

# Audiologists' Guide for Infant Diagnostic Evaluation



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# Early Hearing Detection & Intervention (EHDI)

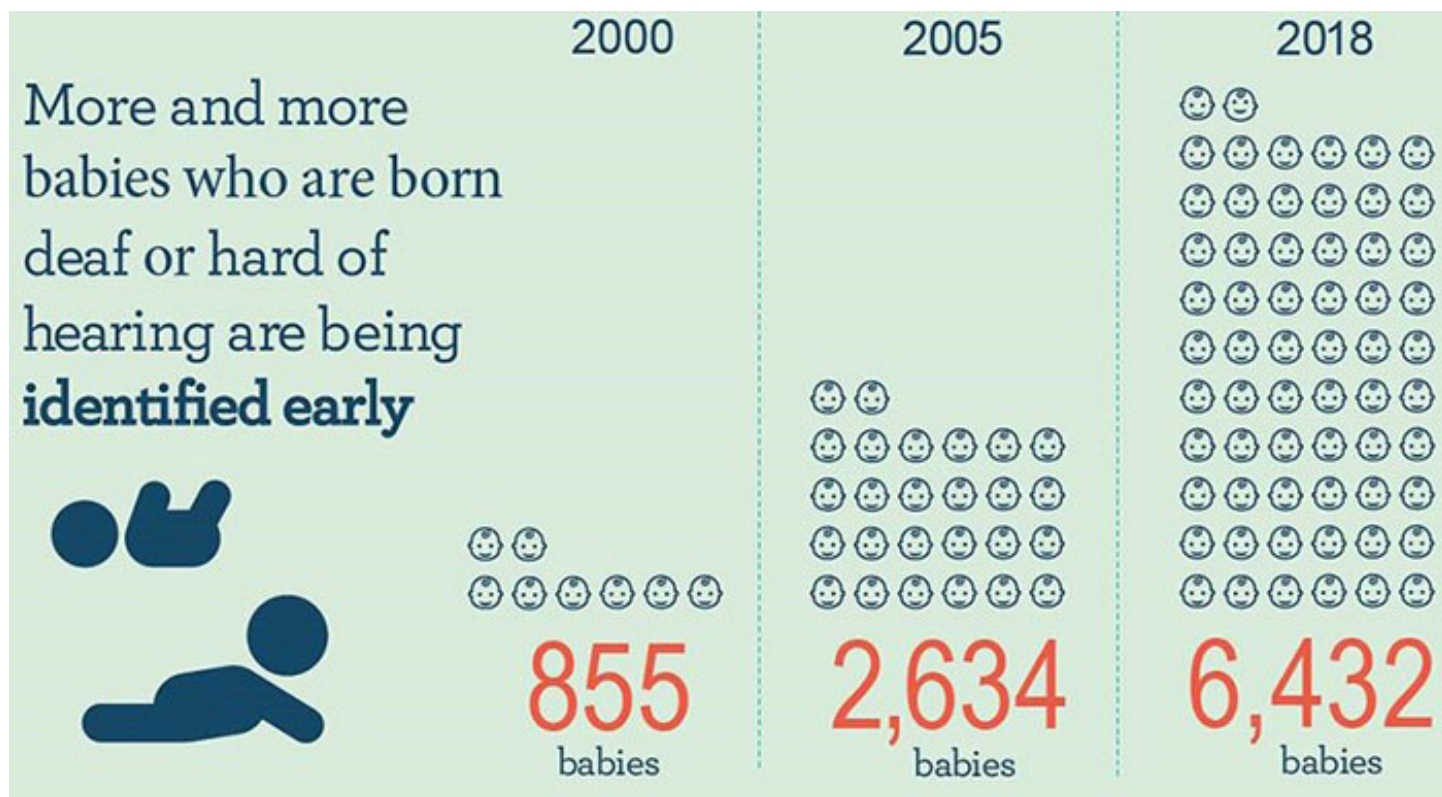
## Who is EHDI?

Each year, an estimated 1 to 3 per 1,000 infants are born with moderate, severe, or profound hearing loss resulting in potentially delayed development in language, learning, and speech. Even more will lose their hearing during infancy or childhood (American Speech-Language-Hearing Association 2020). Children who are deaf or hard of hearing face a neuro-developmental emergency and need to be identified as quickly as possible so appropriate intervention services can begin.

The mission of Arkansas' EHDI program, Arkansas Department of Health's Infant Hearing Program is to ensure all newborns meet the Joint Committee on Infant Hearing (JCIH) EHDI goals:

- Ensure early identification of all deaf and hard-of-hearing children and access to timely intervention enrollment.
- Enhance providers' knowledge about EHDI 1-3-6 guidelines – screening by 1 month of age, diagnosis of hearing loss by 3 months of age, and enrollment in early intervention by 6 months of age if diagnosed with hearing loss.
- Incorporate EHDI into an integrated, medical-home approach to child health.
- Ensure newborn hearing screening results are clearly communicated to parents and reported to EHDI in accordance with state laws and rules.

For tracking and surveillance, the EHDI Program needs hospitals, primary care physicians, pediatricians, audiologists, and interventionists to provide accurate and complete information regarding hearing screening, testing, and intervention next steps on a consistent basis. This also allows the EHDI Program to connect families to appropriate resources and services.



