"Sound Start" of Louisiana
Early Hearing Detection and Intervention Program

Louisiana
Pediatric Diagnostic Audiology Guidelines

Protocols and Standards for Diagnostic Evaluations to Determine Hearing Loss

Department of Health and Hospitals
Office of Public Health
Hearing, Speech and Vision Services

These guidelines were developed in part by funds from grants from the Maternal and Child Health Bureau, and the National Center for Disease Control.

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TABLE OF CONTENTS

GUIDELINE GOALS

The goal of developing this document is to provide guidelines for pediatric diagnostic services that recognize the diversity of individuals and families. These guidelines have been developed specifically for audiological diagnostic services provided to children from **birth-36 months of age**. This is a companion document to the previously created ***Louisiana Hospital Guidelines for Newborn Infant Hearing Screening Programs.***

Audiologists who are not able to provide these services, due to lack of skill, experience, or equipment are ethically obligated by the Louisiana Board of Examiners in Speech Pathology and Audiology Ethics Guidelines to refer families to facilities where the needed services can be obtained.

A well-organized and professional early hearing detection and intervention system can make a significant difference in the lives of children who are deaf or hard of hearing and their families. We hope these guidelines can act as a tool to providing the best services possible.

These guidelines are intended for audiologists who serve infants and young children suspected of having hearing loss. Given the necessity and importance of multi-disciplinary service providers for children and their families, other stakeholders may benefit from these assessment guidelines in the context of early detection and intervention program development.

Who developed these guidelines?

The guidelines on the following pages were developed by a committee of audiologists from the state of Louisiana /Office of Public Health “Sound Start” EHDI Program in collaboration with the Louisiana State Advisory Council on Infant Hearing.

These guidelines are based on the DHH rules and regulations developed to accompany Louisiana Act 653 of 1999 Universal Newborn Screening Legislation and were updated to reflect new recommendations from the Joint Committee on Infant Hearing Position Statement (2007) in November, 2009. See Appendix B for summary on JCIH 2007 Position Paper.

Funding was provided in part by grants from the US Maternal and Child Health Bureau and Center for Disease Control and Prevention.

Status of infants after hospital or private initial screening:

Infants who need further testing due to failed initial hearing screening

- Infants failing the newborn screening are to be referred for follow-up rescreening as soon as possible within two to four weeks from hospital discharge.

- Infants failing the hospital screening must be referred to the primary care physician or to a licensed audiologist for follow-up rescreening**.** See *Louisiana Audiological Guidelines for Hearing Rescreening of Infants Ver. 2.0 (2008)*

Infants who need further testing due to failed follow-up rescreening

- The audiologist or physician should help the parents to make arrangements for the diagnostic testing at the time of the failed rescreening.

- If possible, diagnostic testing should be performed during the same visit as the rescreening or as soon as possible.

- Only appropriately credentialed and qualified audiologists who possess a valid state license should perform follow-up diagnostic testing.

- **Diagnosis and evaluation of the type and degree of hearing loss should be completed by the time the child is <u>3 months of age</u>.**

. The goal for follow up for successful early hearing detection and intervention is***:***

1-3-6

- ***Before <u>1 month</u> old:***
  Complete initial newborn hearing screening

- ***By <u>3 months</u> old:***
  Complete an appropriate audiological diagnostic assessment on infants failing screening or rescreening

- ***Before <u>6 months</u> old:***
  Fit amplification and begin early intervention services

EVALUATION OF NEWBORNS AND INFANTS
0-6 MONTHS OF AGE

Audiological Diagnostic Assessment Protocol

To be considered a diagnostic procedure, ear specific estimates of type, degree, and configuration of the hearing loss must be obtained. This differs from a simple screening.

Adequate confirmation of an infant's hearing status cannot be obtained from a single test measure; rather the initial test battery must include physiologic measures and, if possible, developmentally appropriate behavioral techniques.

1. **Detailed history** should include but is not limited to:
    a. Parental report of auditory and visual behaviors
    b. Motor development
    c. Family history of hearing loss
    d. History of middle ear pathologies
    e. Parental concerns
    f. Prenatal, birth, and neonatal history
    g. Medical history including: Syndromes or other inheritable conditions, craniofacial anomalies, kidney issues, conditions of limbs/digits, pigmentation issues, exposure to ototoxic medications

2. **Otoscopy**
    Visual inspection for obvious structural abnormalities of the pinna and ear canal should be included.

3. **Evoked Otoacoustic Emissions**
    Either Transient or Distortion Products Emissions are acceptable**.**

    TEOAE click stimuli: One level (e.g., 80-85 dB pSPL) should be completed.
    DPOAE stimuli: Use L1/L2 of 65/55 dB SPL.

    Pass criterion: **An emission of 6 dB signal to noise ratio for at least three frequencies in each ear**.

    At least one frequency should be located between **2000 and 3000 Hz**
    A second frequency should be located between **3000 and 4000 Hz**
    The third point could be at any other frequency between **1000 Hz and 6000 Hz**

4. **<u>Acoustic Immitance testing</u>**
    a. Tympanometry - 660 Hz or higher probe tone
    b. Acoustic Reflex- Ipsilateral middle ear muscle reflex thresholds for 500, 1000, 2000, and 4000 Hz if possible. Currently there is insufficient data for routine use of acoustic middle ear muscle reflex in infants younger than 4 months.

Pass criterion: Type A tympanogram

5. **<u>Diagnostic Auditory Evoked Potential Testing</u> (Non-sedated)**

a. ABR to air-conducted clicks:

Diagnostic testing should include Wave V latency-intensity function responses to at least three differing intensity levels ending with at least one tracing at or below threshold.

Pass Criterion: **Normal results consist of Wave V responses for clicks at 25dBnHL within a normal absolute latency range adjusted for the child's corrected gestational age**.

b. Change polarity of clicks

Supra-threshold click testing should also include one average with condensation clicks and another average at the same intensity with rarefaction clicks to rule-out **auditory neuropathy/dys-synchrony**.

