

## PENNSYLVANIA DEPARTMENT OF HEALTH NEWBORN HEARING SCREENING PROGRAM GUIDELINES

The purpose of this document is to provide guidance and recommended procedures for hospitals, physicians, and audiologists in conducting and administering universal newborn hearing screening programs. It is important to recognize that newborn hearing screening is only one component of a comprehensive approach to the management of childhood hearing loss. The process also requires follow-up diagnostic services, counseling, intervention programs, and parental educational programs. This comprehensive process must be administered by a multidisciplinary team consisting of individuals such as audiologists, physicians, educators, speech/language pathologists, nurses, and parents. Both otologic and genetic consultation are recommended in the management of childhood hearing loss.

Early and consistent screening specific to hearing loss and enrollment in early intervention services once infants have been diagnosed is key to achieving normal language development. Therefore, the goal of Pennsylvania's program is to achieve identification of hearing loss in children by three months of age and to assure enrollment in appropriate services by six months of age through the following steps:

- Appropriate follow-up by an audiologist with expertise in the evaluation of infants and young children;
- Medical evaluation of hearing loss; and
- Referral to local Part C Early Intervention as soon as a confirmed diagnosis of hearing loss is made.

### I. DEFINITIONS

- *Audiologist*: A person who is licensed by the Commonwealth of Pennsylvania to provide audiological services.
- *Auditory brainstem response (ABR)*: An objective electrophysiologic measurement of the peripheral and central auditory system to acoustic stimulation of the ear. This test can be automated auditory brainstem response (AABR), obtained with equipment that automatically provides a pass/refer outcome.
- *Department*: The Department of Health of the Commonwealth.
- *Diagnostic audiology evaluation*: An in-depth, age-appropriate evaluation of auditory function using behavioral, electrophysiologic and acoustic immittance measures to determine if a hearing loss exists and if a hearing loss is present, to determine the type, degree, and configuration of the hearing loss.
- *Discharge*: Release from the hospital after birth to care of the parent or legal guardian.
- *Early Intervention Services*: Services and supports designed to help families with children with developmental delays. Early intervention builds upon the natural learning occurring in those first few years.
- *Evoked Otoacoustic Emissions*: An objective physiologic response from the cochlea, used to determine preneural auditory function. The response may be elicited with the use of either click-evoked otoacoustic emission or distortion product otoacoustic emission test procedures. This test can be automated to provide a pass/refer outcome.
- *Follow-up*: Referral for appropriate services and procedures relating to the confirmation of hearing loss and appropriate referrals to an audiologist and primary care provider for infants who did not pass the follow-up rescreening.
- *Referral*: To direct an infant, who does not pass the initial hearing screening, and the follow-up rescreening, to an audiologist for appropriate diagnostic procedures to determine

- the existence and extent of a hearing loss, as well as appropriate habilitation of a hearing loss.
- *Hearing loss*: A dysfunction of the auditory system of any type or degree that is sufficient to interfere with the acquisition and development of speech and language skills.
  - *Hearing screening*: An objective physiological measure to be completed in order to determine the likelihood of hearing loss.
  - *Hearing Screening Facility*: A facility that performs Newborn Hearing Screenings as outlined in the body of this document.
  - *Initial hearing screening*: Procedure(s) employed for the purpose of screening hearing prior to discharge.
  - *Incomplete result*: Result of initial screening which does not yield either a conclusion of normality or possible abnormality. An incomplete result can be due to an uncooperative infant, debris in the ear canal and excess miogenic activity. In such cases, the infant should be referred for another screening within 30 days of birth.
  - *Newborn*: A child up to and including 29 days of age.
  - *Infant*: A child from 30 days of age up to 12 months of age.
  - *Parent*: A biological parent, stepparent, adoptive parent, legal guardian, or other legal custodian of a child.
  - *Primary Care Provider (PCP)*: The licensed caretaker to whom the infant will go for routine medical care following birth.

