
PENNSYLVANIA DEPARTMENT OF HEALTH

**NEWBORN HEARING SCREENING PROGRAM
GUIDELINES**

ISSUED MARCH 2013

CONTENTS

Introduction.....	- 4 -
Recommended Guidelines for Hearing Screening.....	- 6 -
Hospital Births.....	- 6 -
Out-of-Hospital Births.....	- 6 -
Screening Technologies.....	- 7 -
Initial Screening: Protocols and Procedures.....	- 9 -
Repeat Hearing Screening.....	- 12 -
Recommended Guidelines for Primary Care Providers.....	- 14 -
Primary Care Provider (PCP) Role.....	- 14 -
Communicating with Parents.....	- 14 -
Ongoing Surveillance and Risk Indicators for Late Onset Hearing Loss.....	- 15 -
Recommended Guidelines for Audiologists.....	- 17 -
Audiologist Role.....	- 17 -
Audiologic Assessment.....	- 18 -
Assessment Types and Technology.....	- 18 -
Diagnostic Audiologic Evaluation.....	- 19 -
Recommended Guidelines for Early Intervention.....	- 25 -
Referrals to Early Intervention.....	- 25 -
Early Intervention Enrollment Process.....	- 25 -
Parent Rights as Part of the Early Intervention Process.....	- 26 -
Department of Health – Newborn Hearing Screening Program.....	- 27 -
Data Reporting to the Department of Health.....	- 28 -

Hospitals.....- 28 -

Out of Hospital Birth Hearing Screening Program.....- 29 -

Audiologists.....- 29 -

Early Intervention.....- 29 -

References and Resources.....- 30 -

Information Bulletins.....- 30 -

Monthly Hospital and Out-of-Hospital Reporting Templates.....- 30 -

Audiology Diagnostic Reporting Template.....- 30 -

Early Intervention Policy Statement.....- 30 -

Joint Committee on Infant Hearing (JCIH) Position Statement.....- 30 -

PA Early Hearing Detection and Intervention Website.....- 31 -

Definitions.....- 32 -



INTRODUCTION

This document provides guidance and recommended procedures for hospitals, physicians and audiologists in conducting and administering universal newborn hearing screening programs in accordance with the Infant Hearing Education, Assessment, Reporting and Referral (IHEARR) Act—Act 89 of 2001.


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, early intervention programs and parental educational programs. This comprehensive process must be administered by a multidisciplinary team of individuals such as audiologists, parents, physicians, educators, speech/language pathologists, nurses and early interventionists. All professionals involved in the process should have experience, knowledge and skills to provide comprehensive and appropriate services to infants and toddlers who may be deaf or hard of hearing.

Early and consistent monitoring of the hearing loss and enrollment in Early Intervention services once infants have been diagnosed is the key to achieving optimal communication, academic, cognitive and social-emotional development.

The goal of Pennsylvania's Newborn Hearing Screening Program is to provide appropriate and timely screening to improve the quality of life for those infants identified with permanent bilateral or unilateral Sensorineural or conductive hearing loss.

Therefore, consistent with National Early Hearing Detection and Intervention (**EHDI**) initiatives and the recommendations of the Joint Committee on Infant Hearing (**JCIH**) in 2007, the components of Pennsylvania's program are:

- Availability of initial and repeat hearing screenings for all infants before one month of age;
- Appropriate audiological and medical evaluations for all infants to confirm hearing loss before three months of age;
- Immediate access to high-quality technology for all infants with confirmed hearing loss including hearing aids, cochlear implants and other assistive devices when appropriate.

- Referral to local Early Intervention (EI) as soon as a diagnosis of hearing loss is confirmed, before 6 months of age with services to be provided by professionals knowledgeable about childhood hearing loss;
 - Ongoing monitoring for hearing loss in the medical home through continued communication developmental assessments of infants and children both with and without risk indicators for late-onset or progressive hearing loss; and,
 - Family-centered services, access to information through culturally-sensitive approaches, informed choices, parent consent and the guarantee of infant and family rights and privacy.
- 

RECOMMENDED GUIDELINES FOR HEARING 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.

HOSPITAL BIRTHS

Each birth hospital (also known as a hearing screening facility) shall designate a professional to be responsible for the newborn hearing screening program in that facility. Preferably an audiologist (JCIH 2007), this professional will act as a single point of contact between the hearing screening facility and the Department of Health's Newborn Hearing Screening Program. The hearing screening facility is responsible for ensuring that all screening personnel are appropriately trained to carry out newborn hearing screening using appropriate technology. Assessment of competencies should be conducted at least annually for personnel involved in administering the screenings. It is recommended that a licensed audiologist with appropriate training and experience or Audiology Center should advise the hospital about all aspects of the newborn hearing screening program, including screening and tracking. The Department of Health can provide the names of audiologists with expertise in newborn hearing screening to hospitals. Upon request by a hearing screening facility, the Department of Health can have the hospital's protocols for newborn hearing screening reviewed by a licensed audiologist (see page 28 for information on contacting the Department of Health).

OUT-OF-HOSPITAL BIRTHS

Midwives assisting in out-of-hospital births may participate in the out-of-hospital hearing screening program by obtaining screening equipment through the Department of Health. Participation in the out-of-hospital hearing screening program is dependent on birth volume by geographic concentration. The licensed free-standing birth centers are all equipped with hearing screening equipment.

Infants can be referred to participating midwives or the licensed birth centers to have the initial hearing screening. Infants can also be referred to participating midwives or the licensed birth centers to have the repeat hearing screening if the baby does not pass the

initial hearing screening. An infant can also be referred to any local hospital that provides hearing screenings.