In a normal ABR the waveforms will be essentially the same morphology and latency with both polarities. If all waveforms in the tracings at one polarity invert when compared to the other polarity, that represents the presence only of the cochlear microphonic (CM) with no neural response. If only the CM is observed, that is consistent with auditory neuropathy/dys-synchrony.

Even though the ABR is abnormal, in this case toneburst testing is not necessary, as it will not yield any additional information.

c. ABR to tonebursts:

In order to obtain more frequency-specific information, ABR stimuli should include <u>at least one</u> low frequency toneburst (such as 500Hz) in combination with clicks.

For even greater specificity both low and high frequency tonebursts could be used in place of clicks (such as 500Hz, 2000Hz and 4000Hz).

If CONDUCTIVE HEARING LOSS is suspected, testing must also include:

c. Bone conduction ABR:

Stimuli should be bone-conducted clicks; masking of the non-test ear should be applied, as appropriate.

Diagnostic testing should include Wave V latency-intensity function responses to at least three differing intensity levels ending with one tracing below threshold.

Diagnostic testing at minimum should include Wave V latency-intensity function responses to at least three differing intensity levels ending with at least one tracing at or below threshold.

Pass Criterion Normal results would consist of Wave V responses at **25dBnHL** for higher frequencies and **35dBnHL** at lower frequencies.

Audiological Diagnostic Assessment

1. **Detailed history:** including but not limited to:

    a. Parental report of auditory and visual behaviors
    b. Motor development
    c. Family history of hearing loss
    d. History of middle ear pathologies
    e. Parental concerns
    f. Prenatal, birth, and neonatal history
    g. Medical history including: Syndromes or other inheritable conditions, craniofacial anomalies, kidney issues, conditions of limbs/digits, pigmentation issues, exposure to ototoxic medications

2. **Otoscopy:**
   Visual inspection for obvious structural abnormalities of the pinna and ear canal

3. **Evoked Otoacoustic Emissions:** Transients or Distortion Products

   TEOAE click stimuli: One level (e.g., 80-85 dB pSPL) should be completed.
   DPOAE stimuli: Use L1/L2 of 65/55 dB SPL.

   Pass criterion: Emission of **6 dB signal to noise ratio for at least three frequencies** in each ear.
   At least one frequency should be located between **2000 and 3000 Hz**
   A second frequency should be located between **3000 and 4000 Hz**
   The third point could be at any other frequency between **1000 Hz and 6000 Hz**

4. **Acoustic Immitance Testing:**
    a. Tympanometry - 660 Hz or higher probe tone in children under 6-18 months, 220 Hz is acceptable in children 18-36 months

    b. Acoustic Reflex- Ipsilateral middle ear muscle reflex thresholds for 500, 1000, 2000, and 4000

    Pass Criterion: Type A tympanogram and present acoustic reflexes

5. **<u>Behavioral Observation Audiometry (BOA):</u>**
   In soundfield or with earphones using calibrated stimuli. Insert earphones are recommended if possible.

   Pass criterion: minimal and/or startle response at 65 dB.

6. **<u>Visual Reinforcement Audiometry (VRA):</u>** if appropriate due to child's developmental level.

   Stimuli should be speech and also frequency specific tones between 250-6000 Hz. Insert earphones are preferable; sound field may be necessary with some children who will not tolerate earphones.

   Pass Criterion: **20 dB** to speech and threshold responses at 500, 1000, 2000, and 4000 Hz tones.

7. **<u>At least one ABR</u> is recommended as part of a complete audiology diagnostic evaluation for children younger than 3 years old for confirmation of permanent hearing loss.**

   (The same procedures as outlined in newborn-6 months for recommended ABR procedures apply. See previous section.**)**

USE OF SEDATION IN THE EVALUATION PROCESS

No child, especially those under the age of six months of age, should be given medication to sedate for testing unless absolutely necessary. Sedating merely for convenience or to speed testing time in a busy clinic schedule is neither ethical audiological practice nor good medical practice. Most normally developing children from birth to 6 months of age can be tested using sleep deprivation and other techniques to induce natural sleep.

The standard is to begin with less medically invasive procedures (i.e. behavioral) and move to more complex procedures (i.e. electrophysiological) requiring or including the use of sedation only when deemed necessary to complete the evaluation.

Children 6 months and older or children with complex medical conditions may need to be sedated to complete necessary diagnostic procedures when **behavioral audiology is inappropriate due to the child's age or other limitations or attempts at behavioral testing have been made** with no success. Conscious sedation is recommended over deep sedation whenever possible.

Administering the sedation and discharging patients after the procedure is not within the scope of practice of the audiologist.

Both the Louisiana Board of Examiners and the American Speech-Language-Hearing Association have adopted standards for the procedural use of sedation for speech pathology and audiology. Audiologists involved with sedation should be knowledgeable of these position statements. You can view the ASHA Technical Report on Sedation and Topical Anesthetics at http://www.asha.org/docs/pdf/TR1992-00257.pdf

MONITORING CHILDREN AT-RISK FOR PROGRESSIVE HEARING LOSS

Considerations for managing babies with risk factors for late onset or progressive hearing loss are extremely important**. The timing and number of hearing reevaluations for children with these risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss.**

The JCIH 2007 Position Paper recommends an inclusive strategy of surveillance of **all children** within the medical home on the pediatric periodicity schedule.

All families should receive informational materials that discuss major milestones in normal speech and language development and risk factors for hearing loss in their native language.

Families of infants at high risk should receive additional information on late-onset or progressive hearing loss as well as local diagnostic resource centers.

The first monitoring appointment for infants at-risk for progressive loss should be made prior to discharge from the birth hospital or NICU.

See Appendix B for a list of RISK INDICATORS ASSOCIATED WITH PERMANENT CONGENITAL, DELAYED-ONSET OR PROGRESSIVE HEARING LOSS IN CHILDHOOD

Infants who pass the neonatal screening but have any of the risk factors on the list not in bold print should have at least 1 diagnostic audiology assessment by 24 to 30 months of age.