# Arkansas Law: Act 1559 of 1999

## Arkansas Law – Act 1559 of 1999

An Act to ensure early detection of hearing loss for all newborns/infants in Arkansas and to establish the universal newborn/infant hearing screening, tracking, and intervention program and advisory board.

- All birthing hospitals must provide bilateral hearing screening on each birth admission;
- All birthing hospitals, physicians, or other providers administering initial screening or follow-up care must report results to the Arkansas Department of Health's (ADH) Infant Hearing Program (IHP); and
- All birthing hospitals, physicians, or other providers administering initial screening or follow-up care shall inform parents where to obtain medical and audiological follow-up screening/care.

All test results are reported via the Electronic Registry of Arkansas Vital Events (ERAVE), a web-based database that collects and reports hearing, birth, and death records.

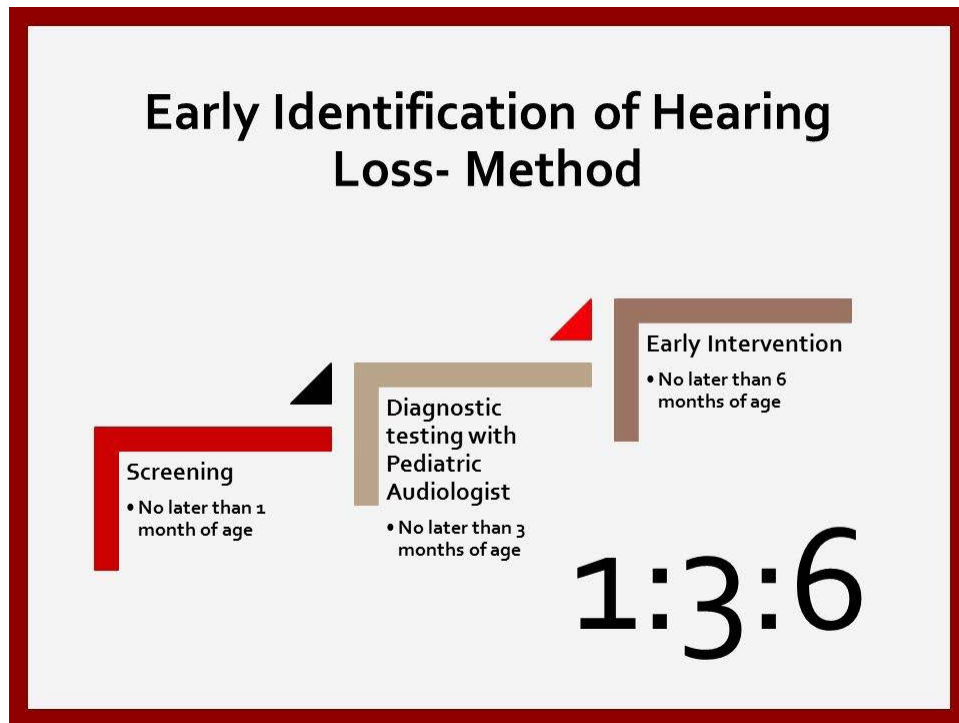


# When Should Infants Be Screened?

## 1-3-6 EHDI Plan

Infant hearing loss has been defined as a neurodevelopmental emergency. Children with hearing loss are at risk not only for lifelong deficits in speech and language acquisition, but poor academic performance, personal-social maladjustments and emotional difficulties. Timing is critical to optimize outcomes. Babies with hearing loss identified in the first weeks of life, and who begin hearing optimally no later than six months, have a good chance of developing neural connections in their auditory brain pathways necessary to lay the foundation for spoken language development.

Newborn Hearing Screening Checklist provided with this toolkit to assist with next steps in the 1-3-6 process, [https://www.healthy.arkansas.gov/images/uploads/pdf/NBHS\\_Checklist.pdf](https://www.healthy.arkansas.gov/images/uploads/pdf/NBHS_Checklist.pdf)