SCREENING TECHNOLOGIES

The Joint Committee on Infant Hearing (**JCIH 2007**) recommends two technologies for use in newborn hearing screening programs: the automated auditory brainstem response (A-ABR) and evoked otoacoustic emissions (OAE). Some technology may be more appropriate for one program than another. Factors to consider include test performance characteristics such as test sensitivity for detecting various permanent forms of hearing loss and auditory disorders, test specificity for identifying infants with normal hearing (low sensitivity in the presence of transient outer ear conditions and middle ear effusion) and the cost of equipment and supplies.

In the well-baby nursery/out-of-hospital births, OAE and A-ABR screening tests may be used as the sole screening technology or in combination: a 'two-stage' screening protocol wherein a 'not pass' outcome of one technology (OAE) results in immediate re-screening using a second technology (A-ABR). The result of the A-ABR screening ('pass'/'not pass') indicates whether there is a need for follow-up.

In the NICU, only A-ABR technology is recommended for use in hearing screening programs because of the high incidence of neural hearing loss in this population (**JCIH 2007**). For these infants, a 'not pass' A-ABR screening requires an immediate referral to an audiologist for further testing.

There are two screening technologies currently available:

A. AUDITORY BRAINSTEM RESPONSE (ABR)

ABR is an electrophysiological measure of the auditory nervous system's response to sound. A sound (e.g., click or tone) is presented to the ear via eartips and electrodes record the response as the nerve impulse travels from the cochlea (inner ear) through the auditory nervous system to the brainstem. ABR tests are best interpreted by an audiologist to determine if further testing is needed.

For the purposes of hearing screening, automated ABR (A-ABR) equipment has been developed. A-ABR screening equipment provides fully automated results interpretation (only a 'pass'/'not pass' outcome). Consequently, the A-ABR allows for others besides audiologists to perform the hearing screening. Other

personnel that can be trained include nurses, midwives, technicians, support staff and volunteers.

- A-ABR may miss a small percentage of hearing losses, such as a mild hearing loss or a low frequency or high frequency hearing loss.
- A-ABR is sensitive to identifying those children at risk for sensory, conductive and neural (auditory neuropathy/auditory dysynchrony) hearing loss/disorders.

B. OTOACOUSTIC EMISSIONS (OAE)

Otoacoustic emissions evaluate the integrity of the outer hair cells in the cochlea. Thus, the test can identify infants at risk for sensory hearing loss. A sound is presented into the ear canal and a small microphone measures the response from the infant's cochlea (inner ear) that is evoked by the test signal(s). The audiologist analyzes the response to determine how well the inner ear sensory cells are functioning. There are two types of OAE technologies: Transient Evoked Otoacoustic Emissions (TEOAE) and Distortion Product Otoacoustic Emissions (DPOAE).

OAE testing can be fully automated for the purposes of hearing screening and providing a 'pass'/'not pass' outcome.

- OAE may miss a mild hearing loss and children that are at risk for auditory neuropathy/dyssynchrony.
- OAE responses indicate the status of the peripheral auditory system extending to the outer hair cells of the cochlea. OAE results are sensitive to sensory hearing loss greater than 40 dB in addition to transient outer ear or middle ear conductive hearing loss.

Both ABR and OAE technologies require that the newborn be asleep or in a quiet state to maximize the probability of a reliable and valid result. If the baby is awake, he/she must be in a quiet state, i.e., not sucking or moving.

Whenever possible, all screening should be conducted in an environment where the acoustical and electrical interference is kept sufficiently low to avoid negatively influencing the test results. Therefore, careful selection of the test room or area may be necessary to achieve satisfactory environmental conditions.

1. HEALTHY NEWBORNS – SCREENING IN THE NEONATAL (WELL-BABY) NURSERY AND OUT-OF-HOSPITAL BIRTHS:

One screening technology alone (either OAE or A-ABR) may be used for hearing screening for well-babies. Programs may also decide to use a two-stage screening protocol (typically, an OAE screening not pass is followed by A-ABR). The decision regarding use of a single technology or a two-stage hearing screening protocol is based on the goals and resources of the individual neonatal hearing screening program.

To prepare for the screening:

- Test the newborn no sooner than 12 hours after birth.
- Choose a time when the infant is not being seen by other healthcare professionals.
- Test when infants are quiet or sleeping, optimally one hour following feeding.
- To help calm a restless infant, swaddle the infant and dim the lights.
- Select a time when the infant is medically stable.
- Ensure a quiet environment for testing, away from background noise.
- Follow standard precautions for infection control (e.g., hand washing, appropriate cleansing of equipment, etc.).

If a newborn does not pass the screening, it is acceptable to repeat the screen one time during the same session with the same technology, especially in those cases when the recording conditions were not optimal either due to timing,

noise conditions or state of the newborn. Multiple screenings (greater than two times) during one session with the same technology are not recommended in order to avoid getting a false positive result i.e., not a true 'pass'. Screening results at individual frequencies should never be combined across screening sessions in order to obtain a 'pass' result.

An infant that does not complete an initial hearing screen for any reason should be scheduled for an appointment to return for completion of the hearing screen.

2. HIGH RISK NEWBORNS - SCREENING IN THE NICU:

Since the incidence of sensory as well as neural (auditory neuropathy/auditory dysynchrony) hearing loss is approximately ten times higher in the NICU versus well-baby nursery, **A-ABR is the recommended screening technology for use in the NICU population (JCIH 2007).**

The neonatologist and/or the PCP should be consulted to determine the most appropriate opportunity for hearing screening. Babies should be screened the week prior to discharge to assure the hearing screening occurs.