Risk indicators that are in **bold** are of greater concern for delayed-onset hearing loss and will need closer audiologic monitoring. Closer monitoring would include audiological testing every 6 months up to age 3 years and annually after that time.

AMPLIFICATION FOR INFANTS AND CHILDREN BIRTH – 3 YEARS

Most infants and children with bilateral hearing loss and many with unilateral loss benefit from some form of personal amplification.

Pediatric Amplification:

a. Should definitely be considered for infants and children when the pure tone average or high frequency pure tone average is greater than 25dB HL in at least one ear.

b. Should be considered when the pure tone average of either ear is greater than 15dB HL and the child is exhibiting speech and language difficulties due to fluctuating or mild hearing loss.

c. Should be assessed for appropriateness for infants and young children with unilateral loss. Depending on the amount of residual hearing, a hearing aid may be indicated. Contra-lateral routing of signals (CROS) is not currently recommended for children.

d. Definitive resolution of otitis media (OM) should never delay the fitting of an amplification device. The infant should be referred to the physician for medical management and the audiologist should monitor the status of the OM when determining appropriate prescriptive targets during the hearing aid fitting.

Timeline for amplification fitting:

If the family chooses personal amplification for their child, hearing aid selection should occur **within 1 month of initial confirmation** of hearing loss even if additional audiologic assessment is ongoing.

Amplification/Hearing Aid fitting Guidelines:

The goal of the amplification device fitting is to provide the infant with maximum access to all of the acoustic features of speech within an intensity range that is safe and comfortable. Amplified speech should be comfortably above the infant's sensory threshold but below the level of discomfort across the speech range.

a. The hearing aid size, make and model should be appropriate for the child's age and development.
b. For infants under 6 months of age, hearing aid fitting will usually be based on physiologic measures alone using real-ear measurements.
c. Behavioral threshold assessment using VRA should be obtained **as soon as possible** to cross-check and augment physiologic findings. (JCIH, 2007). Long term monitoring of the validity of the fitting and refinement of the gain and output targets is necessary.

d. Age appropriate hearing aid prescriptive formulas (such as DSL) that incorporate the use of individual real-ear measurements that account for each infant's ear canal acoustics and hearing loss should be used for fitting infants.

Complementary or alternative technology, such as FM systems or cochlear implants may be recommended as the primary or secondary listening device, depending on the degree of the infant's hearing loss, the goals of auditory habilitation, the infant's acoustic environment, and the family's informed choices.
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Pediatric Referral for Cochlear Implant:
Cochlear implantation (CI) should be given careful consideration for any child. Since the benefit of CI is variable, a trial fitting of a traditional hearing aid at this time is still recommended by JCIH. The decision to discontinue the hearing aid should be made on the basis of the benefit derived from the amplification.

According to the current FDA guidelines, a cochlear implant:

a. Is appropriate for children over 12 months of age with profound bilateral sensorineural hearing loss.

b. May be considered for children 12 months and older with severe bilateral sensorineural hearing loss who are not developing speech and language skills on target after attempting conventional hearing aid use.

c. May be considered for children 12 months and older diagnosed with auditory neuropathy/dys-synchrony who are not developing speech and language skills on target. A hearing aid trial in patients with AN/AD is still indicated in most cases.

d. The presence of developmental conditions (e.g., developmental delay, autism) in addition to hearing loss should not preclude the considerations of a CI for an infant or child who is deaf.

REPORTING RESULTS OF ALL RESCREENING AND DIAGNOSTIC TESTING IS EXTREMELY IMPORTANT.

Testing results should be reported on the EHDI **Follow-up Form** which can be obtained by emailing the EHDI Follow-Up Coordinator. This follow-up from should be emailed, faxed, or mailed within 14 days to the Department of Health and Hospitals EHDI Program at the address and fax listed on the bottom of the form.

Email EHDI Follow-Up Coordinator at:
ehdifollowup@hughes.net

Which results should be reported?

1. Rescreening and/or diagnostic testing results on all **infants who failed** a hospital newborn hearing screening even if the results of your testing are normal.

2. Initial screening or diagnostic testing results on all **infants who never had a hospital newborn hearing screening** even if your results are normal.

3. All results on any child ages birth-5 years of age **identified with a hearing loss for the first time.**

4. All results on any child birth- 5 years of age **fitted with a hearing aid or cochlear implant for the first time**.

5. Every child that is **considered lost to follow-up**. Report information on any child who failed to keep their rescreening or diagnostic testing appointments and who is now considered to be lost to follow-up:

    - It is recommended that at least two attempts be made to schedule the patient for follow-up testing before reporting them as lost to follow-up.

    - Communication should be made by at least two different methods before giving up (i.e.: telephone contact and mail contact).

    - Send information on all possible contact family and phone numbers, and information on all previous contacts (phone disconnected, wrong address, other family members, etc).

REFERRAL TO EARLY INTERVENTION

Early intervention is an integral part of the habilitation process for all children identified with hearing loss. Simply establishing an amplification system is only the first step. It is ethically and legally the responsibility of the audiologist to refer all children identified with hearing loss into an early intervention system.

According to Federal Law IDEA, all children diagnosed with a hearing loss **must be referred to Part C (early intervention) within 48 hours.**

In 2009, a new single point of entry into the system of early intervention was developed. The regional LA-Hear Coordinators from PPEP have now been designated as the single point of entry for children identified with any type of hearing loss.

These changes were made in cooperation with Early Steps, DHH EHDI Program, and the PPEP program. These changes assure that families and their children who have hearing loss will have immediate access to:

- an individual with specialized skills, knowledge, and experience in the areas of early childhood language development, hearing loss, communication methods, and family support
- unbiased information on parent and family choices for communication and intervention services as recommended in 2007 Position Statement: Joint Committee on Infant Hearing.

