Pediatric Diagnostic Audiology Guidelines

Louisiana Early Hearing Detection and Intervention Program

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PARTNERS FOR FAMILY HEALTH LOUISIANA
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LOUISIANA DEPARTMENT OF HEALTH

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INTRODUCTION

The Louisiana Early Hearing Detection and Intervention (LA EHDI) program supports coordinated systems of care that ensure all infants and children receive hearing screenings, and all who are deaf or hard of hearing receive timely diagnosis, referral to early intervention services, and referral to family to family support. The goal of the program is to ensure that children who are deaf or hard of hearing (DHH) have the opportunity for appropriate early intervention to optimize language, literacy, cognitive, social and emotional development.

The following guidelines have been developed specifically for pediatric diagnostic services provided to children from **birth to 3 years of age**. The EHDI program has a number of family-facing resources, developed with the input of both families and providers, to support providers and families through the diagnostic process. These materials are available free of charge, are linked throughout the guidelines, are available electronically through the Louisiana Department of Health website[1], and upon request, LA EHDI will send these to providers for use in their practice.

LA EHDI recognizes the diversity of the individuals and families in Louisiana. The family-facing materials are available in English and Spanish, but do not take the place of offering translation/interpreter services for families whose primary language is not English/spoken English.

JOINT COMMITTEE ON INFANT HEARING (JCIH) BENCHMARKS[2]

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

*JCIH encourages reducing timelines to 1-2-3 as soon as feasible

[1] https://ldh.la.gov/page/LouisianaEHDI
[2] https://www.infanthearing.org/nhstc/docs/Year%202019%20JCIH%20Position%20Statement.pdf

RESCREENING INFANTS WITH "FURTHER TESTING NEEDED" NEWBORN HEARING SCREENING RESULT

- Auditory brainstem response (ABR) **must** be used to rescreen infants screened in the neonatal intensive care unit (NICU).
- Well-babies screened with ABR may be rescreened using otoacoustic emissions (OAE) or ABR.
- Both ears **must** be tested, **even if only 1 ear did not pass the newborn screening**.
- An infant who does not pass the outpatient rescreen in **one or both ears must** be referred to a pediatric audiologist.
- Multiple screening attempts are not warranted and prevent timely diagnosis.
- Regardless of newborn hearing screening results, an infant with any risk factor for delayed-onset hearing loss requires diagnostic assessment by 9 months of age, or sooner dependent upon specific risk factor (see p. 6).
- Share "Your Baby Needs a Follow-Up Hearing Screening: What Families Need to Know[3]"

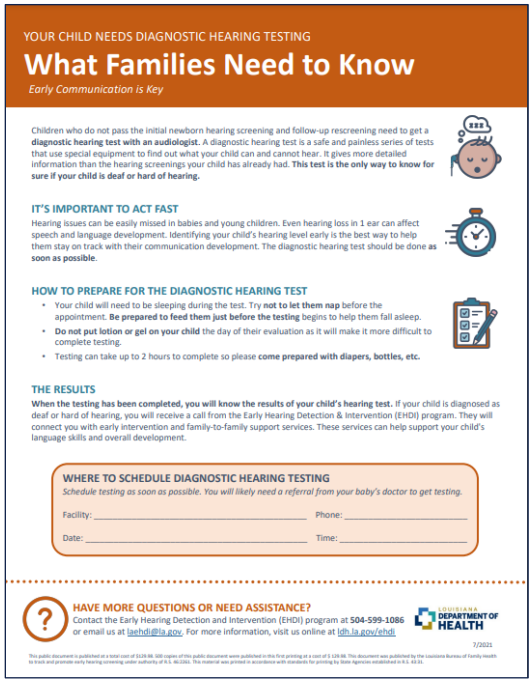
[3] https://ldh.la.gov/assets/oph/Center-PHCH/Center-PH/cshs/EHDI/EHDI_Rescreening_FINAL.pdf

INFANTS WHO DO NOT PASS OUTPATIENT RESCREENING

- Diagnosis and evaluation of the type and degree of hearing loss should be completed before the child is 3 months of age.
- 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.
- 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.
- Share "Your Child Needs Diagnostic Hearing Testing: What Families Need to Know[4]"



YOUR CHILD NEEDS DIAGNOSTIC HEARING TESTING

What Families Need to Know

Early Communication is Key

Children who do not pass the initial newborn hearing screening and follow-up rescreening need to get a **diagnostic hearing test with an audiologist**. A diagnostic hearing test is a safe and painless series of tests that use special equipment to find out what your child can and cannot hear. It gives more detailed information than the hearing screenings your child has already had. **This test is the only way to know for sure if your child is deaf or hard of hearing.**