The A-ABR, however, should not be completed before the infant is 32 weeks gestational age because it tends to generate more referrals for diagnostic testing. Therefore screening should be delayed until the baby has reached this age.

For those infants transferred between facilities, **the facility discharging the baby to home is responsible for completing the screening, arranging any necessary follow-up, and reporting the outcome to the Department of Health.**

Any NICU infant who fails initial A-ABR neonatal hearing screening should receive direct referral to an audiologist for a comprehensive audiological evaluation that includes a diagnostic ABR (JCIH 2007).

3. PARENT INFORMATION AND FOLLOW-UP

BEFORE INITIAL SCREENING:

Families should receive written information regarding the importance of newborn hearing screening prior to testing. Parents should be informed about what test(s) is/are going to be done on their infants. Families have the right to

refuse the hearing screening. The parent should, however, be made aware of the benefits of screening or risk of choosing not to screen. If the parent refuses, the refusal shall be documented in writing, made a part of the medical records of the infant and reported to the Department of Health (see DOH Reporting section on pages 28-29).

The hearing screening facilities (birth hospitals, birth centers and midwives) should identify a primary care physician (PCP) for each newborn by consulting with the family prior to discharge. However, if the family has not chosen/designated a PCP, the hearing screening facility is encouraged to provide the newborn's family with recommendations for designating a PCP.

FOLLOWING THE INITIAL SCREENING:

COMMUNICATION WITH PARENTS

The person performing the hearing screening should give parents the following information in clear, non-technical terms and in the native language of the family:

- Provide results of the test whether the newborn passed or did not pass the hearing screening and explain what the results mean. The results should be conveyed verbally and in writing to the parents using a standard notification form prepared explicitly for the purpose.
- Provide written notification to parents if an infant needs a repeat hearing screening. If the infant does not pass the initial screening the parents should understand the importance of obtaining a repeat hearing screening as soon as possible.
- Schedule the repeat hearing screening appointment with parents prior to hospital discharge if an infant does not pass the initial screening.
- Provide an opportunity for a hearing screening after discharge if the initial screening was not completed (e.g., the baby was missed or results were incomplete) prior to discharge. This includes making an appointment for the

parent prior to discharge to return to the birth center for the hearing screening.

- Provide all parents with information on milestones of normal auditory, speech and language development in children. See reference section for sample brochure.

COMMUNICATION WITH PRIMARY CARE PROVIDER (PCP)

The hearing screening facility (i.e., birth hospital, birth center or midwife) must notify the **primary care provider** via newborn summary, discharge summary sheet, letter or other specific written means that the newborn passed, did not pass or missed the initial hearing screening, that the screening results were incomplete, or that the parent refused the newborn screening and that a repeat hearing screening or referral for diagnostic evaluation by an audiologist is needed.

COMMUNICATION WITH DEPARTMENT OF HEALTH

The hearing screening facility (i.e., birth hospital, birth center or midwife) must provide the **Department of Health** (DOH) with data concerning the number of newborns screened or not screened on the appropriate reporting form (see DOH Reporting section on pages 28-29).

REPEAT HEARING SCREENING

Newborns that did not pass, missed the initial hearing screening or those whose results were considered incomplete should be referred by the hearing screening facility to have a repeat hearing screening performed within **30 days of life**. With respect to procedure and the newborn's mood and disposition, the same conditions that apply to initial hearing screening also apply to the repeat hearing screening (i.e., sleep is needed to maximize the probability of a valid result).

For the repeat hearing screening, a complete screening of both ears is recommended regardless of whether or not a pass was recorded in one ear at the time of the initial screening (**JCIH 2007**).

If a well-baby did not pass the initial screening using A-ABR, the infant should not be re-screened with OAE technology alone because of the risk that a neural hearing loss (auditory neuropathy/dyssynchrony) might be missed by such practice (**JCIH 2007**).

Parents may refuse the repeat 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 of Health on the Newborn Hearing Screening Program Reporting Form (see DOH Reporting section on pages 28-29).

A newborn who does not pass the initial and repeat hearing screenings within 30 days of birth or a NICU infant who fails an initial A-ABR hearing screening should be referred by the hearing screening facility to a pediatric audiologist for a diagnostic evaluation and reported to the Department of Health. NOTE: Insurance companies may require a referral from the PCP for a diagnostic evaluation.

Note that the Joint Committee on Infant Hearing (2007) recommends that a hearing screening program (initial and repeat combined) achieve a **refer rate to comprehensive audiologic assessment of <4 percent**. The Pennsylvania Infant Hearing Screening Advisory Committee recommends that the refer rate should be no lower than 1% percent.

RECOMMENDED GUIDELINES FOR PRIMARY CARE PROVIDERS

It is the PCP's responsibility to ensure that an infant has received a hearing screening and to know whether the infant passed or did not pass the hearing screening. If the infant did not pass, the PCP should make a referral to an audiologist for a comprehensive audiological evaluation. This evaluation should be done by an audiologist capable of performing a full range of diagnostic audiological tests on infants (see pages 18-21 for description of a comprehensive audiological evaluation). If necessary, the Department of Health, in collaboration with the PCP, will contact the family to encourage necessary follow-up and referral to appropriate services.

PRIMARY CARE PROVIDER (PCP) ROLE

The primary care provider in cooperation with the audiologist directs and coordinates, as needed, the evaluation and referral process within the child's medical home by:

- Referring the newborn for comprehensive audiological evaluation;
- Providing a statement to parents stressing the importance of follow-up, the time and location of the follow-up appointment, and the telephone number of the screening audiology center;
- Referring a baby diagnosed with hearing loss 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;
- Monitoring individual cases to assure that the diagnostic hearing evaluation was completed and facilitating the infant's receipt of amplification if needed and linkage to Early Intervention services; and,
- Providing updates regarding the infant's hearing status to the Department of Health upon request.

