Utah EHDI NICU and High-Risk Infant Screening, Diagnostic & Follow-Up Recommended Protocol

RATIONALE: The Utah Early Hearing Detection and Intervention (EHDI) Program adheres to the national EHDI ’1-3-6’ goals:

- all newborns receive a hearing screening before 1 month of age
- for infants that do not pass, complete a diagnostic hearing evaluation before 3 months of age
- for infants who are diagnosed as deaf or hard of hearing (DHH), enroll into early intervention services before 6 months of age

Infants who are deaf or hard of hearing (DHH) who do not meet each of the 1-3-6 milestones are at high-risk for significant language delays when compared to their same-aged peers (Yoshinaga, 2017). Additionally, infants in the Newborn Intensive Care Unit (NICU) are 10 times more likely to have Permanent Childhood Hearing Loss (PCHL) than those in the well-baby nursery. These factors support the need for early and comprehensive hearing evaluations for infants in NICUs and other infants identified as being at high-risk for hearing loss.

While Utah EHDI acknowledges the barriers to meeting the ½-1-3-6 milestones, every effort should be made to meet these milestones when it is medically appropriate and complete a baseline comprehensive diagnostic hearing evaluation, ideally before discharge, after a failed NBHS with any of the risk factors listed in the protocol below and/or Appendix 1. The Joint Committee on Infant Hearing (JCIH) Risk Indicators are listed in Appendix 2.

Once any baby, regardless of nursery level, is discharged with the requirement of follow-up, they are at risk for becoming lost-to-follow-up (LTFU). Tran, Ng, and Webb et al. (2016) found that 56% of babies who had to wait more than 30 days to follow-up never came back. They also found 40% of babies who had two or more outpatient appointments never went back to complete the process.

A baseline diagnostic hearing evaluation is essential to monitoring infants and children at risk for late-onset / progressive hearing loss. Providing an inpatient hearing evaluation will reduce the risk of a medically complex infant becoming LTFU. NICU families are often required to prioritize their child’s care – this can result in delayed diagnosis due to more critical (i.e., cardiology, GI/feeding, etc.) sub-specialty follow-up appointments required after discharge. A

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Utah EHDI “1/2” in ½-3-6, indicates the screening process should be completed before 14 days of age in order for eligible (two failed hearing screenings) babies to have time to complete CMV testing before 21 days of age. If the baby fails one hearing screening after 14 days of age, the baby is eligible for CMV testing (Utah Code 26-10-10).

NOTE: Utah EHDI recognizes a new JCIH Position Statement will be published soon. This protocol will be reviewed and updated to reflect the new JCIH recommendations when they are published.
delayed evaluation can put the infant at risk for requiring sedation, further delaying the
diagnosis. An example of LTFU research for medically complex infants was completed at
Cincinnati Children’s Hospital, showing the importance of completing an inpatient diagnostic
evaluation to reduce LTFU. This can be seen in Appendix 3.

SCOPE: All Utah hospitals with a NICU or special care nursery as well as pediatric
audiologists who evaluate infants and children at high-risk for PCHL.

CONTENT REVIEWERS and CONTRIBUTORS: This protocol was developed by
the Utah Department of Health Early Hearing Detection and Intervention (EHDI) Program using
evidence-based research. Utah EHDI believes it is essential in all protocol/guideline
development for review by a multi-disciplinary team with expertise in meeting the needs of
infants / children and their families. The following individuals/groups have reviewed this
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This protocol has been reviewed and unanimously approved by the Utah Newborn
Hearing Screening Advisory Committee on August 14th, 2018.
NICU Screener Qualifications:

Properly trained screeners are essential to the success of all NBHS programs. Due to the complex medical histories of infants born in the NICU, it is in the best interest of infants/families to have an audiologist complete the screenings of medically complex infants; however, hospitals may utilize screeners. If a NICU program utilizes non-audiologist screening staff, only automated screening equipment is allowed. For hospital programs utilizing diagnostic ABR technology, screening ABR should consist of a 60/30 dBnHL click stimuli and interpretation should be done by an audiologist who adheres to strict operator and interpretation protocols (ASHA Expert Panel on NBHS; ASHA Audiology Assistants; AAA Considerations). Adhering to this best-practice standard allows the audiologist to move into a comprehensive diagnostic evaluation when NICU and high-risk infants do not pass their screening, leading to earlier identification. The presence of middle ear effusion is not a contraindication to testing, and evoked potentials should not be delayed until effusion has cleared (AAA, 2012).