## II. ASSESSMENT AND EDUCATION

### A. Hearing Screening Facility Role:

Each hearing screening facility shall designate a person to be responsible for the newborn hearing screening program in that facility. This person will act as a single point of contact between the hearing screening facility and the Department's Newborn Hearing Screening Program. The hearing screening facility is responsible for ensuring that all screening personnel are appropriately trained to carry out the newborn hearing screening using appropriate technology. A licensed audiologist with appropriate training and experience shall advise the hospital about all aspects of the newborn hearing screening program, including screening and tracking. The Department can provide the names of audiologists with expertise in newborn hearing screening to hospitals that do not have access to audiological personnel.

### 1. Screening Technologies:

Although there are many technologies available to screen newborns, the Year 2000 Joint Committee on Infant Hearing of the American Academy of Pediatrics recommends two technologies: Auditory Brainstem Response and Otoacoustic Emissions. The Joint Committee on Infant Hearing recommends that screening programs achieve a refer rate to audiologists of less than four percent.

#### a. Auditory Brainstem Response (ABR)

ABR is an electrophysiological measure of the auditory system's response to sound. A click is presented to the ear via earphones, and electrodes record the response as the nerve impulse travels from the cochlea (inner ear) through the auditory nervous system to the brain. ABR requires a trained technician or audiologist to perform the evaluation and an audiologist to interpret the screening results. It is recommended that the screening level be no greater than 35 dBnHL.

Automated ABR (AABR) refers to equipment that is fully automated for interpretation and elicits a pass/refer response. Consequently, the AABR allows for a variety of trained hospital personnel to perform the screen such as nurses, technicians, support staff, or volunteers.

- ABR and AABR may miss a small percentage of hearing losses, such as a low frequency or high frequency hearing loss.
- The refer rates at discharge for ABR are typically less than four percent.

#### **b. Otoacoustic Emissions (OAE)**

Otoacoustic emissions measures the integrity of the outer hair cells in the cochlea. A sound is presented and a small microphone, which is placed in the baby's ear canal, measures the response that is made from the baby's ear. The audiologist analyzes the response to determine how well the inner ear is working.

The test can be fully automated for interpretation. If automated equipment is used, trained hospital personnel such as audiologists, nurses, or technicians can perform this procedure.

There are two types of OAE technologies: Transient Evoked Otoacoustic Emissions (TEOAE) and Distortion Product Otoacoustic Emissions (DPOAE). Automated OAE technology is now available for both TEOAE and DPOAE.

- OAE does not identify a disorder called auditory neuropathy. Although this is rare, auditory neuropathy has been diagnosed more frequently in the Neonatal Intensive Care Unit population.
- The refer rates for OAE typically range between five and ten percent.

## **2. Initial Screening**

Early detection of hearing loss in the newborn period is important to minimize its adverse impact on the infant's development. The Infant Hearing, Education, Assessment, Reporting and Referral Act—IHEARR (Act 89 of 2001; 11 P.S. §§876-1 – 876-9) recognizes the importance of completing hearing screening within 30 days of birth. Hearing screening facilities should provide families with written information regarding the importance of newborn hearing screening prior to testing and assure that follow-up rescreening is completed. Both the primary care provider and the Department shall be notified of those not passing hearing screening so that appropriate follow-up and/or tracking is conducted. Families have the right to refuse the hearing screening. Such refusal shall be documented in writing and be made a part of the medical records of the newborn or infant and reported to the Department in a manner described by the Department.

The hearing screening facilities are encouraged to make every effort to identify a primary care provider for each newborn prior to discharge. If one is not designated, the hearing screening facility is encouraged to provide the newborn's family with recommended options for designating a primary care provider able to provide culturally competent care.

The hearing screening facility shall notify the primary care provider with the following information:

- The newborn's screening results and the procedures used for hearing screening, and an explanation of the results of screening for each ear;

- If the initial screening was not completed, whether refused or not; and
- Information on milestones of normal auditory, speech and language development in children.

