

UNIVERSAL NEWBORN HEARING SCREENING PROGRAM GUIDELINES

Public Act 91-0067, the Hearing Screening for Newborns Act, requires all hospitals performing deliveries to conduct hearing screening for all newborns prior to discharge. The following standards are based on current state practice, the recommendations of the Joint Committee on Infant Hearing Year 2000 Position Statement, the Policy Statement of the American Academy of Pediatrics, guidance available from the Resource Center for Universal Newborn Hearing Screening (www.infanthearing.org), guidance available from the Marion Downs National Center for Infant Hearing (www.colorado.edu/slhs/mdnc), and information obtained from state programs in Utah, Colorado, Virginia and Rhode Island.

I. NEWBORN HEARING SCREENING PROGRAM GOALS

- A. **All infants born in Illinois will have their hearing screened.**
- B. **All newborns referred from the Illinois Newborn Hearing Screening Program will have diagnostic testing completed by three (3) months of age.**
- C. **All infants diagnosed with significant hearing loss will receive appropriate treatment, including hearing aids, and Early Intervention services by six (6) months of age.**
- D. **All infants diagnosed with significant hearing loss will be referred to the Illinois Early Intervention System within two (2) days of diagnosis.**

II. DEFINITIONS

- A. **Audiologist:** a person holding an Illinois audiology license
- B. **Auditory Brainstem Response:** electrophysiologic measurement of the brainstem's response to the acoustic stimulation of the ear
- C. **Automated Auditory Brainstem Response (AABR):** objective electrophysiologic measurement of the brainstem's response to acoustic stimulation of the ear, obtained with equipment which automatically provides a pass/refer outcome
- D. **Diagnostic Audiological Evaluation:** for the purposes of this program, it is the physiologic and behavioral procedures required to evaluate and diagnose hearing status
- E. **DSCC:** University of Illinois at Chicago, Division of Specialized Care for Children
- F. **Hospital:** a facility licensed by the State of Illinois for birthing babies
- G. **IDHS:** Illinois Department of Human Services
- H. **IDPA:** Illinois Department of Public Aid
- I. **IDPH:** Illinois Department of Public Health
- J. **Medical Diagnostic Evaluation:** the examination and medical procedures provided by an otolaryngologist, otologist, or other qualified personnel to evaluate otologic status, for the purposes of this program
- K. **Otoacoustic Emissions Testing:** a specific test method which elicits a physiologic response from the outer hair cells in the cochlea, and may include Transient Evoked Otoacoustic Emissions (TEOAE) and/or Distortion Product Otoacoustic Emissions (DPOAE)
- L. **Otologist:** a physician who specializes in treatment of the ear
- M. **Otolaryngologist:** a physician trained in the medical and surgical management and treatment of patients with disease and disorders of the ear, nose, throat (ENT), and related structures of the head and neck

- N. Screening:** the completion of one or more objective, physiologic, electronic tests administered to determine the infant's bilateral hearing status and the need for further diagnostic testing by an audiologist or physician. Such screening is performed by individuals who have been appropriately trained in the procedure and instrumentation used by the hospital.

III. HOSPITAL SCREENING STANDARDS

A. Population

Public Act 91-0067, the Hearing Screening for Newborns Act, Section 5 requires all hospitals performing deliveries to conduct hearing screening of all newborn infants prior to discharge. While this act does not require the screening of infants born outside the hospital, the UNHS Standards Committee recommends that all birthing hospitals provide screening for those infants at the request of the parents or by the child's physician.

1. Beginning December 31, 2002, all hospitals performing deliveries will provide bilateral hearing screening to every newborn born in their institution. This hearing screening shall be provided prior to discharge.
2. Beginning December 31, 2002, if a newborn is transferred without written documentation of a completed hearing screening, such hearing screening will be completed by the receiving hospital prior to discharge.
3. Beginning December 31, 2002, all hospitals performing deliveries will make provisions to screen infants born in the home or other location outside the hospital when requested by the parents or by the child's physician.

B. Parental Information/Consent

The Hearing Screening for Newborns Act, Section 25 allows the parent or guardian to refuse testing on the grounds that the screening conflicts with his or her religious beliefs and practices. The Act requires that this refusal be made in writing. The Act implies that screening may not be refused by the parents for any other reason.

1. All hospitals shall provide information to the parents about newborn hearing screening that shall include: the purposes and benefits of newborn hearing screening; indications of hearing loss; what to do if the parent suspects a hearing loss; and procedures used for hearing screening.
2. Parents must provide written denial for hearing screening on the grounds that such test conflicts with their religious tenets and practices.

C. Documentation

1. The hospital shall provide written information to the parents and to the infant's primary care provider that includes procedures used for hearing screening, limitations of screening procedures, results of the hearing screening, recommendations for further diagnostic testing, where those diagnostic tests may be available, and resources available for these diagnostic tests.
2. The hospital shall maintain written documentation in the infant's clinical record. Such documentation shall include: procedures used for hearing screening, time and location for the screening, individual administering the screening test, screening results per ear for each and every screening or screening attempt and recommendations for further testing.

