early identification of hearing loss required a hearing screening of all newborns prior to discharge (NIH Consensus Statement, 1993 and White, Forsman, Eichwald, & Munoz, 2010).

As a result of NIH endorsement and several concurrent events, the percent of newborns screened for hearing loss in the U.S. rose from under 3 percent in 1993, to 94 percent in 2006 (White et al., 2010). In 2010, ninety-eight percent of babies born in Georgia were screened prior to hospital discharge. Newborn hearing screening is an everyday practice throughout the United States and territories and has been successful. Newborn hearing screening is the first step to determine if hearing loss is a concern. An audiological diagnostic test is required to determine if a permanent hearing loss exists.

For babies who have a "refer" result on the newborn hearing screening and are tracked to an audiological diagnostic evaluation, figures reported in 1989 by the National Institute on Deafness and Other Communication Disorders revealed that 1 in 1,000 infants is born totally deaf, while an additional one to six per 1,000 are born with hearing loss of varying degrees (Kushalnagar et al., 2010). More recent CDC data from 2009 indicated a slightly lower figure of 1.3 in 1,000 newborns is born with a hearing loss in the United States. Hearing loss is one of the most common birth disorders in the United States (Kushalnagar et al., 2010).

Research driven benchmarks were developed for states to meet in reference to initial screening, rescreening, diagnostic testing, and enrollment into early intervention. Current recommendations from the Joint Committee on Infant Hearing (JCIH) (2007) include screening/rescreening by one month of age, comprehensive diagnostic audiological evaluation by three months, and enrollment in early intervention by six months for infants who refer on an initial screen and subsequently diagnosed with permanent hearing loss. These recommendations are supported by the CDC and have been included in the United State Department of Health and Human Services Healthy People 2020 goals. The Healthy People 2020 goal related to this investigation is directed at the completion of audiologic evaluation no later than age three months for infants who did not pass the hearing screening. This specific goal is located within the newborn hearing screening portion of the hearing and other sensory or communication disorders goal. The Healthy People 2020 goal recognizes that "through early diagnosis and intervention, these children (identified with permanent hearing loss) can develop speech and language skills on schedule with their peers".

Georgia's Department of Public Health Vision and Mission

Vision

A Healthy and Safe Georgia

Mission

The mission of the Georgia Department of Public Health is to prevent disease, injury, and disability; promote health and well being; and prepare for and respond to disasters.

Georgia UNHSI Program Goals

To sustain a comprehensive coordinated system for UNHSI in Georgia in which stakeholders work together to ensure that by 2020:

- 99% or more of newborns are documented to have received a screening for hearing loss prior to hospital discharge in Georgia;
- 95% or more of infants who "referred" or "missed" the initial hospital hearing screening are documented to have received a rescreening by 1 month of age (30 days);
- 90% or more of infants who "referred" their rescreening are documented to have received a diagnostic audiological evaluation by 3 months of age (90 days);
- 90% or more of infants who have a diagnosed permanent hearing loss identified through newborn hearing screening are documented to be enrolled in early intervention by 6 months of age;
- All newborns are linked with medical homes and their families and caregivers receive culturally competent support throughout the screening, diagnostic, and intervention processes;
- A data management system is maintained to document hearing screen and diagnostic audiological evaluation results, risk factors for late onset hearing loss, referrals to and enrollment date of intervention services, and follow-up actions related to facilitating the process of screening through intervention;
- Essential information about UNHSI is shared with stakeholders.

History of Newborn Hearing Screening in Georgia

As a result of the passage of the Official Code of Georgia Annotated (O.C.G.A.) 31-1-3.2 in 1999, the Georgia Division of Public Health (DPH) developed and implemented a statewide Universal Newborn Hearing Screening and Intervention (UNHSI) initiative. Georgia law mandates education on the importance of newborn hearing screening, reporting of aggregate hearing screening data on a quarterly basis by birthing facilities, and establishing the State Advisory Committee on Newborn Hearing Screening (SACNHS). DPH developed a "Recommended Guidelines for the UNHSI program" document, hospital reporting system, and implemented the UNHSI Program statewide in 200l. SACNHS dissolved in 2005, but the committee's recommendation remains that all hospitals perform newborn hearing screening on at least 95% of all newborns. Georgia continues to strive to maintain newborn hearing screening rates above 95% with a referral rate of 4% or less. With 130,000 to 140,000 births annually, Georgia's UNHSI Program follows on average 4,500 children who are referred into the system.

In 2007, The Georgia Commission on Hearing Impaired and Deaf Persons was created by House Bill 655. The Commission consists of seven members; five members appointed by the Governor, one member appointed by the Senate, and one member appointed by the Speaker of the House of Representatives. At minimum, two members are hearing impaired and the remaining members of the commission are selected from among parents of children with hearing loss, persons who are involved with hearing impaired persons or programs, and representatives of private providers of services to hearing impaired persons. The Commission assists the UNHSI Program by being an advocate for children with hearing loss. They assist people who are deaf and hard of hearing as well as the parents of children with hearing loss to ensure equal access to services, programs, and opportunities. The Commission also coordinates its efforts with other state and local agencies that are serving children with hearing loss.

In addition to Georgia's Commission on Hearing Impaired and Deaf Persons, the Georgia UNHSI Program has assembled a Stakeholders Committee to replace the dissolved SACNHS committee. The first meeting of the Committee was held in April 2010, with meetings scheduled quarterly. Representatives include individuals from DPH, pediatric audiology and otolaryngology specialists, educators of the deaf and hard of hearing, medical home representatives, parents of children identified with hearing impairment, and community partners. The purpose of the Stakeholders Committee is to advise and assist with UNHSI program enhancements and serve as a resource.

Mandatory Reporting in Georgia

In July 2002, the Board of the Department of Human Resources approved a request to add childhood hearing impairment to the state's Notifiable Disease List. Birth defects are reportable under State Law, Official Code of Georgia Annotated (O.C.G.A.) 31-12-2 and 31-1-3.2, which mandate the reporting of notifiable diseases.

The following conditions related to hearing loss are required to be reported to Public Health:

1. Newborns not passing the initial or follow-up hearing screening (suspected hearing impairment): Newborns that "refer" a newborn hearing screening are to be reported to the Children 1st (C1st) Coordinator in the health district where the child resides, using the Children 1st Screening and Referral Form (Appendix A) immediately following screening or at least within 7 calendar days. All information on the form that is known to

the screener/evaluator/audiologist should be filled out and submitted. The Children 1st Screening and Referral form is completed to best of provider knowledge to identify any possible risk factors for progressive or late onset hearing loss or for other disabilities that may qualify the child for Children1st, Babies Can't Wait (BCW), and/or Children's Medical Services (CMS).

2. Children through the age of five (5) years with the initial confirmation/diagnosis of hearing loss/impairment, which is suspected to be permanent, measured and described by a licensed audiologist is required to be reported within 7 days of diagnosis. The "Surveillance of Hearing Impairment in Infants and Young Children" (Appendix B) form is used by the audiologist and/or physician with knowledge of the hearing impairment and should be sent to the health district where the child resides.
   Hearing loss/impairment is defined as a threshold average of 15 dB or greater between 500Hz - 4000Hz, whether unilateral or bilateral.

The following scenarios are recommended to be reported to Public Health. Reporting the following scenarios is crucial to reduce Georgia's UNHSI lost to follow-up documentation rate.

1. Newborns transferred out or discharged from a birthing facility without screening, which includes parent refusal. Reporting should be completed using a C1st Screening and Referral form and submitted to the health district where the child resides within 7 calendar days of discharge.
2. Newborns transferred to a birthing facility that did not receive a hearing screening and receive a hearing screening with a "pass" result. Reporting should be completed using a C1st Screening and Referral form and submitted to the health district where the child resides within 7 calendar days of screening.
3. Infants, who "refer" on an initial screening from a birthing facility, but "pass" a secondary outpatient screening. Reporting should be completed using a C1st Screening and Referral form and submitted to the health district where the child resides within 7 calendar days of re-screen.
4. Infants who "refer" the newborn hearing screening and receive a diagnostic audiological evaluation and are found to have normal hearing or suspected transient conductive hearing loss. Reporting should be completed using a "Surveillance of Hearing Impairment in Infants and Young Children form" and submitted to the health district where the child resides within 7 calendar days of evaluation.

Georgia law requires this information to be submitted for these infants and children. Information submitted to the Department of Public Health is protected and confidential by law. The reporting of this information is not in violation of consumer privacy and protection regulations, including the Health Insurance Portability and Accountability Act (HIPAA).

Data Management and Tracking

In October 2009, the Georgia UNHSI program implemented the statewide use of the State Electronic Notifiable Disease Surveillance System (SendSS) Newborn module, a web-based child health information system. SendSS Newborn is a population-based surveillance and tracking system that identifies and monitors children throughout Georgia with or at risk for developmental disabilities. SendSS Newborn allows for consistent data entry across the state and also allows for increased programmatic monitoring and quality assurance activities by ensuring

easy access to data and all follow-up actions performed by the UNHSI District Coordinators. Prior to SendSS, each health district developed their own tracking system. The shift from 18 separate public health district databases to one statewide system has eliminated the loss of information for transient families; as families move from one public health district to another, information is passed from one UNHSI District Coordinator to another thus allowing for continued coordination and tracking.

Role of UNHSI District Coordinator

The Department has provided funding to each of the 18 public health districts for a UNHSI District Coordinator as of July 2009. The purpose of the UNHSI District Coordinator is to provide a primary contact to support the UNHSI program at the local level of the Health District. UNHSI District Coordinators meet weekly with district representatives from the other birth to five programs to ensure that all children referred to Public Health are referred to eligible programs. The role of the UNHSI District Coordinator is to:

- Act as a liaison between the hospital(s) within their health district with reference to the UNHSI program
- Work with State UNHSI Program Staff to address hospital performance falling below set benchmarks on screening and referral rates
- Receive referrals on newborns who have "referred" initial or follow up screenings, whose parents refused screenings, or who have been discharged from the hospital without having a newborn hearing screening; and enter information into the SendSS database, and track all Children 1st Screening and Referral forms
- Coordinate with the PCP/Medical Home to facilitate referral of newborns that "refer" screening for rescreening and for diagnostic audiological evaluation if necessary, and linkage to appropriate intervention for those babies diagnosed with hearing loss
- Receive and track all Surveillance of Hearing Impairment forms sent to Public Health and enter the information into the SendSS database
- Receive and track all out of state referrals for children who "refer" on an initial screening or rescreening or those identified with hearing loss; and enter the information into the SendSS database
- Document in SendSS the follow-up activities, as indicated by the Risk Factor Protocol, for newborns (0-6 months) identified with risk factors for hearing loss through the electronic birth certificate or by Children 1st Screening and Referral Form
- Perform tasks as outlined in Georgia's Loaner Hearing Aid Bank protocol
- Identify areas to provide education and awareness of the UNHSI program in their local Public Health District and community

Role of the Birthing Hospital or Facility

Every birthing hospital or facility shall designate an employee to be their Newborn Hearing Screening Program coordinator. It is recommended that this employee be an audiologist. If the program coordinator is NOT an audiologist, then each birthing hospital or birth center should have access to an audiologist for consultation. Each Newborn Hearing Screening Program Coordinator shall act as a liaison between their facility and the UNHSI District Coordinator for their health district.

Equipment

There are two technologies available for automated physiologic screening of hearing in newborns. Automated physiologic hearing screening equipment:

- does not require interpretation by the screener
- is noninvasive, and
- has a history of success in newborn hearing screening programs.

Birthing hospitals and facilities must conduct newborn hearing screening using at least one of the following physiological hearing screening methods. Both technologies are appropriate for screening newborns however there are differences between the two methods.

- Automated Auditory Brainstem Response (aABR) – Objective measurement obtained from surface electrodes that record neural activity generated in the cochlea, auditory nerve, and brainstem in response to low intensity click stimuli delivered through earphones. aABR measurements reflect the status of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway (ASHA, 2007). The screening level may not exceed 35dB HL.
  Note: It is recommended that babies in the neonatal intensive care unit (NICU) be screened with aABR technology
  Limitation: Infant must remain quiet, it is best if baby is asleep
- Automated Otoacoustic Emissions (aOAE) – Objective measurement obtained from the ear canal using a sensitive microphone within a probe assembly that records cochlear responses to low intensity acoustic stimuli. OAEs are a physiologic test that reflects the status of the peripheral auditory system specifically measuring cochlear (outer hair cell) function (ASHA, 2007). There are two types of OAE technologies: Transient Evoked Otoacoustic Emissions (TEOAE) click stimuli and Distortion Product Otoacoustic Emissions (DPOAE) tone pairs.
  Note: aOAEs may miss a disorder called Auditory Neuropathy Spectrum Disorder (ANSD)
  Limitations: Infant must be relatively inactive during the test and aOAEs are very sensitive to middle-ear effusions, and cerumen or vernix in the ear canal.

Despite both technologies being appropriate in detecting cochlear hearing loss, some infants who pass newborn hearing screening will later demonstrate permanent hearing loss. Although this loss may reflect delayed-onset hearing loss, both aABR and aOAE screening technologies will miss some hearing loss (e.g., mild or isolated frequency region losses) (Cone-Wesson et al., 2000: Johnson et al., 2005).

- Hearing screening equipment shall be calibrated in accordance with manufacturer's recommendation and should be monitored with monthly equipment checks to ensure equipment is functioning properly
- Calibration certificates kept on record (annually)
- Log maintained to show monthly equipment checks and any equipment issues with dates and explanations
- Birthing facility should have alternate plans for newborn hearing screening in the event of equipment malfunction
- Equipment should be cleaned with disinfectant wipes before and after every hearing screen

- Disposable components of equipment shall not be re-used

Hearing Screening Personnel

A team of professionals, including audiologists, physicians, and nursing personnel, are needed to establish and maintain the UNHSI program. It is important that all members of the team work together to ensure a successful program. An audiologist should be involved in the newborn hearing screening program. Hospitals and agencies should also designate a physician to oversee the medical aspects of the UNHSI program.

Newborn hearing screening can be conducted by a wide variety of people, including audiologists, nurses, technicians, volunteers and students. The Official Code of Georgia, Section 43-44-7 (h) states that "a person not licensed as an audiologist may perform non-diagnostic electro-physiologic screenings of the auditory system, using automated otoacoustic emissions or automated auditory brainstem response technology as part of a planned and organized screening effort for the initial identification of communication disorders in infants **under the age of three months**, provided that:

1. The person not licensed as an audiologist has completed a procedure-specific training program directed by an audiologist licensed under this chapter;
2. The screening equipment and protocol used are fully automated and the protocol is not accessible for alteration or adjustment by the person not licensed as an audiologist;
3. The results of the screening are determined automatically by the programmed test equipment, without discretionary judgment by the person not licensed as an audiologist, and are only reported as '"pass" or "refer";
4. An audiologist licensed under this chapter is responsible for the training of the person not licensed as an audiologist, the selection of the screening program protocol, the determination of administration guideline the periodic monitoring of the performance of the person not licensed as an audiologist, and the screening program results; and,
5. The participation of the person not licensed as an audiologist in such an automated screening program is limited to the recording of patient demographic information, the application of earphones, electrodes, and other necessary devices; the initiation of the test; the recording of the results; and the arrangement of the referral for those who do not pass the screening to an audiologist licensed under this chapter for follow-up evaluation.

For children over 3 months of age, refer the child to a licensed audiologist or physician for testing. A physician, by law, may delegate aOAE and aABR hearing screenings to staff under their supervision, while they are on the premises, for infants over 3 months of age as stated in O.C.G.A. 43-44-7 (g).

- Screeners should be knowledgeable about screening technologies and competent in performing newborn hearing screening
- Screeners should be comfortable working with newborns and their families
- Screeners should be prepared to educate parents about newborn hearing screening including describing the hearing screen, type of technology, and delivering the results, and follow up recommendations

- Screener scripts are recommended to assist screeners with education and follow-up for newborn hearing screening
- Hospitals may choose to contract with an agency, audiologist, or practice that specializes in newborn hearing screening

The National Center for Hearing Assessment and Management (NCHAM) has an interactive Newborn Hearing Screening Training Curriculum (NHSTC) that is available as a resource and can be accessed online at http://www.infanthearing.org. The NHSTC has seven modules that provide information for a screener on the rationale for newborn hearing screening through all the steps of the hearing screening process including out-patient rescreening. The curriculum includes scripts for the screener on how to convey the hearing screening results in English and in Spanish. The NHSTC also has a resource section that lists supplemental educational material. Participants who register and complete the course will receive a Certificate of Completion from NCHAM. To register for the NHSTC course, please use the following URL: http://www.infanthearing.org/survey/nhstc.

All screeners should complete a hearing screening training curriculum directed by an audiologist licensed in Georgia with initial and annual competency documented.

Parent Education

Provide education to parent(s) in their native language, on the importance of newborn hearing screening to include the following:

- Approximately 3 out of every 1,000 live births are born with permanent hearing loss
- The first 18 months of life are the most critical for speech and language development; therefore it is extremely important to identify hearing loss as soon as possible
- Hearing loss is one of the most common birth defects and most babies who are born with hearing loss are born to hearing parents. 92% of children with permanent hearing loss are born to two hearing parents (Mithchell & Karchmer, 2004)
- Inform parents on the type of hearing screening technology that will be used to screen baby
- Explain the hearing screening process and how the screening technology works, clarifying that the hearing screening will not cause the baby any pain or discomfort, in fact it is best if the baby sleeps during the hearing screening
- Parents should be given a copy of the "Have You Heard?" brochure and informed of the importance of monitoring hearing developmental milestones
- Results of the hearing screen should be delivered to the parents both verbally and in writing, The "Have You Heard?" brochure has a section to document hearing screen results
- A language interpreter should be used if parents do not speak English, hospital protocol is to be followed when screening a baby from a non-English speaking family
- A baby with a "refer" result on the newborn hearing screening does not mean that the baby has a hearing loss; it indicates that further testing is needed to determine the baby's hearing status; **for babies who have a "refer" result on an in-patient hearing screening, it is extremely important that parents understand a follow-up hearing screening needs to be completed before the baby is one month of age**
- Every child with a hearing loss can achieve their fullest potential if they are identified very early and receive comprehensive intervention services

Parent Refusal

The Georgia Informed Consent law does not apply to newborn hearing screening. Written parental consent is not necessary; however, every effort should be made to educate parents about newborn hearing screening prior to conducting the screening. Parents do have the right to refuse the inpatient hearing screen, but must sign a waiver. The waiver must state that they have been educated on the importance of newborn hearing screening and follow-up, but are declining the newborn hearing screen. This waiver must become a permanent part of the infant's medical record. If the hospital does not have a waiver in place, they can use the UNHSI Program's Parent Refusal waiver (Appendix C).

- Original copy is placed in the baby's medical record
- A copy should be given to the parents with the "Have You Heard?" brochure stressing the importance of monitoring hearing developmental milestones
- A copy should accompany the Children 1st Screening and Referral form, sent to the health district where the child resides
- Notify PCP of parent refusal

Newborn Hearing Screening Checklist

It is recommended that all hospitals incorporate a newborn hearing screening checklist (Appendix D) into the baby's medical record for documentation of the following:

- Date of hearing screen
- Technology: aABR or aOAE
- Hearing screen results
- Verify correct primary care physician (PCP)
- PCP notified of results on all hearing screens in writing
    - Hospital should have a process in place where PCP is receiving results on all hearing screens (e.g. progress notes, discharge summary, etc.)
- Verify two primary contacts and numbers for the family
- Results of screen given to the parents verbally
- Results of screen given to the parents in writing
- Parents should be provided the "Have You Heard?" educational brochure
- Results of the screening are delivered semi-scripted to the parents (Appendix E)
- Follow-up appointment scheduled, if available, for an infant with a "refer" hearing screen result

Every hospital should also maintain a hearing screen log on all babies born in the facility documenting date, type of hearing screen, results of final hearing screen, follow-up if appropriate, and reason if baby was not screened (Appendix F).

Mandatory Birthing Facility Reporting

Georgia law 31-1-3.2, "Hearing screenings for newborns", mandates that all hospitals report to the Department of Public Health:

(1) Number of newborn infants born in the hospital
(2) Number of newborn infants screened
(3) Number of newborn infants who "passed" the screening
(4) Number of newborn infants who "referred" the screening
(5) Information is also requested on newborns discharged without the screening to include:

- Number of newborns that died prior to hearing screening
- Number of newborns in NICU and unable to be screened
- Number of newborns where hearing screening was refused
- Number of newborns transferred out prior to hearing screening
- Number of newborns discharged home without hearing screening

Aggregate numbers are to be reported quarterly to UNHSI via SendSS or paper based form (Appendix G). The number of births reported to Vital Records will be pre-populated in the SendSS module and viewable to the person reporting. The report is due 30 days following the end of the reporting quarter to allow more time for babies to be screened and reported in the quarter they were born. Newborns reported to be screened during the quarter should reflect the date of birth, not date of screen. Therefore, a baby can be born in one quarter, screened the subsequent quarter, but reported in the quarter they were born.

Detailed instructions and data definitions are available on the form and in the SendSS module to ensure all hospitals are reporting data accurately and consistently across the state. Any questions or concerns should be addressed by the UNHSI District Coordinator in the birthing hospitals health district.

As stated earlier, in addition to quarterly aggregate data, birthing facilities are required to report to Public Health: Newborns "referring" the initial or follow-up hearing screening (suspected hearing impairment) are to be reported to the Children 1st Coordinator in the health district where the baby resides by completing the Children 1st Screening and Referral Form within 7 days of a "refer" result hearing screen.

Where to Screen

Newborn hearing screening may be performed in the nursery, mother's hospital room, designated quiet room, or neonatal intensive care unit (NICU). It is best to select an area that is quiet and free of electrical interference. Screener should be knowledgeable about optimal screening conditions and troubleshooting techniques to minimize interference and obtain efficient screen.