# 1-3-6 NEWBORN HEARING SCREENING CHECKLIST

Patient Name: \_\_\_\_\_ Patient DOB: \_\_\_\_\_ Date of Visit: \_\_\_\_\_

<b>1 INITIAL SCREENING</b> <i>(by no later than 1 month of age)</i>		
Has the child had a newborn hearing screening?	<b>Yes</b>	<b>No</b> ⇒ Schedule initial screening
Did you obtain the test results from the screening hospital or state EHDI program?	<b>Yes</b>	<b>No</b> ⇒ Contact the hospital or state EHDI program
Are the results recorded in the patient's chart?	<b>Yes</b>	<b>No</b> ⇒ Record test results in patient chart
Did the child pass the newborn hearing screening?	<b>Yes</b>	<b>No</b> ⇒ Schedule rescreening appointment
Have the results been reported to the state EHDI program?	<b>Yes</b>	<b>No</b> ⇒ Confirm results have been reported to state EHDI program within 48 hours of receiving them
Have results been discussed with family?	<b>Yes</b>	<b>No</b> ⇒ <input type="checkbox"/> For a child who passed, stress the importance of ongoing surveillance and risk factors* <input type="checkbox"/> For a child who did not pass, discuss the need for follow-up and assist in arranging a rescreening
Has a rescreening occurred (if the initial screen resulted in "did not pass" or if otherwise necessary)?	<b>Yes</b>	<b>No</b> ⇒ Schedule rescreening appointment
<b>RESCREENING</b> <i>(by no later than 1 month of age)</i>		
Where will the rescreening be performed? ✓ If hospital/outpatient center, when is the rescreening appointment? ✓ If conducted in office: • Determine what screening equipment was used at the hospital. • Follow the AAP office rescreening guidelines.	<input type="checkbox"/> Hospital: _____ <input type="checkbox"/> Office <input type="checkbox"/> Other (specify): _____ Location: _____ Date: _____	
Did the child pass the rescreening?	<b>Yes</b>	<b>No</b> ⇒ Send child to audiologist with pediatric expertise for diagnostic evaluation.
Are the results recorded in the patient chart?	<b>Yes</b>	<b>No</b> ⇒ Record results in patient chart.
Have the results been discussed with the family?	<b>Yes</b>	<b>No</b> ⇒ <input type="checkbox"/> For a child who passed, stress the importance of ongoing surveillance and risk factors* <input type="checkbox"/> For a child who did not pass, discuss the need for follow-up and assist in arranging an audiologic evaluation
Have the results been reported?	<b>Yes</b>	<b>No</b> ⇒ Confirm results have been reported to state EHDI program within 48 hours of receipt

<b>3</b> <b>DIAGNOSTIC EVALUATION</b> <i>(by no later than 3 months of age)</i>		
If the child did not pass the rescreening, was he/she referred to an audiologist with expertise in pediatrics?	<b>Yes</b> Provider: _____ Date of Visit: _____	<b>No</b> ⇒ Refer to audiologist with expertise in pediatrics
Were the results of the diagnostic test normal?	<b>Yes</b>	<b>No</b> ⇒ Discuss early intervention (EI) and need for comprehensive plan
Have the results been discussed with the family?	<b>Yes</b>	<b>No</b> ⇒ <input type="checkbox"/> For a child who passed, stress the importance of ongoing surveillance and risk factors* <input type="checkbox"/> For a child who did not pass, discuss EI and need for comprehensive plan
Have the results been reported?	<b>Yes</b>	<b>No</b> ⇒ Confirm results have been reported back to state EHDl program within 48 hours of receipt
<b>6</b> <b>EARLY INTERVENTION</b> <i>(by no later than 6 months of age)</i>		
If the child was diagnosed with a hearing loss, was he/she referred for early intervention and multidisciplinary evaluation?	<b>Yes</b> Date of visit: _____	<b>No</b> ⇒ Provide referral for EI, ophthalmology, and otolaryngology and offer referral for genetics
<b>ONGOING SURVEILLANCE AND SCREENING</b>		
Continue to perform ongoing surveillance and screening for late-onset hearing loss, particularly children with risk factors.		

\*JCIH Risk Factors



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# Roles and Responsibilities of Audiologists

## The Role of the Audiologist in Arkansas' EHCI Program:

- Provide a seamless progression of hearing healthcare throughout the EHCI process to ensure infants and their families receive appropriate and timely services from screening through evaluation and early intervention.
- Serve as key stakeholder assisting the Infant Hearing Program in developing and implementing comprehensive, community-based protocols to facilitate optimal outcomes for infants and families.
- Provide education to community/hospital personnel identifying the importance of early identification and intervention of hearing loss in infants.