IT'S IMPORTANT TO ACT FAST

Hearing issues can be easily missed in babies and young children. Even hearing loss in 1 ear can affect speech and language development. Identifying your child's hearing level early is the best way to help them stay on track with their communication development. The diagnostic hearing test should be done **as soon as possible.**

HOW TO PREPARE FOR THE DIAGNOSTIC HEARING TEST

- Your child will need to be sleeping during the test. Try **not to let them nap** before the appointment. **Be prepared to feed them just before the testing** begins to help them fall asleep.
- **Do not put lotion or gel on your child** the day of their evaluation as it will make it more difficult to complete testing.
- Testing can take up to 2 hours to complete so please **come prepared with diapers, bottles, etc.**

THE RESULTS

When the testing has been completed, you will know the results of your child's hearing test. If your child is diagnosed as deaf or hard of hearing, you will receive a call from the Early Hearing Detection & Intervention (EHDI) program. They will connect you with early intervention and family-to-family support services. These services can help support your child's language skills and overall development.

WHERE TO SCHEDULE DIAGNOSTIC HEARING TESTING

Schedule testing as soon as possible. You will likely need a referral from your baby's doctor to get testing.

Facility: _____ Phone: _____

Date: _____ Time: _____

HAVE MORE QUESTIONS OR NEED ASSISTANCE?

Contact the Early Hearing Detection and Intervention (EHDI) program at **504-599-1086** or email us at laehdi@la.gov. For more information, visit us online at ldh.la.gov/ehdi

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7/2021

[4] https://ldh.la.gov/assets/oph/Center-PHCH/Center-PH/cshs/EHDI/EHDI_Diagnostics_FINAL.pdf

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

Regardless of newborn hearing screening results, children with risk factors for delayed-onset hearing loss should have a diagnostic assessment by an audiologist at least once by 9 months of age, or sooner, dependent upon specific risk factor(s).

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

Immediate referral
Caregiver concern regarding hearing, speech, language, developmental delay and/or developmental regression
ABR by 1 month
Mother + Zika and infant with laboratory evidence of Zika
No later than 3 months after occurrence
Extracorporeal Membrane Oxygenation (ECMO)
In utero infection with cytomegalovirus (CMV)
Culture positive postnatal infections
Head Trauma
Chemotherapy
By 9 months
Family History of permanent childhood hearing loss
Neonatal Intensive Care More Than 5 Days
Hyperbilirubinemia with exchange transfusion
Aminoglycosides > 5 days
Asphyxia or Hypoxic Ischemic Encephalopathy
In utero infections associated with hearing loss (other than CMV)
Syndromes or birth conditions associated with hearing loss

Share "Your Baby Needs Another Hearing Test By 9 Months of Age: What Families Need to Know[5]"


YOUR BABY NEEDS ANOTHER HEARING TEST BY 9 MONTHS OF AGE

What Families Need to Know

Knowing your child's hearing level is key to language development

Your baby passed the newborn hearing screening. However, they will need to be tested again because they are at risk for hearing loss that can show up later. This means that even though their hearing level is okay today, there is an increased chance that their hearing can change in the future.

For more information about your baby's risk factor(s), review your child's *Newborn Hearing Screening Report Form* (it's a pink form with additional hearing information on the back).

Why Is This Important?

Your baby's communication and language development begins at birth. Children use sights and sounds to take in the world around them and learn how to communicate. It is important to have your baby's hearing retested **by 9 months of age** help them stay on track.

What Are Your Next Steps?

9 MONTHS

- Talk to your baby's doctor to schedule a diagnostic hearing test with an audiologist (a specialist in hearing testing).
- This is a safe and painless test to determine what your baby can and cannot hear.
- If you have any questions, or need help finding an audiologist, just give us a call or visit **ehdipals.org**.

Have Questions or Need Assistance?

Contact the Early Hearing Detection and Intervention (EHDI) program at **504-599-1086** or email us at laehdi@la.gov. For more information, visit us online at ldh.la.gov/ehdi.

