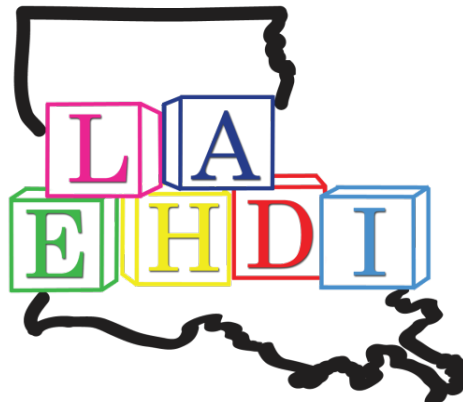


Louisiana Pediatric Diagnostic Audiology Guidelines

LOUISIANA EARLY HEARING DETECTION AND INTERVENTION (EHDI)



Louisiana Early Hearing
Detection and Intervention
Program

**Department of Health
Office of Public Health
Bureau of Family Health
EHDI**

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INTRODUCTION

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**.

The early hearing detection and intervention (EHDI) program supports early diagnosis and timely enrollment into early intervention services. The goal of the EHDI program is to maximize linguistic competence and literacy development for children who are deaf or hard of hearing. Without early diagnosis and appropriate early intervention, these children will fall behind their hearing peers in communication, cognition, reading, and social emotional development.

JOINT COMMITTEE ON INFANT HEARING (JCIH) BENCHMARKS

EHDI 1-3-6 GOALS

- ❖ ***Before 1 month of age:
Complete initial newborn hearing screening***
- ❖ ***Before 3 months of age:
Complete diagnostic assessment***
- ❖ ***Before 6 months of age:
Enroll in early intervention***

INFANTS WITH “FURTHER TESTING NEEDED” RESULT AFTER NEWBORN HEARING SCREENING

- Test both ears, **even if only 1 ear did not pass the hospital screening.**
- If ABR was used for hospital screening, ABR **must** be used for rescreen.
- An infant who does not pass the rescreen in both ears at a single visit must be referred to a pediatric audiologist. Multiple screening attempts are not warranted and often further delay proper diagnosis of hearing loss.
- Regardless of newborn hearing screening results, an infant with risk indicators for delayed onset hearing loss requires diagnostic assessment by 24 to 30 months of age.

INFANTS WHO DID NOT PASS AFTER OUTPATIENT RESCREENING

- **Diagnosis and evaluation of the type and degree of hearing loss should be completed before the child is 3 months of age.**
- If referral to another facility becomes necessary to complete or confirm diagnostic assessment, the audiologist should assist in linking the family to the second facility.
- **DO NOT** complete a second rescreening.
- Repeated rescreens:
 - Delay the confirmation of hearing status
 - Increase the likelihood of obtaining a false negative result
 - Increase the likelihood that sedation will be required to complete the diagnostic evaluation
- Diagnostic audiological evaluations can be completed during medical management of a middle ear problem. **DO NOT delay.**

PROCEDURES FOR INFANTS WITH RISK INDICATORS FOR DELAYED-ONSET OR PROGRESSIVE HEARING LOSS

Regardless of newborn hearing screening results, children with risk indicators for delayed-onset hearing loss should have a diagnostic assessment by an audiologist at least once by 24 to 30 months of age.

Early and more frequent assessment may be needed for children with a high-risk indicator, denoted below with an asterisk (*).

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

Risk Indicators Associated with Delayed-onset or Progressive Hearing Loss in Childhood

- *Family history of permanent childhood hearing loss**
- *Congenital CMV infection**
- *Syndrome associated with hearing loss**
- *Caregiver concern regarding hearing, speech, language, or developmental delay**
- *Extracorporeal membrane oxygenation (ECMO)**
- *Culture-positive postnatal infections, such as viral or bacterial meningitis**
- *Chemotherapy**
- *Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome**
- Neonatal intensive care of more than 5 days**
- Exposure to ototoxic medications (i.e., gentamicin and tobramycin)**
- Exposure to loop diuretics (i.e., furosemide and Lasix)**
- Hyperbilirubinemia that requires exchange transfusion**
- In utero infections, such as herpes, rubella, syphilis, and toxoplasmosis**
- Assisted ventilation**
- Craniofacial anomalies**
- Physical findings, such as white forelock, associated with a syndrome known to include a sensorineural or permanent conductive hearing loss**
- Head trauma, especially basal skull/temporal bone fracture that requires hospitalization**

DIAGNOSTIC ASSESSMENT

Pediatric diagnostic assessment and amplification fitting guidelines were developed through consultation of current clinical practice guidelines published by the American Academy of Audiology and the American-Speech-Language-Hearing Association (see Appendix A).