COMMUNICATING WITH PARENTS

The need for a child to have a repeat hearing screening or a diagnostic audiological evaluation should be conveyed to parents in a clear and easy to understand terms. PCPs should provide this information in a supportive and confidential environment

with time allowed to answer questions. It is also helpful to provide this information in writing in case parents have questions later.

Parent support resources should be provided to parents of children diagnosed with hearing loss. Hands & Voices Guide By Your Side (GBYS) of PA is a specialized parent support program that links infants and toddlers newly identified with deafness and hearing loss throughout Pennsylvania with trained and experienced parent guides. Parent guides are also parents of children with deafness and hearing loss who provide unbiased information, emotional support and a perspective of optimism and hope on a shared journey. Services are free and confidential. GBYS is funded through the PA Department of Health. Contact GBYS at 1-800-360-7282, ext. 3908 or on the web at www.paearlyhearing.org/gbys.php.

ONGOING SURVEILLANCE AND RISK INDICATORS FOR LATE ONSET HEARING LOSS

Many infants in the NICU and some in the neonatal nursery have risk indicators that place them at risk for late onset hearing loss. Some neonates that pass newborn hearing screening may actually have hearing loss, and use of risk indicators (specifically, family history and parent/caregiver concern regarding communication development) may assist in identifying such infants.

The **JCIH (2007)** recommends that the timing and number of hearing evaluations for children who have one or more risk indicators be individualized depending on the likelihood for delayed onset hearing loss. In the table below, those conditions placing infants at increased risk for late onset hearing loss are indicated by an asterisk (*). Regardless of increased risk for late onset hearing loss, the **JCIH (2007)** recommends that infants who have a risk factor should have at least one comprehensive audiologic evaluation by 24 to 30 months of age.

Consistent with the American Academy of Pediatrics (AAP) pediatric periodicity schedule, it is recommended that all infants should have a standardized screening of global development with a validated testing tool at 9, 18, and between 24 to 30 months of age. Testing should also occur in the medical home at any time there is parental or professional concern regarding communication development.

Joint Committee on Infant Hearing (JCIH) 2007 Risk Indicators Associated with Permanent Early Onset and or Late Progressive Hearing Loss in Childhood. *: Indicates risk indicators are of greater concern for delayed onset hearing loss. (Joint Committee on Infant Hearing Year 2007 Position Statement. Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Pediatrics* 2007; 120:898-921).

Caregiver concerns	Any concerns* regarding hearing, speech, language or developmental delays
Family history	Family history* of permanent childhood hearing loss
NICU >5 days	Neonatal intensive care of more than 5 days, or any of the following regardless of length of stay: extracorporeal membrane oxygenation (ECMO)*, assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia requiring exchange transfusion
In-utero infections	In-utero infections such as cytomegalovirus (CMV)*, herpes, rubella, syphilis, and toxoplasmosis
Craniofacial anomalies	Craniofacial anomalies including those involving the pinna, ear canal, ear tags, ear pits and temporal bone anomalies.
Physical findings	Physical findings such as white forelock, associated with a syndrome known to include Sensorineural or permanent conductive hearing loss
Syndromes	Syndromes associated with hearing loss or progressive hearing loss or late onset hearing loss such as neurofibromatosis, osteoporosis, and Usher syndrome. Other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
Neurodegenerative disorders	Neurodegenerative disorders*, such as Hunter syndrome or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome
Postnatal infections	Culture-positive postnatal infections associated with Sensorineural hearing loss*, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
Head trauma	Head trauma, especially basal skull/temporal bone fracture* requiring hospitalization
Chemotherapy	Chemotherapy*

RECOMMENDED GUIDELINES FOR AUDIOLOGISTS

The goal of newborn hearing screening is early detection and intervention. A diagnostic audiologic evaluation, which provides ear-specific information regarding degree, configuration and type of hearing loss, should be completed by 3 months of age or as soon as possible for babies who have been in the NICU. Habilitation, including fitting amplification and initiation of 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 three 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 audiologic tests. The audiologist is also responsible for ongoing 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 the required equipment and facilities to evaluate infants and children.

AUDIOLOGIST ROLE

- Completes initial comprehensive audiologic evaluation using a battery of physiological testing procedures (**JCIH 2007**);
- Reports the results of the diagnostic hearing evaluation to the child's PCP, including information that an infant was not successfully tested after being referred for testing;
- Reports to the Department of Health, 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;
- Makes a referral to Early Intervention for children diagnosed with permanent hearing loss and reports the date of referral to the Department of Health;
- Provides parents information about hearing and hearing loss and communication and education options for children who are deaf or hard of hearing; and,

- Fits children with amplification, including hearing aids or other assistive technology; refers for consideration of cochlear implant candidacy when appropriate.

AUDIOLOGIC ASSESSMENT

The audiologic assessment should include not only the behavioral and physiologic tests of hearing, but also 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 acoustic immittance measurements.

ASSESSMENT TYPES AND TECHNOLOGY

- For very young children, a complete assessment of hearing may not be completed in one session, yet the goal is to minimize delays and time between assessments.
- Incomplete audiologic information should not delay the referral to Early Intervention services or audiologic habilitation. Rather, habilitation, including the fitting of amplification, should be initiated, with refinements and adjustment of the hearing aid fitting 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 (preferably 2000 Hz) stimulus to allow for selection of appropriate amplification. If hearing aids are initially recommended based on thresholds limited to one low and one high frequency ABR threshold, other thresholds should be obtained as soon as possible to ensure appropriate amplification across the speech frequencies.
- Insert earphones or 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. While not sufficient for hearing assessment, this provides valuable information on middle ear function in conjunction with other audiologic results. The use of a 1000 Hz probe tone frequency

has proven a more valid indication of middle ear state for children less than 6 months of age (JCIH 2007).