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[5] https://ldh.la.gov/assets/oph/Center-PHCH/Center-PH/cshs/EHDI/EHDI_Diagnostics_FINAL.pdf

DIAGNOSTIC ASSESSMENT

The following 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

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

Prenatal, birth, and neonatal history	Parental concerns
Hearing screening results	History of middle ear pathologies
Risk factors for early childhood hearing loss	Medical history including: syndromes or other heritable conditions, craniofacial anomalies, kidney issues, conditions of limbs/digits, pigmentation issues, exposure to ototoxic medications
Family history of childhood hearing loss	
Parental report of auditory behaviors	
Motor, cognitive, and visual development	
Emerging communication milestones	

2. Otoscopy

Visual inspection for obvious structural abnormalities of the pinna and ear canal

3. Evoked Otoacoustic Emissions

Either Transient Evoked (TEOAE) or Distortion Product (DPOAE) Otoacoustic 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.

4. Acoustic Immittance Testing

- Tympanometry
    - A higher probe tone frequency (such as 1,000 Hz) should be used in infants younger than 6 months. Continued use through 9 months of age may yield greater sensitivity and specificity for middle ear status for certain populations, such as infants with Down syndrome, (Heeren, 2013).
    - 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).

5. Diagnostic Auditory Evoked Potential Testing

Either ABR (Auditory Brainstem Response) and/or ASSR (Auditory Steady State Response) may be performed.

- ABR to tonebursts:
    - Threshold responses should be obtained via air conduction to tonebursts at a variety of frequencies. If possible, testing should begin with a high frequency (i.e., 2000 Hz) in each ear, followed by a low frequency (i.e., 500 Hz) in each ear. Bone conduction threshold determination at 500 Hz and 2000 Hz should immediately follow, unless air conduction thresholds at these frequencies are unequivocally within normal limits. If time permits, additional air conduction thresholds should be obtained at 4000 Hz and 1000 Hz.
    - Frequency-specific chirp stimuli may be considered for ABR testing, but due to limited published literature on normative data, results should be interpreted with caution.
    - All infants with elevated air conduction thresholds should receive bone conduction assessment via tonal ABR. 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.
    - **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.

- Auditory Neuropathy Spectrum Disorder (ANSD):
    - If there is no ABR response at the limits of the equipment to air conduction at 2000 Hz, or if all ASSR thresholds are abnormal, an assessment for auditory neuropathy spectrum disorder (ANSD) should be initiated. Supra-threshold click testing should include one average with condensation clicks and another average at the same intensity with rarefaction clicks.
    - 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, further threshold testing with ABR or ASSR is not necessary, as it will not yield any additional information.

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

- ASSR (Auditory Steady State Response)
  - Threshold responses should be obtained via air conduction to tonal ASSR at a variety of frequencies. If possible, testing should begin with a high frequency (i.e., 2000 Hz) in each ear, followed by a low frequency (i.e., 500 Hz) in each ear. If time permits, additional air conduction thresholds should be obtained at 4000 Hz and 1000 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, particularly in individuals with hearing loss (Small & Stapells, 2004; Swanepoel et al., 2008).
  - Infants with more than a mild hearing loss should have bone conduction thresholds measured using tonal ABR. Any disparities identified between air-conducted tonal ASSR and bone-conducted tonal ABR should be cross-checked with air-conducted tonal ABR.
  - **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.

6. Behavioral audiometry

- Visual reinforcement audiometry (VRA)/Conditioned Play Audiometry (CPA): if appropriate for 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. Due to ear volume and calibration differences, headphones are preferable over insert earphones for children with patent pressure equalization (PE) tubes.
  - **Pass Criteria**: 20 dB HL to speech and threshold responses at 500, 1000, 2000, and 4000 Hz tones.

References for VRA normative data are located in Appendix B.

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 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 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 TO 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:

- Pure tone average or high frequency pure tone average is greater than 25 dB HL in at least one ear.
- 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.
- 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.
- 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.
- 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).
- Potential cochlear implant candidates. An ABR finding of “no response” does not exclude a child from hearing aid candidacy, as aidable residual hearing may exist beyond the stimulus limits of the ABR test.

Note: **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.

- The hearing aid size, make and model should be appropriate for the child’s hearing loss, age, and development.
- For infants under 6 months of age, hearing aid fitting will usually be based on physiologic measures alone using real-ear measurements.
- 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.

- Age appropriate hearing aid prescriptive formulas, such as DSL (desired sensation level), 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 frequency modulated (FM), digitally modulated (DM), and Bluetooth systems, and/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 (CI):

According to the current Food and Drug Administration (FDA) guidelines, a cochlear implant:

- Is appropriate for children over 9 months of age with profound bilateral sensorineural hearing loss.
- 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.
- 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.
- May be considered for children 5 years and older with single-sided deafness.

