- 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,