Referrals to regional LA-Hear Coordinators can be made by an audiologist through:

Phone: 225-757-3331; Fax: 225-757-3332;
TDD: 225-769-8160x331; Email: ppep@lalsd.org

The following information must be provided when referring a child to PPEP:

- Family contact information- at least two phone numbers if possible
- Documentation of hearing loss i.e. written report, audiogram, or copy of the Office of Public Health/Follow-up Services Report

A regional LA-Hear Coordinator will contact the family within 3 days. Services are provided to families of children who are identified at any age with **any type or degree of either bilateral or unilateral** hearing loss. Additionally, families of infants up to 3 years of age will be referred to Early Steps by the LA-Hear Coordinator in order to receive any other services the child might need. An additional Early Steps referral from the audiologist is not necessary.

PPEP services are statewide, cost-free, home/community-based services provided to families of children. Services are individualized to meet the needs and priorities of the family. Every family should be referred, and it will be the family's choice to determine their need for services.

Parent-Pupil Education Program
Louisiana Deaf & Hard of Hearing Resource [LA-Hear] Coordinators

- Contact a family immediately after diagnosis to offer assistance
- Provide home-based support to families of children with hearing loss
- Ensure referral to Early Steps
- Participate in the development of the Individual Family Services Plan (IFSP)
- Provide information to the family about specialized programs available to children with hearing loss, i.e., Bright School & New Orleans Oral School
- Link the family with parent-to-parent contact and the statewide parent consultant
- Track and monitor all children with ***any type or degree of bilateral or unilateral hearing loss***
- Participate in audiological visits and collaborate with audiologists regarding the child's progress in communication development and hearing aid/cochlear implant use
- Provide the family with unbiased information about all modes of communication so they can make informed decisions for their child
- Help the family to assess, monitor & enhance their child's communication development during daily routines
- Assist the family and/or childcare provider to identify and practice optimal listening environments in the home or in daycare settings
- Provide technical assistance to occupational therapists, physical therapists, speech therapists, special instructors, etc. so that language learning can be incorporated into all activities
- Provide daycare staff and/or other family members with information about the child's hearing loss and how they can assist in the development of communication skills and hearing aid/cochlear implant use
- Assist families with the transition from Early Steps at age 3

The Parent-Pupil Education Program is a free outreach service provided through the Louisiana School for the Deaf for families with children who are deaf or hard of hearing.

Appendix A: Legislation/ Rules and Regulations

1. Louisiana Identification of Hearing Impairment in Infants Law (Act 653 amended statutes July 1, 1999 and Act 417 July, 1992 combined)

2. Louisiana DHH Revised Procedures for Identification of Hearing Impairment in Infants law (April, 2002)

3. Louisiana Insurance Coverage for Hearing Aids (ACT 816 – 2003)

Appendix A1:

ACT No. 653

SENATE Bill NO. 436

BY SENATORS LANDRY AND IRONS AND REPRESENTATIVES DURAND, JETSON, TOOMEY, AND WILKERSON

To amend and reenact R.S. 46:2263 (7)(a)(xi) and (b)(ix) and (8), 2264 (A)(4) and 2267 and to enact R.S. 46:2262 (D), relative to the identification of hearing impairment in infants; to revise certain definitions; to require hospitals to provide screening for hearing impairment to all newborn infants prior to discharge; to provide an effective date for adoption of rules and regulations; to provide for related matters.

Be it enacted by the Legislature of Louisiana:

Section 1. R.S. 46:2263 (7)(a)(xi)and (b)(xi) and (8), 2264 (A)(4) and 2267 are hereby amended and reenacted and R.S. 46:2262 (D) is hereby enacted to read as follows:

2262. Purpose

A. The purpose of the program for early identification of hearing impairment is to identify hearing impaired infants at the earliest possible time so that medical treatment, early audiological evaluation, selection of amplification, and early educational intervention can be provided.

B. Early educational intervention and early audiological services are required under the Education of the Handicapped Act, Amendments of 1986, Public Law 99457.

C. Early identification and management of the hearing impaired infant is essential if that infant is to acquire the vital language and speech skills needed to achieve maximum potential educationally, emotionally, and socially.

D. Appropriate screening and identification of Newborns and Infants with Hearing Loss will therefore serve the public purpose of promoting the healthy development of children, and reducing public expenditure for health care and special education, and related services.

2263. Definitions

Except where the context clearly indicates otherwise, in this Chapter:

(I) "Advisory council" means the advisory council created pursuant to R.S. 46:2265.

(2) "Department" means the Department of Health and Hospitals.

(3) "Hearing impaired infant" means an infant who has a disorder of the auditory system of any type or degree, causing a hearing impairment sufficient to interfere with the development of language and speech skills. The term "hearing impaired infant" includes both deaf and hard-of ·hearing infants.

(4) "Infants at risk" means those infants who are at risk for hearing impairment because they have one or more risk factors.

(5) "Office" means the office of public health within the department.

(6) "Program" means the program that the office of public health establishes to provide for the early identification and follow-up of infants at risk, of hearing impaired infants, and of infants who have a risk indicator for developing a progressive hearing impairment.

(7) "Risk factors" means those criteria or factors, anyone of which identifies an infant as being at risk for hearing impairment.

(a) The risk factors that identify those neonate infants from birth through the first twenty-eight days, who are at risk for sensorineural bearing impairment included the following:

(i) Family history of congenital or delayed onset childhood sensory neural impairment.

(ii) Congenital infection known or suspected to be associated with sensorineural hearing impairment such as toxoplasmosis, syphilis, rubella, cytomegalovirus, and herpes.

(iii) Craniofacial anomalies including morphologic abnormalities of the pinna and ear canal, absent philtrum, low hairline, etc.

(iv) Birth weightless that one thousand five hundred grams or less than three and three tenths pounds.

(v) Hyperbilirubinemia at a level exceeding indication for exchange transfusion.

(vi) Ototoxic medications, including but not limited to the aminoglycosides used for more than five days, such as gentamicin, tobrarnycin, kanamycin, streptomycin, and loop diuretics used in combination with aminoglycosides.

(vii) Bacterial meningitis

(viii) Severe depression at birth, which may include infants with Apgar scores of zero to three at five minutes or those who fail to fully initiate spontaneous respiration by ten minutes or those with hypotonia persisting to two hours of age.