- ✓ Adequate confirmation of an infant's hearing status cannot be obtained from a single test measure.
- ✓ A diagnostic procedure must include ear specific type, degree, and configuration of the hearing loss.
- ✓ The initial test battery must include physiologic measures and, if possible, developmentally appropriate behavioral techniques.
- ✓ When hearing aids and/or cochlear implants are recommended, electrophysiologic measures of threshold prediction are needed for all children under 3 years of age, as well as any child in whom reliable behavioral audiometric tests cannot be obtained.
- ✓ Behavioral threshold measures should be obtained and cross-checked with prior results as soon as the child is able to participate.

Diagnostic Test Battery should include:

- 1) Detailed history
- 2) Otoscopy
- 3) Evoked otoacoustic emissions
- 4) Acoustic immittance testing
- 5) Diagnostic auditory evoked potential testing
- 6) Behavioral audiometry

Detailed history

Including, but not limited to:

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

Otoscopy

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

Evoked Otoacoustic Emissions

Either Transient or Distortion Product Emissions are acceptable.

Typically, a total of 6 to 8 frequencies are tested in the mid- to high-frequency range, though this may not be possible in young infants due to the presence of high levels of physiologic noise below 1500 Hz.

TEOAE

TEOAE click stimuli:

80 dB peak-equivalent SPL +/- 3 dB stimulus level should be used.

Pass criteria: Signal to noise ratio > 3 to 6 dB in the majority of frequency bands assessed, overall wave reproducibility > 70%, and overall response amplitude within the range typical for normal hearing children of comparable age.

References for TEOAE normative data are located in Appendix B.

DPOAE

DPOAE stimuli:

L1/L2 of 65/55 dB SPL should be used.

Pass criteria: Signal to noise ratio > 3 to 6 dB in the majority of frequency bands assessed, and overall response amplitude within the range typical for normal hearing children of comparable age.

References for DPOAE normative data are located in Appendix B.

Acoustic Immittance Testing

Tympanometry

- A higher probe tone frequency (such as 1,000 Hz) should be used in infants younger than 6 months.
- 226 Hz probe tone is acceptable beginning at 6 months of age.

Acoustic Reflex

Ipsilateral middle ear muscle reflex thresholds for 500, 1000, 2000, and 4000 Hz if possible.

Pass criteria: Type A tympanogram and acoustic reflex thresholds ≤ 95 dB HL for 500 and 4000 Hz and ≤ 100 dB HL for 1000 and 2000 Hz.

Note: Maximum stimulus level should not exceed 105 dB HL due to the possibility of a noise-induced hearing loss caused by the reflex stimulus, (Hunter et al., 1999).

Diagnostic Auditory Evoked Potential Testing

ABR (Auditory Brainstem Response) 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 Criteria: Wave V responses for clicks at 25 dB nHL within a normal absolute latency range for the child's age are generally accepted as within normal limits. Appropriate correction factors may be applied as determined by each individual clinic.

References for ABR normative data are located in Appendix B.

ABR with change of polarity of clicks to confirm or rule out ANSD:

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 spectrum disorder (ANSD)**.

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 ANSD.

Note: *A catch trial where the signal is running but not delivered to the ear should be included to rule out stimulus artifact, which can be indistinguishable from the CM. Stimulus artifact may be misinterpreted as the CM. The catch trial consists of running an additional average with the earphone tubing clamped or disconnected. The CM should disappear. If not, the observed response is likely stimulus artifact.*

If a determination of ANSD is made, toneburst testing is not necessary in the presence of an abnormal ABR, as it will not yield any additional information.

Pass Criteria: Presence of similar waveform morphology and latency with both polarities.