Hearing Screening

Perform no more than two completed hearing screens prior to discharge.
When statistical probability is used to make pass/refer decisions, as is the case for (aOAE) and (aABR) screening technologies, the likelihood of obtaining a pass outcome by chance alone is increased when screening is performed repeatedly (Benjamini & Yekutieli, 2005; Hochberg & Benjamini, 1990; Zhang, Chung, & Oldenburg, 1999).

- All infants should have a hearing screen prior to discharge

- Make sure baby is medically eligible and stable for a hearing screening
- Recommend performing the hearing screen after 34 weeks gestational age
- It is advisable to screen after an infant has completed nursing or feeding, to increase the chance of the infant sleeping during the screening
    - Newborn hearing screening is more efficient and accurate when the newborn is quiet and content
    - Swaddling a baby is often helpful so the baby feels secure
- Newborns receiving antibiotic therapy should have a hearing screen prior to discharge. Antibiotic therapy should **not** be a reason for a "missed" screen
- Newborns receiving phototherapy for hyperbilirubinemia should have a hearing screen prior to discharge. Phototherapy should **not** be a reason for a "missed" screen
- Test time will depend on technology used and cooperation of baby
- If a baby "refers" on the initial hearing screening it is recommended that the hearing screen be repeated prior to discharge with at least four hours between screenings if possible

For rescreening, a complete screening on both ears is standard practice, even if only one ear "referred" the initial screening (JCIH Position Statement 2007).
Both ears have to "pass" every hearing screening for a "pass". If a baby has a switched ear result, for example, on initial screening "passed" in the left ear and "referred" in the right and then on the second screen "referred" in the left and "passed" in the right, this is a "REFER" not a "passed" hearing screen. Baby should be scheduled for follow-up.

Infants Identified with Risk Factors for Hearing Loss

Since 1972, the Joint Committee on Infant Hearing (JCIH) has identified specific risk factors that are often associated with infant and childhood hearing loss. Risk factors for hearing loss that are identified by the birthing facility should be reported using the Children 1st Screening and Referral form and submitted to the Children 1st Coordinator in the health district where the baby resides. There is a possibility that some of the important indicators, such as family history of hearing loss, may not be known at the time of screening in a newborn hearing screening program. The identification of risk factors should occur during early well-baby visits by the primary care physician/provider and referred via the Children 1st Screening and Referral form. Infants who are identified with these risk factors need to be monitored closely for normal communication developmental milestones during routine medical care.

Georgia has two protocols for infants and children birth to five who pass the newborn hearing screening but are identified with risk factors based on recommendations from the JCIH Position Statement 2007 (Appendix H). Please refer to *Notification Process for Infants (0-6 months) identified with Risk Factors for Late Onset* or *Progressive Hearing Loss and Notification Process for Children Late-identified with Risk Factors for Hearing Loss (6 months - 5 years)* for a detailed outline on procedures found in the appendix (Appendix I and Appendix J).

Screening Protocols in Well-Baby Nursery

Screening Technologies:

- It is appropriate to use either technology (aOAE) or (aABR) in the Well-Baby Nursery
- A combination of screening technologies, such as a two step protocol using an aOAE for the initial screening and an aABR if the baby "refers" the aOAE screening is acceptable *With this approach, infants who "refer" on an (aOAE) but "pass" an (aABR) are considered a "pass" screening result*

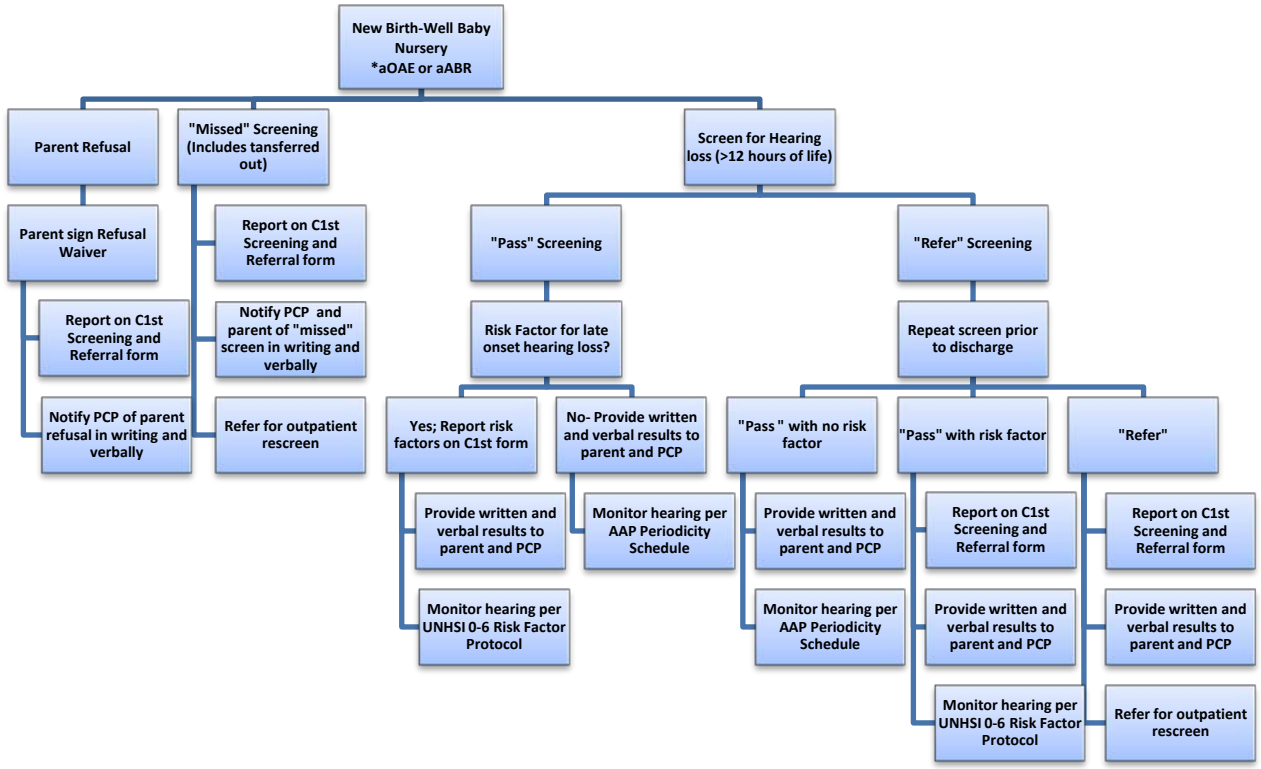
Infants who "refer" on an (ABR) screening should never be rescreened with (OAE) technology and "passed" because these infants are considered to be at risk for auditory neuropathy spectrum disorder (ANSD) (JCIH, 2007).

Time of Screening:

Hearing screens may be performed as early as 6 hours after birth. Waiting at least 12 hours and optimally 24 hours before newborn hearing screening allows more time for birth debris or vernix that may be in the newborns ear canal to clear. The additional time maximizes the opportunity to obtain an efficient newborn hearing screen especially for babies that are delivered by caesarean section.

Delaying a hearing screen 12 hours after birth may not be feasible if early discharge policies are in place; however, screening too early can result in high refer or false positive rates. Repeating the screening prior to discharge will help eliminate this problem (Winston, 2012). Hospital policy should be reviewed with regard to discharge when developing individual hospital newborn screening protocols. It is standard for vaginal deliveries to discharge between 24 to 48 hours and cesarean deliveries between 48 to 72 hours.

Hearing Screening Protocol – Well Baby Nursery:



Recommendations:

Newborn "passes" the hearing screen with no known risk factor(s):

- Parents should be notified of results verbally and in writing
- Information should be provided in the appropriate language for a non-English speaking family
- PCP should be notified of the hearing screen results in writing
- Educate the parent(s) on the importance of the baby achieving hearing developmental milestones; provide the "Have You Heard?" brochure
- Infant should be followed by their medical home/PCP according to the American Academy of Pediatrics (AAP) Periodicity Schedule
- Document the result in the infant's medical record and complete the checklist

Newborn "passes" the hearing screen with risk factor(s):

- Parents should be notified of results verbally and in writing
- Information should be provided in the appropriate language for a non-English speaking family
- PCP should be notified of the hearing screen results in writing
- Educate the parent(s) on the importance of the baby achieving hearing developmental milestones and recommended follow-up for risk factor(s)
- Complete the Children 1st form and forward to the UNHSI District Coordinator

- Document the result in the infant's medical record and complete the checklist
- Infant should be followed by their medical home/PCP according to the AAP Periodicity Schedule

Newborn "refers" the initial hearing screen:

- Rescreen the baby prior to discharge remembering that a "refer" result on an aABR hearing screen must be followed up with an aABR hearing screen
- Both ears must be screened at every screening even if only one ear "refers"
- It is recommended that screens be separated by a minimum of 4 hours if possible

Newborn "passes" second/final screen; follow above protocol for a "passed" screen.

Newborn "refers" the second/final hearing screen:

- Parents should be notified of results verbally and in writing
- Information should be provided in the appropriate language for a non-English speaking family
- Rescreen appointment should be scheduled if possible prior to discharge
- It is recommended to schedule the out-patient appointment within 7 to 14 days after discharge
- Parents should be educated on the importance of keeping the rescreen appointment or scheduling an out- patient rescreen appointment if not made prior to discharge
- The PCP should be informed of the "refer" result on the newborn hearing screen by the hospital in writing
- Document the result in the infant's medical record and complete the checklist
- The baby should be rescreened prior to one month of age
- The hearing rescreen can be completed by the birthing hospital, health department, PCP, ENT, or audiologist
- The Children 1st form must be completed, with risk factors identified and forwarded to the Health District where the child resides within seven days of "referred" hearing screen
- Infant should be followed by the medical home/PCP according to the AAP Periodicity Schedule

<u>Screening Protocols in the Neonatal Intensive Care Unit (NICU)</u>

The NICU is a newborn unit in the hospital where infants who need special care are placed. In the NICU, a neonatologist provides primary care for the infant. Newborn units are divided into the following 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)

Appropriate screening Technologies:

Due to the NICU population being at greater risk for ANSD, the JCIH 2007 position statement recommends that infants in the NICU who are admitted for greater than 5 days should have an auditory brainstem response (ABR) included as part of their hearing screening.

Time of Screening:

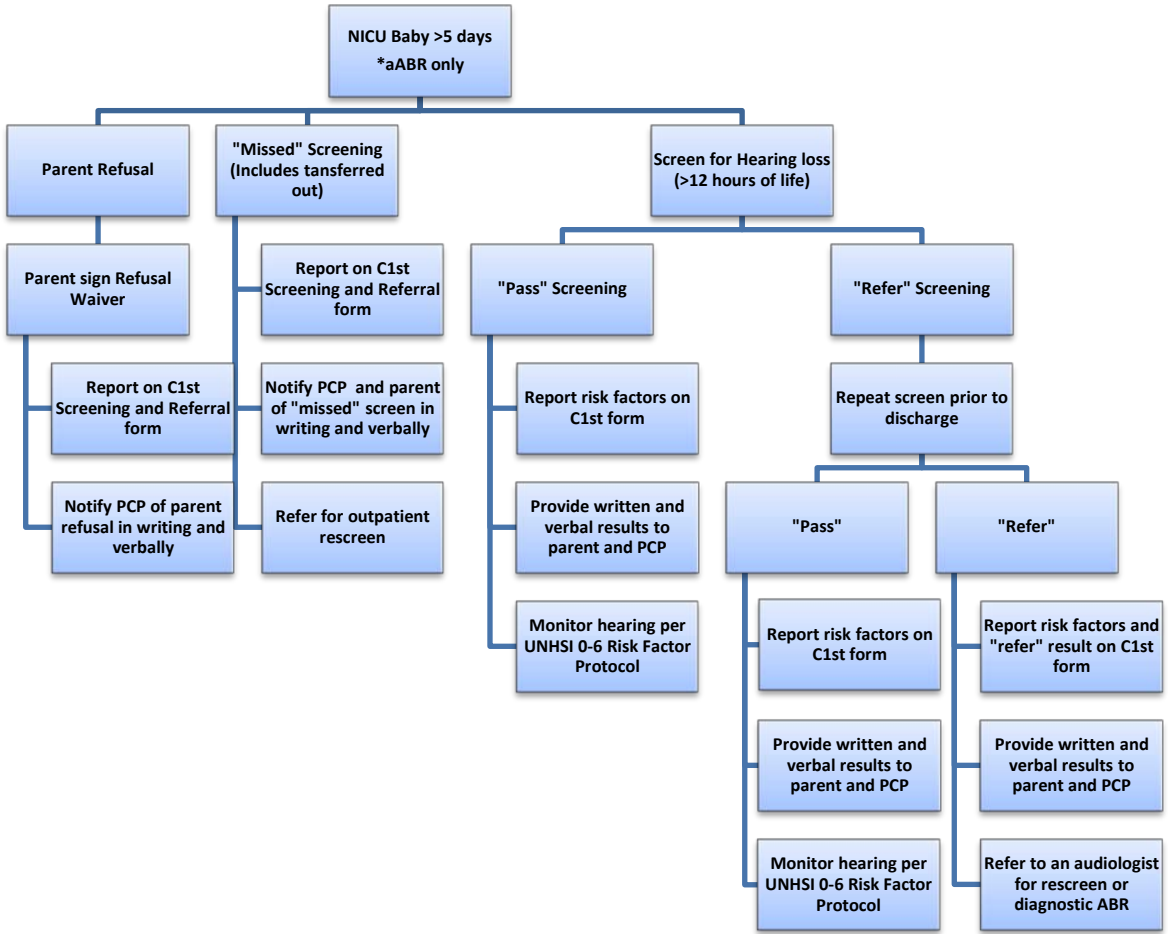
Hearing screening for infants in the NICU should be performed:

- prior to discharge
- after 34 weeks gestational age
- with infant in stable condition and off oxygen and antibiotics for 24 hours

Hearing Screening Protocol - NICU:

As stated previously, The Official Code of Georgia, Section 43-44-7 (h) and (g), a person not licensed as an audiologist may perform (aOAE) or (aABR) hearing screens in infants **under the age of three months**, chronological age. For infants over 3 months of age, refer to a licensed audiologist or physician for testing. A physician, by law, may delegate aOAE and aABR hearing screenings to staff under their supervision, while they are on the premises.

Outlined below are the minimal requirements for newborn hearing screening in the NICU. It is acceptable to perform dual newborn hearing screens which include screening with aOAE and aABR technologies as long as infants who are in the NICU for greater than 5 days receive an aABR hearing screening regardless of aOAE results. It is also appropriate to perform diagnostic ABR or complete diagnostic evaluation on infants in the NICU in place of automated ABR for facilities that have the capability.



Recommendations:

Newborn "passes" the hearing screen with risk factor(s):

- Parents should be notified of results verbally and in writing
- Information should be provided in the appropriate language for a non-English speaking family
- PCP should be notified of the hearing screen results in writing
- Educate the parent(s) on the importance of the baby achieving hearing developmental milestones and recommended follow-up for risk factor(s)
- Complete the Children 1st form and forward to the Health District where the child resides within seven days of hearing screening
- Document the result in the infant's medical record and complete the checklist
- Infant should be followed by their medical home/PCP according to the AAP Periodicity Schedule

Newborn "refers" the initial hearing screen:

- Rescreen the baby prior to discharge, remembering that a "refer" result on an aABR screen must be followed up with an aABR screen
- Both ears must be screened at every screening even if only one ear "refers"
- It is recommended that screens be separated by a minimum of 4 hours if possible

Newborn "passes" second/final screen; follow above protocol for a "passed" screen.

Newborn "refers" the second/final hearing screen:

- Parents should be notified of results verbally and in writing
- Information should be provided in the appropriate language for a non-English speaking family
- Follow-up appointment should be scheduled if possible prior to discharge
- It is recommended to schedule the outpatient appointment within 7 to 14 days after discharge
- Parents should be educated on the importance of keeping the follow-up appointment or scheduling an out- patient follow-up appointment if not made prior to discharge.
- The PCP should be informed of the refer result on the newborn hearing screen by the hospital in writing
- Document the result in the infant's medical record and complete the checklist
- Infant should be rescreened prior to one month of age
- **Refer to a pediatric audiologist for an ABR hearing rescreen or diagnostic evaluation**
- The Children 1st form must be completed, with risk factors identified and forwarded to the Health District where the child resides within 7 days of "referred" hearing screen
- Infant should be followed by the medical home/PCP according to the AAP Periodicity Schedule

Readmissions during first month of life

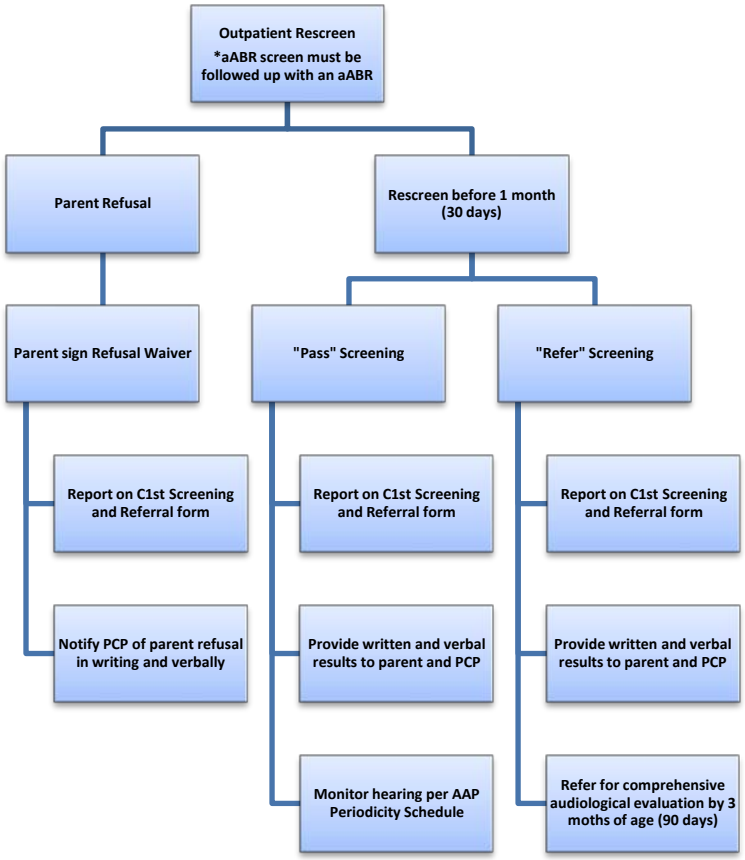
It is recommended that a hearing screening/rescreening be completed on **all** infants readmitted during the first month of life when there are conditions associated with potential hearing loss (JCIH Position Statement 2007).

Out-Patient Hearing Screening/Rescreen and Follow-up

For infants that are "missed" or "refer" on an in-patient newborn hearing screening, an out-patient hearing screening/rescreen may be conducted at the birth facility, health department, audiologist/ENT office, or physician's office. Automated OAE or ABR hearing screens may be conducted by trained screeners in accordance with Georgia Law, (Official Code of Georgia, Section 43-44-7), on infants under 3 months of age. Infants should be referred to a facility that has the appropriate services available in following best practice procedures for an infant who has a "refer" result on the newborn hearing screening.

- Outpatient hearing screening/rescreen should be completed prior to 1 month of age
- Recommend reminder appointment call to parents
- An aOAE may be followed up with an aOAE or aABR
- A "refer" on aABR **MUST** be followed up with an aABR; **patients who received an aABR screening as an inpatient, should not be followed up solely with an aOAE and "passed". Infants "referring" on aABR screening and "passing" a subsequent aOAE screening are assumed to be at risk for a diagnosis of neural hearing loss.**
- A baby who stayed in the NICU greater than five days that "referred" screening should be scheduled with pediatric audiologist for rescreen or diagnostic evaluation

- Both ears should be screened at out-patient hearing screening; **even if infant only "refers" on one ear**
- **Infants are to be screened one time at out-patient hearing screening**
- Infants that "pass" the outpatient hearing screening are monitored audiologically according to presence or absence of risk factors for delayed onset hearing loss or at discretion of PCP or audiologist; *parental concern for hearing loss, regardless of risk factors, is an immediate referral to an audiologist for hearing evaluation*
- **Infants that "refer" out-patient hearing screening should be referred to pediatric audiologist for diagnostic evaluation immediately**
- Diagnostic evaluation should be completed prior to 3 months of age
- Hearing screening results "pass" or "refer", from out-patient hearing screening/rescreening are reported on Children 1st Screening and Referral form to the Health District where the child resides within 7 days of screening/rescreening
- Hearing screening results "pass" or "refer", from out-patient hearing screening/rescreening are to be reported to PCP in writing
- Infant are to be followed by the medical home/PCP according to the AAP Periodicity Schedule



Border Babies

Infants whose parents are residents of Georgia but are born in another state are followed up according to the same procedures as infants that are residents of Georgia and born in Georgia. Neighboring states have differing protocols with regard to notification. Once a referral is received from another state indicating a newborn "referred" on hearing screening or was

identified with hearing impairment, that information is forwarded to the UNHSI District Coordinator for appropriate follow-up, documentation, and referrals to other Georgia public health programs. Referrals of infants that are born in Georgia but reside in another state are forwarded to appropriate state's Early Hearing Detection and Intervention (EHDI) Coordinator immediately for notification and follow-up.