D. Personnel

1. Newborn hearing screening shall be performed by an individual who is appropriately trained and supervised. (This recommendation is not discipline-specific because the key to quality screening is appropriate training and supervision. It was felt that volunteers could be used as long as each one is appropriately trained and supervised. The experiences of other states support this recommendation).
2. Each hospital shall identify a liaison to the Universal Newborn Hearing Screening Program at the Department of Human Services and at the Department of Public Health.

E. Equipment

3. Technology utilized must:
 - a. Measure a physiologic response;
 - b. Be implemented with objective response criteria;
 - c. Measure the status of the peripheral or peripheral and central auditory system that is highly correlated with hearing status.

4. Acceptable methodologies for physiologic screening include evoked otoacoustic emissions (EOAE), either transient or distortion product, and auditory brainstem response (ABR), either automated or non-automated. These techniques can be performed either alone or in combination. Both are noninvasive, quick, and easy to perform, although each assesses hearing differently. The following are guidelines:
 - a. The methodology used should detect, at a minimum, all infants with unilateral or bilateral hearing loss greater than or equal to 35 dB HL.
 - b. The methodology used should have a false-positive rate (the proportion of infants without hearing loss who are identified incorrectly by the screening process as having significant hearing loss) of 3% or less.
 - c. The methodology used ideally should have a false-negative rate (the proportion of infants with significant hearing loss missed by the screening program) of zero.

F. Pass/Refer Criteria

Type of Test	Stimulus	Pass Criteria
Transient Evoked Otoacoustic Emission (TEOAE)	air conduction click	Testing in the 1500 through 3000 Hz range with replicability in the 70% or greater across that frequency range
Distortion Product Otoacoustic Emission(DPOAE)	pure tone complex Intensity = maximum levels < 65 dB SPL	F2=1000,2000,3000 & 4000 Hz 3 of 4 frequencies exceeding the noise floor
Automated Auditory Brainstem Response AABR	air conduction click	ABR response @ 35 dBnHL for both the right and left ears
Auditory Brainstem Response ABR	air conduction click	Replicable wave V thresholds for each ear less than or equal to 35 dBnHL

IV. REPORTING AND TRACKING STANDARDS

The Hearing Screening for Newborns Act, Section 10, requires hospitals to report information about each child with a positive hearing screening result to the Illinois Department of Public Health (IDPH). Section 15 requires the development of a registry of children with positive hearing screening results. Section 20(b) requires the development of a tracking and follow-up program for diagnostic hearing testing for those infants failing hospital-based screening.

A. Hospitals will report screening results to the Illinois Department of Public Health (IDPH).

1. Until electronic reporting is available, hospitals will report all required data on IDPH forms (IDPH Neonatal Hearing Screening Referral and IDPH Neonatal Hearing Screening Monthly Numeric Report).
2. On a monthly basis, every hospital shall report aggregate data on its universal newborn hearing screening activities. Such report shall include: number of live births, number of newborns screened, number of newborns passing screening, number of newborns failing screening, and number of newborns referred for further diagnostic testing.
3. Infant specific information shall be reported within 7 calendar days of the birth of the infant only for infants who fail the hearing screening, miss the hospital screening, or miss the hospital rescreening. The infant specific information will be used by IDPH for follow-up with the infant's family and physician. Each hospital shall provide identifying information including: infant's name, mother's name and address, name and address of infant's physician, and date of referral for further diagnostic.
4. Infants who pass the screening or whose parent refuses the screening shall not be individually reported because there is no need for follow-up.
5. Outpatient screenings, rescreenings, and audiologic follow-up must be completed and reported to IDPH within 30 days of the birth of the infant.

A. Hospitals will report screening results to the Illinois Department of Public Health (IDPH)

1. All infants born at birth hospital or transferred to the facility must be reported via electronic encrypted transfer file. Infant specific information shall be reported within seven (7) calendar days after the hearing rescreening for infants who do not pass the rescreening and for those who miss the hospital screening or rescreening. The infant specific information should be provided for all children who:
 - are missed
 - are referred in one or both ears
 - are transferred
 - are not tested due to written parental refusal (on religious grounds; only)
 - expired
2. The infant specific information will be used by IDPH for matching and follow-up with the infant's family and physician of record as reported by the hospital. Each hospital shall provide identifying information including infant's last name, infant's first name (or "Baby Boy"/"Baby Girl"), gender, mother/contact name, address, and phone number, mother's language, and identify the infant's outpatient physician who will be providing ongoing medical care (the infant's medical home).
3. For those hospitals that schedule and/or perform outpatient screenings or diagnostic testing, the testing should be completed and the results must be reported to IDPH within thirty (30) days after the testing of the infant.

B. IDPH will establish a registry of all infants as a result of the newborn hearing screening program.

The registry will include all infants born in or residing in Illinois.

C. IDPH will notify the infant's physician and parents of the need for follow-up.

1. Written notification will be sent to both the infant's physician listed on the hospital record and the parents within five (5) business days of the receipt of the hospital report.
2. Written notification will be made with prescribed IDPH letters.

D. Upon notification by IDPH, the child's physician will arrange for diagnostic testing.

1. The diagnostic test shall be performed within three (3) months of the infant's date of birth.
2. Written notification of test results shall be reported by the physician to IDPH within five (5) days of diagnosis.