## Responsibilities during the Diagnostic Hearing Evaluation:

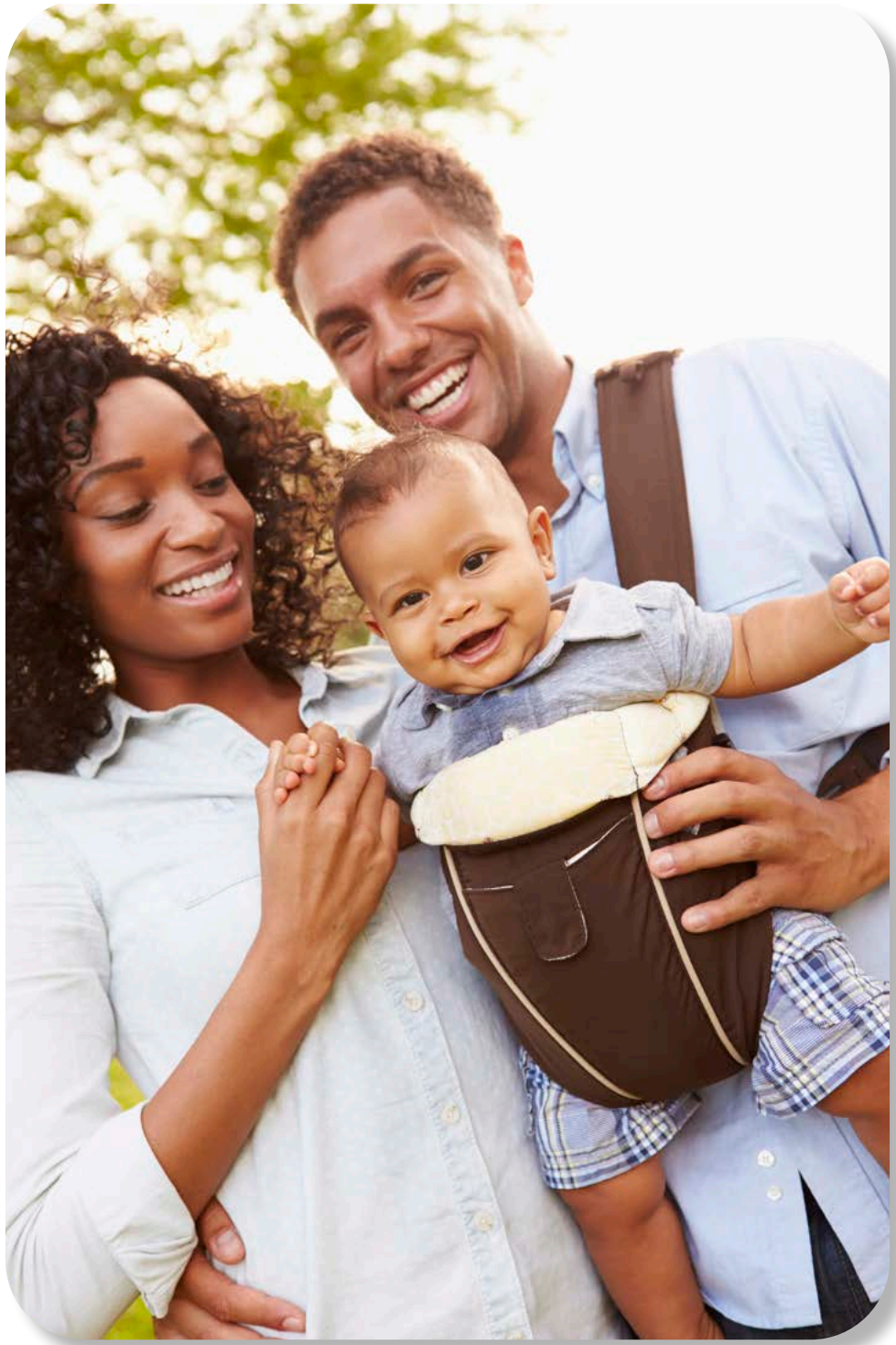
- Conduct appropriate diagnostic testing to determine type and degree of hearing loss.
- Share the results of the diagnostic hearing evaluation both verbally and in writing with the parents/caregivers (in the family's primary language, if possible).
- Ensure the family can restate the next steps following the diagnostic appointment including results and the need for future appointments.
- Provide information about unbiased communication options and resource materials to families.
- Provide information about early intervention services to families.

## Responsibilities after the Diagnostic Evaluation:

- Monitor and evaluate infants and toddlers at risk for hearing loss.
- Report results to the child's Primary Care Provider and the Infant Hearing Program within 7 business days of the evaluation.
- Remind families/caregivers of any upcoming appointments for additional testing.







# Pediatric Audiological Assessment

The following best practice recommendations were developed by the Arkansas Department of Health's Infant Hearing Program in collaboration with the Universal Newborn Hearing Screening, Tracking, and Intervention Advisory Board. The recommendations take into consideration information from national organizations, such as, the National Center for Hearing Assessment and Management (NCHAM), the Joint Committee of Infant Hearing (JCIH), the American Academy of Audiology (AAA) and the American Speech-Language-Hearing Association (ASHA).

This document serves to facilitate the diagnosis of hearing loss in infants. The Arkansas Department of Health recognizes individual cases may present unique challenges influencing the approach to evaluation and management. Therefore, these recommendations should be regarded as a guideline for practice, not as standards, and are not intended or designed to substitute for the reasonable exercise of independent clinical judgment. However, the protocol presented is consistent with generally accepted "standards of care" in pediatric audiology.

## Initial Hearing Screen/Rescreen

Otoacoustic emissions (OAEs) and automated auditory brainstem response (ABRs) are physiologic measures that can be used to screen newborns and infants for hearing loss. The following summarized protocols for the initial hearing screen/rescreen process were obtained from the 2020 JCIH Position Statement.

### ○ **Screening Protocols in the Well-Infant Nursery**

- OAEs and automated ABRs are both acceptable screening measures for infants in the well-infant nursery.
- Infants in the well-infant nursery who fail automated ABR testing should not be rescreened by OAE testing and "passed," because such infants are presumed to be at risk of having a subsequent diagnosis of auditory neuropathy/dyssynchrony.

### ○ **Screening Protocols in the NICU**

- JCIH recommends ABR technology as the only appropriate screening technique for use in the NICU.
- For infants who do not pass automated ABR testing in the NICU, referral should be made directly to an audiologist for rescreening and, when indicated, comprehensive evaluation, including diagnostic ABR testing, rather than general outpatient rescreening.

### ○ **Outpatient Rescreening for Infants Who Do Not Pass the Birth Admission Screening**

- The outpatient rescreening should include the testing of both ears, even if only one ear failed the inpatient screening.
- If the infant does not pass the rescreening, a referral for a diagnostic evaluation should be made to an audiologist.