NOTE: 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

Timely reporting allows all providers to view test results and obtain a comprehensive clinical history. To report screening and diagnostic testing to Louisiana Early Hearing Detection and Intervention (LA EHDI), register to become a LA EHDI-Information System (LA EHDI-IS) user.

This allows you to submit a Follow-up Services Report (FSR) electronically. Register at https://laehdiis.ldh.la.gov. Alternatively, FSR paper forms can be downloaded from ldh.la.gov/index.cfm/page/3533, and scanned to LAEHDI@la.gov or faxed to 504-568-5854.

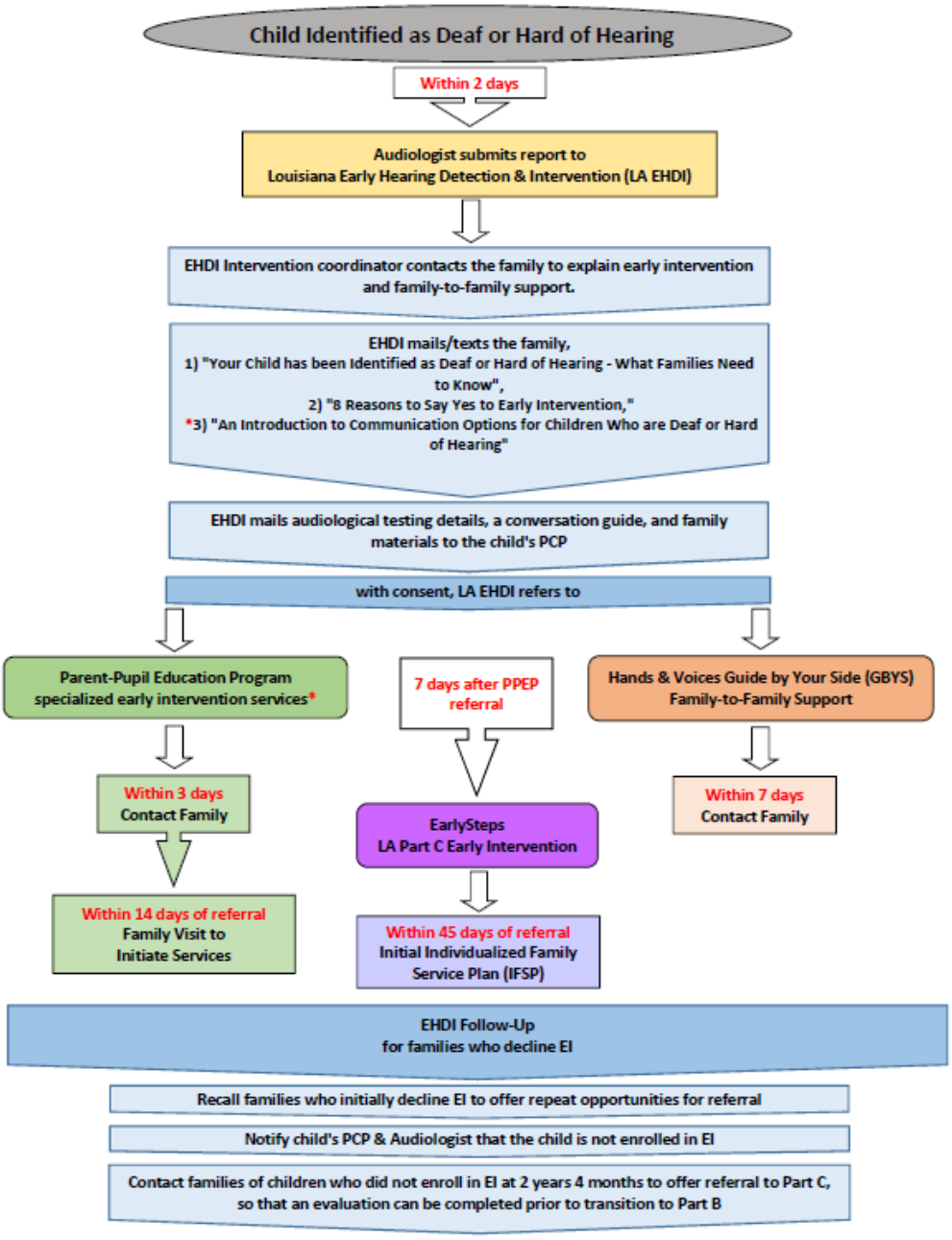
Report children initially diagnosed with permanent childhood hearing loss to LA EHDI **within 2 days.** Submit all other FSRs within 7 days of testing.