Hearing Screening after the Newborn Period

OAE Hearing Screening:

There are national efforts encouraging home visiting and early childhood (0-3 years) programs to conduct Otoacoustic Emissions (OAE) hearing screenings to detect late onset hearing loss and capture children that "referred" on newborn hearing screening but did not receive follow up testing. Although these efforts to monitor for childhood hearing loss are appreciated, programs/providers are governed by Georgia state laws and regulations that define personnel who can conduct hearing screenings and under what conditions. In Georgia, automated OAE and ABR screenings can only be conducted by an audiologist or a physician beyond three months of age. The Georgia Department of Public Health wants to ensure that all programs utilizing automated OAE screening technology operate within the legal requirements imposed by state law. **For children over 3 months of age, refer the child to a licensed audiologist or physician for OAE screening.** A physician, by law, may delegate OAE and ABR hearing screenings to staff under their supervision, while they are on the premises according to O.C.G.A. 43-44-7 (g).

Diagnostic Audiological Assessment

This protocol is intended as a guide for audiologists who are performing diagnostic evaluations for children birth to five, who are at risk of, suspected of, or identified with auditory impairment, disorder, or disability. Audiologists performing audiological evaluations must hold a valid and current Georgia Audiology license. Audiologists designated to provide assessment and management of infants and children with hearing loss must have the commensurate knowledge, skill, and instrumentation necessary for use with current pediatric hearing assessment methods and evaluation procedures (The Pediatric Working Group, 1996).

The goal of the audiological evaluation is to determine degree and type of hearing loss. Hearing loss can be categorized into four types of hearing loss: conductive, sensory, neural, and mixed. Conductive hearing loss occurs when there is an issue between the external and inner ear, which can be transient in nature and may be medically treated (e.g. middle ear effusion or perforated tympanic membrane). Sensory hearing loss is generally permanent and indicates a problem with the inner ear. A neural hearing impairment can be due to a failure in the neural portion of the auditory pathway [e.g. auditory neuropathy spectrum disorder (ANSD)]. A mixed hearing loss is a combination of the types of hearing impairments and occurs when more than one type of hearing impairment contributes to the hearing loss. CDC data (2009) suggest that approximately 85% of babies identified through the newborn screening program are born with some permanent type of hearing loss.

Confirmation of an infant's hearing status requires a battery of audiological tests procedures to assess the integrity of the auditory system in each ear, to estimate hearing sensitivity across the speech frequency range, to determine the type of hearing loss, to establish a baseline for further monitoring, and to provide information needed to initiate amplification-device fitting if appropriate (JCIH, 2007). A comprehensive audiologic assessment includes a detailed case history, otoscopy, and behavioral and physiologic measures. Additionally, the

objective of an audiological evaluation is to obtain ear specific information, even for children referring on only one ear. Recommended testing procedures can be found at http://www.audiology.org/resources/documentlibrary/Documents/201208_AudGuideAssessHear_youth.pdf.

Diagnosis of hearing loss or audiological evaluation should not be delayed due to suspicion of middle ear dysfunction. A full battery of audiometry should be conducted to determine if there is a permanent hearing loss component and if the middle ear dysfunction is affecting hearing at that time. Presence of middle ear dysfunction as identified with tympanometry does not necessarily result in a hearing loss. Vice versa, the presence of middle ear dysfunction with hearing loss may prompt more aggressive treatment by a medical physician to minimize delays in language exposure.

Audiological assessment of infants referring on newborn hearing screening:

All infants who "refer" the initial hearing screening and the subsequent rescreening should have appropriate audiological and medical evaluations to confirm the presence of hearing loss at no later than 3 months of age (JCIH, 2007). For infants, birth to three months of age, the following testing protocol is recommended.

An appointment confirmation call to parents/caregivers of infants who are scheduled for a diagnostic audiological evaluation is recommended When confirming the appointment, convey the importance of follow up and evaluation, recommend an immediate family member to be present at appointment (emotional support), provide pre-test instructions (hungry, sleep-deprived, etc), and address any parental concerns. An audiologist on staff should be made available to talk with scheduled families if requested.

- Case History; a detailed case history should include relevant medical and developmental history, including prenatal and perinatal history, newborn hearing screening results, risk factors for infant hearing loss and progressive/late onset hearing loss, and parent/caregiver's judgments regarding responsiveness to sound in real world environment (e.g. http://www.asha.org/docs/html/GL2004-00002-F1.html)
- Otosopy
- Otoacoustic emissions (DPOAE or TEOAE): *OAEs provide a physiologic means of assessing preneural auditory function (Gorga, Neely, Bergman et al., 1993; Kemp, Ryan, & Bray, 1990) and used in conjunction with ABR, OAEs are not only useful in the differential diagnosis of cochlear hearing loss, but also in the identification of children with neurological dysfunction (ASHA, 2004)*
- Tympanometry: *For infants under 6 months of age, a probe tone of 1000 Hz is the most reliable to determine the presence or absence of middle ear pathology*
- Comprehensive ABR Testing for Threshold Estimation
    - Obtain a threshold response to a tone burst ABR or Auditory Steady State Response (ASSR) for 500, 1000, 2000, and 4000 Hz; at minimum, a low and high frequency response should be obtained
    - If a neural response is not identified, compare responses obtained to rarefaction and condensation clicks presented at 80 to 90 dB nHL using a fast click rate (>30 per second). If a response (e.g., cochlear microphonic) is observed, an auditory neuropathy should be suspected
    - Obtain a bone conduction click threshold if ABR thresholds are elevated (>20 dB nHL)

- Observation of infant and/or behavioral audiometry when developmentally appropriate: *Behavioral testing is recommended as part of the protocol to validate physiological results. 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 (JCIH, 2007)*

Immediately after assessment for infants referred from newborn hearing screening, the following should be completed**:**

- Review results of the audiologic assessment, implications of the audiologic diagnosis, and recommendations for intervention with the parents/caregivers verbally, including:
    - Information about typical speech, language, and listening developmental milestones
    - Information about risk indicators for progressive and delayed-onset hearing loss and
    - Document in chart or on report if the results were provided verbally
- Provide any relevant educational brochures or handouts on diagnosis and other appropriate subject matters to parents and/or immediate caregivers
- Provide a written report to the family/caregiver, to the infant's primary care provider, and to the referral source with consent. Chart should contain documentation for all persons/facilities receiving report
- Complete Georgia's Surveillance of Hearing Impairment form (Appendix B) and submit to the UNHSI District Coordinator for the health district in which child resides
    - For Audiologists registered in SendSS, document in child record audiological evaluation results, testing, notes, and recommendations.
    - Chart should contain documentation that that referral was sent to Public Health and/or entered into SendSS.

Audiological assessment beyond newborn hearing screening and referral:

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

- Case history: *A detailed case history should include relevant medical and developmental history, including prenatal and perinatal history, newborn hearing screening results, risk factors for infant hearing loss and progressive/late onset hearing loss, and parent/caregiver's judgments regarding responsiveness to sound in real world environment (e.g. http://www.asha.org/docs/html/GL2004-00002-F1.html)*
- 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
    - Speech-detection and/or speech recognition measures depending on the child's age and developmental level.
- OAE testing (DPOAE or TEOAE)
- Tympanometry: *For children greater than 6 months of age, a 226 Hz probe tone should be used*
- Acoustic reflex thresholds

- ABR testing if responses to behavioral audiometry are not reliable: *For children greater than 4 months of age, sedation may be required to perform ABR testing; sedation has significant health risks associated with the administration of drugs, and should only be administered and monitored in the presence of a medical professional*

Immediately after audiological assessment, the following should be completed**:**

- Review results of the audiologic assessment, implications of the audiologic diagnosis, and recommendations for intervention with the parents/caregivers verbally, including:
    - Information about typical speech, language, and listening developmental milestones
    - Information about risk indicators for progressive and delayed-onset hearing loss
    - Document in chart or on report if the results were provided verbally
- Provide any relevant educational brochures or handouts on diagnosis and other appropriate subject matters to parents and/or immediate caregivers
- Provide a written report to the family/caregiver, to the infant's primary care provider, and to the referral source with consent; chart should contain documentation for all persons/facilities receiving report
- When a hearing loss is identified, complete Georgia's Surveillance of Hearing Impairment form (Appendix B) and submit to the UNHSI District Coordinator for the health district in which child resides
    - For Audiologists registered in SendSS, document in child record audiological evaluation results, testing, notes, and recommendations
    - Chart should contain documentation that that referral was sent to Public Health and/or entered into SendSS

Parent Support and Education Following Diagnosis of Permanent Hearing Loss

Of the children with permanent congenital hearing loss, 92% are born to two hearing parents and 96% of children with permanent hearing loss are born to two hearing parents or one hearing parent and one parent with hearing loss (Mitchell & Karchmer, 2004). Research suggests that parents experience very powerful emotions at the time of diagnosis including denial, shock and upset, with a great need for emotional support (Russ et. al., 2004). Similarly, communication difficulties between parents and providers exists (Russ et. al., 2004). Research also suggests that parents prefer to have parent to parent support after diagnosis (Fitzpatrick et. al., 2007) for emotional support and information gathering. Furthermore, parents share a desire to connect with other parents who have children diagnosed with a hearing loss for support and information (Jackson, 2011).

Counseling parents of children identified with hearing loss is an ongoing process. Guidelines and information on counseling parents can be found through the NCHAM EHDI E-Book at http://infanthearing.org/ehdi-ebook/2013_ebook/13Chapter12ParentCounseling2013.pdf and through American-Speech-Language-Hearing Association (ASHS) at http://www.asha.org/policy/GL2008-00289.htm. Parents are to be encouraged to reach out to parent and family support networks through Parent to Parent of Georgia and/or Hands & Voices.

Guidelines for Pediatric Amplification

Audiologists providing amplification services must hold a valid and current Georgia Audiology license and must have knowledge, skill, and instrumentation necessary for providing amplification and management for children. Medical clearance must be obtained from an otolaryngologist prior to hearing aid fitting. Amplification decisions should be based on audiological information from ABR and behavioral testing. Electrophysiological results may need to stand alone for a period of time to determine appropriate fitting levels. However, as soon as the child is able to participate, behavioral threshold measures should be obtained and used to cross-check prior results (American Academy of Audiology, 2012). Other factors contributing to fitting include, but are not limited to performance in the home and/or educational environments, family preference, other existing conditions, and speech and language development.

Amplification Options

- Behind-the-ear (BTE) hearing aid
    - Most appropriate for children due to rapid growth of the outer ear
    - Hearing aid features (e.g. directional microphone, volume control, tamper resistant battery door) and processing schemes should be closely considered when choosing an appropriate hearing aid given child's age and hearing loss
    - Earmolds should be made of soft material for safety and retention
- Bone conduction aid
    - Appropriate for conductive hearing loss in cases that a BTE hearing aid cannot be fit
    - A bone anchored hearing aid may be considered, but is not approved for use for children less than five years old by the U.S. Food and Drug Administration (FDA)
- Cochlear Implant
    - Currently not FDA approved until 12 months of age for children with bilateral profound hearing loss
    - Eligibility and candidacy criteria should be carefully considered prior to implantation and should include a team of professionals working with the family and child
- Frequency Modulation (FM) system: *Coupled with personal hearing aid or cochlear implant, which is used to improve the signal to noise ratio*

Verification and Monitoring

Verification of hearing aid fitting should be performed to ensure child has optimal settings of hearing aid. Probe microphone measurements and aided soundfield responses are recommended to be conducted after hearing aid fitting to evaluate the hearing aid output values and the audibility of sounds. Follow up visits, at minimum, should include parent input on child performance with hearing aids (Cochlear implant, FM system), functional auditory skill assessments, verification of proper usage and fit of hearing aid and any necessary troubleshooting. The frequency and scheduling of follow up visits depend on the patient's age and family needs, but should occur more frequently after initial fitting.

Georgia Hearing Aid Loaner Bank – GA HALB

In September 2012, the Georgia Hearing Aid Loaner Bank (GA HALB) began servicing infants/children in need of amplification. The purpose of the program is to provide temporary

hearing aids for children with hearing loss, who are birth to 36 months of age, while they are waiting to receive their personal amplification devices. The GA HALB lends hearing aids for up to six months, on a onetime per child basis, for children newly diagnosed with a hearing loss. Referrals to the program must be initiated through a District UNHSI Follow-up Coordinator, who will oversee the application process and serve as a resource to the family throughout the process (Appendix K).

Role of the Primary Care Physician and Medical Home

The primary care physician's role and support is vital to the success of the UNHSI program. The primary care physician is to be the center of the medical home as they are an active participant in the life of the family during a baby's first year. The medical home is responsible for ensuring appropriate and timely referrals to providers that are capable of performing evaluations and knowledgeable in congenital hearing impairment. Georgia has adapted the AAP Guidelines for Pediatric Medical Home Providers, a flow chart of follow up procedures to newborn hearing screening, which can be found in the appendix (Appendix L) and should serve as resource for primary care physicians.

1. Obtain written results of newborn hearing screening from birthing facility on all newborns.
2. By one month of age, ensure that all newborns have at minimum one hearing screening or a secondary screening if infant "referred" inpatient hearing screening.
3. Refer for audiological diagnostic evaluation for infants "referring" secondary screening before three months of age.
4. Provide referrals to early intervention, otolaryngologist, ophthalmologist and genetics after diagnosis of permanent hearing impairment.
5. Manage otitis media with effusion.
6. Closely monitor for signs of hearing loss for infants who pass newborn hearing screening and refer for audiological evaluation per JCIH recommendations, developmental/speech delay, or parental concern, as hearing loss may develop at any age.

Role of the Otolaryngologist

A child newly diagnosed with a permanent hearing loss is to be referred to an Otolaryngologist (ENT) for a medical evaluation. The ENT's role is to determine the potential etiology of the hearing loss, provide clearance for hearing aids, and make referrals to the appropriate specialists as needed. Additionally, the physician will also determine if the problem is medically or surgically treatable, and if so, provide the necessary medical or surgical treatment. To determine cause of hearing loss or possible treatments, the ENT may refer the child for procedures such as imaging studies (X-rays, CT-scans, MRI scans).

There are many causes of congenital hearing loss. Genetic causes are responsible for hearing loss among 50% to 60% of children with hearing loss, and about 20% of babies with genetic hearing loss have a "syndrome" associated with hearing loss (e.g. Usher Syndrome or Down Syndrome) (Morton and Nance, 2006). As it relates to congenital hearing loss, 30% can be attributed to infections during pregnancy in the mother, other environmental causes, and complications after birth (Morton and Nance, 2006). Three to five percent of infants are exposed to congenital cytomegalovirus (CMV), a preventable infection, develop bilateral moderate to

profound sensorienural hearing loss and 14% of infants exposed to CMV develop some degree of sensorineural hearing loss (Grosse, Ross, and Dollard, 2008).

Referral to Intervention Services

A child, birth to five, diagnosed with permanent hearing loss is reported to Georgia Department of Public Health (DPH) by the audiologist. This referral to DPH serves multiple purposes. One function is to act as the referral source into Georgia early intervention programs. The referral is handled by Children 1st, Georgia's "single point of entry", who links the family with the two state intervention programs: Babies Can't Wait (BCW) and Georgia Parent Infant Network for Education Services (Georgia PINES). Each program has its own eligibility criteria and has different program goals. All families of babies who meet program eligibility criteria, who have a permanent childhood hearing loss will be offered enrollment in State intervention programs, which may either be accepted or refused.

All families are offered an initial early hearing orientation specialist (EHOS) visit conducted by Georgia PINES within 7 days of initial referral to Public Health. The EHOS visit is to provide written and oral information on hearing loss, to give unbiased information on communication options and amplification devices, and to stress the importance and urgency of enrollment in intervention. Additionally, parents and families receive information on local, state, and national resources and services for children with hearing loss. Parents can either accept or refuse the EHOS visit. Families who refuse the EHOS visit are referred back to Children 1st. For families who have an EHOS visit, a report is sent back to Children 1st and BCW, which includes the family's decision whether to enroll or defer services from Georgia PINES.

Families who chose to enroll in Georgia PINES are visited by Parent Advisors, who serve as a resource and outline the curriculum that will be used to promote the child's acquisition of communication and listening skills. Georgia PINES provides family-centered services that include the provision of information and emotional support, a home hearing aid program, preparation for a cochlear implant, communication and auditory and/or visual (e.g. sign language) programs, as well as assisting parents with resources. Parents of children who have hearing loss have options for the SKI-HI or INSITE curriculum, and Signing Deaf Mentors are available as well. Services regarding communication methods are based on the family's priority. The parent advisor works in partnership with the family to set goals/outcomes for the child specific to hearing loss. Home visits focus on strategies to obtain the goals and assess progress. The goal of Georgia PINES is to have the child demonstrate 12 months language advancement in one calendar year. Visits are conducted at minimum bi-monthly and last 60 minutes in duration. Each visit includes a greeting, follow up and review of previous activities, check hearing aid, cochlear implant, and/or FM system when applicable, presentation of new curriculum activities, discuss challenges, summary, and future planning. For more information on Georgia PINES services visit http://www.gapines.info/.

BCW provides intervention for children with bilateral, mild to profound hearing loss, or any children with any type of documented developmental delay. BCW offers service coordination that assists the family and other professionals in developing a plan to enhance the child's development. Services, provided in natural environments, are provided by agencies and individuals from both public and private sectors. Intervention services, training, resources and referrals in the community are made to help meet the developmental needs of the child. More information can be found at www.georgiafamiliesmatter.org and http://dph.georgia.gov/Babies-Cant-Wait.

There are a variety of private resources available across the state, including audiologists, speech-language pathologists, and programs that specialize in the development of a particular mode of communication. A brief description with contact information for statewide programs serving children with hearing loss is provided to all families during the EHOS visit described above. For more information about hearing impairment-related resources available in a particular community, you may wish to contact the Georgia Council for the Hearing Impaired (1-800-541-0710 [V/TTY] or 404-292-5312 [V/TTY] in metro Atlanta, www.gachi.org) and Parent to Parent of Georgia (1-800-229-2038 or 770-451-5484 in metro Atlanta, www.parenttoparentofga.org). You may also visit the UNHSI web page at http://dph.georgia.gov/universal-newborn-hearing-screening-unhsi.

Additional Resources

Alexander Graham Bell Association for the Deaf and Hard of Hearing
www.agbell.org

American Academy of Audiology
www.audiology.org

American Academy of Otolaryngology – Head and Neck Surgery
www.entnet.org

American Academy of Pediatrics, National Headquarters
http://www.aap.org

Americian Academy of Pediatrics Bright Futures Guidelines
http://brightfutures.aap.org/pdfs/Preventive%20Services%20PDFs/Screening.PDF

American Academy of Pediatrics, Georgia Chapter
www.gaaap.org

American Society for Deaf Children
www.deafchildren.org

American Speech-Language-Hearing Association
www.asha.org

Boys Town National Research Center
http://www.boystownhospital.org/pages/default.aspx

Early Hearing Detection Intervention Program
http://www.cdc.gov/ncbddd/hearingloss/ehdi-programs.html

Educational Audiology Association
www.edaud.org

Georgia Council for the Hearing Impaired
www.gachi.org

Georgia PINES
www.gapines.info

Hands & Voices-National
http://www.handsandvoices.org

Hands & Voices-Georgia Chapter
http://www.gahandsandvoices.org

Hearing, Speech, & Deafness Center
http://www.hsdc.org

John Tracy Clinic
http://www.jtc.org

Joint Committee on Infant Hearing
www.jcih.org

Marion Downs Hearing Center
http://www.mariondowns.com

National Association of the Deaf
www.nad.org

National Center for Hearing Assessment and Management (NCHAM)
www.infanthearing.org

National Cued Speech Association
www.cuedspeech.org

National Institute on Deafness and Other Communication Disorders
www.nidcd.nih.gov

Oberkotter Foundation (Private Oral Schools)
www.oraldeafed.org

Parent to Parent of Georgia
http://p2pga.org

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Appendix A Children1st_Screening Referral Form

DPH Georgia Department of Public Health

Children 1st
Screening and Referral Form

DIRECTIONS: Please complete form on every child, birth to age 5, having any of the conditions listed on 1st or 2nd page. Check or fill in as much information as possible. Send form to local Children 1st Coordinator.