## Professional qualifications for Providers Completing Diagnostic Assessments

Any audiologist accepting infants for diagnostic audiological evaluations should be prepared with the equipment, training and skills to complete the procedures recommended by (JCIH). A licensed audiologist with experience in the pediatric population is the professional best qualified to perform diagnostic audiological assessments for infants. If the audiologist does not have the expertise and/or equipment necessary to evaluate and manage infants/young children, consideration for a referral should be made to professionals and facilities that provide pediatric diagnostic services.

## Recommended Pediatric Audiologic Assessment

### Birth to 6 Months of Age

When testing infants birth to 6 months of age, the confirmatory audiological test battery includes:

- Child and family history (including an evaluation of risk factors for congenital hearing loss).
- Assessment of parental report of the infant's response to environmental sounds.
- Clinician observation of the infant's auditory behavior as a crosscheck in conjunction with electrophysiological measures.
- Confirm middle ear status with one or more of the following techniques:
  1. High frequency tympanometry.
  2. Pneumatic otoscopy by a physician experienced in its application.
  3. Acoustic reflex threshold testing should be considered especially with concern of auditory neuropathy.
  4. Distortion product and/or transient evoked OAEs (TEOAE and/or DPOAE).
  5. Frequency specific assessment of hearing sensitivity using ABR air-conduction toneburst at 500Hz, 1000Hz, 2000Hz, and 4000Hz with bone-conduction toneburst utilized when indicated.
  6. When permanent hearing loss is detected, frequency-specific ABR testing is needed to determine the degree and configuration of hearing loss in each ear for fitting of amplification devices.
  7. Click-evoked ABR testing using both condensation and rarefaction single-polarity stimulus, if there are risk indicators for neural hearing loss (auditory neuropathy/auditory dyssynchrony) such as hyperbilirubinemia or anoxia, to determine if a cochlear microphonic is present.
  8. Because some infants with neural hearing loss have no risk indicators, any infant who demonstrates "no response" on ABR elicited by tone-burst stimuli must be evaluated by a click-evoked ABR.
  9. Identify cases of non-permanent hearing loss (non-congenital origin, such as fluctuating conclusive loss).

### Six Months to 36 Months of Age

- For subsequent testing of infants and toddlers at developmental ages of 6 to 36 months, the confirmatory audiological test battery includes:
  - Child and family history including an evaluation of risk factors for congenital hearing loss and parental report of auditory and visual behaviors and communication milestones.
  - Confirm middle ear status with one or more of the following techniques:
    - Tympanometry.
    - Pneumatic otoscopy by a physician experienced in its application.
    - Acoustic reflex threshold testing should be considered especially when concern is present of auditory neuropathy.
    - Distortion product and/or transient evoked OAEs (TEOAE and/or DPOAE), when indicated.
    - Behavioral audiometry (either visual reinforcement or conditioned-play audiometry, depending on the child's developmental level), including pure-tone audiometry across the frequency range for each ear.
    - ABR testing if responses to behavioral audiometry are not reliable.

### Once Assessment is Completed

- It is important to discuss the results of the evaluation and follow-up recommendations with parents.
- The audiologist is responsible for entering the initial diagnostic and any subsequent audiological evaluation results into ERAVE within 2 days following the appointment.
- Notify the infant's Medical Home Provider (MHP) of results.

### Normal Hearing

- Provide information to the parents about hearing, speech and language milestones and information regarding risk indicators for progressive hearing loss. Encourage the parents to schedule the infant for a follow-up evaluation in the future should concerns arise.

## **Normal Hearing (Identified as “At Risk” for Acquired or Late Onset Hearing Loss)**

- The JCIH Year 2020 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs has outlined those conditions that put infants at greater risk for late onset/ progressive or progressive hearing loss. These indicators include:
  - Caregiver concern regarding hearing, speech, language or developmental delay.\*
  - Family history of permanent childhood hearing loss.\*
  - Neonatal intensive care admission of more than 5 days or any of the following, regardless of length of stay: ECMO, assisted ventilation,\* exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.\*
  - In utero infections, such as CMV, herpes, rubella, syphilis, and toxoplasmosis.\*
  - Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits and temporal bone anomalies.
  - Physical findings, such as white forelock, that are associated with a syndrome known to include sensorineural or permanent conductive hearing loss.
  - 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.
  - Neurodegenerative disorders, such as Hunter syndrome\* or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
  - Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.\*
  - Head trauma, such as basal skull/temporal bone fracture, \* that requires hospitalization.
  - Chemotherapy\*
- \*Greater concern for delayed onset hearing loss
- 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. According to the JCIH 2020 Position Statement, all infants with a risk factor for hearing loss should be referred for an audiological assessment at least once by 24 to 30 months of age. Children with risk indicators that are highly associated with delayed-onset hearing loss, such as having received ECMO or CMV infection, should have more frequent audiological assessments.
- The Arkansas Infant Hearing Program collects risk factor data from the birth facilities in the ERAVE system. The birth facility has the responsibility of assessing all newborns for risk factors, and for reporting any recognized risk factors to the IHP through ERAVE. The birth facility should notify the newborn’s Medical Home Provider (MHP) of any identified risk factors associated with the potential for late onset/progressive or progressive hearing loss that warrants the need for ongoing audiological evaluations.