DIAGNOSTIC AUDIOLOGIC EVALUATION

1. 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.

2. OTOSCOPY

This should be performed to ensure ear canals are clear of occlusion.

3. PHYSIOLOGIC TESTING

a. Acoustic Immittance

I. Tympanometry -- should be conducted to obtain information on middle ear status while using a 1000 Hz probe tone frequency for children less than 6 months of age. Ear canal volumes should be measured using 226 Hz, but the tympanogram should be interpreted using the higher frequency probe tone only.

II. Acoustic reflexes -- Acoustic reflexes can provide additional information regarding possible pathology, e.g., middle ear fluid, auditory neuropathy. If acoustic reflexes are evaluated in an infant less than 6 months of age, a 1000-Hz probe frequency should be used for the measure. Care should be taken to consider the unique outer ear acoustic characteristics of the infant ear in the selection of the maximum sound pressure level of the evoking stimulus.

b. Auditory Evoked Potentials – are collected when the infant is in natural or sedated sleep. Sedation should only be used in those facilities that have a comprehensive written sedation policy that outlines the steps required to ensure patient safety. If sedation is used, necessary equipment for life support (e.g., pediatric equipped crash cart), equipped facilities (e.g., oxygen and suction-trained certified personnel), and

appropriate monitoring are required. Sedation should only be administered at the testing facility and should never be administered outside of the medical facility.

I. ABR -- The goal for the ABR is to provide sufficient information to estimate the audiogram. Responses to frequency specific stimuli and at minimum, 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 due to child awakening during testing.

1.) Click-evoked ABR via air conduction using insert earphones

Testing multiple intensities is needed to plot the latency-intensity function. Threshold interpretation is based on individual clinic normative data. Absolute and interwave latencies and response morphology should be evaluated. Specifically the absolute latencies for waves I, III, and V at 70 dBnHL or higher should be evaluated. In addition, waveform morphology and interpeak latencies for waves I-V at 70 dBnHL or higher should be evaluated to assess retrocochlear function. If no reproducible waveform is present, attempts should be made to record the cochlear microphonic. The cochlear microphonic can be recorded by obtaining separate responses to rarefaction and condensation and overlapping the tracings to identify the cochlear microphonic (CM) [out of phase]. The CM needs to be differentiated from stimulus artifact by clamping the sound tube.

2.) Frequency-specific air-conduction testing

The goal for using tone burst ABRs is to get sufficient information to be able to fit a hearing aid and, at minimum, include obtaining responses to low frequencies and a higher frequency. The most important frequencies to obtain are 500 Hz and 2000 Hz. If either of these response levels is elevated, obtain thresholds at 1000 Hz and 4000 Hz.

3.) Bone-conducted testing, if indicated

When air-conducted thresholds are elevated, bone conduction testing should be completed.

II. Auditory Steady State Responses (ASSR) can be completed as an adjunct to tone burst ABRs, to supplement information regarding frequency specific information or in lieu of tone bursts to help to determine the degree of hearing loss. At this time, ASSR should not be used as the sole measure for estimating auditory threshold sensitivity in infants for air- or bone-conducted stimuli.

III. Evoked Otoacoustic Emissions (TEOAE OR DPOAE) Evoked otoacoustic measures should be made in conjunction with all ABRs to assess both cochlear and neural response.

4. BEHAVIORAL AUDIOLOGIC EVALUATION

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

For children less than 6 months developmental age, reliable behavioral hearing assessment procedures are not clinically available. For ages less than 6 months, behavioral observation audiometry (BOA) can provide information on the type of auditory response a child makes, as well as his or her auditory development, but should not be used to estimate thresholds. At this age, physiological test measures only should be used to establish thresholds with behavioral data supplementing the physiologic data.

For children older than 6 months developmental 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 if a hearing loss exists. 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 and physiologic findings i.e., click-evoked and tone-evoked ABR thresholds, acoustic immittance measures, evoked otoacoustic emission, and bone conduction thresholds (behavioral and/or ABR) are essential.

5. OUTCOMES AND CONFIRMED HEARING LOSS FOLLOW-UP

If a hearing loss is diagnosed, the following steps should be taken:

- a. Counsel parent(s): Counseling should include informational and emotional support counseling to families (ASHA 2008). This includes a review of the results of the diagnostic audiologic assessment, implications of the audiologic diagnosis and information to parents on the following:
 - Amplification options and initiation of the amplification fitting process;
 - Communication and educational options;
 - Funding assistance and assistance in the completion of an application to Medical Assistance to obtain hearing aids;
 - Importance of Early Intervention and the CONNECT Helpline number: 1-800-692-7288;
 - Explanation of various medical specialty evaluations that might be recommended; and,
 - Parent resources, especially resources that connect them to other parents of children with hearing loss. Hands & Voices Guide By Your Side (GBYS) of PA is a specialized parent support program that links infants and toddlers newly identified with deafness and hearing loss throughout Pennsylvania with trained and experienced parent guides. Parent guides are also parents of children with deafness and hearing loss who provide unbiased information, emotional support and a perspective of optimism and hope on a shared journey. Services are free and confidential. GBYS is funded through the PA Department of Health. Contact the GBYS Program at 1-800-360-7282, ext. 3908.
- b. Provide medical follow-up (after explaining to parents the various medical specialty evaluations that might be recommended), including:
 - Medical clearance from an otolaryngologist
 - Recommendations with Medical Home approval for appropriate specialty evaluations:

- Pediatric otolaryngologist/otologist
 - Medical geneticist
 - Pediatric ophthalmologist
 - Cardiologist for some syndromes
- c. Refer to Early Intervention Services: Once a hearing loss has been identified, a referral should be made immediately, with parental permission, to Early Intervention services. Habilitation and Early Intervention should proceed concomitantly with the medical evaluation of the hearing loss and should not be delayed by completion of the medical evaluation and findings.
- d. Submit DOH form “Results of Diagnostic Audiologic Evaluation” for all infants seen for diagnostic evaluation.
- e. Refer parents to parent support group or communication with other parents of children with hearing loss including Hands & Voices Guide By Your Side of PA (1-800-360-7282, ext. 3908).

6. HEARING AID EVALUATION

The goal is to fit infants who have permanent sensory hearing loss with personal amplification within one month of confirmed diagnosis. An infant is considered to be a candidate for amplification if a permanent hearing loss equal to or greater than 25 dBnHL estimated by the ABR exists in one or both ears in the frequency regions critical for speech understanding (1000-4000 Hz). Although little empirical data exists regarding the degree of hearing loss at which there is a definitive need for amplification, the acoustic spectrum of speech at normal conversational levels would suggest that estimated hearing threshold levels of 25 dBnHL/(eHL) or greater can impede a child’s ability to perceive acoustic features of speech necessary for optimal aural/oral language development.

Selection and fitting of amplification for infants and children requires real-ear measurement equipment and the use of a pediatric prescriptive procedure for determining gain and output (e.g., Desired Sensation Level [DSL] or National Acoustic Laboratory-Revised [NAL-R]). Selection of the hearing aid instrument gain and output characteristics should be based on the

prescriptive targets. Use of manufacturer's proprietary algorithms for this purpose is discouraged.

Custom ear molds should be available at the time of the hearing aid selection and performance verification in order to measure the Real-Ear to Coupler Difference (RECD). The RECD allows 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. RECDs should be reassessed as the infant grows or whenever new ear molds are made.

Verification of the hearing aid settings should be completed using probe microphone measurements. Once the targets are verified (DSL or NAL-R) for the individual infant and the device is fitted, ongoing monitoring of hearing levels, real-ear measures and the amplification targets should take place, e.g., at least every three months during the first two years of life, semi-annually until age 3 and annually thereafter.

Hearing aids options for most children should be in keeping with the accepted recommendations for pediatric amplification fitting (AAA 2004). These include Direct Audio Input (DAI), telecoil (T) and microphone-telecoil (M-T) switches. Devices should also be flexible and have safety-related features such as tamper-resistant batteries 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.

RECOMMENDED GUIDELINES FOR EARLY INTERVENTION

REFERRALS TO EARLY INTERVENTION

1. Referrals from the follow-up audiologist, initial care physician or any other source must be made no more than two business days after the child has a confirmed hearing loss. Call **1-800 CONNECT** line (1-800-692-7288) with parental consent for direct referral.
2. Once the Early Intervention Program receives the referral, it appoints 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/case manager will make contact with the child's family to set up the initial home visit (intake).

EARLY INTERVENTION ENROLLMENT PROCESS

3. The initial home visit is used to determine the existence of previous evaluations and to discuss parent concerns. The service coordinator will review all pertinent records and information on the child (i.e., written professional reports such as audiologist reports, hospital neonatal discharge information, physician's 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. Within 45 days of the referral, a multidisciplinary evaluation (MDE) will be conducted by an independent evaluator and a written report provided to the parent. The report will include a determination of eligibility for Early Intervention services and recommendations for supports.
5. When the child is found eligible for Early Intervention services, a meeting is scheduled to develop an Individualized Service Plan (IFSP/IEP) for the child and family. The plan is

developed by a team, which includes the parent and may include the audiologist or PCP as appropriate.

A description of the frequency, duration and location of services needed to support the family's outcomes for their child is included in the plan. The service coordinator will continue to coordinate all activities with the family's PCP and will ask parents for consent to send copies of pertinent information to their PCP and for signed consent to send confirmation of Early Intervention linkage to the Department of Health (e.g., copy of the plan, MDE).

- In the event that the parent chooses not to enroll their child in Early Intervention services, a tracking/monitoring program may be developed between the family and the Early Intervention service coordinator.

PARENT RIGHTS AS PART OF THE EARLY INTERVENTION PROCESS

- Access to information including potential benefits and risks, in the family's native language;
- Input into decision making, creating a plan and confidentiality of the family and child's information;
- Right to accept or decline Early Intervention services and supports for each child within statutory regulations;
- Right to allow or refuse permission to release all communications regarding each child's test results, including medical home and Early Intervention; and,
- Right to problem-solve with the Early Intervention program if there is a dispute through a complaint resolution process, IFSP facilitation, mediation or due process hearing.

DEPARTMENT OF HEALTH – NEWBORN HEARING SCREENING PROGRAM

The responsibilities of the Newborn Hearing Screening and Intervention Program at the Department of Health are as follows:

- Collects and maintains data from hearing screening facilities, as well as the results of repeat hearing screenings and audiologic diagnostic testing for specific infants;
- 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 infants diagnosed with hearing loss to Early Intervention services;
- Provides training and technical assistance to hospital staff conducting the newborn hearing screening;
- Monitors hospital newborn hearing screening not pass rates;
- Conducts program 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 hearing screening performance result data to hospitals on a yearly basis;
- 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; and,
- Provides informational materials to hearing screening facilities, PCPs, and families.

