



## Indiana's Early Hearing Detection and Intervention Program Why Universal Newborn Hearing Screening?

- UNHS has become the standard of care with all states and territories participating in this program. The American Academy of Pediatrics, as well as other hearing health organizations, advocate for universal newborn hearing screening.
- Technology now exists to provide safe, cost effective and reliable methods to assess hearing in newborns, making UNHS possible and practical. Screening involves the use of non-invasive, objective physiologic measures.
- Screening procedures for newborns can lead to detection of significant bilateral or unilateral hearing loss. **All children can be evaluated for hearing loss, regardless of their age.**
- Hearing loss is invisible, but the effects can lead to lack of exposure to language and can cause lifelong cognitive, educational and vocational challenges. **This is preventable with early intervention and family education.**
- With one out of every 300 infants born with significant hearing loss, it is one of the most common health conditions found in newborns. In Indiana approximately 250-300 babies will be identified annually.
- If only high-risk babies were screened, half of all babies with hearing loss would be missed.
- The incidence of congenital hearing loss is greater than the sum total of all other conditions detected by newborn metabolic blood screening tests.
- More than 90 percent of infants who are born with or develop early onset hearing loss have parents and families with normal hearing.
- Without universal newborn hearing screening, the average age of diagnosis is over two years of age.
- Recent research indicates that children identified with hearing loss, who receive intervention before six months of age, develop language (spoken or signed) comparable with their hearing peers.
- **Don't wait for signs of hearing loss to appear.** Many children with hearing loss will not have an obvious speech/language delay until 2, 3, or even 4 years of age.
- **There is help for children who are deaf or hard of hearing.** Research indicates that early hearing detection of hearing loss and early enrollment in quality early intervention services specific to the needs of children with hearing loss make a huge difference. Because of early hearing detection and intervention, many children with hearing loss enter kindergarten with age-appropriate communication skills.



## Indiana's Early Hearing Detection and Intervention Program Referral for Delayed Onset Risk Factors

Indiana's UNHS Policy Manual identifies four risk factors for delayed onset hearing loss that require referral to Indiana State Department of Health. Babies who pass the screening but have one of the following risk factors need to be referred. Risk factors include:

1. A family history of permanent childhood hearing loss
2. Exposure to in-utero infection (See Newborn Risk Criteria)
3. Bad jaundice (hyperbilirubinemia) that needed a special procedure (exchange transfusion)
4. Ears that are formed in a different way than usual (no ear, partial ear or no ear canal opening) Ears that are formed in a different way than usual (no ear, partial ear or no ear canal opening). If ear anomalies prevent hearing screening from being completed, children should be reported as not passing UNHS, reported to ISDH, and referred for immediate testing with an audiologist.
5. Babies with other craniofacial anomalies who are screened and pass, should be referred to the Primary Care Provider (PCP) for follow-up diagnostic testing at 9-12 months of age.

Families of babies who pass the screening, but have one of these risk factors should:

1. Be informed about the risk factors identified
2. Be provided a copy of the hearing and language developmental milestones and told to monitor the child's progress
3. Be referred to their PCP on the discharge summary
4. Be referred to ISDH on the Monthly Summary Report and to the PCP for follow-up evaluation at 9-12 months of age with further testing as recommended by the audiologist
5. Be informed of the results and the importance of follow-up testing

### **Tips for UNHS Screeners Referral for Other Risk Indicators**

Other risk factors do exist (as identified below). For babies who pass the screen but have one of these additional risk factors, a referral to the child's primary care physician should be completed on the discharge summary.

- Syndromes commonly associated with hearing loss (Down, Usher, Waardenburg and Neurofibromatosis Type 2)
- Neonatal intensive care stay with any of the following regardless of length of stay: ECMO, assisted ventilation, exposure to oto-toxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/lasix.) If uncertain about the risk, please check with the neonatologist.
- Any infant not passing two newborn hearing screenings
- Parental concern
- An infection around the brain and spinal cord caused by bacteria (bacterial meningitis)
- Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome
- Chemotherapy

Families of babies who pass the screening, but are identified with one of the above risk factors, should:

1. Be informed about the risk factors identified
2. Be referred to their PCP on the discharge summary for follow-up testing
3. Be made aware of the hearing and language developmental milestones on the screening certificate that is given to the family and told to monitor the child's progress
4. Be informed of the results and the importance of follow-up testing



## Indiana's Early Hearing Detection and Intervention Program **Newborn At-Risk Criteria for Delayed Onset/Progressive Hearing Loss Recommended Follow-up**

### **Family History of Congenital Childhood Hearing Loss**

- Family members born with hearing loss in one or both ears
- A family member with a hearing loss that was identified in childhood
- Hearing loss not caused by a medical condition like ear infections
- Does not include family members with known causes of hearing loss like rubella, meningitis, loud noise exposure &/or trauma