The hearing screening facility shall notify the newborn's family in writing of the results of their newborn's initial screening using a standard notification form prepared explicitly for the purpose. The notification form indicates whether the newborn passed or did not pass the screening. For newborns not passing the initial screening, the form explains the importance of obtaining follow-up rescreening as soon as possible. Information is conveyed in non-technical language that is both easily understood and culturally competent.

Hearing screening facilities shall provide data to the Department concerning the number of newborns screened in a manner to be determined by the Department (see attachment).

### **3. Follow-up Rescreening**

Newborns who did not pass the initial screen shall be referred by the hearing screening facility to have a rescreening test performed. Parents may refuse the follow-up rescreening. Such refusal shall be documented in writing and be made a part of the medical records of the newborn or infant and reported to the Department in a manner described by the Department. Hearing screening facilities are responsible for the following events:

- Providing written notification to parents that an infant needs follow-up rescreening;
- Scheduling the follow-up rescreening appointment with parents prior to discharge;
- Notifying the newborn's primary care provider, via discharge summary sheet, letter, or other specific means, that the newborn did not pass the initial screening and informing the physician of the need for follow-up.

### **4. Data Reporting to the Department**

- a. The IHEARR Act mandates that hearing screening facilities provide the following data to the Department:
  - *Newborn Hearing Screening Program Screening Reporting Form*—to be submitted weekly for each newborn not passing follow-up rescreening within 30 days of birth (see attached).
  - *Monthly Report*—to be submitted by the 15<sup>th</sup> of every month by fax, mail or e-mail, covering births and screenings for the preceding month (see attached). Birthing facilities should update any incomplete data submitted on previous Monthly Reports with the submission of each new Monthly Report.
- b. Audiologists shall send data to the Department concerning the results of diagnostic testing of infants, who did not pass two independent screens within 30 days of birth, in a manner to be prescribed by the Department.

### **B. Primary Care Provider Role**

The primary care provider directs and coordinates the evaluation and referral process within the child's medical home by:

- Referring the newborn for audiologic diagnostic testing and evaluation.
- Providing referral to appropriate agencies capable of providing intervention services and to appropriate medical specialists (i.e., otolaryngologist and geneticist) as may be indicated by the diagnostic hearing evaluation.

- Confirming the infant's receipt of intervention services, including amplification, if appropriate, by the age of 6 months. The primary care provider monitors individual cases to assure that the diagnostic hearing evaluation was completed.

The Department, in collaboration with the primary care provider, will contact the family to encourage follow-up on results of the diagnostic evaluation and referral to appropriate services.

### **1. Diagnostic Referral and Evaluation**

A newborn who does not pass two independent screens within 30 days of birth shall be referred to the primary care provider for further diagnostic testing and be reported to the Department. The primary care provider must make recommendations for audiologic diagnostic testing and provide the parents with a referral for a diagnostic evaluation of the newborn.

### **2. Information to Parents**

The primary care provider shall provide the following information to parents:

- A statement stressing the importance of follow-up; the time and location of the follow-up appointment; the telephone number of the screening or diagnostic center; and a list of Department-approved diagnostic centers.
- Recommended interventions and modalities based on the outcome of audiological testing.

### **C. Pennsylvania Department of Health Role**

- Collects and maintains data from hearing screening facility screening and the results of audiological follow-up diagnostic testing.
- Tracks children needing follow-up; communicates with parents to assure they have the information needed to seek timely and appropriate follow-up services.
- Assures appropriate linkage of diagnosed infants to Early Intervention services.
- Provides training and technical assistance to hospital staff.
- Monitors hospital referral rates.
- Conducts review and evaluation of the Newborn Hearing Screening statewide program, including follow-up rates, false-positive rates, false-negative rates, referral mechanisms and effectiveness of tracking.
- Conducts epidemiological analysis of the data for planning and program management purposes.
- Communicates screening performance result data to hospitals on a yearly basis;
- Approves centers/providers of diagnostic audiological services for infants and young children.
- Provides monthly reports to the Department of Public Welfare's Office of Mental Retardation on those children identified and referred to Early Intervention for service.
- Consults with the Infant Hearing Advisory Committee on issues related to, but not limited to, program regulation and administration, diagnostic testing, technical support and follow-up.
- Provides informational materials to hearing screening facilities, primary care providers, and families.