Referral Source: _____ **Date Received:** _____

SECTION A — CHILD AND FAMILY INFORMATION

CHILD'S INFORMATION

Child: _____
Last Name First MI

Date of Birth: _____ Birth weight: _____

Sex: ❑ Male ❑ Female ❑ Unknown Gestational Age: _____

Select race: (Mark all that apply)

❑ White ❑ Black or African American
❑ Asian ❑ American Indian or Alaska Native
❑ Unknown ❑ Hawaiian/ Other Pacific Islander

Latino/Hispanic: ❑Yes ❑No ❑ Unknown

Hospital: _____ Discharge Date: _____

Transfer Hospital: _____ Discharge Date: _____

Type of Insurance: ❑ Medicaid ❑ PeachCare ❑ Private
❑ WellCare CMO ❑ Tri-Care
❑ Amerigroup CMO ❑ None
❑ PeachState CMO ❑ Unknown

Child's Insurance #: (if known) _____

LANGUAGE NEEDS

Primary Language: _____ Translator/Interpreter Needed: ❑ Y ❑ N

CHILD'S PRIMARY MEDICAL/HEALTH CARE PROVIDER

_____
Name

_____
Street or Route

_____
City State Zip

_____
Phone Fax

MOTHER'S INFORMATION

Mother: _____
Last Name First MI Maiden

Age: _____ Date of Birth: _____

Education: (last grade completed)

Marital Status: ❑ M ❑ NM ❑ SEP ❑ D ❑ W

Live in Partner: ❑ Yes ❑ No

Prenatal Care: ❑ 1st ❑ 2nd ❑ 3rd ❑ None

Parity G:____ P:____ Pre-Term:____ AB: Elective/Spontaneous ____ /____

Parent's Medicaid #: _____

FATHER'S INFORMATION

_____
Last Name First MI

GUARDIAN/FOSTER CARE REFERRALS

_____
Guardian/Foster Parent Last Name First Phone Number

_____
DFCS Case Worker Last Name First Phone Number Fax Number

CONTACT INFORMATION

Child Lives with: ❑ Mother ❑ Father ❑ Guardian ❑ Foster Parent

Child's Address: _____
Street /Route Apt Complex # / Mobile Hm Park#

_____
City County Zip

Phone #: _____ Emergency Contact #: _____

Caregiver email address: _____

SECTION B — HOSPITAL INFORMATION

Newborn Hearing Screening: ❑ **Not Screened** ❑ **Family Refused Screening**

Inpatient: Date: ____/____/____ **Left:** ❑ Pass ❑ Refer **Right:** ❑ Pass ❑ Refer ❑ AOAE ❑ AABR ❑ Other

Outpatient: Date: ____/____/____ **Left:** ❑ Pass ❑ Refer **Right:** ❑ Pass ❑ Refer ❑ AOAE ❑ AABR ❑ Other

Newborn Bloodspot Metabolic Screening: ❑ Not Screened ❑ Family Refused Screening

Equipment:

Vaccines Given During Hospital Stay:

Hepatitis B Vaccine: (date) _____

HBIG: (date) _____

SECTION C — LEVEL 2 RISK CONDITIONS (3 OR MORE MUST BE PRESENT FOR ELIGIBILITY)

Conditions Identified at Birth

655.4 ❑ Suspected damage to fetus (Mother Smoked and/or Drank, > 7 drinks/week, during Pregnancy)

765.16-765.18 ❑ Disorders r/t other preterm infants <2500 Grams (5 lbs. 8 oz.) and > 1500 Grams

V23.7 ❑ Insufficient Prenatal Care (Little or no prenatal care)

V23.83-V23.84 ❑ Young Prima-/Multi-gravida (Maternal Age <18 years)

V62.3 ❑ Education Circumstances (Maternal Education <12 Years)

Child Abuse Prevention Treatment Act (CAPTA)

All CAPTA referrals are automatic referral (Child age birth to 3 years)

V60.81 ❑ *Foster Care*

995.5 ❑ *Child Maltreatment Syndrome (Substantiated Case)*

DFCS Referrals (no CAPTA)

V60.81 ❑ *Foster Care (over age 3)*

995.5 ❑ *Child Maltreatment (Substantiated Case) (over age 3)*

V61.05 ❑ *Unsubstantiated or sibling of victim of substantiated case (birth to 5)*

C1MD.1 ❑ *Child under age 5 exhibiting physical or developmental delay*

Socio-Environmental Conditions Present in the Family

V17.0 ❑ Psychiatric condition (Parental Mental Illness, Depression)

V60.0 ❑ Lack of Housing (Homelessness)

V61.05 ❑ Family disruption due to child in welfare custody

V61.5 ❑ Multiparity - in Mother (<20 Years of age, >3 pregnancies)

V62.5 ❑ Legal Circumstances (Parental Incarceration)

V16-V19 ❑ Family History of (**Specify**)_____ (Illness/disability affecting care of child)

C1SEC.1 ❑ Child Injuries (>3 in 1 Year) Requiring Medical Attention **Specify**: _____

V18.4 ❑ Mental Retardation (Parental Mental Retardation)

V60.2 ❑ Inadequate Material Resources (Affecting Care of Child)

V61.2 ❑ Parent-Child Problems (Questionable Mother/Child Attach)

V62.0 ❑ Parental Unemployment

V62.8 ❑ Other Psych. or Physical Stress, (History of Family Violence)

SECTION D — SIGNATURES

_____
Name of Person Completing Form Agency Email Address Phone Date

Parent Signature (Encouraged but not required for referral) _____ Parent Informed of Referral? ❑ Yes ❑ No

Form #3267 Page 1 of 2

Child's Name:	Mother's Name:

SECTION E (check all that apply) **LEVEL 1 RISK CONDITIONS**

(Medical/Biological Conditions Present in Child Indicating Referral to Public or Private Sector Care)

Infectious and Parasitic Diseases

- 042 ❑ HIV
- 090 ❑ Syphilis

Mental Disorders

- 299.00-299.01 ❑ Autistic disorder
- 315.3 ❑ Developmental speech or language disorder
- 315.9 ❑ Unspecified delay in development
- C1MD.1 ❑ Suspected Developmental Delay

Endocrine, Nutritional & Metabolic Diseases, and Immunity Disorders

- 243 ❑ Congenital hypothyroidism
- 27X.X X ❑ Disturbances of amino-acid metabolism (Metabolic disease)
  Specify(code, diagnosis):_____

Diseases of the Blood and Blood-Forming Organs

- 282.X ❑ Hereditary hemolytic anemias
  Specify(code, diagnosis): _____

Diseases of the Nervous System and Sense Organs

- 320 ❑ Meningitis, Bacterial
- 321 ❑ Meningitis, All Other
- 323.9 ❑ Encephalitis
- 343.1-343.9 ❑ Infantile cerebral palsy
- 345 ❑ Epilepsy/Seizure Disorder
- 348.3 ❑ Encephalopathy
- 356-359 ❑ Neuromuscular Disorder
- 362.26 or 362.27 ❑ Retinopathy of Prematurity (Grades 4 or 5)
- 369.XX ❑ Blindness and low vision
  Specify (code, diagnosis):_____
- 382.9 ❑ Unspecified otitis media – chronic (recurrent or persistent)
- 389.XX ❑ Hearing Loss
  Specify(code, diagnosis): _____
- C1DNS.1 ❑ Suspected Hearing Impairment

Serious Problems or Abnormalities of Body Systems

- 390 – 459 ❑ Heart/Circulatory System
- 460 – 519 ❑ Respiratory System
- 493 ❑ Asthma
- 520 – 579 ❑ Digestive System
- 580 – 629 ❑ Genito-Urinary System
- 710 – 739 ❑ Musculoskeletal System and Connective Tissue
- 740 – 759 ❑ Congenital anomalies
- 749 ❑ Cleft Palate/Lip

Specify Conditions for All Above (include Diagnosis Code):_____

_____

_____

_____

_____

_____

Conditions Originating in the Perinatal Period

- 760.71 ❑ Fetal Alcohol Syndrome
- 764.00 ❑ Light-for-dates infant without fetal malnutrition unspecified (birth weight < 10% for gestational age)
- 764.9 ❑ Fetal Growth Retardation (Intrauterine Growth Reduction-IUGR)
- 765.01-765.03 ❑ Disorders r/t extreme immaturity of infant (BW < 999 gms)
- 765.14-765.15 ❑ Disorders r/t other preterm infants (BW 1000-1500 gms)
- 767.0 ❑ Subdural and cerebral hemorrhage due to birth trauma
- 768.5 ❑ Severe birth asphyxia (APGAR < 3 at 5 Minutes)
- 770.7 ❑ Chronic Respiratory Disease in perinatal period (Broncho-pulmonary Dysplasia)
- 770.81 or 770.82 ❑ Primary apnea or other apnea in newborn
- 770.9 ❑ Unspec. Respir. Condition of fetus/newborn (vent > 48hrs)
- 771.0 ❑ Congenital Rubella
- 771.1 ❑ Congenital cytomegalovirus infection (CMV)
- 771.2 ❑ Other congenital infection in perinatal period (Herpes Simplex-congenital, Toxoplasmosis)
- 772.13 or 772.14 ❑ Intraventricular Hemorrhage (IVH), Grade III or IV
- 774.4 ❑ Perinatal jaundice d/t hepatocellular damage (NB Hepatitis)
- 774.6 ❑ Neonatal jaundice (requiring exchange transfusion)
- 777.53 ❑ Stage III necrotizing enterocolitis in newborn
- 779.0 ❑ Convulsions in newborn
- 779.3 ❑ Feeding Problems in newborn (severe reflux/feeding tube)
- 779.5 ❑ Drug Withdrawal Syndrome in Newborn
- 779.7 ❑ Periventricular/Preventricular Leukomalacia (PVL)
- C1COP.1 ❑ NICU Stay > 5 days

Symptoms, Signs and Ill-Defined Conditions

- 783.4 ❑ Failure to Thrive/Growth Deficiency (growth below 5th %)
- 796.4 ❑ Other abnormal clinical findings
  Specify(code, diagnosis):_____

Injury and Poisoning

- 959.01 ❑ Other and unspecified injury to head
- 984 .0-984.9 ❑ Toxic effect of lead and its compounds, including fumes
  Lead Level > 20 µg/dl (Venous)
  Specify:_____
  Lead Level > 10 <20 µg/dl (Venous)
  Specify:_____
- C1INJ.1 ❑ Ototoxic medications including chemotherapy

Other Significant Conditions

- V02.6 ❑ Carrier/suspected carrier of viral hepatitis (Hep. B in Mom)
- V19.2 ❑ Family history of deafness or hearing loss
- V61.41 or V61.42 ❑ Alcoholism or Substance Abuse in Family (Maternal use of street, prescription or OTC drugs via self-report, drug screen or court record)
- 237.70-237.79 ❑ Neurofibromatosis

SECTION F **REFERRAL CRITERIA LEGEND**

Health Department Staff: Please see eligibility lists for Babies Can't Wait, Children's Medical Services, 1st Care, Universal Newborn Hearing Screening, Genetics, and Lead Programs in order to appropriately refer children.

SECTION G **COMMENTS**

Has child received a recent developmental screening ?: ❑ Not screened ❑ Yes, screened by _____ (Please attach results)

Measure used: _____ Date screening completed_____ Scores _____

Appendix B Surveillance Form

Surveillance of Hearing Impairment in Infants and Young Children

Georgia's Universal Newborn Hearing Screening and Intervention (UNHSI) Program

Confirmation of hearing loss for children, birth to five (OCGA 31-12-2)

Reportable hearing impairment as measured and described by an audiologist or physician as a permanent (or suspected to be permanent) hearing loss, averaging 15 dB HL or greater in either or both ears, in the frequency region 500 Hz – 4000 Hz. Cases of hearing loss in newborns and children through 5 years, must be reported to Public Health within 7 days of diagnosis.

Child: Last:_____ First:_____ Sex:☐M ☐F DOB:____/____/____

Address:_____ City:_____ Zip:_____

Parent: Last_____ First:_____ Relationship: _____

Phone:_____ Alt. Phone:_____ Language:_____

PCP:_____ Clinic:_____ Phone:_____

Complete the section(s) appropriate for your evaluation. If performing re-screening due to referral from newborn hearing screening, please complete the Children 1st Screening and Referral Form. Do NOT delay complete diagnosis solely due to middle ear dysfunction

Diagnostic Hearing Results **Date of Evaluation:____/____/____**

Tympanometry: ☐ 226 Hz ☐ 1000 Hz

Right Ear: ☐ Normal ☐Abnormal

Left Ear: ☐ Normal ☐Abnormal

☐ABR Click ☐ VRA

☐ Freq. Specific ABR ☐Pure Tone Threshold

☐ Bone Conduction ABR

☐ Other: _____

OAE:

Right Ear: ☐Present ☐Absent

Left Ear: ☐Present ☐Absent

Degree of Hearing Impairment is based on a four frequency pure tone average, if available

Left Ear		Right Ear	
Degree (in dB HL)	**Type**	**Degree (in dB HL)**	**Type**
☐ Normal	☐ Normal	☐ Normal	☐ Normal
☐ Slight (15-25)	☐ Conductive	☐ Slight (15-25)	☐ Conductive
☐ Mild (26-40)	☐ Transient Conductive	☐ Mild (26-40)	☐ Transient Conductive
☐ Moderate (41-55)	☐ Mixed	☐ Moderate (41-55)	☐ Mixed
☐ Moderately-Severe (56-70)	☐ Sensorineural	☐ Moderately Severe (56-70)	☐ Sensorineural
☐ Severe (71-90)	☐ Auditory Neuropathy	☐ Severe (71-90)	☐ Auditory Neuropathy
☐ Profound (>90)		☐ Profound (>90)	

Recommended Follow Up:

☐ Refer to ENT, name if known: _____ ☐ Refer to Part C Program (BCW) and GA PINES

☐ Repeat Audiological Testing, Date:_____ ☐ Refer for Speech and Language Evaluation

☐ Hearing Aid Evaluation, Date:_____ ☐ Other: _____

Audiologist: _____ Clinic:_____ Phone:_____

Additional Comments:

Georgia Department of Public Health
UNHSI Program
2 Peachtree, NW, 11th Floor
Atlanta, GA 30303
Phone: (404) 657-4143
Fax: (404) 657-2773

Case Definition of Confirmed Hearing Impairment in Newborns and Children through Age 5 Years:
Reportable hearing impairment is defined as hearing impairment measured and described by an audiologist or physician as a permanent (or suspected to be permanent) hearing loss, averaging 15 dB HL or greater in either or both ears (unilateral or bilateral), in the frequency region important for speech recognition (500 Hz – 4000 Hz). Severity of the hearing impairment shall be defined on the basis of the following measured or described hearing threshold levels:

Slight Hearing Loss: 15 to 25 dB	Moderate Hearing Loss: 41 to 55 dB	Severe Hearing Loss: 71 to 90 dB
Mild Hearing Loss: 26 to 40 dB	Moderately Severe Hearing Loss: 56 to 70 dB	Profound Hearing Loss: > 90 dB

Case Status:
Confirmed: A diagnosis of hearing impairment (ICD-9 codes 389.0 through 389.9) confirmed by a licensed audiologist or physician according to the case definition above is reportable.
Suspected: A 'refer' result on an initial newborn hearing screening or a follow-up hearing screening with a newborn (by either aABR or aOAE) in either ear or both ears is reportable. **Screening test results should be reporting using the Children 1st Screening and Referral Form**.

Reporting Procedures: The initial diagnosis of hearing loss that is determined or suspected to be permanent and/or progressive in nature in children up to age 5 is reportable by law (OCGA 31-12-2) to Public Health. Suspected and confirmed cases of hearing loss in newborns must be reported to Public Health within 7 days of screening or confirmatory diagnosis. Initial diagnosis of hearing loss that is determined or suspected to be permanent and/or progressive in nature in children through age 5 years is also reportable within the 7-day time frame. The Surveillance of Hearing Impairment in Infants and Young Children Form should be completed for each diagnosed case of hearing loss in children through age 5 years and mailed or faxed directly to the District UNHSI Coordinator within 7 days of confirmatory diagnosis. If the county of residence of the child is unknown, the form should be mailed or faxed to the state UNHSI program office.
Forms may be reproduced or downloaded from the web at http://health.state.ga.us/programs/unhs/.

Form Definitions and Completion Guidelines:
Child Information: Complete all fields and demographic information for the child. If an alternative phone number is known, please list to assist in follow up.

PCP: Please provide the name of the current Primary Care Physician (PCP) for the child, including clinic name and phone number.

Diagnostic Hearing Results: Diagnostic hearing results must contain a threshold search, either by ABR or behavioral. If partial testing completed, which indicates a permanent hearing impairment, report degree of hearing impairment based on threshold testing completed. Please complete the entire diagnostic section and include test results to assist in appropriate follow up and referrals to state programs.

Degree of Hearing Impairment: Degree of hearing impairment is based on a four frequency pure tone average (500, 1000, 2000, and 4000 Hz), if available. If partial testing completed that indicates a permanent hearing impairment, report hearing impairment based on threshold testing completed. Example, if click threshold obtained for both ears at 50 dB with absent OAEs and normal tympanometry; report degree of hearing impairment as Moderate. If type of hearing loss is "Auditory Neuropathy", a degree of hearing impairment does not have to be selected.

Type of Hearing Impairment: Select type of hearing impairment, normal, conductive, transient conductive, mixed, sensorineural, or auditory neuropathy, based on comprehensive audiometric testing.

Recommended Follow Up: Please indicate all necessary follow up, based on comprehensive audiometric findings and child/family needs. If dates of future service known, either appointment date and/or time frame (i.e. 4-6 weeks), please report.

Audiologist: Provide the name of the Audiologist submitter, clinic name, and clinic phone number.

Comments Section: Provide any necessary comments not addressed on the form that are essential for adequate follow up by Public Health.

Georgia Department of Public Health
UNHSI Program
2 Peachtree, NW, 11th Floor
Atlanta, GA 30303
Phone: (404) 657-4143
Fax: (404) 657-2773

Appendix C Parent Refusal Final

Georgia Department of Public Health

Brenda Fitzgerald, MD, Commissioner | Nathan Deal, Governor

2 Peachtree Street NW, 15th Floor
Atlanta, Georgia 30303-3142
www.health.state.ga.us

Georgia Universal Newborn Hearing Screening and Intervention (UNHSI): Hearing Screening Refusal Form

Hearing loss is one of the most common birth defects and most babies who are born with hearing loss are born to hearing parents. It is through hearing that your child will learn to talk. ***Listening in the first few months of life prepares a baby to learn language, and develop speech. The only way to ensure your baby can hear is to have the hearing screen.***

Newborn hearing screening is safe and painless. Most babies sleep through the screening.

I, _____ (Parent/ Legal Guardian) of

_____ (child's name and date of birth),

REFUSE to have my child's hearing screened/rescreened or a diagnostic evaluation.

I understand that there are services to assist with paying for hearing screening/rescreening or a diagnostic evaluation if there is a financial concern.

I have been advised of the importance of having my baby's hearing tested. I have read and fully understand the brochure "Have You Heard". I will contact my physician if I decide to have my baby's hearing tested at a future date.

I release _____ (hospital/midwife/birthing coach/state) of any liability by requesting not to have the screening test done. I accept full responsibility for choosing not to have this test performed.

Underlying Reason for Refusal_____

_____

Signature of Parent/Legal Guardian _____**Date**_____

Original copy is placed in baby's medical record. A copy should be given to parents and a copy should accompany the Children 1st Screening and Referral form, sent to the health district the child resides in.

Tammy Uehlin, Au.D.
UNHSI Program Coordinator
Phone: 404-657-4143
http://www.health.state.ga.us/programs/unhs/index.asp

We Protect Lives.

Appendix D Newborn Hearing Screening Checklist - Hospital Chart

Hospital:

Hearing Screen completed by:

Name:

Date of Birth:

Date of Hearing Screen:

Technology Used: aABR or aOAE

Right Ear Pass/Refer:

Left Ear Pass/Refer:

Not Screened/Reason:

PCP verified:

PCP notified of results:

2 Family Contact numbers:

Have You Heard? brochure provide:

Results given verbally:

Results given in writing:

Results delivered semi-scripted:

***Complete only if infant "refers"last screen:**

- **Follow-up appointment schedule:**
- **Results sent to public health on C1st Screening & Referral Form:**
- **PCP notified of results:**

Appendix E NCHAM Screener Scripts

Passing Script for Babies:
"Congratulations on the birth of your baby. We just finished your baby's hearing screening and your baby passed the screen today. Here's a brochure that talks about development of speech and language. It's always important to monitor the progress of your baby's development, because your baby's hearing can change at any time. If you're 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's skilled at testing infants and young children."

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 normal "on time" development to occur. Your doctor can help you to monitor your baby's hearing development and tell you when your baby should have further tests with an audiologist that's skilled at testing infants and young children."

Not Passing Script:
"Congratulations on the birth of your baby. We just finished screening your baby's hearing and your baby did not pass the screen today. This doesn't necessarily mean that your baby has a permanent hearing loss, but without additional testing we can't be sure. We'll provide the screening results to your baby's doctor. Please be sure you make or keep the appointment for further hearing testing."

Not 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 did not pass the screen today. There can be simple reasons for this, but without further testing with an audiologist I can't tell you what your baby hears. Because your baby has had some medical problems at birth, your baby is at greater risk for hearing loss. Talk to your baby's doctor about the results and ask for help to schedule an audiological diagnostic test as soon as possible. Finding out about hearing issues early is going to help to make sure your baby has the best chance of normal "on time" development."

Incomplete or Missed Results Script:
"Although we attempt to provide newborn hearing screening to all babies born at our hospital, we were unable to complete the screening on your baby. It is important that your baby be screened while he or she is a newborn to identify a possible hearing loss as soon as possible. Let's schedule a time for the screening to be completed within the next week."

Not Passing Outpatient Rescreen:
Your baby did not pass the second screen. The screening does not tell us whether your baby has a hearing loss; it just tells us that further testing should be done as soon as possible. The next step is to get a diagnostic ABR as soon as possible. This should be discussed immediately with your baby's doctor who may need to help you with obtaining a referral to a pediatric audiologist.

Appendix F Hospital Newborn Hearing Screening Log

Hospital Newborn Hearing Screening Log

Hospital:

Hearing Screener:

Name/Hospital Label	Screen Date	Technology Used	Right Ear Result	Left Ear Result	Not Screened (Reason)	Refer at Discharge	Date of Rescreen appointment	Location of Rescreen appointment

Hospital Newborn Hearing Screening Log

Appendix G GA_Hospital_Form_2013

GEORGIA UNIVERSAL NEWBORN HEARING SCREENING & INTERVENTION
2013 Hospital Report Form

Directions: As required under Georgia law please report hearing screening information for all newborns born in your hospital or birthing facility during the specified quarter. All data reported should only be based on actual, non-estimated information. Please see the "Data Definitions" on page 2 for additional information about this form.