## **Confirmed Hearing Loss**

- If a permanent hearing loss is present, the audiologist should make the necessary accommodations for the infant to be fit with amplification and be seen for follow up care, as needed. This includes a referral to Early Intervention (EI), medical clearance from an ENT, and all results/recommendations sent to the Medical Home Provider (MHP). The diagnosing audiologist should provide the parent/responsible party with the information they need to make informed decisions regarding early intervention options.
- If the presence of middle-ear fluid or pathology is noted with a hearing loss, the infant should be referred to the Medical Home Provider (MHP) for diagnosis, treatment, and potential referral to ENT. A follow-up appointment should be within 4-6 weeks to determine middle ear status and complete testing to rule out sensorineural hearing loss within 4-6 weeks. If middle ear fluid persists, every effort should be made to assess hearing status within the three months time window.

# Reporting Results to EHDI

## Introduction

The Electronic Registration of Arkansas Vital Records (ERAVE) system is provided by the Arkansas Department of Health. The ERAVE system provides authorized users a secure, online method for submitting and managing reports of vital events including deaths, infant hearing screenings, births, and fetal deaths. The Arkansas Infant Hearing Program utilizes ERAVE for reporting infant hearing screening results and tracking infants with known or suspected hearing loss (follow up). ERAVE allows hospital staff, audiologists, Primary Care Physicians (PCP), Early Intervention Specialists, and IHP staff to facilitate children with hearing loss are receiving the timely follow-up and specialized care they need.

## Evaluation Status

- **Completed** – Evaluation obtained, results may be abnormal, normal, or unspecified.
- **Could Not Test** – Child present for evaluation, but no information obtained.
- **Inconclusive** – Evaluation attempted, but minimal data obtained preventing the evaluation from being completed or determining if hearing loss exists.
- **Previously Passed** – Evaluation not conducted due to child previously passing evaluation.

## Type of HL

- **Conductive Permanent** – Child has a permanent conductive hearing loss preventing sound from reaching the outer or middle ear; includes atresia, microtia, etc. Definite EI referral.
- **Conductive Undetermined** – Child has abnormal evaluation results, but further testing is required to confirm permanent loss or resolve the issue causing the loss at the time of evaluation. Child will be re-evaluated and additional information submitted. EI referrals will be made on a case-by-case basis if signs of developmental delay present or if the case is unresolved for a long period of time. Unknown if hearing loss is permanent or not.
- **Mixed** – Child has a permanent hearing loss including both a conductive and a sensorineural loss. Definite EI referral.
- **Sensorineural** – Child has a permanent hearing loss due to a problem in the inner ear or hearing nerve. Definite EI referral.
- **Neural** – Child has a permanent hearing loss due to damage to the inner ear or Auditory nerve (Auditory Neuropathy Spectrum Disorder - ANSD). Definite EI referral.
- **Transient** – Child has a fluctuating conductive condition indicating hearing loss of a non-congenital origin. The hearing loss is lasting only for a short time; non-permanent. EI referral is likely not required.

## Hearing Loss

- **Yes** – Evaluation completed. Results are abnormal and hearing loss is likely permanent.
- **No** – Evaluation completed. Results are normal.
- **Undetermined** – Evaluation completed, but not complete enough to confirm hearing loss or normal hearing. Results are inconclusive or hearing loss is likely temporary.

## Special Reminders

- Audiologist should include notations in the notes tab if there are details not otherwise addressed by the standard menu item response (i.e. details about future appointments and related information to guide the IHP in next steps for every diagnosed child).
- Each diagnosis should identify when non-permanent hearing loss is suspected.

# Communicating Results To Families

## Suggestions for Communicating Results to Families



**DO** give a **POSITIVE** message:

- “Your baby did not pass the hearing screening.”
- “Your baby did not pass on (the left/right or either ear), which means more information is needed about your baby’s hearing”
- “The next step is an outpatient follow-up rescreen for your baby when they are at least 10 days old”.



**DO** give the “**AR Specific Roadmap**” handout to explain processes for rescreen and audiological diagnostic evaluations.



**DO** discuss why an outpatient follow-up rescreen is needed. According to your hospital’s newborn hearing screening procedures, possibly offer to assist with getting an appointment with another facility if your hospital does not perform follow-up rescreens.



**DO NOT** say:

- Your baby failed or referred.
- A lot of babies don’t pass. (minimizes need to attend rescreen)
- Your baby doesn’t need follow-up testing.
- Your baby was fussy. (Then it was an invalid screening)
- The equipment was not working right. (Then it was an invalid screening)



**DO NOT** assume:

- It’s just because of fluid or vernix.
- Your baby is deaf or hard of hearing.
- Probably nothing is wrong.



**DO NOT** perform multiple screens in an attempt to get a pass:

- Complete two screen attempts, one inpatient and one outpatient rescreen, then refer for diagnostic testing.

# Has your child been diagnosed as deaf or hard of hearing?



Organizations who will contact you:

How they can help:

## Infant Hearing Program

(Arkansas Department of Health)  
501-280-4740  
[www.arhealthyhearing.com](http://www.arhealthyhearing.com)

Supply information for parents on hearing screenings and connect parents to available resources before and after diagnosis to help you understand the process and make informed decisions. This is your first step towards getting early intervention services for your child.