### III. REFERRAL AND INTERVENTION

#### A. Audiologist Role

- Reports the results of each diagnostic hearing evaluation to the referring primary care provider, including information that an infant was not successfully tested after being referred for testing.
- Reports to the Department, using a reporting form provided by the Department, the names of all children who received a diagnostic evaluation and those who were not successfully tested after being referred for a diagnostic evaluation.
- The audiologist makes a referral to Early Intervention and reports the date of referral to the Department.
- Provides parents information about hearing loss, including choices about communication and education options for children who are deaf or hard of hearing.
- Fits children with amplification including hearing aids or other assistive technology.

#### 1. Principles

The goal of newborn hearing screening is early detection and intervention. Thus, diagnostic audiological evaluations, which provide ear-specific information regarding degree, configuration, and type of hearing loss, should be completed by three months of age or three months post-discharge for babies who have been in the Neonatal Intensive Care Unit (NICU). Habilitation, including fitting amplification and receiving Early Intervention Services, should occur no later than 6 months of age.

Regardless of prior hearing screening outcomes, all infants who demonstrate risk indicators for delayed onset or progressive hearing loss should receive ongoing audiologic and medical monitoring for 3 years and at appropriate intervals thereafter to ensure prompt identification and intervention.

The audiologist is responsible for the administration and interpretation of behavioral and physiologic tests. The audiologist is also responsible for audiologic follow-up and management including candidacy for use, fitting, and dispensing of amplification and/or other communication devices.

The audiologist should be experienced and skilled in pediatric assessment and have access to the required equipment and facilities to evaluate infants and children.

The assessment should include not only the behavioral and physiologic tests of hearing, but also include the case history, parent/caregiver counseling and referrals to other professionals. All assessments and interventions should be family-centered, interdisciplinary, culturally competent and be built on informed choice for families.

Each evaluation should include an assessment protocol that is timely, uses frequency-specific stimuli, is ear-specific and includes a determination of middle ear status by bone conduction testing, otoscopy and immittance measurements.

- For very young children, a complete assessment of hearing is unlikely to be completed in one session, yet the goal is to minimize delays and time between assessments. Incomplete audiologic information should not delay the initiation of habilitation. Rather, habilitation, including the fitting of amplification, should be initiated,

with refinements and adjustment of the hearing aid fitting occurring as more and more precise information is obtained.

- Both behavioral and physiologic thresholds should be obtained for each ear.
- Assessment should include, at minimum, a low frequency (500 Hz) and a high frequency (4000 Hz or click) stimulus to allow for selection of appropriate amplification.
- Insert earphones are the transducers of choice.
- When air conduction thresholds (behavioral or physiologic methods) are abnormal, testing by bone conduction should be completed.
- Acoustic immittance should be done during each test session and, while not sufficient for middle ear assessment, it does provide valuable information in conjunction with other audiologic results. The use of higher probe tone frequency (660/678/1000 Hz) has proven a more valid indication of middle ear state for children less than 6 months of age.

## 2. Diagnostic Audiologic Follow-up Evaluation

a. **Case History** -- The evaluation should start with a comprehensive case history. At a minimum, this history should include information about congenital hearing loss in the family, medical factors and risk indicators for hearing loss, responses to sound observed by parents at home, and information on overall health and development.

b. **Physiologic Testing** – Sedation should only be used in those facilities that have a comprehensive sedation policy that outlines the steps required to ensure patient safety. Sedation medications should only be administered at the testing facility by trained personnel and should never be administered at home.