### **In-utero Infection (TORCH) for this pregnancy:**

- **Toxoplasmosis** – infected during or just before pregnancy, especially 1<sup>st</sup> trimester
- **Group Beta Strep (GBS)** – sick infant with positive GBS culture
- **Syphilis** – infected during pregnancy, baby can be treated prior to delivery
- **Rubella** – infected primarily during the first trimester
- **Cytomegalovirus (CMV)** – can be transmitted through the placenta, birth canal or postnatally through breast milk
- **Herpes Simplex Virus (HSV)** –
  - Yes if: Baby is diagnosed with neonatal herpes
  - Active infection during vaginal delivery
  - Active infection during cesarean delivery with a premature membrane
  - No if: Mother had a cesarean delivery with no membrane rupture
  - No active infection was present at birth

### **Hyperbilirubinemia (Jaundice)**

- At levels exceeding indication for exchange transfusion

### **Ear Malformations/Cranio-facial Anomalies**

- Babies who cannot be screened at the hospital due to no ear, partial ear or no ear canal opening should be immediately referred to an audiologist (Level1) and to their physician (PCP)
- Babies with craniofacial anomalies who can be screened and pass should be referred for follow-up at 9-12 months of age.

**Other at-risk factors for hearing loss** in infants exist and would routinely be investigated by the infant's primary care physician. These factors include:

- Syndromes that are commonly associated with hearing loss (Down, Usher, Waardenburg and Neurofibromatosis Type 2)
- Neonatal intensive care stay with any of the following regardless of length of stay: ECMO, assisted ventilation, exposure to oto-toxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/lasix) (If uncertain about the risk, please check with the neonatologist)
- Any infant not passing two newborn hearing screenings
- Parental concern



## Indiana's Early Hearing Detection and Intervention Program **Minimizing Referral Rates**

Test while the baby is quiet, relaxed (preferably sleeping), well fed and comfortable. Swaddling the infant often helps.

If a second screen is necessary, wait a few hours. This can significantly reduce the referral rates. Always re-screen both ears.

Screening will be faster and more effective if you minimize noise and distraction before screening. Testing area should be quiet (avoid talking, ringing phones, running water, etc.).

It is important to have a backup equipment plan in the event of a breakdown.  
\*See Tips for Screeners / Back-up Equipment for suggestions on a plan.

### **OAE specific tips:**

For OAE screening, the single most important factor in reducing referral rates is achieving a good probe fit:

- Visually inspect the ear canal for debris (wax, blood, vernix);
- Seat the earphone probe by gently pulling the ear up and out: this will open up the canal;
- Begin the test once the probe is placed and baby has quieted;
- If the baby does not pass on the first try,
  - Remove the probe and check for debris
  - Replace the tip if needed
  - Clean probe if needed
  - Reposition the probe and repeat the screen



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**Back-up Equipment**

**If the equipment malfunctions, have a plan in place for a back-up unit. Babies who miss the screening due to equipment problems must be brought back for screening when the equipment is repaired. Being prepared for equipment problems will decrease the delays in screening all of your newborns.**

Possible suggestions include:

1. Many hospitals have "sister" facilities that may be able to loan equipment to each other.
2. Some manufacturers offer loaner equipment—arrangements can be made for equipment to be sent immediately for loan until repairs are completed. Check with your manufacturer's sales representative regarding this possibility.
3. Check with local audiologists or ENT practices to see if they have equipment that they could loan or contract the service for a fee.



## Indiana's Early Hearing Detection and Intervention Program **Sharing Hearing Screening Results with Parents**

### Why do we screen babies?

Parents need to know that the UNHS screening is designed to find babies who may have a hearing loss, and therefore, need further testing. It is a screening only, not a definitive test.

### Communication of Results - Your words matter

- The importance of communicating the results to both families and physicians is often underestimated.
- Results need to be conveyed in a manner that is respectful, thoughtful, sensitive and consistent.
- The person conveying results should be able to discuss the screening and answer questions that parents may have or know who to refer them to if they don't know the answers to the family's questions.
- Screeners should provide information on normal hearing development, risk factors and screening results. This information is available on the hearing screening certificate provided by ISDH.

### ***Give results both verbally and in written form.***

***Babies who pass:*** Use the Hearing Screening Results (on back of Who, What Why brochure) to discuss the results. Provide the hearing/speech/language milestones that parents may use to monitor progress.

***Babies who pass, but have risk factors:*** Provide the Hearing Screening Results and Hearing/Speech milestones. Note the risk factors and the hearing/speech/language milestones. Tell parents their baby needs to be seen for a hearing evaluation by an audiologist at 9 to 12 months of age.

***Babies who do not pass:*** Common reasons babies do not pass include an ear canal blocked with debris, the presence of fluid in the middle ear, or permanent hearing loss (approximately 1 per 300 births). Do not indicate that "lot's of babies do not pass". In fact, less than 2% of all Indiana babies do not pass UNHS.

Provide the brochure "What if Your Baby Needs More Hearing Tests". Choose your words carefully. Inappropriate communication of screening results may cause stress and anxiety for families. Do not use the word "fail" or "deaf". If the baby doesn't pass, use the word "refer" or "did not pass". Be careful to not minimize the screening while at the same time not unduly worrying the family.