ABR to tonebursts:

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

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

Pass Criteria: Wave V responses are typically attempted down to 20 dB nHL for tonebursts at each frequency tested (ideally 500, 1000, 2000, and 4000 Hz). Normal results would consist of Wave V responses at 20-35 dB nHL, depending on frequency, (Elsayed et al., 2015). Appropriate correction factors may be applied as determined by each individual clinic.

References for ABR normative data are located in Appendix B.

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

Bone conduction ABR:

Stimuli should be bone-conducted clicks; masking of the non-test ear should be applied, as appropriate. Due to differences in skull structure of infants compared to adults, forehead placement of the bone oscillator should be avoided because it yields smaller response amplitudes and higher thresholds than mastoid and temporal placements.

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 Criteria: Normal results would consist of Wave V responses at 25-35 dB nHL, depending on frequency, (Elsayed et al., 2015). Appropriate correction factors may be applied as determined by each individual clinic.

References for ABR normative data are located in Appendix B.

ASSR (Auditory Steady State Response)

ASSR may be used to supplement ABR results. Thresholds should be obtained at frequencies of 500, 1000, 2000, and 4000 Hz. Use of ASSR to measure bone conduction thresholds is not advised in infants and young children due to the increased risk of detecting stimulus artifact (Small & Stapells, 2004; Swanepoel et al., 2008).

Pass Criteria: Results should correlate with behavioral and ABR thresholds. There are no accepted normative standards for ASSR in young children. Correction factors are typically applied to ASSR thresholds to obtain an estimate of behavioral thresholds. A 10 dB correction factor is widely used, but many have noted correlations to behavioral thresholds to vary based on ASSR carrier frequency and test duration. See Dimitrijevic et al. (2002), Francois et al. (2016), and Small and Stapells (2006) for more information on ASSR air and bone conduction correction factors.

References for ASSR normative data are located in Appendix B.

Behavioral audiometry

Visual reinforcement audiometry (VRA)/Conditioned Play Audiometry (CPA):
if appropriate due to child's developmental level.

Stimuli should be speech as well as frequency-specific tones from 250-6000 Hz. Insert earphones or supra-aural/circumaural headphones are preferable; sound field may be necessary with some children who will not tolerate earphones or headphones.

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

USE OF SEDATION IN THE EVALUATION PROCESS

Sound sleep during electrophysiologic testing is desired to obtain clean recordings with low noise. Natural sleep is preferred, but when this is not possible, sedation may be needed to complete testing. **No child, especially those under 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 typically developing children from birth to 6 months of age can be tested using sleep deprivation and other techniques to induce natural sleep.

Each facility should develop its own protocols surrounding sedation and anesthesia in accordance with institutional guidelines. Consultation with the American Academy of Pediatrics Guidelines for Monitoring and Management of Pediatric Patients During and After Sedation for Diagnostic and Therapeutic Procedures (2006) is advised when developing these protocols.

AMPLIFICATION FOR INFANTS AND CHILDREN BIRTH – 3 YEARS

Timeline for amplification fitting:

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

Pediatric Amplification should be considered for the following:

- a. Pure tone average or high frequency pure tone average is greater than 25 dB HL in at least one ear.
- b. Pure tone average of either ear is greater than 15 dB HL and the child is exhibiting speech and language difficulties due to fluctuating or mild hearing loss.
- c. Unilateral loss. Fitting with a standard air conduction hearing aid is optimal if sufficient residual hearing exists. For children with severe or profound losses, Contralateral routing of the signal (CROS) or bone conduction devices may be considered.
- d. Mild or greater conductive hearing loss due to chronic otitis media with effusion, especially if medical treatment is being deferred or delayed for several months.
- e. Permanent conductive hearing loss. Air conduction hearing aids are the preferred amplification treatment when anatomically possible. Bone conduction hearing aids may be warranted in cases where coupling is not possible (atresia, malformation of the pinna, or chronic drainage).
- f. Potential cochlear implant candidates. An ABR finding of “no response” does not exclude a child from hearing aid candidacy, as audible residual hearing may exist beyond the stimulus limits of the ABR test.

Never delay the fitting of an amplification device until resolution of otitis media (OM). Refer to a physician for medical management and monitor the status of the OM when determining appropriate prescriptive targets during the hearing aid fitting.