- **Otoscopy** -- Perform otoscopy to ensure ear canals are clear of occlusion.
- **Infant Tympanometry** -- Perform tympanometry to obtain immediate information on middle ear status using a higher probe tone frequency (678/1000 Hz) in children less than 6 months of age. Ear canal volumes should be measured using 226 Hz, but the tympanogram “shape” should be assessed using the higher frequency probe tone only. Whenever possible, acoustic reflex testing should be completed. The use of a probe frequency higher than 220/226 to obtain acoustic reflexes should be considered in infants under 6 months of age.
- **Click-Evoked ABR via Air Conduction using insert earphones**
  - Obtain latency intensity function. Threshold interpretation is based on individual clinic normative data.
  - Evaluate absolute latencies for waves I, III, and V at 70 dBnHL or higher.
  - Evaluate waveform morphology and interpeak latencies for waves I-V at 70 dBnHL or higher to assess retrocochlear function .
  - If ABR response is absent, evaluate for presence of cochlear microphonic.

- **Frequency-specific air-conduction testing** -- Responses to clicks and low-frequency stimuli (500 Hz tonebursts/pips) should be obtained to provide an estimate of audiometric configuration. Ideally, as many frequency-specific thresholds should be obtained as time and patient tolerance allows, keeping in mind that testing may need to be completed in multiple sessions. The most important frequencies to obtain are 500 Hz and 4000 Hz. If either of these response levels is elevated, consider obtaining 1k and 2k Hz.
- **Bone-conducted click testing, if indicated** -- When air-conducted thresholds are elevated, bone conduction testing should be completed.
- **Evoked Otoacoustic Emissions (TEOAE or DPOAE)** -- should be obtained to further evaluate cochlear function, and to rule out cases of auditory neuropathy.

The audiologic evaluation of infants and children is an ongoing process. Behavioral testing should be attempted as soon as possible to supplement physiologic data.

### c. Behavioral Audiologic Evaluation

For children less than 6 months of age, reliable behavioral hearing assessment procedures are not clinically available. Behavioral observation audiometry can provide information on the type of auditory response the child makes and about the auditory development of the child. Physiological test measures should supplement behavioral data.

For children older than 6 months of age, visual reinforcement audiometry (VRA) should be employed to assess hearing sensitivity for speech and frequency-specific stimuli. Ear-specific threshold information using insert earphones should be sought with this technique. The goal is to fit a hearing aid should a hearing loss exist. Consequently, both high and low frequencies should be used (i.e., 500 through 4000 Hz in octave intervals). If air conduction thresholds are elevated, bone conduction thresholds should be obtained.

Consistencies among several audiometric measures, behavioral findings, click-evoked and tone-evoked ABR thresholds, acoustic immittance measures, evoked otoacoustic emission, and bone conduction thresholds (behavioral and/or ABR) are essential.

### d. Outcomes and Confirmed Hearing Loss Follow-up

- **Complete and Submit DOH form “Results of Diagnostic Audiological Evaluation”** for all infants seen for diagnostic evaluation.
- **Counsel parent(s):** Review results of the diagnostic audiology assessment, implications of the audiologic diagnosis, and recommendations for intervention with the parents, including information on the following:
  - amplification options
  - importance of early intervention (see below for information on referral)
  - medical follow-up (including explaining the various medical specialty evaluations which might be recommended)
  - funding assistance (assist with medical assistance application process)

- parent support group or communication with other parents of children with hearing loss.
- **Initiate the amplification process if appropriate.**
- **Obtain MEDICAL CLEARANCE from the infant's medical home prior to fitting hearing aids.**
- **Assist in application to Medical Assistance** to obtain hearing aids.
- **Specialty Evaluations:** Recommend, with Medical Home approval, appropriate specialty evaluations:
  - Pediatric otolaryngologist
  - Medical geneticist
  - Pediatric ophthalmologist
- **Referral to Early Intervention:** Once hearing loss has been identified, a referral should be made immediately, with parental permission if making direct referral, to Early Intervention services. Habilitation and intervention should proceed concomitantly with the medical evaluation of the hearing loss and should not wait for completion of the medical evaluation and findings.