E. Audiologists providing diagnostic evaluation must report findings to IDPH.

1. Persons who conduct any procedure necessary to complete an infant's hearing screening or diagnostic testing shall report this information to IDPH. Diagnostic testing results for each audiological visit shall be reported within thirty (30) days after testing.
2. Report must contain the information included on the IDPH Neonatal Hearing Screening Follow-up Services Report form.
3. Infants with confirmed hearing loss must be referred to the Early Intervention Program and to the Division of Specialized Care for Children (DSCC) within two (2) days of diagnosis.

F. When hearing loss is confirmed, IDPH will ensure that referral is made to the Early Intervention Program, to DSCC, and to the MCH Family Case Management agency.

G. IDPH will notify the local Perinatal follow-up agency, in writing, of infants with no reported diagnostic testing sixty (60) days after hospital report.

H. Local Perinatal follow-up agency will contact family.

1. Family will be provided information about the importance of early identification of hearing loss.
2. Assistance will be provided, as needed, in arranging for diagnostic testing.
3. Report will be made to IDPH of the results of the follow-up contact.
4. Infants with confirmed hearing loss will be referred to the Early Intervention Program and to DSCC within forty-eight (48) hours of receipt of the confirming report.

V. ACCESS TO DIAGNOSTIC TESTING STANDARDS

The Hearing Screening for Newborns Act, Section 20(d) requires an application process for financial assistance by the Division of Specialized Care for Children for follow-up diagnostic hearing testing of newborns failing hospital-based screening.

A. DSCC will provide assistance to families of infants referred from the UNHS Program to help them obtain diagnostic testing.

1. DSCC financial assistance for diagnostic services is available to families without regard to income. Families with insurance coverage are expected to use insurance benefits prior to DSCC payment. DSCC will coordinate benefits with insurance plans. Medicaid and KidCare will pay for children eligible for those benefits. DSCC cannot supplement Medicaid or KidCare.
2. Family use of DSCC is voluntary.
3. Otolaryngologists and audiologists meeting DSCC training and experience criteria will be used by DSCC to provide diagnostic testing. Lists of DSCC approved otolaryngologist and audiologists are available from DSCC Regional Offices.
4. DSCC will provide all agency-approved otolaryngologists and audiologists with the forms necessary to report findings and bill for payment of diagnostic services.
5. DSCC approved otolaryngologists and audiologists must report and bill on agency provided form (DSCC form 03.44, Diagnostic Evaluation Authorization for Genetic Diseases Program/Newborn Hearing Screening Program). Bills for children with Medicaid or KidCare must be billed to the Illinois Department of Public Aid.
6. Infants with confirmed hearing loss will be referred to the Early Intervention Program within two (2) days of the receipt of the confirming report.
7. Families of infants with confirmed hearing loss will be provided with medical care coordination and, if eligible, financial assistance for medical treatment and hearing aids.

B. IDPA will support diagnostic testing for Medicaid and KidCare eligible children.

Audiologists can bill directly for audiological services.

VI. ACCESS TO EARLY INTERVENTION (EI) SERVICES STANDARDS

Hearing Screening for Newborns Act, Section 20(c) requires the development of a referral system to early intervention services and hearing aids for those infants diagnosed with hearing loss. It should be noted, however, that family participation in the Early Intervention Program is voluntary.

A. IDEA Part C Referral Requirements

1. In order to comply with Individuals with Disabilities Education Act (IDEA), Part C, referrals of children with hearing loss potentially eligible for early intervention services must be made within two (2) days of confirmation of hearing loss.
 - a. Information must be provided to all audiologists and otolaryngologists about both their responsibility to refer children diagnosed with hearing loss to the Illinois Early Intervention System and the procedure for making such a referral.
 - b. All infants identified with hearing loss will be referred to the Child and Family Connections Offices, which are established to provide access to early intervention services for Illinois children with disabilities under the age of three.

B. Payment Responsibility

1. The federal hierarchy of payment will be followed to provide hearing aids or other needed services to infants identified with hearing loss.
2. Medicaid or KidCare is first payer for Medicaid supportable services.
3. Title V (DSCC) is second payer for DSCC supportable services.
4. Part C (EI Program) is the payer of last resort.

VII. PROGRAM EVALUATION AND PLANNING STANDARDS

Hearing Screening for Newborns Act, Section 20 (f) requires the Hearing Screening Advisory Committee to monitor any reports available to the State with respect to the hearing screening status of all newborns. Additionally, hospitals and local communities will need reports to evaluate and plan.

A. IDPH will provide reports for program evaluation and planning.

1. Using the software provided to them by IDPH, screening hospitals will be taught how to run numeric reports of the screening activity at their hospital.
2. UNHS Advisory Committee will be provided numeric reports at scheduled Advisory Committee meetings.
3. IDHS and DSCC will be provided numeric reports of program activity quarterly.

B. DSCC will provide aggregate reports on referrals received and children served as a result of the Universal Newborn Hearing Screening Program.

VIII. TRAINING AND EDUCATION STANDARDS

Hearing Screening for Newborns Act, Section 20(a) and (e) require the development of educational materials and training programs for hospital staff, health care professionals, and parents about the newborn hearing screening program and the follow-up procedures for infants referred from the program.