Submission: This form can be completed online using the GA SendSS system (*insert URL*) or by emailing the form to (*insert email address*)

Date: _____ Hospital Code: _____ Facility Name: _____

Name/Title of Person Completing Report _____

Contact Phone Number: _____ E-Mail Address: _____

Reporting Schedule: January 1, 2013 (12:00 AM) – December 31, 2013 (11:59 PM)

X	Reporting Period –Year 2012	Due Date	X	Reporting Period –Year 2012	Due Date
	Jan 1 - March 31, 2013	May 1, 2013		July 1 – Sept. 30, 2013	Nov. 1, 2013
	April 1 - June 30, 2013	August 1, 2013		October 1 - Dec 31, 2013	Feb. 1 2014

I. Births In The Current Reporting Period

Quarterly Births	
1) Total # Live Births in the Quarter (*according to State Office of Vital Records*)	*(Populated by the State)*
2) Total # Live Births in the Quarter (*according to hospital/facility*)	
Screened for Hearing Loss^ (***Note:*** *Do not include any newborns not born in the facility, such as transfers)*	
3) Total # of newborns screened for hearing loss	
4) # Newborns who *Passed* the hearing screening	
5) # Newborns who did *Not Pass* the hearing screening	
Not Screened for Hearing Loss (**Note:** *Referral to Children 1st is required for all newborns reported in fields # 8 – 11)*	
6) # Newborns not screened because they died before discharge^	
7) # Newborns in NICU and unable to be screened in current reporting quarter	
8) # Newborns where screening was refused	
9) # Newborns referred to private practice for initial screening	
10) # Newborns transferred out to another hospital that were not screened	
11) # Newborns discharged home *without* screening – *Please list all reasons below*	
Reasons for not screening:	
Quarterly Totals*	

^If a newborn was screened but died before discharge please report this in field #6 and do not include in fields #3, 4, & 5.

II. Previous Quarter NICU Cases *(These cases should not be included in any fields in Section 1)*

NICU	
12) # Newborns in NICU that were unable to be screened in previous reporting quarter	*From Previous Quarter*
13) # Newborns from #12 that were screened and *Passed*	
14) # Newborns from #12 that were screened and did *Not Pass*	

III. Comments

1

Data Definitions

Notes

- The value for the "Quarterly Totals" field is calculated based on adding the numbers reported in field #3, "Screened for Hearing Loss," + the total of the numbers reported as "Not Screened for Hearing Loss" (fields #6, 7, 8, 9, 10, & 11). The total of these fields **must be the same** as the number reported in field #2 ("Total # Live Births in the Quarter").
- A completed hearing screening is defined as the automated screening equipment (either aABR or aOAE) generating a "Pass" or "Refer" result.

I. Births In The Current Reporting Period

Quarterly Births

1) **Total # Live Births in the Quarter (*according to State Vital Records*):** The number of live births occurring in this reporting period for your hospital or facility according to the State Office of Vital Records. This field is included for reference and will be completed by the State. The number of births in this field should be the same as the number of births reported in field #2, "Total # Live Births in the Quarter."

    - If the numbers of births in these fields do not match please verify that the data being reported is complete and accurate before submitting this form.
    - If the data has been verified but the numbers of births still do not match please explain the reason why these numbers do not match in the "Comments" section.

2) **Total # Live Births in the Quarter** (*according to hospital/facility*): The number of live births occurring in reporting period which begins at 12:00 AM of the first day of the quarter and ends at 11:59 PM the last day of the quarter. A live birth is defined as the complete expulsion or extraction of a product of conception from its mother, irrespective of the duration of the pregnancy, which after such separation, breathes or shows any other evidence of life; beating of the heart, pulsation of the umbilical cord, or definite movement of voluntary muscle, whether or not the umbilical cord has been cut or the placenta is attached. GA Code (31-10)

Screened for Hearing Loss

3) **Total # of newborns screened for hearing loss:** The total number of newborns born in the current reporting period and screened for hearing loss prior to discharge from the hospital or birthing facility.

    - **Example:** This applies even if the infant was born on the last day of the reporting quarter and screened on the following day.
    - This should be an unduplicated count of all newborns where each newborn is only counted once, regardless of the number of hearing screening attempts.
    - If a newborn was screened for hearing loss but died before being discharge from the hospital or birthing facility please do not report or include these cases in any of the "Screened for Hearing Loss^" fields (i.e., # 3, 4, and 5). These cases should **ONLY** be reported in field #6 ("# Newborns not screened because they died before discharge").

4) **# Newborns who Passed the hearing screening**: The number of newborns in the reporting quarter whose last/most recent hearing screening was a "Pass."

5) **# Newborns who did Not Pass the hearing screening**: The number of newborns in the reporting quarter whose last screening before discharge was a "Not Pass."

    - Please note that no more than two completed hearing screenings should be performed prior to discharge.

Not Screened for Hearing Loss

6) **# Newborns not screened because they died before discharge:** The number of newborns who were not screened for hearing loss because they died before being discharged from the hospital or birthing facility.

   - If a newborn was screened for hearing loss but died before being discharge from the hospital please do not report or include these cases in any of the "Screened for Hearing Loss^" fields (i.e., # 3, 4, and 5). These cases should **ONLY** be reported in this field.

7) **# Newborns in NICU and unable to be screened in current reporting quarter:** The number of newborns who were unable to be screened for hearing loss in this current reporting quarter because they were in the NICU.

8) **# Newborns where screening was refused:** The number of newborns who are not screened for hearing loss because the newborn's parent or guardian refused the screening. Documentation of the refusal should be included in the newborn's medical record.

9) **# Newborns referred to private practice for initial screening:** The number of newborns that were not screened for hearing loss because they were referred to a private pediatrician or Otolaryngologist (ENT) for their initial hearing screening.

   - This field is different and separate from a parent refusing the hearing screening and only applies to those newborns that were not screened for hearing loss because they were referred to a private practice.
   - **Note:** A referral to Children 1st is required for all newborns reported in this field.

10) **# Newborns transferred to another hospital that were not screened:** The number of newborns transferred to another hospital or facility before a hearing screening was completed,

   - **Note:** The birth hospital or facility is responsible for notifying the receiving hospital that a hearing screening was not completed.
   - **Note:** A referral to Children 1st is required for all newborns reported in this field.

11) **# Newborns discharged *without* screening:** The number of newborns born in the reporting quarter that were not screened for hearing loss for any reason other than those stated in fields # 6, 7, 8, 9, and 10.

   - The reasons newborns reported in this field were not screened should be included in the space immediately below this field.
   - This should not include cases of newborns that were transferred out of the hospital or birthing facility.
   - **Note:** A referral to Children 1st is required for all newborns reported in this field.

II. Previous Quarter NICU Cases

NICU

12) **# Newborns in NICU that were unable to be screened in previous reporting quarter:** The number reported in field #7 in the report for the previous quarter. This represents the number of newborns in the NICU that were unable to be screened for hearing loss in the previous quarter.

   - The sum of fields #13 and #14 must not be larger than the value reported in field #12.

13) # Newborns from #12 that were screened and *Passed*: The number of newborns reported in the previous quarter that were unable to be screened because they were in the NICU that have now been screened for hearing loss in the current reporting quarter and passed.

14) # Newborns from #12 that were screened and *Not Passed:* The number of newborns reported in the previous quarter that were unable to be screened because they were in the NICU that have now been screened for hearing loss in the current reporting quarter and did not pass.

Appendix H JCIH 2007

PEDIATRICS®
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Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs

Joint Committee on Infant Hearing

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American Academy of Pediatrics
DEDICATED TO THE HEALTH OF ALL CHILDREN™

POLICY STATEMENT

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

Joint Committee on Infant Hearing

THE POSITION STATEMENT

The Joint Committee on Infant Hearing (JCIH) endorses early detection of and intervention for infants with hearing loss. 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.[1] 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 health care 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.[2] EHDI systems should guarantee seamless transitions for infants and their families through this process.

2007 JCIH POSITION STATEMENT UPDATES

The following are highlights of updates made since the 2000 JCIH statement[3]:

1. Definition of targeted hearing loss
   - The definition has been expanded from congenital permanent bilateral, unilateral sensory, or permanent conductive hearing loss to include neural hearing loss (eg, "auditory neuropathy/dyssynchrony") in infants admitted to the NICU.
2. Hearing-screening and -rescreening protocols
   - Separate protocols are recommended for NICU and well-infant nurseries. NICU infants admitted for more than 5 days are to have auditory brainstem response (ABR) included as part of their screening so that neural hearing loss will not be missed.
   - For infants who do not pass automated ABR testing in the NICU, referral should be made directly to an audiologist for rescreening and, when indicated, comprehensive evaluation including ABR.
   - For rescreening, a complete screening on both ears is recommended, even if only 1 ear failed the initial screening.
   - For readmissions in the first month of life for all infants (NICU or well infant), when there are conditions associated with potential hearing loss (eg, hyper-

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All policy statements from the American Academy of Pediatrics automatically expire 5 years after publication unless reaffirmed, revised, or retired at or before that time.

Key Word
hearing screening

Abbreviations
JCIH—Joint Committee on Infant Hearing
EHDI—early hearing detection and intervention
ABR—auditory brainstem response
CMV—cytomegalovirus
ECMO—extracorporeal membrane oxygenation
AAP—American Academy of Pediatrics
MCHB—Maternal and Child Health Bureau
HRSA—Health Resources and Services Administration
NIDCD—National Institute on Deafness and Other Communication Disorders
CDC—Centers for Disease Control and Prevention
UNHS—universal newborn hearing screening
OAE—otoacoustic emission
IFSP—individualized family service plan
OME—otitis media with effusion
FM—frequency modulation
DSHPSHWA—Directors of Speech and Hearing Programs in State Health and Welfare Agencies
GPRA—Government Performance and Results Act
OMB—Office of Management and Budgets

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bilirubinemia that requires exchange transfusion or culture-positive sepsis), a repeat hearing screening is recommended before discharge.

3. Diagnostic audiology evaluation
   - Audiologists with skills and expertise in evaluating newborn and young infants with hearing loss should provide audiology diagnostic and auditory habilitation services (selection and fitting of amplification device).
   - At least 1 ABR test is recommended as part of a complete audiology diagnostic evaluation for children younger than 3 years for confirmation of permanent hearing loss.
   - The timing and number of hearing reevaluations for children with risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss. 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 extracorporeal membrane oxygenation (ECMO) or chemotherapy; and when there is caregiver concern or a family history of hearing loss.
   - For families who elect amplification, infants in whom permanent hearing loss is diagnosed should be fitted with an amplification device within 1 month of diagnosis.
4. Medical evaluation
   - For infants with confirmed hearing loss, a genetics consultation should be offered to their families.
   - Every infant with confirmed hearing loss should be evaluated by an otolaryngologist who has knowledge of pediatric hearing loss and have at least 1 examination to assess visual acuity by an ophthalmologist who is experienced in evaluating infants.
   - The risk factors for congenital and acquired hearing loss have been combined in a single list rather than grouped by time of onset.
5. Early intervention
   - All families of infants with any degree of bilateral or unilateral permanent hearing loss should be considered eligible for early intervention services.
   - There should be recognized central referral points of entry that ensure specialty services for infants with confirmed hearing loss.
   - Early intervention services for infants with confirmed hearing loss should be provided by professionals who have expertise in hearing loss, including educators of the deaf, speech-language pathologists, and audiologists.
   - In response to a previous emphasis on "natural environments," the JCIH recommends that both home-based and center-based intervention options be offered.
6. Surveillance and screening in the medical home
   - For all infants, regular surveillance of developmental milestones, auditory skills, parental concerns, and middle-ear status should be performed in the medical home, consistent with the American Academy of Pediatrics (AAP) pediatric periodicity schedule. All infants should have an objective standardized screening of global development with a validated assessment tool at 9, 18, and 24 to 30 months of age or at any time if the health care professional or family has concern.
   - Infants who do not pass the speech-language portion of a medical home global screening or for whom there is a concern regarding hearing or language should be referred for speech-language evaluation and audiology assessment.
7. Communication
   - The birth hospital, in collaboration with the state EHDI coordinator, should ensure that the hearing-screening results are conveyed to the parents and the medical home.
   - Parents should be provided with appropriate follow-up and resource information, and hospitals should ensure that each infant is linked to a medical home.
   - Information at all stages of the EHDI process is to be communicated to the family in a culturally sensitive and understandable format.
   - Individual hearing-screening information and audiology diagnostic and habilitation information should be promptly transmitted to the medical home and the state EHDI coordinator.
   - Families should be made aware of all communication options and available hearing technologies (presented in an unbiased manner). Informed family choice and desired outcome guide the decision-making process.
8. Information infrastructure
   - States should implement data-management and -tracking systems as part of an integrated child health information system to monitor the quality of EHDI services and provide recommendations for improving systems of care.

- An effective link between health and education professionals is needed to ensure successful transition and to determine outcomes of children with hearing loss for planning and establishing public health policy.

BACKGROUND

It has long been recognized that unidentified hearing loss at birth can adversely affect speech and language development as well as academic achievement and social-emotional development. Historically, moderate-to-severe hearing loss in young children was not detected until well beyond the newborn period, and it was not unusual for diagnosis of milder hearing loss and unilateral hearing loss to be delayed until children reached school age.

In the late 1980s, Dr C. Everett Koop, then US Surgeon General, on learning of new technology, encouraged detection of hearing loss to be included in the *Healthy People 2000*[4] goals for the nation. In 1988, the Maternal and Child Health Bureau (MCHB), a division of the US Health Resources and Services Administration (HRSA), funded pilot projects in Rhode Island, Utah, and Hawaii to test the feasibility of a universal statewide screening program to screen newborn infants for hearing loss before hospital discharge. The National Institutes of Health, through the National Institute on Deafness and Other Communication Disorders (NIDCD), issued in 1993 a consensus statement on early identification of hearing impairment in infants and young children.[5] In the statement the authors concluded that all infants admitted to the NICU should be screened for hearing loss before hospital discharge and that universal screening should be implemented for all infants within the first 3 months of life.[4] In its 1994 position statement, the JCIH endorsed the goal of universal detection of infants with hearing loss and encouraged continuing research and development to improve methods for identification of and intervention for hearing loss.[6,7] The AAP released a statement that recommended newborn hearing screening and intervention in 1999.[8] In 2000, citing advances in screening technology, the JCIH endorsed the universal screening of all infants through an integrated, interdisciplinary system of EHDI.[3] The *Healthy People 2010* goals included an objective to "increase the proportion of newborns who are screened for hearing loss by one month, have audiological evaluation by 3 months, and are enrolled in appropriate intervention services by 6 months."[9]

The ensuing years have seen remarkable expansion in newborn hearing screening. At the time of the National Institutes of Health consensus statement, only 11 hospitals in the United States were screening more than 90% of their newborn infants. In 2000, through the support of Representative Jim Walsh (R-NY), Congress authorized the HRSA to develop newborn hearing screening and follow-up services, the Centers for Disease Control and Prevention (CDC) to develop data and tracking systems, and the NIDCD to support research in EHDI. By 2005, every state had implemented a newborn hearing-screening program, and approximately 95% of newborn infants in the United States were screened for hearing loss before hospital discharge. Congress recommended cooperation and collaboration among several federal agencies and advocacy organizations to facilitate and support the development of state EHDI systems.

EHDI programs throughout the United States have demonstrated not only the feasibility of universal newborn hearing screening (UNHS) but also the benefits of early identification and intervention. There is a growing body of literature indicating that when identification and intervention occur at no later than 6 months of age for newborn infants who are deaf or hard of hearing, the infants perform as much as 20 to 40 percentile points higher on school-related measures (vocabulary, articulation, intelligibility, social adjustment, and behavior).[10–13] Still, many important challenges remain. Despite the fact that approximately 95% of newborn infants have their hearing screened in the United States, almost half of newborn infants who do not pass the initial screening do not have appropriate follow-up to either confirm the presence of a hearing loss and/or initiate appropriate early intervention services (see www.infanthearing.org, www.cdc.gov/ncbddd/ehdi, and www.nidcd.nih.gov/health).

State EHDI coordinators report system-wide problems including failure to communicate information to families in a culturally sensitive and understandable format at all stages of the EHDI process, lack of integrated state data-management and -tracking systems, and a shortage of facilities and personnel with the experience and expertise needed to provide follow-up for infants who are referred from newborn screening programs.[14] Available data indicate that a significant number of children who need further assessment do not receive appropriate follow-up evaluations. However, the outlook is improving as EHDI programs focus on the importance of strengthening follow-up and intervention.

PRINCIPLES

All children with hearing loss should have access to resources necessary to reach their maximum potential. The following principles provide the foundation for effective EHDI systems and have been updated and expanded since the 2000 JCIH position statement.

1. All infants should have access to hearing screening using a physiologic measure at no later than 1 month of age.
2. All infants who do not pass the initial hearing screening and the subsequent rescreening should have appropriate audiological and medical evaluations to

confirm the presence of hearing loss at no later than 3 months of age.

3. All infants with confirmed permanent hearing loss should receive early intervention services as soon as possible after diagnosis but at no later than 6 months of age. A simplified, single point of entry into an intervention system that is appropriate for children with hearing loss is optimal.
4. The EHDI system should be family centered with infant and family rights and privacy guaranteed through informed choice, shared decision-making, and parental consent in accordance with state and federal guidelines. Families should have access to information about all intervention and treatment options and counseling regarding hearing loss.
5. The child and family should have immediate access to high-quality technology including hearing aids, cochlear implants, and other assistive devices when appropriate.
6. All infants and children should be monitored for hearing loss in the medical home.[15] Continued assessment of communication development should be provided by appropriate professionals to all children with or without risk indicators for hearing loss.
7. Appropriate interdisciplinary intervention programs for infants with hearing loss and their families should be provided by professionals who are knowledgeable about childhood hearing loss. Intervention programs should recognize and build on strengths, informed choices, traditions, and cultural beliefs of the families.
8. Information systems should be designed and implemented to interface with electronic health charts and should be used to measure outcomes and report the effectiveness of EHDI services at the patient, practice, community, state, and federal levels.

GUIDELINES FOR EHDI PROGRAMS

The 2007 guidelines were developed to update the 2000 JCIH position statement principles and to support the goals of universal access to hearing screening, evaluation, and intervention for newborn and young infants embodied in *Healthy People 2010*.[9] The guidelines provide current information on the development and implementation of successful EHDI systems.

Hearing screening should identify infants with specifically defined hearing loss on the basis of investigations of long-term, developmental consequences of hearing loss in infants, currently available physiologic screening techniques, and availability of effective intervention in concert with established principles of health screening.[15–18] Studies have demonstrated that current screening technologies are effective in identifying hearing loss of moderate and greater degree.[19] In addition, studies of children with permanent hearing loss indicate that moderate or greater degrees of hearing loss can have significant effects on language, speech, academic, and social-emotional development.[20] High-risk target populations also include infants in the NICU, because research data have indicated that this population is at highest risk of having neural hearing loss.[21–23]

The JCIH, however, is committed to the goal of identifying all degrees and types of hearing loss in childhood and recognizes the developmental consequences of even mild degrees of permanent hearing loss. Recent evidence, however, has suggested that current hearing-screening technologies fail to identify some infants with mild forms of hearing loss.[24,25] In addition, depending on the screening technology selected, infants with hearing loss related to neural conduction disorders or "auditory neuropathy/auditory dyssynchrony" may not be detected through a UNHS program. Although the JCIH recognizes that these disorders may result in delayed communication,[26–28] currently recommended screening algorithms (ie, use of otoacoustic emission [OAE] testing alone) preclude universal screening for these disorders. Because these disorders typically occur in children who require NICU care,[21] the JCIH recommends screening this group with the technology capable of detecting auditory neuropathy/dyssynchrony: automated ABR measurement.

All infants, regardless of newborn hearing-screening outcome, should receive ongoing monitoring for development of age-appropriate auditory behaviors and communication skills. Any infant who demonstrates delayed auditory and/or communication skills development, even if he or she passed newborn hearing screening, should receive an audiological evaluation to rule out hearing loss.

Roles and Responsibilities

The success of EHDI programs depends on families working in partnership with professionals as a well-coordinated team. The roles and responsibilities of each team member should be well defined and clearly understood. Essential team members are the birth hospital, families, pediatricians or primary health care professionals (ie, the medical home), audiologists, otolaryngologists, speech-language pathologists, educators of children who are deaf or hard of hearing, and other early intervention professionals involved in delivering EHDI services.[29,30] Additional services including genetics, ophthalmology, developmental pediatrics, service coordination, supportive family education, and counseling should be available.[31]

The birth hospital is a key member of the team. The birth hospital, in collaboration with the state EHDI coordinator, should ensure that parents and primary health care 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.[2] The hospital ensures that hearing-screening information is transmitted promptly to the medical home and appropriate data are submitted to the state EHDI coordinator.

The most important role for the family of an infant who is deaf or hard of hearing is to love, nurture, and communicate with the infant. From this foundation, families usually develop an urgent desire to understand and meet the special needs of their infant. Families gain knowledge, insight, and experience by accessing resources and through participation in scheduled early intervention appointments including audiological, medical, habilitative, and educational sessions. This experience can be enhanced when families choose to become involved with parental support groups, people who are deaf or hard of hearing, and/or their children's deaf or hard-of-hearing peers. Informed family choices and desired outcomes guide all decisions for these children. A vital function of the family's role is ensuring direct access to communication in the home and the daily provision of language-learning opportunities. Over time, the child benefits from the family's modeling of partnerships with professionals and advocating for their rights in all settings. The transfer of responsibilities from families to the child develops gradually and increases as the child matures, growing in independence and self-advocacy.

Pediatricians, family physicians, and other allied health care professionals, working in partnership with parents and other professionals such as audiologists, therapists, and educators, constitute the infant's medical home.[2] A medical home is defined as an approach to providing health care services with which care is accessible, family centered, continuous, comprehensive, coordinated, compassionate, and culturally competent. The primary health care professional acts in partnership with parents in a medical home to identify and access appropriate audiology, intervention, and consultative services that are needed to develop a global plan of appropriate and necessary health and habilitative care for infants identified with hearing loss and infants with risk factors for hearing loss. All children undergo surveillance for auditory skills and language milestones. The infant's pediatrician, family physician, or other primary health care professional is in a position to advocate for the child and family.[2,16]

An audiologist is a person who, by virtue of academic degree, clinical training, and license to practice, is qualified to provide services related to the prevention of hearing loss and the audiological diagnosis, identification, assessment, and nonmedical and nonsurgical treatment of persons with impairment of auditory and vestibular function, and to the prevention of impairments associated with them. Audiologists serve in a number of roles. They provide newborn hearing-screening program development, management, quality assessment, service coordination and referral for audiological diagnosis, and audiological treatment and management. For the follow-up component, audiologists provide comprehensive audiological diagnostic assessment to confirm the existence of the hearing loss, ensure that parents understand the significance of the hearing loss, evaluate the infant for candidacy for amplification and other sensory devices and assistive technology, and ensure prompt referral to early intervention programs. For the treatment and management component, audiologists provide timely fitting and monitoring of amplification devices.[32] Other audiologists may provide diagnostic and auditory treatment and management services in the educational setting and provide a bridge between the child/family and the audiologist in the clinic setting as well as other service providers. Audiologists also provide services as teachers, consultants, researchers, and administrators.