(ix) Prolonged mechanical ventilation for a duration equal to or greater than ten days, such as persistent pulmonary hypertension.

(x) Stigmata or other findings associated with a syndrome known to include sensorineural hearing loss, such as Waardenburg or Usher's syndrome.

(xi) Other risk factors added or deleted by the office of public health upon recommendation of the advisory council for early identification of hearing impaired children

(b) The factors that identify those infants aged twenty-nine days to two years who are at-risk for sensorineural hearing impairment include the following:

(i) Parent or caregiver concerns regarding hearing, speech, language and/or developmental delay,

(ii) Bacterial meningitis.

(iii) Neonatal risk factors that may be associated with progressive sensorineural hearing loss, such as cytomegalovirus. prolonged mechanical ventilation, and inherited disorders.

(iv) Head trauma especially with either longitudinal or transverse fracture of the temporal bone.

(v) Stigmata or other findings associated with syndromes known to include sensorineural hearing loss, such as Waardenburg or Usher's Syndrome,

(vi) Ototoxic medications, including but not limited to the aminoglycosides used for more than five days, such as gentamicin, tobramycin, kanamycin, streptomycin, and loop diuretics used in combination with aminoglycosides.

(vii) Children with neurodegenerative disorders such as neurofibrornatosis, myoclonic epilepsy, Werdnig-Hoffman disease, Tay-Sach's disease, infantile Gaucher's disease, Nieman-Pick disease, any metachromatic leukodystrophy, or any infantile demyelinating neuropathy.

(viii) Childhood infectious diseases known to be associated with sensorineural hearing loss, such as mumps or measles.

(ix) Other risk factors added or deleted by the office of public health upon recommendation of the advisory council for early identification of hearing impaired children.

(8) "Screening for hearing impairment" means employing a device for identifying whether an infant has a disorder of the auditory system, but may not necessarily provide a comprehensive determination of hearing thresholds in the speech range. Procedures may include auditory brainstem response (ABR) screening or evoked Otoacoustic emission (OAE) or other devices approved by the office upon recommendation of the advisory council.

2264. Identification of hearing impairment in infants.

A. The office of public health in the Department of Health and Hospitals shall establish, in consultation with the advice of the Louisiana Commission for the Deaf and advisory council created in R.S. 46:2265, a program for the early identification and follow-up of infants at risk , hearing impaired infants, and infants at risk of developing a progressive hearing impairment. That program shall, at a minimum:

(l) Develop criteria or factors to identify those infants at risk for hearing impairment and infants at risk of developing a progressive hearing impairment including the risk factors set forth in this Chapter, and develop an at-risk questionnaire for infant hearing loss.

(2) Create an at-risk registry 10 include, but not be limited 10, the identification of infants at risk for hearing impairment, hearing impaired infants, and infants at risk of developing a progressive hearing impairment.

(3) Provide to the hospitals and other birthing sites the at-risk questionnaire for infant hearing loss and require that the form be completed for any newborn prior to discharge from the hospital or other birthing site. As to infants at risk, copies of the completed at-risk questionnaire shall be distributed to the at-risk registry of the office, the parent or
guardian, and. if known, the infant's primary care physician and the provider of audiological services.

(4) Require for all newborn infants that the hospital of birth or that hospital to which the newborn infant may be transferred provides screening for hearing impairment by auditory brainstem response (ABR) screening, or evoked Otoacoustic emission (OAE) *or* other screening device approved by the office before discharge. The results of that screening for hearing impairment shall be provided to the at risk registry of the office of public health, the parent
or guardian, and if known, the primary physician and the provider of audiological services.

(5) Develop and provide to the hospitals or other birthing sites appropriate written materials regarding hearing impairment, and require that the hospitals or other birthing sites provide this written materials all parents or guardians of
newborn infants.

(6) Develop methods to contact parents or guardians of infants at risk, of hearing impaired infants, and of infants at risk of developing a progressive hearing impairment.

(7) Establish a telephone hotline to communicate information about hearing impairment, hearing screening, audiological evaluation, and other services for hearing impaired infants.

(8) Provide that when screening for hearing impairment indicates a hearing loss, audiological evaluation shall be done as soon as practical. The parents or guardians of the infant shall be provided with information on locations at which medical and audiological follow up can be obtained.

B. The office shall consult with the advisory council and implement the program.

C. The office shall develop a system for the collection of data, determine the cost-effectiveness of the program and disseminate statistical reports to the Louisiana Commission for the Deaf,

D. The office, in cooperation with the state Department of Education, shall develop a plan to coordinate early educational and audiological services for infants identified as bearing impaired.

E. The office shall follow current practices and applicable guidelines that are currently utilized in Louisiana and will consider practices and guidelines that may be established by the National Institute of Deafness and other Communication Disorders (NIDCD).

2265. Advisory council creation; membership; terms; quorum; compensation

A. There is hereby created an advisory council for the program of early identification of hearing impaired infants. The council shall consist of fourteen members as follows:

(I) An otolaryngologist or otologist.
(2) An audiologist with extensive experience in evaluating infants.
(3) A neonatologist.
(4) A pediatrician.
(5) A deaf person
(6) A hospital administrator
(7) A speech and language pathologist
(8) A school teacher or administrator certified in education of the deaf.
(9) A parent of an oral hearing impaired child.
(10) A parent of a hearing impaired child utilizing total communication.
(11) A representative of the state Department of Education designated by the superintendent of education.
(12) A representative of the office designated by the assistant secretary of the office.
(13) A representative from the Louisiana Commission for the Deaf.
(14) A representative from the Louisiana Association of the Deaf.

B. Members of the council in accordance with R.S. 46;2265 (A)(1) through (10) and RS. 46;2265 (A)(l3) through (14) shall be appointed by the governor, subject to Senate Confirmation, Other members are not subject to senate confirmation.