Contact the Department of Health:

Pennsylvania Newborn Hearing Screening and Intervention Program
Bureau of Family Health
Division of Newborn Screening and Genetics
625 Forster Street
7th Floor, East Wing
Harrisburg, PA 17120-0701
717-783-8143 or 1-877-HEALTH (1-877-724-3258)
nbhs@pa.gov

DATA REPORTING TO THE DEPARTMENT OF HEALTH

System development for electronic tracking and reporting of newborn hearing screening are currently in process and ongoing. Until such time that an electronic reporting system is available for use, data reporting to the Department of Health will continue to follow the paper process described below.

HOSPITALS

Individual reporting

Newborn Hearing Screening Program - Screening Reporting Form—to be submitted immediately for each newborn not passing repeat hearing screening, and/or for each newborn who does return for repeat hearing screening within 30 days of birth. The Screening Reporting Form should be submitted to the Department of Health by the time the child is 30 days of age. Hospitals that do not see return appointments for repeat hearing screenings should submit the Screening Reporting Form for newborns not passing the initial hearing screening as soon as the child is discharged from the hospital.

Monthly reporting

Birth Hospital Monthly Reporting—to be submitted by the 15th of every month by fax, mail or email, covering births and initial screenings for the preceding month. Birth hospitals should update any incomplete data submitted on previous monthly reports with the submission of each new Monthly Reporting Form

OUT-OF-HOSPITAL BIRTH HEARING SCREENING PROGRAM

Individual reporting - should be submitted immediately for each newborn not passing repeat hearing screening and/or for each newborn who does return for repeat hearing screening within 30 days of birth (sample form in Reference section). The Screening Reporting Form should be submitted to the Department of Health by the time the child is 30 days of age.

Monthly reporting - Birth Center and Midwife Monthly Reporting Form—should be submitted by the 15th of every month by fax, mail or email, covering births and initial screenings for the preceding month. Birth centers and midwives should update any incomplete data submitted on previous monthly reports with the submission of each new monthly reporting form

AUDIOLOGISTS

Diagnostic reporting

State law mandates that audiologists send data to the Department of Health concerning the results of diagnostic testing for all infants who did not pass two independent hearing screens within 30 days of birth (for neonatal nursery graduates) or did not pass the initial A-ABR screening (for NICU graduates). Results are reported using the Diagnostic Evaluation Reporting Form.

EARLY INTERVENTION

Announcement EI-09#01, issued in January 2009, provides Early Intervention program administrators with guidance on services to children who are deaf or hard of hearing. The announcement reviews the recommendations related to services for deaf/hard of hearing infants/toddlers, preschool children with hearing loss and their families. A child is considered eligible for Early Intervention services if the child has any degree of diagnosed permanent hearing loss, including mild and unilateral losses and is in need of Early Intervention services. Every family should be asked if their child participated in newborn hearing screening and, if so, what the results indicated. A report of the results should be shared with the Early Intervention program. Every family should be asked if they give consent to share information about their child's eligibility for Early Intervention with referral sources. The Department of Health has developed an intake form to assist in the completion of sharing information. See the references and resources section for more information on the announcement.

REFERENCES AND RESOURCES

INFORMATION BULLETINS

The Department of Health provides guidance concerning both newborn hearing screening referrals and monthly reports in the form of Information Bulletins. To obtain a copy of these bulletins, please contact the Newborn Hearing Screening Program by telephone at 717-783-8143; or by email at nbhs@pa.gov.

MONTHLY HOSPITAL AND OUT-OF-HOSPITAL REPORTING TEMPLATES

To obtain a copy of monthly newborn hearing screening reporting templates please contact the Newborn Hearing Screening Program by telephone at 717-783-8143; or by email at nbhs@pa.gov

AUDIOLOGY DIAGNOSTIC REPORTING TEMPLATE

To obtain a copy of audiology diagnostic reporting template please contact the Newborn Hearing Screening Program by telephone at 717-783-8143; or by email at nbhs@pa.gov

EARLY INTERVENTION POLICY STATEMENT

Announcement EI-09 #01 was issued by the Office of Child Development and Early Learning. The purpose of this announcement is to review recommendations related to services for Infants/Toddlers and Preschool children with hearing loss and their families. The announcement can be viewed on the PA Department of Education website at: http://www.portal.state.pa.us/portal/server.pt/community/early_intervention/8710/p/1133294

JOINT COMMITTEE ON INFANT HEARING (JCIH) POSITION STATEMENT

Throughout its over 30-year history, the Committee explored the complexities of hearing loss and its effect on a child's development, seeking to find newer and better methods to identify and serve infants and their families. Today, the Joint Committee is comprised of representatives from the American Academy of Pediatrics, the American Academy of Otolaryngology and Head and Neck Surgery, the American Speech Language Hearing

Association, the American Academy of Audiology, the Council on Education of the Deaf, and Directors of Speech and Hearing Programs in State Health and Welfare Agencies.

The Committee's primary activity has been publication of position statements summarizing the state of the science and art in infant hearing, and recommending the preferred practice in early identification and appropriate intervention of newborns and infants at risk for or with hearing loss. The current position statement can be viewed on the JCIH website at www.jcih.org.

PA EARLY HEARING DETECTION AND INTERVENTION WEBSITE

The PA Early Hearing Detection and Intervention website contains information on newborn hearing screening, early hearing and speech in children from birth to 3 years of age. Information is provided for families, primary care physicians, birthing facilities, audiologists and others. The website address is www.paearlyhearing.org.