Amplification/Hearing Aid fitting Guidelines:

The goal of the amplification device fitting is to provide the child 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 child'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. 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) which incorporate the use of individual real-ear measurements that account for each infant's ear canal acoustics and hearing loss must 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 child's hearing loss, the goals of auditory habilitation, the child's acoustic environment, and the family's informed choice.

Pediatric Referral for Cochlear Implant:

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 spectrum disorder (ANSD) who are not developing speech and language skills on target. A hearing aid trial in patients with ANSD is still indicated in most cases.

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 RESCREENING AND DIAGNOSTIC RESULTS

Proper reporting is crucial to the success of the EHDI program! Timely reporting allows future providers to view previous test results and obtain a more comprehensive clinical history.

Report screening and diagnostic testing to Louisiana Early Hearing Detection and Intervention (LA EHDI) on a Follow-up Services Report (FSR) form ldh.la.gov/index.cfm/page/3533. To register to submit forms electronically, go to <https://ehdi.opd.dhh.la.gov>. FSR paper forms can be faxed to 504-568-5854.

Submit FSRs for children diagnosed with permanent childhood hearing loss **within 2 days of diagnosis**. Submit all other FSRs within 7 days of testing.

Which results should be reported?

1. Rescreening and/or diagnostic testing results on all **infants with a “further testing needed” result** from 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. Any child birth-5 years of age with a change in hearing status (ex. original diagnosis - unilateral, now bilateral, or original diagnosis - mild, now severe)
5. Any child birth-5 years of age **fitted with a hearing aid or cochlear implant for the first time.**
6. Every child that is considered **lost to follow-up** for your facility. This is any child who failed to keep their rescreening, diagnostic testing or hearing aid fitting appointment.

REFERRAL TO EARLY INTERVENTION

Early intervention is an integral part of the habilitation process for all children identified as deaf or hard of hearing. Simply establishing an amplification system is only the first step.

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

In Louisiana, the Parent Pupil Education Program (PPEP) is the designated single point of entry into early intervention for children identified with any type or degree of hearing loss.

What happens when a child is identified as deaf or hard of hearing?

Within 48 hours of diagnosis:

- The Audiologist submits a Follow-up Services Report (FSR) to EHDH

Upon receipt of FSR, EHDH immediately makes referrals to:

- The Parent Pupil Education Program (PPEP), for home-based early intervention services from a teacher specializing in the education of young deaf/hard-of-hearing children alsd.org/outreach
- Hands & Voices (H&V) Guide by Your Side (GBYS) for parent to parent support from the parent of a child who is deaf or hard of hearing ahandsandvoices.org/guide-by-your-side

Two weeks after receipt of FSR:

- EHDH makes a referral to LA EarlySteps dh.la.gov/earlysteps
A child with any degree of unilateral or bilateral permanent hearing loss from birth to 36 months of age qualifies for services.

Appendix A: REFERENCES

American Academy of Audiology Practice Guidelines

audiology.org/sites/default/files/publications/resources/Assessment%20of%20Hearing%20in%20Infants%20and%20Young%20Children_FINAL%208.2012.pdf

audiology.org/sites/default/files/publications/PediatricAmplificationGuidelines.pdf

American Academy of Pediatrics, American Academy of Pediatric Dentists. (2006). Guidelines for Monitoring and Management of Pediatric Patients During and After Sedation for Diagnostic and Therapeutic Procedures: An Update. *Pediatrics* 118:6 2587-2602.

American Speech-Language-Hearing Association: Permanent Childhood Hearing Loss

asha.org/PRPSpecificTopic.aspx?folderid=8589934680§ion=Assessment

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Joint Committee on Infant Hearing (JCIH) 2007 Position Statement

pediatrics.aappublications.org/content/120/4/898.full?ijkey=oj9BAleq21OIA&keytype=ref&siteid=aapjournals

Joint Committee on Infant Hearing (JCIH) 2013 Supplement

jcih.org/JCIH-2007-Position-Statement-Supplement.pdf

Louisiana Insurance Coverage for Hearing Aids, ACT 816-2003

new.dhh.louisiana.gov/assets/oph/Center-PHCH/Center-PH/hearingspeechvision/Hearing_Aid_Legislation.pdf

Appendix B: NORMATIVE DATA REFERENCES

TEOAE normative data:

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DPOAE normative data:

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ASSR normative data:

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