A. IDHS will coordinate the development of educational materials and training programs.

B. Interagency training programs will be provided about the tracking and follow-up procedure.

IX. DIAGNOSTIC TESTING GUIDELINES

It is the experience of other states that as many as 80% of the newborns referred from newborn hearing screening will pass a rescreen at a later date. Based on this information, it is recommended that all infants who are referred from a newborn hearing screening program because they failed two (2) hearing screenings in one or both ears should be seen for an outpatient ABR rescreen or an Evoked Otoacoustic Emissions rescreen. If the infant doesn't pass the outpatient rescreen, then a full diagnostic audiological assessment should be completed at the same appointment. It is suggested that the rescreen be conducted with a methodology different than that used in the hospital. For example, if the infant is screened with ABR equipment in the hospital, the outpatient rescreen should be done with Evoked Otoacoustic Emissions equipment.

Within the first two (2) months of life, the following procedures should be completed on all infants referred from the newborn hearing screening process.

A. Auditory Brainstem Response or Otoacoustic Emissions Rescreen Guidelines

Depending on the availability of equipment one of the following should be conducted:

1. Rescreen with an Auditory Brainstem Response Screen.
 - a. Obtain a 70 or 75 dBnHL response to click stimulus to assess the latency and morphology of waves I, III, V, I-III, and I-V.
 - b. Obtain a 30 or 35 dBnHL response to click stimulus to access latency and morphology of wave V.
2. Rescreen with an Automated Auditory Brainstem Response Test (AABR).
3. Rescreen with Evoked Otoacoustic Emissions.
 - a. Obtain Transient Evoked Emissions and/or
 - b. Obtain Distortion Product Otoacoustic Emissions.
4. Infants who do not pass the rescreen in one or both ears should receive an audiological diagnostic assessment at the same appointment.

B. Audiologic Diagnostic Assessment Guidelines

In 1999, the Illinois legislature passed the Hearing Screening for Newborns Act. By December 31, 2002, all hospitals delivering babies will be required to provide hearing screening to all babies born in their facility. The goals of the Illinois program are:

1. All infants born in Illinois will have their hearing screened.
2. All newborns referred from the Illinois Newborn Hearing Screening Program will have diagnostic testing completed by three (3) months of age.
3. All infants diagnosed with significant hearing loss will receive appropriate treatment, including hearing aids and Early Intervention Program services by six (6) months of age.

As of May, 2000, ninety-three (93) Illinois hospitals are providing universal newborn hearing screening. This is only the first step in the identification of babies with hearing loss, however. Screening is not very useful if those who are referred for diagnostic testing are not able to receive appropriate diagnostic services.

It is essential that audiologists to whom babies are referred for diagnostic evaluations have experience using age-appropriate diagnostic techniques for such infants. Various organizations such as the American Speech-Language-Hearing Association have produced guidelines that can be helpful. In Illinois, the Department of Human Services Newborn Hearing Screening Advisory Committee has recommended diagnostic audiological guidelines for the state. The purpose of this document is to inform audiologists testing Illinois infants of those guidelines.

It is also important for families of babies with suspected hearing loss have access to financial assistance so that no infant goes without diagnostic testing, medical treatment, or early intervention services due to financial reasons. All audiologists have encouraged to become providers for the Illinois Department of Public Aid (IDPA), the University of Illinois Division of Specialized Care for Children (DSCC), and the Illinois Early Intervention Program (EI).

All audiologists working with infants referred from newborn hearing screening programs are encouraged to adopt the following guidelines in their practice.

Significant Hearing Loss means a dysfunction of the auditory system of any type or degree that is sufficient to interfere with the acquisition of speech and language skills. The methodology used to evaluate infant hearing should detect, at a minimum, unilateral or bilateral hearing loss > 35 dB HL or > 30 dBnHL.

The use of a test battery is strongly recommended in the evaluation of infants and young children in order to provide a cross-check system for accurate diagnosis. It is preferred that infants be tested without sedation to the extent possible. If sedation is required, the audiologist, the patient's physician and the patient's family should discuss whether or not to sedate and should ensure that the appropriate protocol and monitoring is employed.

The use of a test battery is strongly recommended in the evaluation of infant and young children in order to provide a crosscheck system for accurate diagnosis. It is preferred that infants be tested without sedation to the extent possible. For infants under 12 months of age, the use of sedation is absolutely not appropriate to determine unilateral loss. Sedation is appropriate for infants under 12 months of age only when bilateral hearing loss is suspected.