Otolaryngologists are physicians whose specialty includes determining the etiology of hearing loss; identifying related risk indicators for hearing loss, including syndromes that involve the head and neck; and evaluating and treating ear diseases. An otolaryngologist with knowledge of childhood hearing loss can determine if medical and/or surgical intervention may be appropriate. When medical and/or surgical intervention is provided, the otolaryngologist is involved in the long-term monitoring and follow-up with the infant's medical home. The otolaryngologist provides information and participates in the assessment of candidacy for amplification, assistive devices, and surgical intervention, including reconstruction, bone-anchored hearing aids, and cochlear implantation.

Early intervention professionals are trained in a variety of academic disciplines such as speech-language pathology, audiology, education of children who are deaf or hard of hearing, service coordination, or early childhood special education. All individuals who provide services to infants with hearing loss should have specialized training and expertise in the development of audition, speech, and language. Speech-language pathologists provide both evaluation and intervention services for language, speech, and cognitive-communication development. Educators of children who are deaf or hard of hearing integrate the development of communicative competence within a variety of social, linguistic, and cognitive/academic contexts. Audiologists may provide diagnostic and habilitative services within the individualized family service plan (IFSP) or school-based individualized education plan. To provide the highest quality of intervention, more than 1 provider may be required.

The care coordinator is an integral member of the EHDI team and facilitates the family's transition from screening to evaluation to early intervention.[33] This person must be a professional (eg, social worker, teacher, nurse) who is knowledgeable about hearing loss. The care coordinator incorporates the family's preferences for outcomes into an IFSP as required by federal legisla-

tion. The care coordinator supports the family members in their choice of the infant's communicative development. Through the IFSP review, the infant's progress in language, motor, cognitive, and social-emotional development is monitored. The care coordinator assists the family in advocating for the infant's unique developmental needs.

The deaf and hard-of-hearing community includes members with direct experience with signed language, spoken language, hearing-aid and cochlear implant use, and other communication strategies and technologies. Optimally, adults who are deaf or hard-of-hearing should play an integral part in the EHDI program. Both adults and children in the deaf and hard-of-hearing community can enrich the family's experience by serving as mentors and role models. Such mentors have experience in negotiating their way in a hearing world, raising infants or children who are deaf or hard of hearing, and providing families with a full range of information about communication options, assistive technology, and resources that are available in the community.

A successful EHDI program requires collaboration between a variety of public and private institutions and agencies that assume responsibility for specific components (eg, screening, evaluation, intervention). Roles and responsibilities may differ from state to state. Each state has defined a lead coordinating agency with oversight responsibility. The lead coordinating agency in each state should be responsible for identifying the public and private funding sources available to develop, implement, and coordinate EHDI systems.

Hearing Screening

Multidisciplinary teams of professionals, including audiologists, physicians, and nursing personnel, are needed to establish the UNHS component of EHDI programs. All team members work together to ensure that screening programs are of high quality and are successful. An audiologist should be involved in each component of the hearing-screening program, particularly at the level of statewide implementation and, whenever possible, at the individual hospital level. Hospitals and agencies should also designate a physician to oversee the medical aspects of the EHDI program.

Each team of professionals responsible for the hospital-based UNHS program should review the hospital infrastructure in relationship to the screening program. Hospital-based programs should consider screening technology (ie, OAE or automated ABR testing); validity of the specific screening device; screening protocols, including the timing of screening relative to nursery discharge; availability of qualified screening personnel; suitability of the acoustical and electrical environments; follow-up referral criteria; referral pathways for follow-up; information management; and quality control and improvement. Reporting and communication protocols must be well defined and include the content of reports to physicians and parents, documentation of results in medical charts, and methods for reporting to state registries and national data sets.

Physiologic measures must be used to screen newborns and infants for hearing loss. Such measures include OAE and automated ABR testing. Both OAE and automated 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 for UNHS.[19,34–37] However, there are important differences between the 2 measures. OAE measurements are obtained from the ear canal by using a sensitive microphone within a probe assembly that records cochlear responses to acoustic stimuli. Thus, OAEs reflect the status of the peripheral auditory system extending to the cochlear outer hair cells. In contrast, ABR measurements are obtained from surface electrodes that record neural activity generated in the cochlea, auditory nerve, and brainstem in response to acoustic stimuli delivered via an earphone. Automated ABR measurements reflect the status of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway.

Both OAE and ABR screening technologies can be used to detect sensory (cochlear) hearing loss[19]; 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 "failed" screening-test result in the presence of normal cochlear and/or neural function.[38] Moreover, because OAEs are generated within the cochlea, OAE technology cannot be used to detect neural (eighth nerve or auditory brainstem pathway) dysfunction. Thus, neural conduction disorders or auditory neuropathy/dyssynchrony without concomitant sensory dysfunction will not be detected by OAE testing.

Some infants who pass newborn hearing screening will later demonstrate permanent hearing loss.[25] Although this loss may reflect delayed-onset hearing loss, both ABR and OAE screening technologies will miss some hearing loss (eg, mild or isolated frequency region losses).

Interpretive criteria for pass/fail outcomes should reflect clear scientific rationale and should be evidence based.[39,40] Screening technologies that incorporate automated-response detection are necessary to eliminate the need for individual test interpretation, to reduce the effects of screener bias or operator error on test outcome, and to ensure test consistency across infants, test conditions, and screening personnel.[41–45] When statistical probability is used to make pass/fail decisions, as is the case for OAE and automated ABR screening devices, the likelihood of obtaining a pass outcome by chance alone is increased when screening is performed repeatedly.[46–48]

This principle must be incorporated into the policies of rescreening.

There are no national standards for the calibration of OAE or ABR instrumentation. Compounding this problem, there is a lack of uniform performance standards. Manufacturers of hearing-screening devices do not always provide sufficient supporting evidence to validate the specific pass/fail criteria and/or automated algorithms used in their instruments.[49] In the absence of national standards, audiologists must obtain normative data for the instruments and protocols they use.

The JCIH recognizes that there are important issues differentiating screening performed in the well-infant nursery from that performed in the NICU. Although the goals in each nursery are the same, numerous methodologic and technological issues must be considered in program design and pass/fail criteria.

Screening Protocols in the Well-Infant Nursery

Many inpatient well-infant screening protocols provide 1 hearing screening and, when necessary, a repeat screening no later than at the time of discharge from the hospital, using the same technology both times. Use of either technology in the well-infant nursery will detect peripheral (conductive and sensory) hearing loss of 40 dB or greater.[19] When automated ABR is used as the single screening technology, neural auditory disorders can also be detected.[50] Some programs use a combination of screening technologies (OAE testing for the initial screening followed by automated ABR for rescreening [ie, 2-step protocol[5]]) to decrease the fail rate at discharge and the subsequent need for outpatient follow-up.[34,35,37,51–53] With this approach, infants who do not pass an OAE screening but subsequently pass an automated ABR test are considered a screening "pass." Infants in the well-infant nursery who fail automated ABR testing should not be rescreened by OAE testing and "passed," because such infants are presumed to be at risk of having a subsequent diagnosis of auditory neuropathy/dyssynchrony.

Screening Protocols in the NICU

An NICU is defined as a facility in which a neonatologist provides primary care for the infant. Newborn units are divided into 3 categories:

- Level I: basic care, well-infant 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)

A total of 120 level-II NICUs and 760 level-III NICUs have been identified in the United States by survey, and infants who have spent time in the NICU represent 10% to 15% of the newborn population.[54]

The 2007 JCIH position statement includes neonates at risk of having neural hearing loss (auditory neuropathy/auditory dyssynchrony) in the target population to be identified in the NICU,[55–57] because there is evidence that neural hearing loss results in adverse communication outcomes.[22,50] Consequently, the JCIH recommends ABR technology as the only appropriate screening technique for use in the NICU. For infants who do not pass automated ABR testing in the NICU, referral should be made directly to an audiologist for rescreening and, when indicated, comprehensive evaluation, including diagnostic ABR testing, rather than for general outpatient rescreening.

Conveying Test Results

Screening results should be conveyed immediately to families so that they understand the outcome and the importance of follow-up when indicated. To facilitate this process for families, primary health care professionals should work with EHDI team members to ensure that:

- communications with parents are confidential and presented in a caring and sensitive manner, preferably face-to-face;
- educational materials are developed and disseminated to families that provide accurate information at an appropriate reading level and in a language they are able to comprehend; and
- parents are informed in a culturally sensitive and understandable manner that their infant did not pass screening and informed about the importance of prompt follow-up; before discharge, an appointment should be made for follow-up testing.

To facilitate this process for primary care physicians, EHDI systems should ensure that medical professionals receive:

- the results of the screening test (pass, did not pass, or missed) as documented in the hospital medical chart; and
- communication directly from a representative of the hospital screening program regarding each infant in its care who did not pass or was missed and recommendations for follow-up.

Outpatient Rescreening for Infants Who Do Not Pass the Birth Admission Screening

Many well-infant screening protocols will incorporate an outpatient rescreening within 1 month of hospital discharge to minimize the number of infants referred for follow-up audiological and medical evaluation. The out-

patient rescreening should include the testing of both ears, even if only 1 ear failed the inpatient screening.

Outpatient screening at no later than 1 month of age should also be available to infants who were discharged before receiving the birth admission screening or who were born outside a hospital or birthing center. State EHDI coordinators should be aware of some of the following situations under which infants may be lost to the UNHS system:

- Home births and other out-of-hospital births: states should develop a mechanism to systematically offer newborn hearing screening for all out-of-hospital births.
- Across-state-border births: states should develop written collaborative agreements among neighboring states for sharing hearing-screening results and follow-up information.
- Hospital-missed screenings: when infants are discharged before the hearing screening is performed, a mechanism should be in place for the hospital to contact the family and arrange for an outpatient hearing screening.
- Transfers to in-state or out-of-state hospitals: discharge and transfer forms should contain the information of whether a hearing screening was performed and the results of any screening. The recipient hospital should complete a hearing screening if one was not previously performed or if there is a change in medical status or a prolonged hospitalization.
- Readmissions: for 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.

Additional mechanisms for states to share hearing-screening results and other medical information include (1) incorporating the hearing-screening results in a statewide child health information system and (2) providing combined metabolic screening and hearing-screening results to the primary care physician.

Confirmation of Hearing Loss in Infants Referred From UNHS

Infants who meet the defined criteria for referral should receive follow-up audiological and medical evaluations with fitting of amplification devices, as appropriate, at no later than 3 months of age. Once hearing loss is confirmed, coordination of services should be expedited by the infant's medical home and Part C coordinating agencies for early intervention services, as authorized by the Individuals With Disabilities Education Act, following the EHDI algorithm developed by the AAP (Appendix 1).

Audiological Evaluation

Comprehensive audiological evaluation of newborn and young infants who fail newborn hearing screening should be performed by audiologists experienced in pediatric hearing assessment. The initial audiological test battery to confirm a hearing loss in infants must include physiologic measures and, when developmentally appropriate, behavioral methods. Confirmation of an infant's hearing status requires a test battery of audiological test procedures to assess the integrity of the auditory system in each ear, to estimate hearing sensitivity across the speech frequency range, to determine the type of hearing loss, to establish a baseline for further monitoring, and to provide information needed to initiate amplification-device fitting. A comprehensive assessment should be performed on both ears even if only 1 ear failed the screening test.

Evaluation: Birth to 6 Months of Age

For infants from birth to a developmental age of approximately 6 months, the test battery should include a child and family history, an evaluation of risk factors for congenital hearing loss, and a parental report of the infant's responses to sound. The audiological assessment should include:

- Child and family history.
- A frequency-specific assessment of the ABR using air-conducted tone bursts and bone-conducted tone bursts when indicated. When permanent hearing loss is detected, frequency-specific ABR testing is needed to determine the degree and configuration of hearing loss in each ear for fitting of amplification devices.
- Click-evoked ABR testing using both condensation and rarefaction single-polarity stimulus, if there are risk indicators for neural hearing loss (auditory neuropathy/auditory dyssynchrony) such as hyperbilirubinemia or anoxia, to determine if a cochlear microphonic is present.[28] Furthermore, 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.[55]
- Distortion product or transient evoked OAEs.
- Tympanometry using a 1000-Hz probe tone.
- Clinician observation of the infant's auditory behavior as a cross-check 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.

Evaluation: 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.

Other Audiological Test Procedures

At this time, there is insufficient evidence for use of the auditory steady-state response as the sole measure of auditory status in newborn and infant populations.[58] Auditory steady-state response is a new evoked-potential test that can accurately measure auditory sensitivity beyond the limits of other test methods. It can determine frequency-specific thresholds from 250 Hz to 8 kHz. Clinical research is being performed to investigate its potential use in the standard pediatric diagnostic test battery. Similarly, there are insufficient data for routine use of acoustic middle-ear muscle reflexes in the initial diagnostic assessment of infants younger than 4 months.[59] Both tests could be used to supplement the battery or could be included at older ages. Emerging technologies, such as broad-band reflectance, may be used to supplement conventional measures of middle-ear status (tympanometry and acoustic reflexes) as the technology becomes more widely available.[59]

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, and 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.[60–62]

Pediatrician/Primary Care Physician

The infant's pediatrician or other primary health care professional is responsible for monitoring the general health, development, and well-being of the infant. In addition, the primary care physician must assume responsibility to ensure that the audiological assessment is conducted on infants who do not pass screening and must initiate referrals for medical specialty evaluations necessary to determine the etiology of the hearing loss. Middle-ear status should be monitored, because the presence of middle-ear effusion can further compromise hearing. The primary care physician must partner with other specialists, including the otolaryngologist, 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.[63] The medical home algorithm for management of infants with either suspected or proven permanent hearing loss is provided in Appendix 1.[15]

The pediatrician or primary care physician should 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 should ensure that an audiological evaluation is completed for children at risk of hearing loss at least once by 24 to 30 months of age, regardless of their newborn screening results.[25] Infants with specific risk factors, such as those who received ECMO therapy and those with CMV infection, are at increased risk of delayed-onset or progressive hearing loss[64–67] and should be monitored closely. In addition, the primary care physician is responsible for ongoing surveillance of parent concerns about language and hearing, auditory skills, and developmental milestones of all infants and children regardless of risk status, as outlined in the pediatric periodicity schedule published by the AAP.[16]

Children with cochlear implants may be at increased risk of acquiring bacterial meningitis compared with children in the general US population.[68] The CDC recommends that all children with, and all potential recipients of, cochlear implants follow specific recommendations for pneumococcal immunization that apply to cochlear implant users and that they receive age-appropriate *Haemophilus influenzae* type b vaccines. Recommendations for the timing and type of pneumococcal vaccine vary with age and immunization history and should be discussed with a health care professional.[69]

Otolaryngologist

Otolaryngologists are physicians and surgeons 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. Otolaryngologists work closely with other dedicated professionals, including physicians, audiologists, speech-language pathologists, educators, and others, in caring for patients with hearing, balance, voice, speech, developmental, and related disorders.

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 having received ECMO (see Appendix 2).[70,71]

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.[72] 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.[72,73] 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[72,74–76] (Appendix 2). As the widespread use of newly developed conjugate vaccines decreases the prevalence of infectious etiologies such as measles, mumps, rubella, *H 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.[77]

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. 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.[78]

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.[70,71,79]

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, however, may remain uncertain in as many as 30% to 40% of children. Once hearing loss is confirmed, medical clearance for hearing aids and initiation of early intervention should not be delayed while this diagnostic evaluation is in process. Careful longitudinal monitoring to detect and promptly treat coexisting middle-ear effusions is an essential component of ongoing otologic management of these children.

Other Medical Specialists

The medical geneticist is responsible for the interpretation of family history data, the clinical evaluation and diagnosis of inherited disorders, the performance and assessment of genetic tests, and the provision of genetic counseling. Geneticists or genetic counselors are qualified to interpret the significance and limitations of new tests and to convey the current status of knowledge during genetic counseling. All families of children with confirmed hearing loss should be offered, and may benefit from, a genetics evaluation and counseling. This evaluation can provide families with information on etiology of hearing loss, prognosis for progression, associated disorders (eg, renal, vision, cardiac), and likelihood of recurrence in future offspring. This information may influence parents' decision-making regarding intervention options for their child.

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.[1,80] 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.[81] 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 intervention have significantly better outcomes than later-identified infants and children in vocabulary development,[82,83] receptive and expressive language,[12,84] syntax,[85] speech production,[13,86–88] and social-emotional development.[89] Children enrolled in early intervention within the first year of life have also been shown to have language development within the normal range of development at 5 years of age.[31,90]

Therefore, according to federal guidelines, once any degree of hearing loss is diagnosed in a child, a referral should be initiated to an early intervention program within 2 days of confirmation of hearing loss (CFR 303.321d). The initiation of early intervention services should begin as soon as possible after diagnosis of hearing loss but at no later than 6 months of age. Even when the hearing status is not determined to be the primary disability, the family and child should have access to intervention with a provider who is knowledgeable about hearing loss.[91]

UNHS programs have been instituted throughout the United States for the purpose of preventing the significant and negative effects of hearing loss on the cognitive, language, speech, auditory, social-emotional, and academic development of infants and children. To achieve this goal, hearing loss must be identified as quickly as possible after birth, and appropriate early intervention must be available to all families and infants with permanent hearing loss. Some programs have demonstrated that most children with hearing loss and no additional disabilities can achieve and maintain language development within the typical range of children who have normal hearing.[12,13,85,90] Because these studies were descriptive and not causal studies, the efficacy of specific components of intervention cannot be separated from the total provision of comprehensive services. Thus, the family-centered philosophy, the intensity of services, the experience and training of the provider, the method of communication, the curricula, the counseling procedures, the parent support and advocacy, and the deaf and hard-of-hearing support and advocacy are all variables with unknown effects on the overall outcomes of any individual child. The key component of providing quality services is the expertise of the provider specific to hearing loss. These services may be provided in the home, a center, or a combination of the 2 locations.

The term "intervention services" is used to describe any type of habilitative, rehabilitative, or educational program provided to children with hearing loss. In some cases of mild hearing losses, amplification technology may be the only service provided. Some parents choose only developmental assessment or occasional consultation, such as parents with infants who have unilateral hearing losses. Children with high-frequency losses and normal hearing in the low frequencies may only be seen by a speech-language pathologist, and those with significant bilateral sensorineural hearing losses might be seen by an educator of the deaf and receive additional services.

Principles of Early Intervention

To ensure informed decision-making, parents of infants with newly diagnosed hearing loss should be offered opportunities to interact with other families who have infants or children with hearing loss as well as adults and children who are deaf or hard of hearing. In addition, parents should also be offered access to professional, educational, and consumer organizations and provided with general information on child development, language development, and hearing loss. A number of principles and guidelines have been developed that offer a framework for quality early intervention service delivery systems for children who are deaf or hard of hearing and their families.[92] Foundational characteristics of developing and implementing early intervention programs include a family-centered approach, culturally responsive practices, collaborative professional-family relationships and strong family involvement, developmentally appropriate practice, interdisciplinary assessment, and community-based provision of services.

Designated Point of Entry

States should develop a single point of entry into intervention specific for hearing impairment to ensure that, regardless of geographic location, all families who have infants or children with hearing loss receive information about a full range of options regarding amplification and technology, communication and intervention, and accessing appropriate counseling services. This state system, if separate from the state's Part C system, should integrate and partner with the state's Part C program. Parental consent must be obtained according to state and federal requirements to share the IFSP information with providers and transmit data to the state EHDI coordinator.

Regular Developmental Assessment

To ensure accountability, individual, community, and state health and educational programs should assume the responsibility for coordinated, ongoing measurement and improvement of EHDI process outcomes. Early intervention programs must assess the language, cognitive skills, auditory skills, speech, vocabulary, and social-emotional development of all children with hearing loss at 6-month intervals during the first 3 years of life by using assessment tools that have been standardized on children with normal hearing and norm-referenced assessment tools that are appropriate to measure progress in verbal and visual language.

The primary purpose of regular developmental monitoring is to provide valuable information to parents about the rate of their child's development as well as programmatic feedback concerning curriculum decisions. Families also become knowledgeable about expectations and milestones of typical development of hearing children. Studies have shown that valid and reliable documentation of developmental progress is possible through parent questionnaires, analysis of videotaped conversational interactions, and clinically administered assessments.* Documentation of developmental progress should be provided on a regular basis to parents and, with parental release of information, to the medical home and audiologist. Although criterion-referenced checklists may provide valuable information for establishing intervention strategies and goals, these assessment tools alone are not sufficient for parents and intervention professionals to determine if a child's developmental progress is comparable with his or her hearing peers.

Opportunities for Interaction With Other Parents of Children With Hearing Loss

Intervention professionals should seek to involve parents at every level of the EHDI process and develop true and meaningful partnerships with parents. To reflect the value of the contributions that selected parents make to development and program components, these parents should be paid as contributing staff members. Parent representatives should be included in all advisory board activities. In many states, parents have been integral and often have taken leadership roles in the development of policy, resource material, communication mechanisms, mentoring and advocacy opportunities, dissemination of information, and interaction with the deaf community and other individuals who are deaf or hard of hearing. Parents, often in partnership with people who are deaf and hard of hearing, have also participated in the training of professionals. They should be participants in the regular assessment of program services to ensure ongoing improvement and quality assurance.