C. Members of the council representing offices and departments of state government shall serve a four-year term concurrent with that of the governor. Other members shall serve three-year terms, except that in making the initial appointments, four members shall be appointed for a one-year term. Four shall be appointed for two-year terms. and four shall be appointed for three-year terms. No member may serve more than two consecutive terms.

D. Each member shall serve without compensation.

E. A majority of the members of the council shall constitute a quorum for the transaction of all business.

F. The members of the committee shall elect from their membership a chairman and a vice chairman.

2266. Powers. duties, functions of the advisory council

The advisory council shall:

(I) Advise and recommend risk factors or criteria for infants who are at risk of bearing impairments and infants at risk of developing a progressive bearing impairment.

(2) Advise the office as to bearing screening, setting standards for the program, monitoring and reviewing the program, and providing quality assurance for the program.

(3) Advise the office as to integrating the program for early identification of bearing impaired infants with existing medical, audiological, and early infant education programs.

(4) Advise the office as to materials to be distributed to the public concerning hearing

(5) Advise the office on the implementation of the program for early identification and follow up of infants at risk. heating impaired infants, and infants who are at risk of developing a progressive hearing impairment.

2267. Effective dale; rules and regulations

The office of public health shall, by July 1, 2000 adopt rules and regulations necessary to implement the program in accordance with the Administrative Procedure Act.

Approved by the Governor.

A true copy;

W. Fox McKeithen; Secretary of State

[DHH/OPH Rules and Regulations relevant to ACT 653 of 1999- approved 2002] Universal Newborn Hearing Screening

Department of Health and Hospitals
Office of Public Health

Identification of Hearing Impairment in Infants
(LAC 48:V.Chapter 22)

In accordance with the applicable provision of the Administrative Procedure Act, R.S. 49:950 et seq. and the Identification of Hearing Impairment in Infants R.S. 46:2261 et seq., notice is hereby given that the Department of Health and Hospitals, Office of Public Health intends to adopt procedures for the screening of infants to identify hearing impairment, testing of all newborns and referral of newborns failing screening for appropriate follow-up services and ensure proper information distribution to parents, primary care physicians and interested groups.

Louisiana's Act 417 of 1992 mandated hearing screening of all HIGH-RISK infants and rules and regulations were adopted to implement the program in accordance with the Administrative Procedure Act. **On July 1, 1999, Act 417 was amended by Act 653 of 1999 to require UNIVERSAL newborn hearing screening or the hearing screening of all newborn infants, rather than only the infants with high-risk factors**.

It is necessary that new rules be adopted to allow for the proper implementation of the amended legislation of Act 653, to allow for implementation of statewide, universal newborn hearing screening.

Title 48
PUBLIC HEALTH-GENERAL
Part V. Preventive Health Services
Subpart 7. Maternal and Child Health Services
Chapter 22. Identification of Hearing Impairment in Infants
2201. Definitions

Advisory Council--the 14 member council created pursuant to R.S. 46:2265.

*Audiologis*t--an individual licensed to practice audiology by the Louisiana Board of Examiners for Speech Pathology and Audiology.

Auditory Brainstem Response (ABR)--the synchronous electrical response elicited from the auditory nervous system within 20 msec after stimulation and its measurement as used for the detection of hearing loss.

Department--the Department of Health and Hospitals.

Discharge--release from the premises of a medical care facility.

Evoked Otoacoustic Emissions (EOAE)--acoustic echoes, evoked in response to acoustic stimuli, produced by the inner ear and measured by a microphone in the ear canal for the detection of hearing loss.

Hearing Screening--using procedures approved by the office to identify infants in need of diagnostic audiological assessment.

Infants at Risk--those infants who are at risk for hearing loss because they have one or more risk factors as indicated in R.S. 46:2263.

Office--the Office of Public Health within the department.

Other Birthing Site--any site of birth other than a hospital.

Other Risk Factors--any other condition(s) in addition to the factors cited in R.S. 46:2263 added by the office upon recommendation of the advisory council.

Other Screening Device--a device pre-approved in writing by the office, comparable to or better than auditory brainstem response testing.

Program--The Hearing, Speech and Vision Program within the office.

Risk Registry--will be the data base kept by the office of all infants identified as high risk for hearing loss.

AUTHORITY NOTE: Promulgated in accordance with R.S. 46:2261-2267.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 19: 1430 (November 1993). amended LR 28:0000 (April 2002).

2203. Program for Identification of Hearing Loss in Infants

A. The program will include the following:

1**.** The office will require a newborn hearing screening report to be used by the hospitals to report hearing screening results and risk status on all newborns to the risk registry. This form will include written material regarding hearing loss and a toll-free hotline phone number (V/TDD).

2. The office will maintain a risk registry to include information reported on the newborn hearing screening report.

3. The office will notify parents of infants at risk of available follow-up services.

4. The risk registry will include periodic notification to parents of recommended procedures for infants and children at risk for progressive hearing loss.

5. The risk registry will include information on infants diagnosed with hearing loss.

6. The office will provide for a toll-free hotline service for parents and professionals to utilize to obtain information about the program and related services. This hotline will be accessible by voice or TDD.

B. Implementation

1. All birthing sites in Louisiana must be in compliance with this act by March 1, 2002

AUTHORITY NOTE: Promulgated in accordance with R.S. 46:2261-2267.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 19: 1431 (November 1993), amended LR 28:0000 (April 2002).

2205. Procedures for Hospitals

A. Hospitals shall complete the newborn screening report, using the at risk criteria provided by the office on all live births.

B. Hospitals shall conduct hearing screening on all newborn before discharge.

C. Hospitals shall record the results of the hearing screening on the newborn hearing screening report.

D. Hospitals shall disseminate copies of the newborn hearing screening report to the parent, the office (within 14 calendar days of discharge), and the infant's primary health care provider.

E. If an infant is born in one hospital and transferred to one or more hospital(s), the last hospital to which the infant is transferred before being discharged into the care of a parent, or guardian for purposes other than transport, must complete the newborn infant hearing report and perform the hearing screening.