1. Perform an otoscopic evaluation.
 1. Perform otoscopy.
2. Perform Auditory Brainstem Response/Auditory Evoked Potential (ABR/AEP):
 - a. Obtain threshold of each ear with air conductive click-evoked ABR utilizing <20 dB step threshold search. Including:
 - 1) Evaluate response at a high level (i.e., 70 to 80 dBnHL)
 - 2) Evaluate absolute latencies for waves I, III, and V, compare to established norms.
 - 3) Evaluate interpeak latencies for waves I to III, III to V, and I to V.
 - 4) Evaluate waveform morphology relative to normal neonatal waveforms.
 - 5) Consider measuring wave V/I amplitude ratio.
 - 6) Consider evaluating click-rate functions.
 - 7) Each clinic will need to establish its own normative values for infants. As a general guideline, normal sensitivity for clicks may be defined as a repeatable wave V or 30 dBnHL or less in each ear.
 - b. If the air conduction click ABR is abnormal, then perform a toneburst ABR at 500 Hz or 4000 Hz. Also obtain a bone conduction click ABR. Air bone gaps of 20dB or more or absent bone conduction response should be considered significant.
2. Perform Auditory Brainstem Response/Auditory Evoked Potential (ABR/AEP): Obtain ABR threshold in each ear using a click or high frequency (e.g. 4000 Hz) tone-burst stimuli. Actual thresholds should be determined in steps no greater than 10 dB, although larger step sizes may be appropriate early in the evaluation to initially bracket threshold region.

If ABR thresholds in response to click or high frequency tone-bursts are abnormal, ABR thresholds in response to tone-burst stimuli centered at 1000 Hz or lower should be assessed. An abnormal ABR threshold in response to click stimuli is defined as > 30 dBnHL. In some cases, the use of bone conducted stimuli may also be appropriate.

When appropriate, analysis of ABR morphology and waveform latencies in response to high intensity stimuli may be performed. Latencies may be compared to established norms. Additional measures may also be utilized as a part of an assessment for auditory neuropathy or other neurologic dysfunction of the auditory pathway.

3. Evaluate evoked otoacoustic emissions in each ear (TEAOE and/or DPOAE) to further evaluate cochlear function. OAEs may be evaluated even when click ABR thresholds are normal since it is possible to obtain frequency specific information for the low frequencies.
4. Behavioral testing should be conducted, as well, for normally developing infants over 6 months of age. Behavioral procedures should include separate-ear visual reinforcement procedures.
4. In addition, behavioral testing and audiometry should be considered, particularly in infants with suspected hearing loss.
5. If the click-evoked ABR thresholds are normal and OAEs are normal, the parents should receive written information about hearing, speech, and language developmental milestones and information regarding risk indicators for progressive hearing loss. If questions arise about the infant's hearing, speech, and/or language development, the infant should be referred for an age appropriate audiologic assessment.
5. All parents should receive written information about hearing, speech, and language developmental milestones and information regarding risk indicators for progressive or late onset hearing loss. If questions arise about the infant's hearing, speech, and/or language development, the infant should be referred for an age appropriate audiologic assessment.
6. Incorporate an immittance battery with caution for infants less than four months of age. Multi-component/multi-frequency testing is the only appropriate immittance test for this age group.
7. Complete the "Neonatal Hearing Screening Follow-up Services Report" and mail it to the Illinois Department of Public Health - Newborn Screening Program (attached).
8. If family does not have Medicaid or KidCare coverage for the infant and the family needs financial assistance to pay for the diagnostic evaluation, complete DSCC form 3.44 (Diagnostic Evaluation Authorization for Genetic Diseases Program/Newborn Hearing Screening Program) to obtain reimbursement from DSCC for diagnostic evaluation. If hearing loss has been confirmed, DSCC staff will contact the family to invite them to apply for assistance from DSCC.
9. If hearing loss is confirmed, refer the family to the appropriate Child and Family Connections office for early intervention services within 2 days of diagnosis.

C. Confirmed Hearing Loss Follow-Up Guidelines

The following should be completed by three (3) months of age:

1. If a bilateral/unilateral hearing loss of > 35 dB HL or 30 dBnHL in either ear is detected, refer the infant to an otolaryngologist for an examination and medical clearance (See Recommended Guidelines for Medical Protocol for Infants With Confirmed Hearing Loss). Begin the process of fitting amplification, if appropriate.

* Hearing loss is defined as an average of the frequencies 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz or if the hearing level of any two (2) of these frequencies is > 35 dB HL or 30 dBnHL.
2. If significant air-borne gaps are present or if other evidence of middle ear disease is seen, i.e., click-evoked ABR is normal, but OAE results are abnormal, refer the infant to an otolaryngologist for evaluation and treatment. Repeat the diagnostic evaluation following medical treatment.
2. If evidence of middle ear disease is present, refer the infant to an otolaryngologist for evaluation and treatment. Repeat the diagnostic evaluation following medical treatment.
3. If results indicate a mixed hearing loss, refer the infant to an otolaryngologist for evaluation, treatment, and medical clearance if appropriate. Begin the process of fitting amplification if appropriate and repeat the diagnostic evaluation following medical treatment.
3. If the ABR threshold is significantly elevated or the morphology is abnormal (i.e., prolonged interpeak latencies) and OAE results are normal, refer the infant to an otolaryngologist for evaluation of possible retrocochlear

dysfunction (auditory neuropathy). Refer to local Child and Family Connections and initiate appropriate intervention for auditory neuropathy.