Opportunities for Interaction With Individuals Who Are Deaf or Hard of Hearing

Intervention programs should include opportunities for involvement of individuals who are deaf or hard of hearing in all aspects of EHDI programs. Because intervention programs serve children with mild-to-profound, unilateral or bilateral, permanent conductive, and sensory or neural hearing disorders, role models who are deaf or hard of hearing can be significant assets to an intervention program. These individuals can serve on state EHDI advisory boards and be trained as mentors for families and children with hearing loss who choose to seek their support. Almost all families choose at some time during their early childhood programs to seek out both adults and child peers with hearing loss. Programs should ensure that these opportunities are available and can be delivered to families through a variety of communications means, such as Web sites, e-mail, newsletters, videos, retreats, picnics and other social events, and educational forums for parents.

Provision of Communication Options

Research studies thus far of early-identified infants with hearing loss have not found significant differences in the developmental outcomes by method of communication when measured at 3 years of age.† Therefore, a range of options should be offered to families in a nonbiased manner. In addition, there have been reports of children with successful outcomes for each of the different methods of communication. The choice is a dynamic process on a continuum, differs according to the individual needs of each family, and can be adjusted as necessary on the basis of a child's rate of progress in developing communication skills. Programs need to provide families with access to skilled and experienced early intervention professionals to facilitate communication and language development in the communication option chosen by the family.

Skills of the Early Intervention Professional

All studies with successful outcomes reported for early-identified children who are deaf or hard of hearing have intervention provided by specialists who are trained in parent-infant intervention services.[12,90,97] Early intervention programs should develop mechanisms to ensure that early intervention professionals have special skills necessary for providing families with the highest quality of service specific to children with hearing loss. Professionals with a background in deaf education, audiology, and speech-language pathology will typically have the skills needed for providing intervention services. Professionals should be highly qualified in their respective fields and should be skilled communicators who are knowledgeable and sensitive to the importance of en-

*Refs 10–13, 51, 85, 87–90, and 93–96.

†Refs 10–13, 85, 87, 88, 90, 93, and 96.

hancing families' strengths and supporting their priorities. When early intervention professionals have knowledge of the principles of adult learning, it increases their success with parents and other professionals.

Quality of Intervention Services

Children with confirmed hearing loss and their families have the right to prompt access to quality intervention services. For newborn infants with confirmed hearing loss, enrollment into intervention services should begin as soon after hearing-loss confirmation as possible and no later than 6 months of age. Successful early intervention programs (1) are family centered, (2) provide families with unbiased information on all options regarding approaches to communication, (3) monitor development at 6-month intervals with norm-referenced instruments, (4) include individuals who are deaf or hard of hearing, (5) provide services in a natural environment in the home or in the center, (6) offer high-quality service regardless of where the family lives, (7) obtain informed consent, (8) are sensitive to cultural and language differences and provide accommodations as needed, and (9) conduct annual surveys of parent satisfaction.

Intervention for Special Populations of Infants and Young Children

Developmental monitoring should also occur at regular 6-month intervals for special populations of children with hearing loss, including those with minimal and mild bilateral hearing loss,[98] unilateral hearing loss,[99,100] and neural hearing loss,[22] because these children are at risk of having speech and language delay. Research findings indicate that approximately one third of children with permanent unilateral loss experience significant language and academic delays.[99–101]

Audiological Habilitation

Most infants and children with bilateral hearing loss and many with unilateral hearing loss benefit from some form of personal amplification device.[32] If the family chooses personal amplification for its infant, hearing-aid selection and fitting should occur within 1 month of initial confirmation of hearing loss even when additional audiological assessment is ongoing. Audiological habilitation services should be provided by an audiologist who is experienced with these procedures. Delay between confirmation of the hearing loss and fitting of an amplification device should be minimized.[51,102]

Hearing-aid fitting proceeds optimally when the results of physiologic audiological assessment including diagnostic ABR, OAE, and tympanometry and medical examination are in accord. For infants who are below a developmental age of 6 months, hearing-aid selection will be based on physiologic measures alone. Behavioral threshold assessment with visual reinforcement audiometry should be obtained as soon as possible to cross-check and augment physiologic findings (see www.audiology.org).

The goal of amplification-device fitting is to provide the infant with maximum access to all of the acoustic features of speech within an intensity range that is safe and comfortable. That is, amplified speech should be comfortably above the infant's sensory threshold but below the level of discomfort across the speech frequency range for both ears. To accomplish this in infants, amplification-device selection, fitting, and verification should be based on a prescriptive procedure that incorporates individual real-ear measures that account for each infant's ear-canal acoustics and hearing loss.[32] Validation of the benefits of amplification, particularly for speech perception, should be examined in the clinical setting as well as in the child's typical listening environments. Complementary or alternative technology, such as frequency modulation (FM) systems or cochlear implants, may be recommended as the primary and/or secondary listening device depending on the degree of the infant's hearing loss, the goals of auditory habilitation, the infant's acoustic environments, and the family's informed choices.[3] Monitoring of amplification, as well as the long-term validation of the appropriateness of the individual habilitation program, requires ongoing audiological assessment along with electroacoustic, real-ear, and functional checks of the hearing instruments. As the hearing loss becomes more specifically defined through audiological assessments and as the child's ear-canal acoustics change with growth, refinement of the individual prescriptive hearing-aid gain and output targets is necessary. Monitoring also includes periodic validation of communication, social-emotional, and cognitive development and, later, academic performance to ensure that progress is commensurate with the child's abilities. It is possible that infants and young children with measurable residual "hearing" (auditory responses) and well-fit amplification devices may fail to develop auditory skills necessary for successful spoken communication. Ongoing validation of the amplification device is accomplished through interdisciplinary evaluation and collaboration with the early intervention team and family.

Cochlear implantation should be given careful consideration for any child who seems to receive limited benefit from a trial with appropriately fitted hearing aids. According to US Food and Drug Administration guidelines, infants with profound bilateral hearing loss are candidates for cochlear implantation at 12 months of age and children with bilateral severe hearing loss are eligible at 24 months of age. The presence of developmental conditions (eg, developmental delay, autism) in addition to hearing loss should not, as a rule, preclude the consideration of cochlear implantation for an infant or child who is deaf. Benefits from hearing aids and cochlear implants in children with neural hearing loss

have also been documented. The benefit of acoustic amplification for children with neural hearing loss is variable.[28,103] Thus, a trial fitting is indicated for infants with neural hearing loss until the usefulness of the fitting can be determined. Neural hearing loss is a heterogeneous condition; the decision to continue or discontinue use of hearing aids should be made on the basis of the benefit derived from amplification. Use of cochlear implants in neural hearing loss is growing, and positive outcomes have been reported for many children.[28]

Infants and young children with unilateral hearing loss should also be assessed for appropriateness of hearing-aid fitting. Depending on the degree of residual hearing in unilateral loss, a hearing aid may or may not be indicated. Use of "contralateral routing of signals" amplification for unilateral hearing loss in children is not recommended.[104] Research is currently underway to determine how to best manage unilateral hearing loss in infants and young children.

The effect of otitis media with effusion (OME) is greater for infants with sensorineural hearing loss than for those with normal cochlear function.[73] Sensory or permanent conductive hearing loss is compounded by additional transient conductive hearing loss associated with OME. OME further reduces access to auditory cues necessary for the development of spoken English. OME also negatively affects the prescriptive targets of the hearing-aid fitting, decreasing auditory awareness and requiring adjustment of the amplification characteristics. Prompt referral to either the primary care physician or an otolaryngologist for treatment of persistent OME is indicated in infants with sensorineural hearing loss.[105] Definitive resolution of OME should never delay the fitting of an amplification device.[73,106]

Medical and Surgical Intervention

Medical intervention is the process by which a physician provides medical diagnosis and direction for medical and/or surgical treatment options for hearing loss and/or related medical disorder(s) associated with hearing loss. Treatment varies from the removal of cerumen and the treatment of OME to long-term plans for reconstructive surgery and assessment of candidacy for cochlear implants. If necessary, surgical treatment of malformation of the outer and middle ears, including bone-anchored hearing aids, should be considered in the intervention plan for infants with permanent conductive or mixed hearing loss when they reach an appropriate age.

Communication Assessment and Intervention

Language is acquired with greater ease during certain sensitive periods of infant and toddler development.[107–109] The process of language acquisition includes learning the precursors of language, such as the rules that pertain to selective attention and turn taking.[20,110,111] Cognitive, social, and emotional development are influenced by the acquisition of language. Development in these areas is synergistic. A complete language evaluation should be performed at regular intervals for infants and toddlers with hearing loss. The evaluation should include an assessment of oral, manual, and/or visual mechanisms as well as cognitive abilities.

A primary focus of language intervention is to support families in fostering the communication abilities of their infants and toddlers who are deaf or hard of hearing.[20] Spoken- and/or sign-language development should be commensurate with the child's age and cognitive abilities and should include acquisition of phonologic (for spoken language), visual/spatial/motor (for signed language), morphologic, semantic, syntactic, and pragmatic skills, depending on the family's preferred mode of communication.

Early intervention professionals should follow family-centered principles to assist in developing communicative competence of infants and toddlers who are deaf or hard of hearing.[112–114] Families should be provided with information specific to language development and access to peer and language models as well as family-involved activities that facilitate language development of children with normal hearing and children who are hard of hearing or deaf.[115,116] Depending on family choices, families should be offered access to children and adults with hearing loss who are appropriate and competent language models. Information on spoken language and signed language, such as American Sign Language[117] and cued speech, should be provided.

Continued Surveillance, Screening, and Referral of Infants and Toddlers

Appendix 2 presents 11 risk indicators that are associated with either congenital or delayed-onset hearing loss. A single list of risk indicators is presented in the current JCIH statement, because there is significant overlap among those indicators associated with congenital/neonatal hearing loss and those associated with delayed-onset/acquired or progressive hearing loss. Heightened surveillance of all infants with risk indicators, therefore, is recommended. There is a significant change in the definition of risk-indicator 3, which has been modified from NICU stay more than 48 hours to NICU stay more than 5 days. Consistent with 2000 JCIH position statement,[3] the 2007 position statement recommends use of risk indicators for hearing loss for 3 purposes. Historically, the first use of risk indicators is for the identification of infants who should receive audiological evaluation but who live in geographic locations (eg, developing nations, remote areas) where universal hearing screening is not yet available.‡ This use has become less common as a result of the expansion of

‡Refs 3, 19, 21, 24, 25, 64, and 118–124.

UNHS. The second purpose of risk-indicator identification is to help identify infants who pass the neonatal screening but are at risk of developing delayed-onset hearing loss and, therefore, should receive ongoing medical, speech and language, and audiological surveillance. Third, the risk indicators are used to identify infants who may have passed neonatal screening but have mild forms of permanent hearing loss.[25]

Because some important indicators, such as family history of hearing loss, may not be determined during the course of UNHS,[14,72] the presence of all risk indicators for acquired hearing loss should be determined in the medical home during early well-infant visits. Risk indicators that are marked with a section symbol in Appendix 2 are of greater concern for delayed-onset hearing loss. Early and more frequent assessment may be indicated for children with CMV infection,[118,125,126] syndromes associated with progressive hearing loss,[72] neurodegenerative disorders,[72] trauma,[127–129] or culture-positive postnatal infections associated with sensorineural hearing loss[130,131]; for children who have received ECMO[64] or chemotherapy[132]; and when there is caregiver concern or a family history of hearing loss.[16]

For all infants with and without risk indicators for hearing loss, developmental milestones, hearing skills, and parent concerns about hearing, speech, and language skills should be monitored during routine medical care consistent with the AAP periodicity schedule.

The JCIH has determined that the previously recommended approach to follow-up of infants with risk indicators for hearing loss only addressed children with identifiable risk indicators and failed to consider the possibility of delayed-onset hearing loss in children without identifiable risk indicators. In addition, concerns were raised about feasibility and cost associated with the 2000 JCIH recommendation for audiological monitoring of all infants with risk indicators at 6-month intervals. Because approximately 400 000 infants are cared for annually in NICUs in the United States, and the 2000 JCIH recommendation included audiology assessments at 6-month intervals from 6 months to 36 months of age for all infants admitted to an NICU for more than 48 hours, an unreasonable burden was placed on both providers of audiology services and families. In addition, there was no provision for identification of delayed-onset hearing loss in infants without an identifiable risk indicator. Data from 2005 for 12 388 infants discharged from NICUs in the National Perinatal Information Network indicated that 52% of infants were discharged within the first 5 days of life, and these infants were significantly less likely to have an identified risk indicator for hearing loss other than NICU stay. Therefore, the 2007 JCIH recommends an alternative, more inclusive strategy of surveillance of all children within the medical home based on the pediatric periodicity schedule. This protocol will permit the detection of children with either missed neonatal or delayed-onset hearing loss irrespective of the presence or absence of a high-risk indicator.

The JCIH recognizes that an optimal surveillance and screening program within the medical home would include the following:

- At each visit, consistent with the AAP periodicity schedule, infants should be monitored for auditory skills, middle-ear status, and developmental milestones (surveillance). Concerns elicited during surveillance should be followed by administration of a validated global screening tool.[133] A validated global screening tool is administered to all infants at 9, 18, and 24 to 30 months or, if there is physician or parental concern about hearing or language, sooner.[133]
- If an infant does not pass the speech-language portion of the global screening in the medical home or if there is physician or caregiver concern about hearing or spoken-language development, the child should be referred immediately for further evaluation by an audiologist and a speech-language pathologist for a speech and language evaluation with validated tools.[133]
- Once hearing loss is diagnosed in an infant, siblings who are at increased risk of having hearing loss should be referred for audiological evaluation.[14,75,134,135]
- All infants with a risk indicator for hearing loss (Appendix 2), regardless of surveillance findings, should be referred for an audiological assessment at least once by 24 to 30 months of age. Children with risk indicators that are highly associated with delayed-onset hearing loss, such as having received ECMO or having CMV infection, should have more frequent audiological assessments.
- All infants for whom the family has significant concerns regarding hearing or communication should be promptly referred for an audiological and speech-language assessment.
- A careful assessment of middle-ear status (using pneumatic otoscopy and/or tympanometry) should be completed at all well-child visits, and children with persistent middle-ear effusion that last for 3 months or longer should be referred for otologic evaluation.[136]

Protecting the Rights of Infants and Families

Each agency or institution involved in the EHDI process shares responsibility for protecting infant and family rights in all aspects of UNHS, including access to information including potential benefits and risks in the family's native language, input into decision-making, and confidentiality.[77] Families should receive information about childhood hearing loss in easily understood language. Families have the right to accept or decline hearing screening or any follow-up care for their newborn

infant within the statutory regulations, just as they have for any other screening or evaluation procedures or intervention.

EHDI data merit the same level of confidentiality and security afforded all other health care and education information in practice and law. The infant's family has the right to confidentiality of the screening and follow-up assessments and the acceptance or rejection of suggested intervention(s). In compliance with federal and state laws, mechanisms should be established that ensure parental release and approval of all communications regarding the infant's test results, including those to the infant's medical home and early intervention–coordinating agency and programs. The Health Insurance Portability and Accountability Act (Pub L No. 104-191 [1996]) regulations permit the sharing of health information among health care professionals.

Information Infrastructure

In its 2000 position statement,[3] the JCIH recommended development of uniform state registries and national information databases that incorporate standardized methodology, reporting, and system evaluation. EHDI information systems are to provide for the ongoing and systematic collection, analysis, and interpretation of data in the process of measuring and reporting associated program services (eg, screening, evaluation, diagnosis, and/or intervention). These systems are used to guide activities, planning, implementation, and evaluation of programs and to formulate research hypotheses.

EHDI information systems are generally authorized by legislators and implemented by public health officials. These systems vary from a simple system that collects data from a single source to electronic systems that receive data from many sources in multiple formats. The number and variety of systems will likely increase with advances in electronic data interchange and integration of data, which will also heighten the importance of patient privacy, data confidentiality, and system security. The appropriate agencies and/or officials should be consulted for any projects regarding public health surveillance.[69]

Federal and state agencies are collaborating in the standardization of data definitions to ensure the value of data sets and to prevent misleading or unreliable information. Information management is used to improve services to infants and their families; to assess the quantity and timeliness of screening, evaluation, and enrollment into intervention; and to facilitate collection of demographic data on neonatal and infant hearing loss.

The JCIH endorses the concept of a limited national database to permit documentation of the demographics of neonatal hearing loss, including prevalence and etiology across the United States. The information obtained from the information-management system should assist both the primary health care professional and the state health agency in measuring quality indicators associated with program services (eg, screening, diagnosis, and intervention). The information system should provide measurement tools to determine the degree to which each process is stable and sustainable and conforms to program benchmarks. Timely and accurate monitoring of relevant quality measures is essential.

Since 1999, the CDC and the Directors of Speech and Hearing Programs in State Health and Welfare Agencies (DSHPSHWA) have collected annual aggregate EHDI program data needed to address the national EHDI goals. In 1999, a total of 22 states provided data for the DSHPSHWA survey. Participation had increased to 48 states, 1 territory, and the District of Columbia in 2003. However, many programs have been unable to respond to all the questions on the survey because of lack of a statewide comprehensive data-management and reporting system.

The Government Performance and Results Act (GPRA) of 1993 (Pub L No. 103-62) requires that federal programs establish measurable goals approved by the US Office of Management and Budget (OMB) that can be reported as part of the budgetary process, thus linking future funding decisions with performance. The HRSA has modified its reporting requirements for all grant programs. The GPRA measures that must be reported to the OMB by the MCHB annually for the EHDI program are:

- the number of infants screened for hearing loss before discharge from the hospital;
- the number of infants with confirmed hearing loss at no later than 3 months of age;
- the number of infants enrolled in a program of early intervention at no later than 6 months of age;
- the number of infants with confirmed or suspected hearing loss referred to an ongoing source of comprehensive health care (ie, medical home); and
- the number of children with nonsyndromic hearing loss who have developmentally appropriate language and communication skills at school entry.

One GPRA measure that must be reported to the OMB by the CDC annually for the EHDI program is the percentage of newborn infants with a positive screening result for hearing loss who are subsequently lost to follow-up.

EHDI programs have made tremendous gains in their ability to collect, analyze, and interpret data in the process of measuring and reporting associated program services. However, only a limited number of EHDI programs are currently able to accurately report the number of infants screened, evaluated, and enrolled in intervention, the age of time-related objectives (eg, screening by 1 month of age), and the severity or laterality of hearing loss. This is complicated by the lack of data standards and

by privacy issues within the regulations of the Family Educational Rights and Privacy Act of 1974 (Pub L No. 93-380).

Given the current lack of standardized and readily accessible sources of data, the CDC EHDI program, in collaboration with the DSHPSHWA, developed a revised survey to obtain annual EHDI data from states and territories in a consistent manner to assess progress toward meeting the national EHDI goals and the *Healthy People 2010* objectives. In October 2006, the OMB, which is responsible for reviewing all government surveys, approved the new EHDI hearing screening and follow-up survey. To facilitate this effort, the CDC EHDI Data Committee is establishing the minimum data elements and definitions needed for information systems to be used to assess progress toward the national EHDI goals.

The JCIH encourages the CDC and HRSA to continue their efforts to identify barriers and explore possible solutions with EHDI programs to ensure that children in each state who seek hearing-related services in states other than where they reside receive all recommended screening and follow-up services. EHDI systems should also be designed to promote the sharing of data regarding early hearing loss through integration and/or linkage with other child health information systems. The CDC currently provides funds to integrate the EHDI system with other state/territorial screening, tracking, and surveillance programs that identify children with special health care needs. Grantees of the MCHB are encouraged to link hearing-screening data with such child health data sets as electronic birth certificates, vital statistics, birth defects registries, metabolic or newborn dried "blood-spot" screenings, immunization registries, and others.

To promote the best use of public health resources, EHDI information systems should be evaluated periodically, and such evaluations should include recommendations for improving quality, efficiency, and usefulness. The appropriate evaluation of public health surveillance systems becomes paramount as these systems adapt to revise case definitions, address new health-related events, adopt new information technology, ensure data confidentiality, and assess system security.[69]

Currently, federal sources of systems support include Title V block grants to states for maternal and child health care services, Title XIX (Medicaid) federal and state funds for eligible children, and competitive US Department of Education personnel preparation and research grants. The NIDCD provides grants for research related to early identification and intervention for children who are deaf or hard of hearing.[137]

Universities should assume responsibility for special-track, interdisciplinary, professional education programs for early intervention for infants and children with hearing loss. Universities should also provide training in family systems, the grieving process, cultural diversity, auditory skill development, and deaf culture. There is a critical need for in-service and preservice training of professionals related to EHDI programs, which is particularly acute for audiologists and early interventionists with expertise in hearing loss. This training will require increased and sustained funding for personnel preparation.

Benchmarks and Quality Indicators

The JCIH supports the concept of regular measurements of performance and recommends routine monitoring of these measures for interprogram comparison and continuous quality improvement. Performance benchmarks represent a consensus of expert opinion in the field of newborn hearing screening and intervention. The benchmarks are the minimal requirements that should be attained by high-quality EHDI programs. Frequent measures of quality permit prompt recognition and correction of any unstable component of the EHDI process.[138]

Quality Indicators for Screening

- Percentage of all newborn infants who complete screening by 1 month of age; the recommended benchmark is more than 95% (age correction for preterm infants is acceptable).
- Percentage of all newborn infants who fail initial screening and fail any subsequent rescreening before comprehensive audiological evaluation; the recommended benchmark is less than 4%.

Quality Indicators for Confirmation of Hearing Loss

- Of infants who fail initial screening and any subsequent rescreening, the percentage who complete a comprehensive audiological evaluation by 3 months of age; the recommended benchmark is 90%.
- For families who elect amplification, the percentage of infants with confirmed bilateral hearing loss who receive amplification devices within 1 month of confirmation of hearing loss; the recommended benchmark is 95%.