#### **e. Hearing Aid Evaluation**

The child is considered to be a candidate for amplification if a permanent hearing loss of greater than 20 dB HL exists in one or both ears in the frequency regions critical for speech understanding (1000-4000 Hz).

Hearing aids for most children should include Direct Audio Input (DAI), telecoil (T) and microphone-telecoil (M-T) switches, should be flexible, and should have safety-related features such as tamper resistant battery and volume controls. Binaural amplification should always be provided unless there are clear contraindications for fitting an ear. In general, BTEs are the hearing aid style of choice.

Custom earmolds should be available at the time of the hearing aid performance verification in order to measure the Real-Ear to Coupler Difference (RECD). The RECD will allow the hearing aid gain and maximum output characteristics of the hearing aid to be preset in the hearing aid test box prior to the evaluation of the hearing aid on the child. Use of a prescriptive program for gain and output (e.g., DSL[i/o]) is essential. Choice of the hearing aid instrument should be based on the targets. Once the targets are verified (DSL and SHARP) and the device is fitted, ongoing monitoring of hearing levels and of the amplification should take place.

Whenever possible, verification of the hearing aid settings should be completed using probe microphone measurements.

RECDs should be reassessed as the infant grows or whenever new earmolds are made.

### **B. Early Intervention Referral Guidelines**

1. Referrals from primary referral sources must be made no more than two days after the child has been identified as needing Early Intervention Services. 1-800 CONNECT LINE (1-800-692-7288) with parental consent for direct referral.
2. Once the legal authority that is administering the local Early Intervention Service agency (County MH/MR) receives the referral, it shall appoint a service coordinator,

who is responsible for coordinating all services across agency lines, and serves as a single point of contact in helping parents to obtain the services and assistance they need. The service coordinator shall make contact with the family as soon as possible but no later than two business days after receiving the referral. The service coordinator will make contact with the child's family to set up the initial home visit (intake).

3. The initial home visit shall be completed to determine the existence of previous evaluations and to recommend the need for referral for a Multi-disciplinary Evaluation (MDE) to determine or confirm eligibility and access early intervention services or tracking. The service coordinator will review all pertinent records and information on the child (i.e., review of written professional reports such as audiologist reports, hospital neonatal discharge information, physician reports etc.). The service coordinator will also interview the family or caregiver and review parental report information, identify concerns of the family, identify strengths and needs of the family, and determine the family's routines in order to identify the supports and services necessary to enhance the family's capacity to meet the developmental needs of the child.
4. In the event that the child is referred for an MDE and is found eligible for early intervention services, a meeting is scheduled to develop an Individualized Family Service Plan (IFSP) for the child. The IFSP is developed by the MDE team and authorizes services, as well as a description of the frequency, duration and location of such services. The service coordinator will continue to coordinate all activities with the family's primary care provider by asking parents for consent to send copies of pertinent information to their primary care provider (e.g., copy of the IFSP, MDE).

#### **IV. Data Reporting to the Department**

**A.** The IHEARR Act requires that hearing screening facilities provide the following data to the Department:

- *Newborn Hearing Screening Program Screening Reporting Form*—to be submitted weekly for each newborn not passing follow-up rescreening within 30 days of birth (see attached).
- *Monthly Report*—to be submitted by the 15<sup>th</sup> of every month by fax, mail or e-mail, covering births and screenings for the preceding month (see attached). Birthing facilities should update any incomplete data submitted on previous Monthly Reports with the submission of each new Monthly Report.

**B.** Audiologists shall send data to the Department concerning the results of diagnostic testing for all infants, who did not pass two independent screens within 30 days of birth, in a manner to be prescribed by the Department.