4. If a unilateral hearing loss is detected with normal sensitivity in the “good” ear, provide the family with information regarding the effects of unilateral hearing loss on auditory, speech, and language skills, as well as the importance of hearing conservation for the “good” ear. Continued audiologic monitoring of the child’s hearing and speech and language development is recommended every six (6) months until the child is three (3) years of age. Also, consider amplification options and refer the infant to an otolaryngologist for evaluation, treatment, and medical clearance, if appropriate.
4. Defer the fitting of hearing aids, but consider referral to the local CFC office for early intervention services and consider the use of assistive listening devices (e.g., an FM system). Repeat the diagnostic evaluation at three to six month intervals to monitor neuromaturation.

D. Pediatric Amplification Guidelines

The following should be completed by six **three** months of age for infants with confirmed hearing loss.

1. Qualifications for Pediatric Hearing Aid Services

- a. A Medical Clearance must be obtained from an otologist, a pediatric otolaryngologist, or a general otolaryngologist prior to hearing aid fitting.
- b. An audiologist must complete the diagnostic audiologic assessment recommended in the Audiologic Diagnostic Assessment Guidelines section of this document.
- c. An audiologist is the professional singularly qualified to select and fit all forms of amplification for infants and children, including personal hearing aids, FM systems, cochlear implants and assistive listening devices.
- d. An audiologist must have the appropriate Illinois licensure in Audiology issued by the Department of Professional Regulation or must be appropriately licensed in the state in which audiology is practiced.
- e. It is preferred that an audiologist working with infants and children have experience in the management and fitting of amplification in infants and children with hearing loss. The audiologist must also have the equipment necessary to complete the tests required for hearing aid selection and evaluation procedures.

2. Criteria for Determining Candidacy for Amplification

Infants should be fit to the “best estimate” audiogram based on the completion of the physiological assessment techniques outlined in the “Audiologic Diagnostic Assessment Guidelines” section of this document. Final amplification decisions should be based on information obtained from on-going audiologic re-evaluation, performance of the infant in the home and/or educational environment, existence of other special needs, speech, language and auditory developmental milestones, and the family’s preferences.

- 1) Estimates from electrophysiologic correlates of hearing sensitivity (i.e., click or frequency-specific auditory evoked potentials, OAE frequency-specific results), OR WHEN POSSIBLE.
- 2) Ear specific behavioral thresholds obtained by standard audiometric techniques appropriate to the child’s developmental level (i.e., visual reinforcement audiometry, conditional play audiometry, or standard behavioral audiometry).
- 3) Bilateral mixed hearing loss greater than 30 dB HL. The degree and type of hearing loss should be determined by the factors listed above.
- 4) Unilateral hearing loss greater than 30 dB. The degree and type of hearing loss should be determined by the factors listed above.

- a. A child is considered a candidate for amplification when a permanent, bilateral hearing loss of > 35 dB HL for behavioral testing or when a hearing loss equals 30 dBnHL for click ABR threshold in the better ear is present. The degree of hearing loss may be determined by either estimates from electrophysiologic correlates of hearing sensitivity (i.e. click or frequency-specific auditory evoked potentials, OAE frequency-specific results). When possible, ear specific behavioral testing should also be considered.

3. Pre-selection: Physical Characteristics of Amplification

Note: With bilateral hearing loss, all amplification ordinarily should be binaural unless contraindicated.

- a. Amplification options:
 - i. Behind-the-ear (BTE) aids are appropriate for most infants and children. In-the-ear (ITE) hearing aids are not recommended for use with infants and young children due to the growth of the outer ear and problems with increased feedback and safety issues.
 - ii. A bone conduction hearing aid may be appropriate if the hearing loss is conductive and BTE hearing aids cannot be worn due to medical or physical contraindications.
 - iii. Body aids should only be used when BTE hearing aids cannot be fit due to medical or physical contraindications.
 - iv. A cochlear implant may be appropriate if the child has a bilateral profound/severe sensorineural hearing loss, has used appropriate binaural hearing aids, has been enrolled in an appropriate early intervention program, and exhibits minimal benefit from the hearing aids. An FM system coupled to the infant's personal hearing aids should always be considered. Hearing aids with digital processing, dual microphones, multiple channels and directional microphones should be considered for their flexibility, improved signal-to-noise ratio, and other specific features.
- 5) An FM system coupled to the infant's personal hearing aids should be considered when the child becomes mobile and needs to listen to a caretaker/teacher at a greater distance.
- 6) Hearing aids with digital processing, including an FM system and dual microphones, should be considered for their flexibility and their noise reduction algorithms.
- 7) Hearing aids with multiple channels should be considered when the audiometric configuration require the shaping of gain or output in specific frequency regions.
- 8) Directional microphones should be considered only for older children with mild to sever hearing losses to improve signal-to-noise ratio when FM technology, the system of choice to improve signal-to-noise ration, is not being used.
- b. Amplification requirements for FM system and assistive device compatibility:
 - 1) Direct audio-input capabilities (DAI).
 - 2) A telecoil (determine the minimum standard of amplification required for sensitivity re: some telecoils are ineffective.
 - 3) A microphone-telecoil switching option (M - T switch)
- c. Amplification safety feature requirements
 - 1) Tamper resistant battery doors
 - 2) Volume control covers
- d. All amplification fittings should be biaural in children, unless contraindicated.