Quality Indicators for Early Intervention

- For infants with confirmed hearing loss who qualify for Part C services, the percentage for whom parents have signed an IFSP by no later than 6 months of age; the recommended benchmark is 90%.
- For children with acquired or late-identified hearing loss, the percentage for whom parents have signed an IFSP within 45 days of the diagnosis; the recommended benchmark is 95%.
- The percentage of infants with confirmed hearing loss who receive the first developmental assessment with

standardized assessment protocols (not criterion reference checklists) for language, speech, and nonverbal cognitive development by no later than 12 months of age; the recommended benchmark is 90%.

CURRENT CHALLENGES, OPPORTUNITIES, AND FUTURE DIRECTIONS

Despite the tremendous progress made since 2000, there are challenges to the success of the EHDI system.

Challenges

All of the following listed challenges are considered important for the future development of successful EHDI systems:

- Too many children are lost between the failed screening and the rescreening and between the failed rescreening and the diagnostic evaluation.
- There is a shortage of professionals with skills and expertise in both pediatrics and hearing loss, including audiologists, deaf educators, speech-language pathologists, early intervention professionals, and physicians.
- There is often a lack of timely referral for diagnosis of, and intervention for, suspected hearing loss in children.
- Consistent and stable state and federal funding is needed for program sustainability.
- When compared with services provided for adults, pediatric services in all specialties are poorly reimbursed.
- Access to uniform Part C services is inadequate among states and within states.
- There is a lack of integrated state data-management and -tracking systems.
- Demographics and cultural diversity are changing rapidly.
- Funding for hearing aids, loaner programs, cochlear implants, and FM systems is needed.
- There is a lack of specialized services for children with multiple disabilities and hearing loss.
- Children may not qualify for services (state Part C guidelines) before demonstrating language delays (prevention model versus deficit model).
- Children may not qualify for assistive technology (prevention model versus deficit model).
- There is a lack of in-service education for key professionals.
- There are regulatory barriers to sharing information among providers and among states.
- No national standards exist for the calibration of OAE or ABR instrumentation, and there is a lack of uniform performance standards.

Opportunities for System Development and Research

- Establish programs to ensure the development of communication for infants and children with all degrees and types of hearing loss, allowing them access to all educational, social, and vocational opportunities throughout their life span.
- Develop improved, rapid, reliable screening technology designed to differentiate specific types of hearing loss.
- Develop and validate screening technologies for identifying minimal hearing loss.
- Develop state data-management systems with the capacity for the accurate determination of the prevalence for delayed-onset or progressive hearing loss.
- Develop state data-tracking systems to follow infants with suspected and confirmed hearing loss through individual state EHDI programs.
- Track the certification credentials of the service providers for children with confirmed hearing loss who are receiving Part C early intervention services and early childhood special education.
- Track genetic, environmental, and pharmacologic factors that contribute to hearing loss, thus allowing for tailored prevention and intervention strategies.
- Continue to refine electrophysiologic diagnostic techniques, algorithms, and equipment to enable frequency-specific threshold assessment for use with very young infants.
- Continue to refine techniques to improve the selection and fitting of appropriate amplification devices in infants and young children.
- Conduct translational research pertaining to young children with hearing loss, in particular, genetic, diagnostic, and outcomes studies.
- Initiate prospective population-based studies to determine the prevalence and natural history of auditory neural conduction disorders.
- Conduct efficacy studies to determine appropriate early intervention strategies for infants and children with all degrees and types of hearing loss.
- Conduct additional studies on the efficacy of intervention for infants and children who receive cochlear implants at younger than 2 years.
- Conduct additional studies on the efficacy of hearing-aid use in infants and children younger than 2 years.

- Conduct additional studies of the auditory development of children who have appropriate amplification devices in early life.
- Expand programs within health, social service, and education agencies associated with early intervention and Head Start programs to accommodate the needs of the increasing numbers of early-identified children.
- Adapt education systems to capitalize on the abilities of children with hearing loss who have benefited from early identification and intervention.
- Develop genetic and medical procedures that will determine more rapidly the etiology of hearing loss.
- Ensure transition from Part C (early intervention) to Part B (education) services in ways that encourage family participation and ensure minimal disruption of child and family services.
- Study the effects of parents' participation in all aspects of early intervention.
- Test the utility of a limited national data set and develop nationally accepted indicators of EHDI system performance.
- Encourage the identification and development of centers of expertise in which specialized care is provided in collaboration with local service providers.
- Obtain the perspectives of individuals who are deaf or hard of hearing in developing policies regarding medical and genetic testing and counseling for families who carry genes associated with hearing loss.[139]

CONCLUSIONS

Since the 2000 JCIH statement, tremendous and rapid progress has been made in the development of EHDI systems as a major public health initiative. The percentage of infants screened annually in the United States has increased from 38% to 95%. The collaboration at all levels of professional organizations, federal and state government, hospitals, medical homes, and families has contributed to this remarkable success. New research initiatives to develop more sophisticated screening and diagnostic technology, improved digital hearing-aid and FM technologies, speech-processing strategies in cochlear implants, and early intervention strategies continue. Major technological breakthroughs have been made in facilitating the definitive diagnosis of both genetic and nongenetic etiologies of hearing loss. In addition, outcomes studies to assess the long-term outcomes of special populations, including infants and children with mild and unilateral hearing loss, neural hearing loss, and severe or profound hearing loss managed with cochlear implants, have been providing information on the individual and societal impact and the factors that contribute to an optimized outcome. It is apparent, however, that there are still serious challenges to be overcome and system barriers to be conquered to achieve optimal EHDI systems in all states in the next 5 years. Follow-up rates remain poor in many states, and funding for amplification in children is inadequate. Funding to support outcome studies is necessary to guide intervention and to determine factors other than hearing loss that affect child development. The ultimate goal, to optimize communication, social, academic, and vocational outcomes for each child with permanent hearing loss, must remain paramount.

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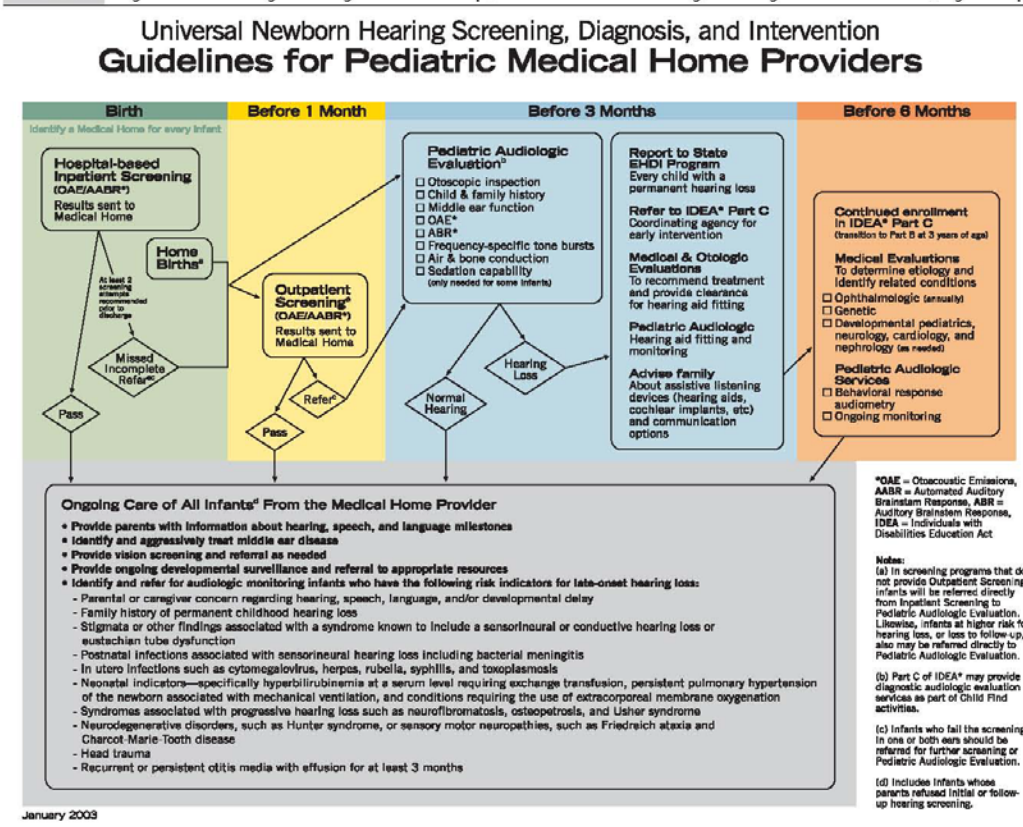
APPENDIX 1 Algorithm for Hearing Screening. Available at: http://www.medicalhomeinfo.org/screening/Screen%20Materials/Algorithm.pdf



APPENDIX 2: RISK INDICATORS ASSOCIATED WITH PERMANENT CONGENITAL, DELAYED-ONSET, OR PROGRESSIVE HEARING LOSS IN CHILDHOOD

Risk indicators that are marked with a "§" are of greater concern for delayed-onset hearing loss.

1. Caregiver concern§ regarding hearing, speech, language, or developmental delay.[62]
2. Family history§ of permanent childhood hearing loss.[24,140]
3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO,§ assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.[64,131]
4. In utero infections, such as CMV,§ herpes, rubella, syphilis, and toxoplasmosis.[64–67,125,126]
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.[24]
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.[24]
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss,§ such as neurofibromatosis, osteopetrosis, and Usher syndrome[131]; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.[72]
8. Neurodegenerative disorders,§ such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.[131]
9. Culture-positive postnatal infections associated with sensorineural hearing loss,§ including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.[130,131,141]
10. Head trauma, especially basal skull/temporal bone fracture§ that requires hospitalization.[127–129]
11. Chemotherapy.§[132]

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

Joint Committee on Infant Hearing

Pediatrics 2007;120;898
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Appendix I Protocol_for_Infants with_Risk_Factors

Notification Process for Infants (0-6 months) Identified with Risk Factors for Late Onset or Progressive Hearing Loss

Policy: Caregivers and primary care physicians of children identified with risk factors during infancy, birth through six months, for late onset or progressive hearing loss will be notified of recommended audiological follow up, so that if hearing loss develops, it will be detected as early as possible and delays in access to intervention and communication options will be minimized.

Background: The Georgia notification process for children identified with risk factors for late onset or progressive hearing loss is based on the Joint Committee on Infant Hearing (JCIH) 2007 position statement:

- The purpose of risk-indicator identification is: (1) to help identify infants who pass the neonatal screening but are at risk of developing delayed-onset hearing loss and, therefore, should receive ongoing medical, speech and language, and audiological surveillance, and (2) to identify infants who may have passed neonatal screening but have mild forms of permanent hearing loss.

- "The timing and number of hearing reevaluations for children with risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss. 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 extracorporeal membrane oxygenation (ECMO) or chemotherapy; and when there is caregiver concern or a family history of hearing loss."

This protocol outlines the notification process for children, birth through six months, with risk factors. A list of risk factors for late onset or progressive hearing loss can be found in the appendix.

PROCEDURES AND IMPLEMENTATION

Reporting risk factors to Public Health
Children 1st provides a population-based system of screening young children for specific risk conditions which place the child at risk for adverse health and/or developmental outcomes.

Risk factors for late-onset or progressive hearing loss are to be reported to the Children 1st program using the Children 1st Screening and Referral form or through the Electronic Birth Certificate.

Primary Care Providers (PCPs), parents, and other care providers should refer an infant for screening any time they suspect a hearing loss. Audiologists who later identify infants and children with a hearing loss, up to age 5 years, should report it to the UNHSI program via Children 1st.

Birth to Five Review Team
According to the Comprehensive Child Find policy for Child Health Programs (ABCH - 911-001), a representative from each child health program to include Babies Can't Wait (BCW), Children 1st, Universal Newborn Health Screening & Intervention (UNHSI), and Children's Medical Services (CMS) shall meet at minimum once per week to review referrals and the disposition of the referrals, including referrals for risk factors for late-onset or progressive hearing loss.

UNHSI Coordinators should be informed of all referrals with risk factors for late-onset or progressive hearing loss for infants birth through 6 months. UNHSI Coordinators will enter referrals into SendSS Newborn in order to document outcomes of the referrals reported to UNHSI Coordinator by the medical home and/or audiologist.

Recommended Follow-up
The risk factor(s) will determine the frequency and timing of follow up hearing evaluations.

The child should see an audiologist for a hearing evaluation by 24 to 30 months of age if one or more of the following risk factors (Tier 1) are present:

- Cranio-facial anomalies
- Exchange transfusion for elevated bilirubin
- Herpes infection confirmed in infant
- NICU stay longer than five days (included extreme prematurity and/or low birth weight <1500 grams)
- Other congenital infection
- Ototoxic medications administered
- Rubella infection confirmed in infant
- Syphilis infection confirmed in infant
- Toxoplasmosis infection confirmed in infant

Refer to the “Child Health Programs Eligible Medical Conditions List” for specific conditions under these broader categories of risk factors.

The child should see an audiologist for a hearing evaluation by six (6) months of age with more frequent audiological assessments as determined by the child’s medical home if one or more of the following risk factors (Tier 2) are present:

- cytomegalovirus (CMV) infection,
- syndromes associated with progressive hearing loss,
- neurodegenerative disorders,
- head trauma
- culture-positive postnatal infections associated with sensorineural hearing loss (i.e. bacterial meningitis);
- extracorporeal membrane oxygenation (ECMO)
- Persistent Pulmonary Hypertension (PPHN) associated with mechanical ventilation
- chemotherapy;
- caregiver concern
- family history of hearing loss.

Refer to the “Child Health Programs Eligible Medical Conditions List” for specific conditions under these broader categories of risk factors.

Children having a co-existing Tier 1 and Tier 2 risk factor should follow the Tier 2 recommendation.

Notification Letters

The UNHSI Coordinator will send a risk factor notification letter to the parent or guardian of the child indicating the risk factor(s) identified the need for follow-up testing and the recommended type of testing necessary.

- A notification letter will also be sent to the primary care provider (PCP) and, as appropriate, to the Children 1st Coordinator and the BCW Service Coordinator.
- Babies having a co-existing Tier 1 and Tier 2 risk factor will only receive a Tier 2 notification letter.

Responsibility for Follow-up
Children 1st will contact the family to offer enrollment and referrals to BCW, CMS, and UNHSI programs. Parents may choose to enroll in or refuse Children 1st and these other public health program services.

- **If the child is eligible for BCW and enrolls in BCW,** the BCW Service Coordinator will assume responsibility for following this family. The UNHSI Coordinator will assist families in locating service providers and funding sources (if meet eligibility criteria) as needed. If there is a developmental concern that requires a diagnostic audiological evaluation, which results in the diagnosis of a permanent hearing loss, this child should be reported to the UNHSI Coordinator for documentation of the diagnosis.

    - The BCW Service Coordinator will notify the UNHSI Coordinator when the follow-up hearing evaluation is scheduled and outcome of the evaluation.
    - Test results should be reported by the Audiologist or PCP to the BCW Service Coordinator once testing occurs.
    - The UNHSI Coordinator and an Audiologist should be included as support personnel on the Individual Family Service Plan (IFSP) for infants or children eligible for BCW identified with or at risk for hearing loss.
    - The BCW Service Coordinators will notify and invite the UNHSI Coordinators to IFSP meetings for children with confirmed hearing loss and/or risk factors for late onset or progressive hearing loss enrolled in BCW.
    - The UNHSI Coordinators may add to the development of goals, supply resources, referrals or supports and provide information to the IFSP, Primary Service Provider (PSP) team and family during the IFSP processes.
        - IFSP meetings are held at initial development of the IFSP, at six (6) month intervals, annually, interperiodic (as needed), and at the time of transition out of BCW services.
            - UNHSI Coordinator participation in these meetings will be optional and not mandatory.

If family is not eligible for or refuses BCW services, the UNHSI Follow-up Coordinator will be responsible for contacting the family to provide education regarding the importance of compliance with the follow-up recommendation.

Documentation of Risk Factor Outcomes
The outcome of testing is to be documented in the child's record in the Children 1st and/or BCW information system, and in the UNHSI module of SendSS NB by the UNHSI Coordinator.

Appendix J Protocol late identified Risk Factors

Notification Process for Children Late-identified with Risk Factors for Hearing Loss

Policy: Caregivers of children, 6 months to 5 years, identified with risk factors beyond infancy for hearing loss will be notified of recommended audiological follow up, so that if hearing loss develops, it will be detected as early as possible and delays in access to intervention and communication options will be minimized.

Background: The Georgia notification process for children identified with risk factors for hearing loss is based on the Joint Committee on Infant Hearing (JCIH) 2007 position statement:

- The purpose of risk-indicator identification is: (1) to help identify infants who pass the neonatal screening but are at risk of developing delayed-onset hearing loss and, therefore, should receive ongoing medical, speech and language, and audiological surveillance, and (2) to identify infants who may have passed neonatal screening but have mild forms of permanent hearing loss.
- "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."

This protocol outlines the process for children, 6 months to 5 years, late-identified with risk factors for hearing loss. A list of risk factors for late onset or progressive hearing loss can be found in the appendix.

PROCEDURES AND IMPLEMENTATION

Children 1st Reporting and Hearing Loss Risk Factors

Children 1st provides a population-based system of screening young children for specific risk conditions which place the child at risk for adverse health and/or developmental outcomes. Primary Care Providers (PCPs), parents, and other care providers may refer a child to Children 1st outside of the newborn period for a condition, which may be a risk factor for hearing loss using the Children 1st Screening and Referral form.

Birth to Five Review Team

According to the Comprehensive Child Find policy for Child Health Programs (ABCH - 911-001), a representative from each child health program to include Babies Can't Wait (BCW), Children 1st, Universal Newborn Health Screening & Intervention (UNHSI), and Children's Medical Services (CMS) shall meet at minimum once per week to review referrals and the disposition of the referrals.

Recommended Follow-up
Caregivers of parents with children who are late-identified with risk factors will be educated regarding the importance of obtaining a hearing screening or evaluation based on the risk factor identified through the referral and the importance of compliance with the follow-up recommendation.

Responsibility for Follow-up
Children 1st will contact the family to offer enrollment and referrals to BCW, and CMS programs. Parents may choose to enroll in or refuse Children 1st and these other public health program services.

- **If the child is eligible for BCW and enrolls in BCW,** the BCW Service Coordinator will assume responsibility for educating and following this family. The UNHSI Coordinator will assist families in locating service providers as needed. If the family follows up with a diagnostic audiological evaluation, which results in the diagnosis of a permanent hearing loss, this child should be reported to the UNHSI Coordinator for documentation of the diagnosis.

    - At initial visit, Service Coordinators inform parents of risk factor(s), ask hearing health history questions, and stress importance of obtaining an audiological screening or evaluation. The family should be provided a speech and language developmental checklist, which can be found in the appendix.
    - At ongoing visits, Service Coordinators follow up with parents regarding status of hearing evaluation.
    - For parents who obtain testing, audiological results should be reported by the parents to the BCW Service Coordinator following testing and entered into the child's record.
    - For parents who chose not to follow through with audiological testing, Service Coordinator should again stress importance of early identification of hearing loss and document non-compliance in child's chart.
    - The BCW Service Coordinators will notify and invite the UNHSI Coordinators to IFSP meetings for children who are enrolled in BCW and identified with late-onset and/or acquired permanent hearing loss.
    - The UNHSI Coordinators may add to the development of goals, supply resources, referrals or supports and provide information to the IFSP, Primary Service Provider (PSP) team and family during the IFSP processes.
        - IFSP meetings are held at initial development of the IFSP, at six (6) month intervals, annually, interperiodic (as needed), and at the time of transition out of BCW services.
            - UNHSI Coordinator participation in these meetings will be optional and not mandatory.

If family is not eligible for or refuses BCW services, the Children 1st Coordinator will be responsible for educating the family to provide information regarding the importance of compliance with the follow-up recommendation. The UNHSI Coordinator will assist families in locating service providers as needed. If the family follows up with a diagnostic audiological evaluation, which results in the diagnosis of a permanent hearing loss, this child should be reported to the UNHSI Coordinator for documentation of the diagnosis.

For children enrolled in Children 1st:

- At initial visit, Service Coordinators inform parents of risk factor(s), ask hearing health history questions, and stress importance of obtaining an audiological screening or evaluation. The family should be provided a speech and language developmental checklist, which can be found in the appendix.
- At ongoing visits, Service Coordinators follow up with parents regarding status of hearing evaluation.
- For parents who obtain testing, audiological results should be reported by the parents to the Service Coordinator following testing and entered into the child's record.
- For parents who chose not to follow through with audiological testing, Service Coordinator should again stress importance of early identification of hearing loss and document non-compliance in child's chart.

If the family refuses Children 1st and BCW: No further notification action at this time is completed. Refusal of enrollment is to be documented in the child's record in the Children 1st information system

Documentation of Risk Factor Outcomes
The outcome of testing, if completed, is to be documented in the child's record in the Children 1st and/or BCW information system. Results will be entered into the UNHSI module of SendSS NB by the UNHSI Coordinator only if a hearing loss is confirmed.

Suggested Items/Questions for Service Coordinators to cover when discussing late-identified risk factor for hearing loss:

1. You're child was referred to Children 1st because of _____, which is/are a risk factor(s) for hearing loss. This does not mean that you're child has a hearing loss, but should be referred for a hearing screening/evaluation if not completed recently. Early identification of hearing loss is important to minimize language and developmental delays.
2. Do you have any concerns regarding your child's hearing or speech and language development?
3. Has your child had a hearing screening or evaluation since their newborn hearing screening? If so, what was the result of that evaluation?

Appendix K UNSHI Medical Home Provider Guide