DEFINITIONS

- **Audiologist:** A healthcare professional specializing in the diagnosis and treatment of hearing and balance disorders. In Pennsylvania, an audiologist must be licensed by the commonwealth's Department of State.
- **Auditory brainstem response (ABR) and automated ABR (A-ABR):** An electrophysiological measurement of the electrical potential produced along the auditory nerve and brainstem in response to sound stimuli. The ABR test requires an audiologist for threshold interpretation. The A-ABR is an automated ABR that compares the response to a normal template and yields a 'pass' or 'not pass' result. The A-ABR test requires no interpretation and is frequently used to screen the hearing of newborn infants.
- **Comprehensive audiological evaluation:** An in-depth, age-appropriate evaluation of auditory function using behavioral, electrophysiologic and middle ear function measures to determine if a hearing loss exists. If a hearing loss is confirmed, the evaluation determines the type, degree and configuration of the hearing loss.
- **Conductive hearing loss:** A type of hearing loss that occurs when sound is not conducted efficiently through the outer ear canal to the eardrum and the tiny bones, or ossicles, of the middle ear. Is typically due to an obstruction. Conductive hearing loss usually involves a reduction in perceived sound level or the ability to hear faint sounds. This type of hearing loss can often be medically or surgically corrected.
- **Early Intervention (EI):** A program of services and supports designed to assist families in helping their children who have, or are at risk for, developmental delays and disabilities. The program builds upon family-centered, natural learning opportunities for infants, toddlers and preschool children who have special needs. The Office of Child Development and Early Learning (OCDEL) through the Bureau of EI, administers the program for the PA Departments of Public Welfare and Education.
- **Evoked otoacoustic emissions (OAE):** This is a hearing test appropriate for newborn infants. It measures a physiologic response from the sensory cells of the inner ear (cochlea) that is used to determine the integrity of the peripheral auditory system. The response may be elicited with the use of either click (transient evoked otoacoustic

emission) or tones (distortion product otoacoustic emission). This test can be automated to objectively provide a 'pass'/'do not pass' outcome.

- **Hearing loss:** A dysfunction of the auditory system of any type or degree. The target population of the EHDI program is infants with permanent forms of hearing loss sufficient to interfere with the acquisition and development of speech and language skills.
- **Hearing screening:** An objective physiological test performed to identify infants at increased risk for hearing loss. This is seen as a first step in testing and as a consequence it should capture most infants with hearing deficits. A small number of hearing infants will necessarily be falsely identified as having a hearing loss (i.e. it is expected that there will be false positives). Infants who do not pass the initial hearing screening in one or both ears should receive a repeat hearing screening. A 'not pass' outcome of the repeat hearing screening requires a referral for a comprehensive audiological evaluation. A 'not pass' outcome does not mean permanent hearing loss exists, only that specific physiologic response criteria for a 'pass' outcome were not met at the time of testing.
- **Hearing screening facility:** A facility such as the birth hospital or birth center that provides hearing screenings to newborns and reports the results of the hearing screenings to the Department of Health.
- **Infant:** A child age 30 days to 12 months.
- **JCIH:** Joint Committee on Infant Hearing. A national committee of professional organizations which, since 1969, proffers position statements and guidelines in support of Early Hearing Detection and Intervention (EHDI) Programs. Member organizations include, but are not limited to, Representatives from (in alphabetical order) the Alexander Graham Bell Association for the Deaf and Hard of Hearing; American Academy of Audiology; American Academy of Otolaryngology-Head and Neck Surgery; American Speech-Language-Hearing Association; Council on Education of the Deaf; Directors of Speech and Hearing Programs in State Health and Welfare Agencies (DSHPSHWA).
- **Midwife:** An individual educated and/or experienced to provide the necessary supervision, care and advice to women during pregnancy, labor and postpartum period, as well as to conduct deliveries and care for the newborn infant.

- **Newborn:** A child age 0 to 29 days.
- **NICU:** Neonatal Intensive Care Unit.
- **Neonatologist:** A pediatrician specializing in the diagnosis and treatment of disorders in newborns; one who works in neonatal intensive care units, providing care for premature infants or those born with infections or other health problems.
- **Neural hearing loss:** A form of hearing loss in which the root cause lies in the vestibulocochlear nerve (cranial nerve VIII) or central processing centers of the brain. Sensorineural hearing loss may include damage to the inner hair cells in the cochlea as well as the vestibulocochlear nerve. Neural or Sensorineural hearing loss is permanent and cannot be medically or surgically corrected. These hearing losses not only involve a reduction in perceived sound level, or ability to hear faint sounds, but may also affect speech understanding or ability to hear clearly. Causes of these types of hearing loss include genetic syndromes, drugs that are toxic to the auditory system, head trauma, and viruses.
- **Otolaryngologist (also known as ear, nose and throat or “ENT”):** A physician specializing in disorders of the ears, nose and throat.
- **Out-of-Hospital Birth:** A birth that takes place in a location other than a hospital, including but not limited to home births, free standing birthing facilities, etc.
- **Primary care provider (PCP):** The licensed healthcare provider to whom the infant will go for ongoing pediatric medical services.
- **Screener:** An individual trained to perform automated hearing screening tests (OAE and A-ABR).
- **Sensorineural hearing loss:** A type of hearing loss stemming from a lesion in the cochlea or in adjacent parts of the vestibulocochlear nerve. This type of hearing loss not only involves a reduction in perceived sound level or ability to hearing faint sounds, but may also affect speech understanding or ability to hear speech clearly. Causes of this type of hearing loss include genetic syndromes, drugs that are toxic to auditory system, head trauma and viruses.