- e. Ear mold requirements
 - 1) Should be a soft material
 - 2) Should be replaced whenever feedback occurs at recommended settings or when retention becomes a problem.
 - 3) May use with "Ototease" to help reduce feedback.
- f. Retention devices can be used to aid in full-time use.
 - 1) "Huggies"
 - 2) "Critter" clips, with appropriate safety warnings for strangulation possibility
 - 3) Two sided tapes
- g. It is recommended that families should be provided with maintenance kit that includes:
 - 1) Dry aid kit
 - 2) Battery tester
 - 3) Listening tube/stethoscope
 - 4) Extra batteries

4. Hearing Aid Selection and Verification

- a. The pediatric hearing aid should be selected and fitted according to procedures that are especially designed for pediatrics (e.g., DSL methodology). The preferred verification method is to use probe microphone measurements and the child's ear, ear mold, and amplification system. The procedure should be combined with a prescriptive technique, which estimates target responses appropriate for the characteristics of the amplification system (linear vs. non-linear, analog vs. digital). This should always include direct measurement of the real-ear saturation response (RESR) and target maximum output values.

Ear mold requirements:

- 1) Should be a soft material
- 2) Should be replaced whenever feedback occurs at recommended settings or when retention becomes a problem

Amplification safety requirements:

- 1) Tamper resistant battery doors
- 2) Volume control covers or ability to disable volume control

It is recommended that families purchase a maintenance kit that includes:

- 1) Dry aid kit
- 2) Battery tester
- 3) Listening tube/stethoscope
- 4) Extra batteries

5. Validation of aided auditory function may be on-going and may include:
 - a. Probe microphone measurements to assess output of hearing aid at the tympanic membrane (TM).
 - b. Audiologic assessment directly measuring the child's performance including aided sound field responses to speech and frequency specific stimuli.
 - c. Functional auditory skill assessment obtained by the audiologist and early interventionist.
 - d. Speech, communication, and language skill assessment obtained by the early interventionist and a speech language pathologist.
 - e. Parent input as well as input from other professionals involved with the child.
6. Counseling and Follow-Up:
 - a. Information about all appropriate amplification options should be given to the parents prior to final purchase of amplification.
 - b. Parents and other family members or individuals that will assist in the insertion of and maintenance of the amplification system should receive orientation and ongoing support.
7. Suggested frequency of audiologic re-evaluation/follow-up:
 - a. At least every three (3) months during the first two (2) years of amplification use.
 - b. Every three (3) to six (6) months after the first two (2) years of amplification use.
8. Audiologic re-evaluation and/or follow-up may include:
 - a. Behavioral audiometric evaluations including air and bone conduction (obtain separate ear information as soon as possible);
 - a. Behavioral audiometric evaluations including air and bone conduction (obtain separate ear information as soon as possible; at least by nine months developmental age).
 - b. Immitance measurements to evaluate middle ear function;
 - c. Adjustment of the amplification system based on updated audiometric information;
 - d. Electroacoustic evaluations of the hearing aids;
 - e. Listening checks of the hearing aids;
 - f. Evaluation of ear mold fit;

- g. Probe microphone measurements, which are very important as changes in the child's outer ear occur as growth takes place;
 - h. Functional gain measurements to document the development of auditory skills.
9. The infant/young child should be enrolled in an Early Intervention Program which includes:
- a. Home visits
 - b. A professional with extensive and in-depth knowledge of, and experience with, children with varying degrees of hearing loss, their families and all the attendant issues

DISCLAIMER: Early Intervention Services, Medicaid or DSCC may not pay for all services or amplification options.

E. Medical Guidelines for Infants with Confirmed Hearing Loss

In 1999, the Illinois legislature passed the Hearing Screening for Newborns Act. By December 31, 2002, all hospitals delivering babies will be required to provide hearing screening to all babies born in their facility. The goals of the Illinois program are:

All infants born in Illinois will have their hearing screening at birth.

All newborns referred from the Illinois Newborn Hearing Screening Program will have diagnostic testing completed by three (3) months of age.

All infants diagnosed with significant hearing loss will receive appropriate treatment, including hearing aids and Early Intervention Program services by six (6) months of age.

Universal newborn hearing screening has quickly become the standard of care in Illinois. The Illinois chapter of the American Academy of Pediatrics and the Illinois Academy of Family Physicians endorses universal newborn hearing screening as an important strategy to identify infants with significant hearing loss. Early identification and intervention have been shown to dramatically impact the development of communication skills in young children with hearing loss. In fact, when appropriate diagnosis and treatment can be implemented prior to six months of age, babies with hearing loss can acquire speech and language much like that of babies with normal hearing.

Your involvement as a physician is critical to ensure early identification and intervention. This brochure has been developed to inform you of the most current thinking of many who have worked closely with early hearing identification and intervention programs throughout the country. This thinking has been adopted by the Illinois Universal Newborn Hearing Screening Advisory Committee as guidelines for medical follow-up in the state of Illinois. We hope this will be useful to you in the care of infants referred from universal newborn hearing screening programs. It includes information for primary care physicians, otolaryngologists, and medical geneticists, all of whom are important to the accomplishment of the goal of early intervention and intervention.

