RISK FACTORS FOR LATE-ONSET AND PROGRESSIVE HEARING LOSS

2007 Joint Committee on Infant Hearing Position Statement

1. Caregiver concern* regarding hearing, speech, language, or developmental delay
2. Family history* of permanent childhood hearing loss
3. NICU stay of more than 5 days, or any of the following regardless of length of stay: ECMO*, assisted ventilation, ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia requiring transfusion.
4. In-utero infections, such as CMV*, herpes, rubella, syphilis, and toxoplasmosis.
5. Postnatal infections associated with hearing loss, including confirmed bacterial and viral meningitis.
6. Craniofacial anomalies of pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
7. Findings of syndrome associated with hearing loss (Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson)
8. Syndromes associated with progressive or delayed-onset hearing loss* (neurofibromatosis, osteopetrosis, Usher Syndrome)
9. Neurodegenerative disorders* (such as Hunter Syndrome) or sensory motor neuropathies (such as Friedreich ataxia & Charcot-Marie-Tooth syndrome).
10. Head trauma, especially basal skull/temporal bone fracture that requires hospitalization.
11. Chemotherapy*  

*Greater concern for delayed-onset hearing loss

www.jcih.org

MEDICAL HOME INSTRUCTIONS FOR INFANTS WITH RISK FACTORS

- The medical home is responsible for monitoring risk factors for late onset and progressive hearing loss.
- Infants who pass the neonatal screening but have a risk factor should have at least 1 diagnostic audiology assessment by 24 to 30 months of age.
- Audiologist should be able to perform:
  - ABR - Frequency-specific tone bursts
  - Air & bone conduction
  - OAE
  - Middle ear assessment
  - Evaluation with sedation (if needed)

AVERAGE DEVELOPMENTAL MILESTONES

<table>
<thead>
<tr>
<th>Age</th>
<th>Milestones</th>
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<tbody>
<tr>
<td>0-3 Months</td>
<td>Quiets when hearing familiar voice, Sthalts to loud sounds, Makes vowel sounds like ahh, ohh</td>
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<tr>
<td>3-6 Months</td>
<td>Looks for sounds with eyes, Uses sounds such as squeals, whimpers, chuckles, Vocalizes excitement and displeasure</td>
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<tr>
<td>6-9 Months</td>
<td>Turns hearing toward sounds, Babbles sounds like ba-ba, ma-ma, da-da, Looks for quiet sounds made out of sight</td>
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<tr>
<td>9-12 Months</td>
<td>Imitates speech sounds made by others, Understands no-no or bye-bye, Correctly uses ma-ma or da-da, Turns to own name</td>
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<tr>
<td>12-24 Months</td>
<td>Conversation babbling to self and others, Follows simple directions &quot;get your shoes&quot;, Knows and uses more than six (6) words, Uses 2-3 word sentences</td>
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This is not a screening tool and does not replace objective testing. Inform parents on hearing, speech, and language milestones.

Funding for the Infant Hearing Guide for Providers provided by Maternal and Child Health Bureau grant

ARIZONA RESOURCES:

- Ear Foundation of Arizona  
  www.earfoundationaz.org
- AZ Hands & Voices Family Support - Guide By Your Side  
  www.azhv.org
- Arizona Chapter of American Academy of Pediatrics  
  www.azaap.net

Arizona EHDI  
Early Hearing Detection and Intervention Program  
Arizona Department of Health Services  
Office of Newborn Screening  
www.arnewborn.com  
(602) 364-1409 • Toll Free: (800) 548-8381  
711 for Relay Service  
Fax: (602) 364-1495

Arizona Department of Health Services  
Office of Newborn Screening  
www.arnewborn.com

NBS-007 (REV. 2/12)
**MEDICAL HOME INSTRUCTIONS**

- At 1st well-baby visit, check for hearing screening results on back of immunization card or hospital discharge summary
- Ensure all screening, audiology evaluations, and/or ENT evaluations are maintained in medical record
- Infants who have a missed or incomplete screening should be referred for further testing
- Infants who fail in only one ear should be referred for further testing of both ears

**MEDICAL HOME REFERRALS**

Every child diagnosed with a confirmed hearing loss should receive timely referral to:

**REQUIRED:**
- Audiology
- Otology/ENT
- Early Intervention (EI)

**RECOMMENDED:**
- Ophthalmologist
- Genetics
- Other medical specialty

**LEGAL INSTRUCTIONS**

- Newborn Objective by standard testing method
- 2, 4, 6, 9, 12, 15, 18, 24 (Age in months) Subjective by history
- 3, 4, 5 (Age in Years) Objective by standard testing method

**MANDATORY REPORTING - (UP TO AGE 3)**

A.R.S 36-694, A.A.C. R9-13-207(E)

Medical Home needs to complete referrals

Medical home needs to monitor for risk factors for late set or progressive hearing loss [www.jcih.org](http://www.jcih.org)

**LEGEND**

- Red: Medical Home needs to complete referrals
- Blue: Medical home needs to monitor for risk factors for late set or progressive hearing loss

**SCREENING**

Hearing screening results must be reported to ADHS

**DIAGNOSTIC**

Diagnostic hearing evaluations, including normal results must be reported

Submit to ADHS within one week following test / Fax to ADHS: (602) 364-1495

For requirements & forms, visit www.aznewborn.com

**HEARING PERIODICITY SCHEDULE**

- Newborn Objective by standard testing method
- 2, 4, 6, 9, 12, 15, 18, 24 (Age in months) Subjective by history
- 3, 4, 5 (Age in Years) Objective by standard testing method

**AHCCCS EPSDT Periodicity Schedule** [www.azahcccs.gov](http://www.azahcccs.gov)