F. If an infant is to be placed for adoption and is to be transferred to another hospital for adoption, the hospital at which the infant is born is to complete the newborn hearing screening report and perform the hearing screening (unless 2205. E above applies). The parent copy of the newborn hearing screening report shall be sent to the guardian.

G. Referrals for infants failing the hospital screening process must be made within seven days of discharge to the infant's primary health care provider and a licensed audiologist.

AUTHORITY NOTE: Promulgated in accordance with R.S. 46:2261-2267.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 19: 1431 (November 1993) amended LR 28:0000 (April 2002).

2207. Procedures for Other (Alternative) Birthing Sites

A. When the infant is born outside the hospital, the person filling out the birth certificate shall complete the newborn hearing screening report.

B. Hearing screening shall be performed at the alternative birthing site before discharge. The results of the screening shall be recorded on the newborn hearing screening report.

C. The person completing the newborn hearing screening report shall disseminate the copies to the parent, primary health care provider, and the office (within 14 calendar days).

D. Referrals for infants who are unable to be tested at the birthing site or who fail the alternative birthing site screening process must be made within seven days of discharge to the infant's primary health care provider and a licensed audiologist.

AUTHORITY NOTE: Promulgated in accordance with R.S. 46:2261-2267.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 19: 1431 (November 1993) amended LR 28:0000 (April 2002).

2209. Hearing Screening Procedures

A. Personnel. Hearing screening will only be performed by:

1. Board eligible or board certified physicians with special training in auditory brainstem response testing and/or otoacoustic emissions and in infant hearing testing. Evidence of training must be submitted to the office.

2. Audiologists licensed by the Louisiana Board of Examiners for Speech Pathology and Audiology with special training in auditory brainstem response testing and/or otoacoustic emissions testing and in infant hearing testing. Evidence of training must be submitted to the office.

3. Persons trained and supervised by personnel meeting requirements for 2209.A.1 or 2 above.

A. A board-certified or board-eligible physician or licensed audiologist who is supervising another individual performing hearing screening must at least be accessible by telephone while the screenings are being performed, review a percentage of the screening documentation and copies of the newborn hearing screening report and perform periodic direct observation of each individual at least once per month as they perform hearing screenings. After an individual supervised by an audiologist or physician has performed hearing screening under the above supervision for one year, direct observation every three months is required.

NOTE: To minimize liability it is recommended that the standard for special training be by an accredited medical or educational institution and include sufficient practicum for proficiency. Any deviation from this recommended standard may increase liability.

B. Test Procedures. The following test procedures are the only acceptable methods for use in infant hearing screening:

1. Auditory Brainstem Response (ABR) either automated or non-automated.
2. Evoked Otoacoustic Emission (EOAE)
3. Test levels, failure criteria and all other test parameters are set by protocols established by the office, upon recommendations of the State Advisory Council.

C. Test Environment. The facility providing the hearing screening tests shall make all efforts possible to insure testing is conducted in a quiet environment.

D. Calibration of Equipment, Hearing screening equipment shall be calibrated annually and documentation maintained at the screening site.

AUTHORITY NOTE: Promulgated in accordance with R.S. 46:2261-2267.
HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 19: 1431 (November 1993) amended LR 28:0000 (April 2002).

2210. REFERRAL AND FOLLOW-UP

A. Referrals for infants failing screening must be made to the infant's primary care physician and a licensed audiologist within 7 days of discharge by the birthing center.

B. Appropriate protocols and standards for diagnostic evaluations to determine hearing loss shall be established by the office, upon recommendations of the State Advisory Council.
AUTHORITY NOTE: Promulgated in accordance with R.S. 46:2261-2267.
HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 19: 1431 (November 1993) amended LR 28:0000 (April 2002).

2211. Confidentiality of Information

All information on the individual newborn hearing screening report is considered confidential and cannot be released by the office, the hospital or the primary health care facility without the parent or guardian's written informed consent.

AUTHORITY NOTE: Promulgated in accordance with R.S. 46:2261-2267.

HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 19: 1431 (November 1993) amended LR 28:0000 (April 2002).

2213. Risk Registry and Tracking

A. The office will maintain a risk registry to include information on all live births and infants identified as at risk for hearing loss.

B. The office will track at-risk infants who fail or do not receive hearing screening prior to hospital discharge. Assistance will be provided for service referrals when necessary.

C. The office will track and notify parents of infants and children at risk for progressive loss of the appropriate procedures for follow-up testing and monitoring of their child's hearing until age 5.

D. The office will develop a system of reporting diagnosis of hearing loss by primary healthcare providers, audiologists, and parents for children up to age 5.

E. The office will disseminate statistical reports regarding the number of infants tested and the number with diagnosed hearing loss to the Louisiana commission for the Deaf, the Louisiana School for the Deaf, the Department of Education, and other interested parties on an annual basis.

F. Infants and children diagnosed with hearing loss shall be referred to appropriate agencies for rehabilitation and education services parental/caregiver consent. For infants and toddlers, up to age 3 with diagnosed hearing loss, referral to Early Steps shall be made for early intervention services.

G. Non-compliance and penalties:

1.. The State Advisory Council shall recommend to the office methods of monitoring hospitals, physicians and audiologists for compliance with all sections of this statute.

2. The State Advisory Council shall report any hospital, physician or audiologist found to be non-compliant to the appropriate licensing, regulatory or other appropriate agency.

AUTHORITY NOTE: Promulgated in accordance with R.S. 46:2261-2267.
HISTORICAL NOTE: Promulgated by the Department of Health and Hospitals, Office of Public Health, LR 19:1432 (November 1993) amended LR 28:0000 (April 2002).

David W. Hood
Secretary

Appendix A3:
Hearing Aid Legislation
Enrolled as ACT No. 816
Regular Session, 2003
SENATE BILL NO. 408
BY SENATORS GAUTREAUX AND HINES AND REPRESENTATIVES FAUCHEUX, MURRAY AND PEYCHAUD

AN ACT
To enact R.S. 22:215.24, relative to health insurance; to provide hearing aid coverage for minor children; and to provide for related matters.
Be it enacted by the Legislature of Louisiana: Section 1. R.S. 22:215.24 is hereby enacted to read as follows:

§215.24. Hearing aid coverage for minor child **R.S. 22:215.24 is all new law.**

A. As used in this Section, "hearing aid" shall mean a non-disposable device that is of a design and circuitry to optimize audibility and listening skills in the environment commonly experienced by children.