Further audiological testing will confirm if the baby has normal hearing or a hearing loss. If a hearing loss is found, testing will determine the degree and type of loss and the next steps in follow-up.

You can also have the family contact the Indiana State Department of Health's (ISDH) Early Hearing Detection and Intervention Program (EHDI). Parents may contact Gayla Hutsell Guignard, Indiana State EHDI Coordinator (317) 234-3358 or the EHDI Parent Consultant (317) 233-1269 or call toll free (888) 815-0006.



## Indiana's Early Hearing Detection and Intervention Program **Tips for Blood Spot Card Completion**

1. **Every effort should be made to have the UNHS results as well as the blood spot sample entered on the blood spot card before it is sent to the lab** (A complete set of data will be entered into the system for each infant).
2. To facilitate data entry, the UNHS screening should be completed and entered on to the blood spot card immediately upon completion. Complete the information requested:
  - a. Check the appropriate box for initial (one screen and passes both ears) or re-screen (if the baby requires a second test for one or both ears);
  - b. Enter the date of reported screen;
  - c. Indicate whether pass or refer for each ear;
  - d. If risk factors are present please indicate the risk factor (Risk factors include: family history, congenital infection, hyperbilirubinemia requiring exchange transfusion, and craniofacial anomalies);
  - e. If not screened, check the appropriate box; (Deceased, Transferred, Hospital error, NICU, Unauthorized Refusal, Religious Refusal, Equipment Problems, and Other).
3. **Do not delay in sending the blood spot card** if hearing screening is not yet completed. All newborn screen blood samples should be sent within 24 hours of collection, even if the UNHS screen has not been done. A delay in sending the blood spot info could result in a delay in diagnosis.
4. If hearing screening has not been completed (due to transfer, NICU, or other reason) retain the pink "hearing" pull out sheet and keep until hearing screen is completed. When the hearing screening has been completed, enter the information on the copy and forward to the IU lab for data entry. Newborn Screening Laboratory, P. O. Box 770, Indianapolis, IN 46206.
5. Report results to ISDH through EARS.



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## **TIPS FOR COMPLETING THE UNHS/EHDI Hearing Screening Results**

Complete the child's name, birth date, date of screen and facility on the certificate.

1. Circle the correct screening result for that child.
2. If passed but has a risk factor, check the appropriate risk factor.
3. If did not pass. Put an\* next to the "did not pass" box and write the referral information (i.e. audiology appointment date, time and place) at the bottom of the page.



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**Risk Factors for Long Stay Babies in Neonatal Intensive  
Care Unit (NICU)**

- Very Low Birth Weight (<1500 grams)
- Ototoxic Drugs
  - Aminoglycosides can damage hair cells in the cochlea resulting in sensorineural hearing loss. Some babies with mitochondrial DNA mutations may be more susceptible to these effects. Common ones include: Streptomycin, Neomycin, Kanamycin, Amikacin, Viomycin, Vancomycin, Gentamycin, Tobramycin
  - Salicylates in large doses may cause reversible hearing loss and tinnitus
  - Quinine can cause hearing loss. If taken by the mother during pregnancy it may result in inner ear abnormalities in the fetus
- Low Apgar Scores (0-4@ 1 minute or 0-6 @ 5 minutes)
- Postnatal infections associated with hearing loss (Sepsis, Meningitis)
- Hyperbilirubinemia requiring Exchange Transfusion can cause damage to the nervous system if severe
- Mechanical Ventilation may cause loss of hearing due to oxygen deprivation or cause other problems associated with hearing loss ( bronchiopulmonary dysplasia, persistent pulmonary hypertension)
- Extracorporeal Membrane Oxygenation (ECMO) may cause hearing loss due to oxygen deprivation and the loss may be progressive



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### Gentamicin as a Risk Factor:

1. All babies should be screened as mandated by Indiana law.
2. Ototoxic Drugs
  - Aminoglycosides can damage hair cells in the cochlea resulting in sensorineural hearing loss. Some babies with mitochondrial DNA mutations may be more susceptible to these effects. Common ones include: Streptomycin, Neomycin, Kanamycin, Amikacin, Viomycin, Vancomycin, Gentamycin, Tobramycin
  - Salicylates in large doses may cause reversible hearing loss and tinnitus
  - Quinine can cause hearing loss. If taken by the mother during pregnancy it may result in inner ear abnormalities in the fetus
3. If other factors exist (i.e. exceeded trough lines, long duration, or in combination with loop diuretics) the physician may order a repeat screen prior to discharge. Literature suggests that these drugs are slow to clear the body. If a screen or re-screen is ordered following administration of these drugs, it is "best practice" to wait 24 hours after the medication has been stopped. Birthing facilities need to determine their own protocols.
4. It is "best practice" to make sure parents are aware of developmental milestones and monitor their baby closely. If concerns develop, they should seek a referral for an audiologic diagnostic evaluation from the physician.