## Guide By Your Side

(Arkansas Chapter of Hands and Voices)  
501-492-9162  
[www.arhandsandvoices.org](http://www.arhandsandvoices.org)

Connect a parent guide (parent of a deaf or hard of hearing child) to other parents of children with hearing loss to provide support, share real life experiences and help them utilize a network of available services.

## First Connections

(Arkansas Department of Human Services)  
1-800-643-8258  
<https://dhs.arkansas.gov/dds/firstconnectionsweb>

Help families with a child with special needs to secure services that will help the child reach their fullest potential, and to enhance the abilities of the family to assist their child.

## ASD Outreach Services

(Arkansas School for the Deaf)  
501-324-9522 or 501-246-8452 Video Phone  
<http://www.arschoolforthe deaf.org>

Offer outreach services to children who are deaf and hard of hearing, their families and service providers.

## Children with Chronic Health Conditions

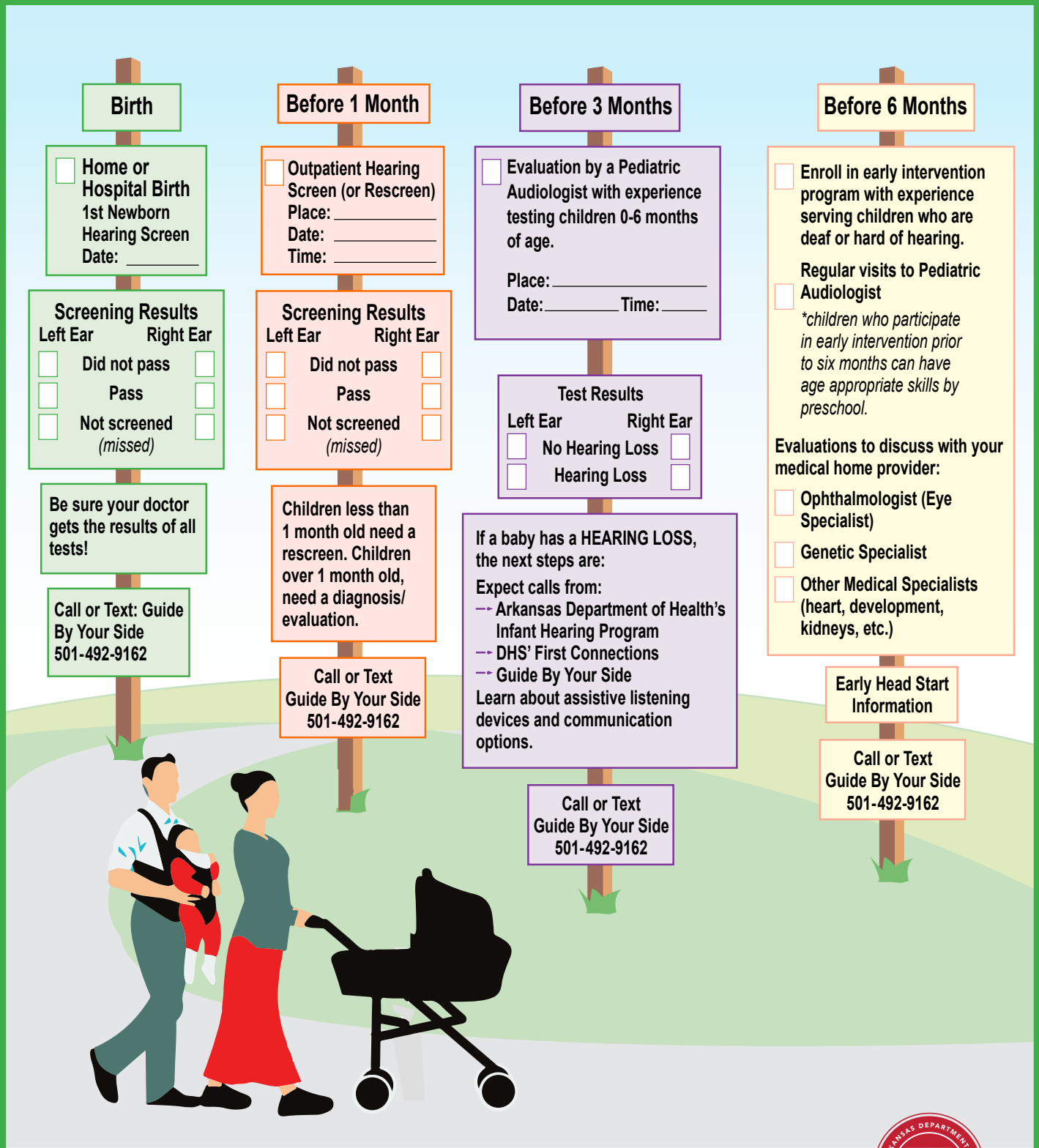
(Arkansas Department of Human Services)  
1-800-482-5850 ext. 22277  
<https://humanservices.arkansas.gov/about-dhs/ddds/childrens-services-information/title-v-children-with-special-health-care-needs-cshcn>

Provide targeted case management services to assist families in accessing all medical, social, education, and other services appropriate to the child's special health care needs.

For more information, call the Arkansas Department of Health Infant Hearing Program at 501-280-4740.