B. This Section shall apply to the following entities:
(1) Insurers and nonprofit health service plans, including the office of group benefits, that provide hospital, medical, or surgical benefits to individuals or groups on an expense-incurred basis under health insurance policies or contracts that are issued or delivered in this state.
(2) Health maintenance organizations as defined and licensed pursuant to Part XII of Chapter 2 of this Title that provide hospital, medical, or surgical benefits to individuals or groups under contracts that are issued or delivered in this state.

C.(1) Notwithstanding the provisions of Act No. 1115 which originated as House Bill No. 1606 of the 2003 Regular Session of the Louisiana Legislature to the contrary, an entity subject to this Section shall provide coverage for hearing aids for a child under the age of eighteen who is covered under a policy or contract of insurance if the hearing aids are fitted and dispensed by a licensed audiologist or licensed hearing aid specialist following medical clearance by a physician licensed to practice medicine and an audiological evaluation medically appropriate to the age of the child.

(2)(i) An entity subject to this Section may limit the benefit payable under Paragraph (1) of this Subsection to **one thousand and four hundred dollars per hearing aid for each hearing-impaired ear** every thirty-six months.
(ii) An insured or enrolled individual may choose a hearing aid that is priced higher than the benefit payable under this Subsection and may pay the difference between the price of the hearing aid and the benefit payable under this Subsection without financial or contractual penalty to the provider of the hearing aid.

(iii) In the case of a health insurer or health maintenance organization that administers benefits according to contracts with health care providers, hearing aids covered pursuant to this Section shall be obtained from health care providers contracted with the health insurer or health maintenance organization. Such providers shall be subject to the same contracting and credentialing requirements that apply to other contracted health care providers.

D. This Section does not prohibit an entity subject to the provisions of this Section from providing coverage that is greater or more favorable to an insured or enrolled individual than the coverage required under this Section.

E. The provisions of this Section shall apply to any new policy, contract, program, or plan issued by an entity subject to the provisions of this Section on or after January 1, 2004. Any such policy, contract, program, or plan in effect prior to January 1, 2004 shall convert to the provisions of this Section on or before the renewal date thereof but in no event later than January 1, 2005. Any policy affected by the provisions of this Section shall apply to an insured or participant under such policy, contract, program or plan whether or not the hearing impairment is a pre-existing condition of the insured or participant.

F. The provisions of this Section shall not apply to individually underwritten, guaranteed renewable limited benefit health insurance policies.

Appendix B: Joint Committee on Infant Hearing

1. Joint Committee on Infant Hearing 2007 Position Paper Summary

2. Risk Factors Associated with Permanent Congenital, Delayed-onset or Progressive Hearing loss in Childhood

Appendix B1:

Joint Committee on Infant Hearing 2007 Summary

Following are the principles of the Joint Committee on Infant Hearing (JCIH) 2007 Position Statement:

- *All infants will have access to a newborn hearing screening test using physiologic measures before 1 month of age.*
- *All infants who do not pass the initial hearing screening and the subsequent rescreening should have appropriate diagnostic audiologic evaluation by an audiologist trained in infant testing and a medical evaluation to confirm the presence of hearing loss before 3 months of age.*
- *All infants with confirmed permanent hearing loss should receive intervention service including amplification if warranted before 6 months of age.*
- *A simplified, single point of entry into an intervention system appropriate to children with hearing loss is optimal.*
- *The EHDI system should be family-centered with infant and family rights and privacy guaranteed through informed choice, shared decision making, and parental consent.*
- *Families should have access to information about all intervention and treatment options and counseling regarding hearing loss.*
- *The child and family should have immediate access to high-quality technology, including hearing aids, cochlear implants, and other assistive devices when appropriate.*
- *All infants and children should be monitored for hearing loss in the medical home. Continued assessment of hearing and communication development should be provided by appropriate providers to all children with or without risk indicators for hearing loss at specified intervals.*
- *Appropriate interdisciplinary intervention programs for deaf and hard of hearing infants and their families should be provided by professionals knowledgeable about childhood hearing loss. Intervention programs should recognize and build on strengths, informed choices, traditions, and cultural beliefs of the families.*

Appendix B2:

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

(Joint Committee on Infant Hearing Position Statement, 2007)

The timing and number of hearing reevaluations for children with risk factors should be **customized and individualized** depending on the relative likelihood of a subsequent delayed-onset hearing loss.

> **Risk indicators that are in bold print** are of greater concern for delayed-onset hearing loss and will need close audiologic monitoring. The previous 2000 JCIH position statement **recommended every 6 months for the first three years.**
>
> Infants who pass the neonatal screening but have any of the other risk factor should have **at least 1 diagnostic audiology assessment by 24 to 30 months of age**.

Risk Indicators:

1. **Caregiver concern** regarding hearing, speech, language, or developmental delay.

2. **Family history** of permanent childhood hearing loss. If a blood relative of the infant had a permanent hearing loss from birth or which began in early childhood and needed a hearing aid or special schooling for the hearing-impaired. This DOES NOT include hearing loss due to illness, ear infections, or aging.

3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: **ECMO,** assisted ventilation, exposure to ototoxic medications (gentamicin and tobramicin) or loop diuretics (Furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.

4. In utero infections, such as **CMV,** herpes, rubella, syphilis, and toxoplasmosis.

5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.

6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.

7. **Syndromes associated with hearing loss or progressive or late-onset hearing loss,** such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.

8. **Neurodegenerative disorders,** such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.

9. **Culture-positive postnatal infections associated with sensorineural hearing loss,** including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.

10. **Head trauma, especially basal skull/temporal bone fracture** that requires hospitalization.

11. **Chemotherapy.**

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

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