Report **all** results for children birth to 5 years of age:

- With a "further testing needed" result from a newborn hearing screening.
- Who have never been screened.
- Identified as deaf or hard of hearing for the first time.
- With a change in hearing status (ex. original diagnosis - unilateral, now bilateral, or original diagnosis - mild, now severe).
- Fitted with a hearing aid or cochlear implant for the first time.
- Lost to follow-up for your facility. This is any child who failed to keep their rescreening, diagnostic testing or hearing aid fitting appointment(s).

REFERRAL TO EARLY INTERVENTION

Early intervention is an integral part of ensuring support for all children identified as deaf or hard of hearing and their families. According to the Individuals with Disabilities Education Act (IDEA) Federal Law, all children identified as deaf or hard of hearing, **must be referred to early intervention.**



*Children with unilateral losses do not receive the Communication Options booklet, nor referral to the PPEP program, except in cases where there may be progression of loss.

LA EHDI ID to EI Flowchart 4.26.2024

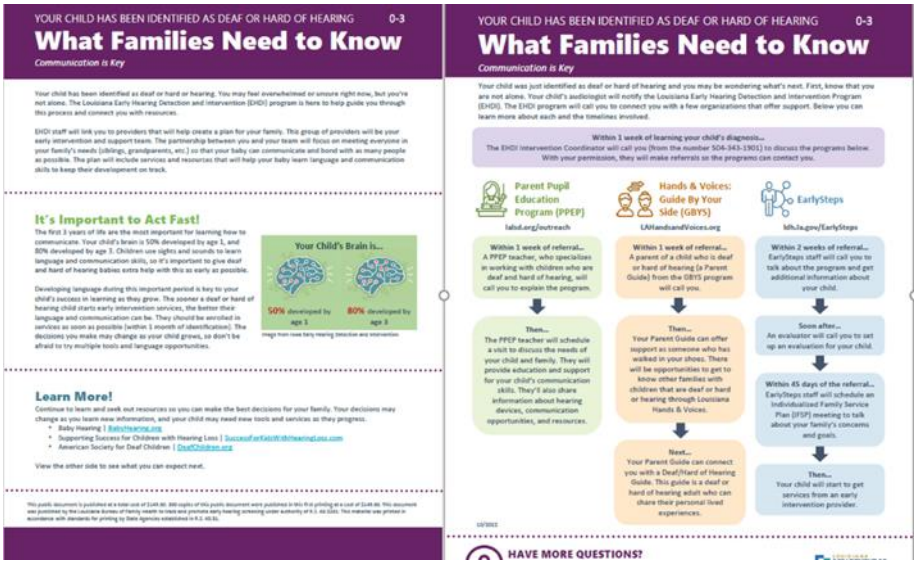
Families are referred to the following supports (outlined in flowchart on page 15):

- EarlySteps[6], for community-based early intervention services for children birth-36 months of age. Any child with a unilateral or bilateral permanent childhood hearing loss, 25 dB or greater is eligible.
- The Parent Pupil Education Program (PPEP)[7], for community- based early intervention services from a teacher specializing in the education of young deaf/hard-of-hearing children
- Hands & Voices (H&V) Guide by Your Side (GBYS)[8] for parent to parent support from the parent of a child who is deaf or hard of hearing

Share "An Introduction to Communication Options for Children Who Are Deaf or Hard or Hearing[9]"

[6] https://ldh.la.gov/page/early-steps
[7] https://lsdvi-lalsd.ss18.sharpschool.com/outreach
[8] https://www.lahandsandvoices.org/
[9] https://ldh.la.gov/assets/oph/Center-PHCH/Center-PH/cshs/EHDI/EHDI_Commmunications_Guide_BMAC_RESIZED.pdf

Share ["Your Child Has Been Identified as Deaf or Hard of Hearing: What Families Need to Know[10]"](#)



Share ["Your Child Has Been Identified as Deaf or Hard of Hearing In One Ear: What Families Need to Know[11]"](#)

[10] https://ldh.la.gov/assets/oph/Center-PHCH/Center-PH/cshs/EHDI/IdentifiedAsDeafOrHardOfHearingRoadmap.pdf

[11] https://ldh.la.gov/assets/oph/Center-PHCH/Center-PH/cshs/EHDI/IdentifiedAsDeafOrHardOfHearingRoadmap_Unilateral.pdf

APPENDIX A: REFERENCES

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APPENDIX B: NORMATIVE DATA REFERENCES

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