It is recommended that infants referred from a universal newborn hearing screening program and identified with a confirmed hearing loss in one or both ears should receive the following evaluations, as indicated by three months of age. It is critical to complete all diagnostic evaluations as quickly as possible so that treatment and intervention such as hearing aids, aural habilitation, and child development therapy can begin before the child is six months of age. "Wait and see" is never recommended.

Primary Care Medical Evaluation

The following should be included in the medical evaluation conducted by the baby's primary care physician who is responsible for initiating and supervising the evaluation and referral process to otolaryngology, genetics, ophthalmology, neurology, and other professionals, as appropriate.

Infants referred from a universal newborn hearing screening program and identified with a confirmed hearing loss in one or both ears should receive the following evaluations, as indicated, by three (3) months of age, but no later than six (6) months of age.

The following should be included in the medical evaluation conducted by the baby's primary care physician who is responsible for initiating and supervising the evaluation and referral process to otolaryngology, genetics, ophthalmology, neurology, and other professionals, as appropriate.

History

Prenatal Information

- Ototoxic medication exposure
- Any significant complications during pregnancy
- Any possibility of parental consanguinity

Perinatal information

- Indicators associated with sensorineural and/or conductive hearing loss
- In utero infection, such as cytomegalovirus, rubella, syphilis, herpes, and toxoplasmosis
- Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal
- Birth weight of less than 1,500 grams (3.3 lbs).
- Hyperbilirubinemia with risk of kernicterus
- Ototoxic medications, including but not limited to the aminoglycosides, used in multiple courses or in combination with loop diuretics
- Bacterial meningitis
- Apgar scores of 0-4 at one (1) minute or 0-6 at five (5) minutes
- Mechanical ventilation lasting five (5) days or longer
- Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss
- Intraventricular hemorrhage, seizures or inborn errors of metabolism

Family History

- Other family members with hearing loss with onset at an early age
- Family's desire to have additional children

Physical Examination

Minor anomalies

- Unusual morphologic features occurring in less than 5% of the population with no cosmetic or functional significance

Major anomalies

- Those causing cosmetic and/or functional abnormality, e.g., cleft palate, cardiac, limb, skeletal deformities

Lab

- Urine culture for CMV prior to three (3) weeks of age, if possible

Referrals

- Audiological evaluation if appointment has not been scheduled
- Ophthalmologic examination
- Genetics evaluation
- Early Intervention
- Referral to local Child and Family Connections office if not yet done

F. Otolaryngological Medical Evaluation

The following should be included in the medical evaluation conducted by an otolaryngologist:

History

Prenatal
Perinatal
Family
Behavioral

Physical Examination

Head and neck examination
Head circumference

Review of Audiological Test Results

Evaluations

Infectious diseases: CMV, Rubella, Syphilis, and Toxoplasmosis
Urinalysis
CT with high resolution of temporal bones (if clinically indicated)

G. Genetic Medical Evaluation

An estimated 60% of prelingual hearing loss is due to genetic causes and more than half of these cases are nonsyndromic. Appropriate genetic consultation serves to confirm diagnosis, identify additional medical risks and communicate genetic risks. The following should be included in the medical evaluation conducted by a Medical Specialist:

History

Pregnancy
Family Pedigree

Developmental

Physical Examination

General pediatric examination
Careful dysmorphicologic examination
Neurologic/developmental evaluation

Diagnostic Tests

Hearing tests on first-degree relatives (parents and siblings)

Ophthalmologic examination by six (6) months of age

TORCH titers or CMG IgG and IgM if less than six (6) months of age

All other laboratory tests depend upon clinical evaluation and history but may include the following:

- Chromosomes if dysmorphic
- EKG
- Skeletal survey if there is short stature or disproportional growth
- Evaluation of other systems: renal, cardiac, skin
- CT or MRI of brain if neurologically abnormal
- Specialized genetic studies, e.g., molecular, gene testing

X. NEWBORN HEARING SCREENING ADVISORY COMMITTEE

PA 91-0067 requires the organization of an advisory committee.

A. Members

- Department of Human Services
- Department of Public Health
- University of Illinois at Chicago, Division of Specialized Care for Children
- Public and private hospitals
- Pediatric Associations
- Audiologists
- Health Insurance Plans
- Hearing-impaired persons
- Parents of hearing-impaired children
- Early Intervention services

B. Responsibilities

1. Develop and conduct training for hospitals implementing newborn hearing screening.
 2. Develop a tracking and follow-up program for diagnostic hearing testing for those infants failing hospital-based screening, in order to diagnose congenital hearing loss.
 3. Develop a referral system to early intervention services and hearing aids for those infants diagnosed with hearing loss.
 4. Develop an application process for financial assistance by the Division of Specialized Care for Children for follow-up diagnostic hearing testing of newborns failing hospital-based screening.
 5. Develop educational and informational materials for hospital personnel, health care professionals, and parents on appropriate follow-up procedures for infants failing hospital-based screening.
 6. Monitor any reports made available to the State with respect to the hearing screening status of all newborns.
1. Monitor any reports made available to the state with respect to the hearing screening status of all newborns.
 2. Review administrative rules and make recommendations to the Department regarding such rules.