# Arkansas Specific Roadmap

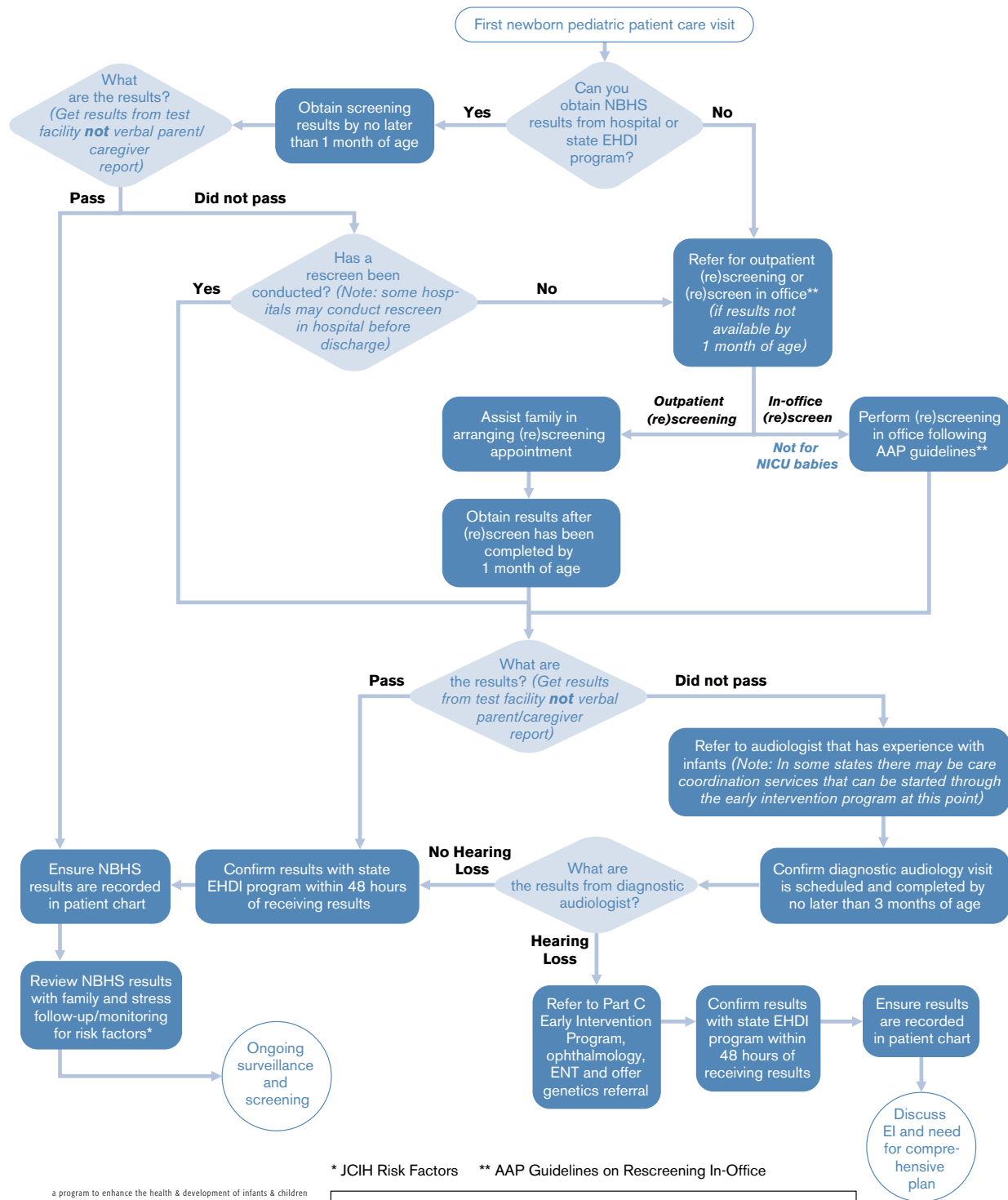


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# REDUCING LOSS TO FOLLOW-UP/DOCUMENTATION IN NEWBORN HEARING SCREENING: GUIDELINES FOR MEDICAL HOME PROVIDERS



\* JCIH Risk Factors \*\* AAP Guidelines on Rescreening In-Office



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## Resources for Audiologists

- Arkansas Infant Hearing Program - <https://www.healthy.arkansas.gov/programs-services/topics/infant-hearing>
- ERAVE - [https://adherave.arkansas.gov/erave/servlet/search\\_hitlist](https://adherave.arkansas.gov/erave/servlet/search_hitlist)
- American Academy of Pediatrics PEHDIC - <https://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Pages/Early-Hearing-Detection-and-Intervention.aspx>
- EHDI-PALS - <http://www.ehdi-pals.org/Default.aspx>
- NCHAM - <http://www.infanthearing.org/index.html>
- Pediatric Audiology Specialty Certification - <http://www.boardofaudiology.org/pediatric-audiology-specialty-certification/>
- American Speech-Language Hearing Association – <http://www.asha.org/default.htm>
- JCIH 2019 Position Statement: principals and Guidelines for Early Hearing Detection and Intervention Programs – <https://digitalcommons.usu.edu/jehdi/vol4/iss2/1/>







For more information:

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