- Qualifications
  - Communication and interpersonal skills necessary for the tasks assigned
  - A basic understanding of the needs of the population being served
  - Successful completion of the training requirements and competency-based skills necessary for the performance of specific, assigned tasks

- Scope of practice (non-audiologist)
  - Performing equipment maintenance and biological checks on screening equipment
  - Troubleshooting of screening equipment
  - Performing newborn hearing screening (automated equipment only)
    - Otocoustic emissions
    - Automated ABR
    - **NOTE:** Automated equipment means the test results will only provide a pass/refer result. No test interpretation is necessary.
  - Assisting audiologists with setup and technical tasks
  - Maintaining and restocking test and treatment rooms
  - Assisting audiologists in hearing testing of pediatric patients
  - Performing infection control duties
  - Assisting patients in completing case history or other relevant forms approved by supervising audiologist
  - Sending written clinical reports and documents approved by the supervising audiologists to other approved providers (Primary Care Physicians, EHDG, Early Intervention, ENT, etc.)
  - Provide screening results (pass or refer only) verbally and written to families prior to discharge
  - Scheduling screening and/or diagnostic follow-up appointments

- Screeners (non-audiologists) are **not to perform** the following:
  - Determining case selection or evaluation protocols
  - Screening cannot include tympanometry or any conventional ABR testing, including bone-conduction
o Interpreting observations or data into diagnostic statements of clinical management strategies or procedures

o Transmitting or disclosing clinical information beyond the screening result “pass” or “refer” from automated screening equipment, to anyone such as the family, medical provider, or others, without the approval of the supervising audiologist

o Counseling or consulting with the patient, family, or others regarding the patient status or service or make referrals for additional services
  ▪ Screeners may not indicate the cause of any abnormal hearing screening result (i.e., ‘it’s probably just fluid in the ear’). This requires diagnostic testing to confirm the presence of fluid and often results in poor follow-up.
Utah EHDI Screening, Assessment and Follow-up Protocol

For infants in the NICU, the screening and/or diagnostic protocols will vary based on the child’s risk factor(s). The audiologist should use an individualized approach in determining each stage of the EHDI process. Some infants may only need a screening with follow-up “as concerns arise”, while others may require an inpatient diagnostic evaluation with more frequent follow-up. The following table provides a recommended protocol:

A comprehensive diagnostic hearing evaluation should include:

1. Auditory Brainstem Response
   a. Click
   b. Frequency-specific (500, 1k, 2k, 4k) - complete as much as possible
   c. Bone-conduction, as indicated
2. DPOAEs and/or TEOAEs (to assess outer hair cell function)
3. Tympanometry (to determine middle ear function), as indicated
   a. 1000 Hz (< 6 months adjusted age)
   b. 226 Hz (> months adjusted age)
4. Acoustic reflexes (test for neural function), as indicated

NOTE: Utah Administrative Code (UAC 26-10-6; Rule 398-2) requires all diagnostic reports to be sent to Utah EHDI within 7 days of the completed evaluation.

If a diagnostic hearing evaluation is ABNORMAL, but cannot be confirmed as Permanent Childhood Hearing Loss (PCHL), follow-up with 1) appropriate audiological follow-up recommendations and 2) provide referral for Early Intervention (EI) services.

If a child has CONFIRMED hearing loss, refer to 1) Early Intervention; 2) ENT; 3) Ophthalmology; 4) Parent Consultants; 5) Continued audiological care; 6) Genetics; 7) Other providers, as needed.

**NS = normal hearing screening; IP = Inpatient; OP = Outpatient; AC = air conduction; BC = bone-conduction; AABR = automated or screening ABR completed by audiologists using conventional ABR technology as defined above**

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Screening Procedure</th>
<th>Hearing Evaluation</th>
<th>Follow-Up Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital Cytomegalovirus</td>
<td>None / complete diagnostic at IP</td>
<td>IP Diagnostic Eval</td>
<td>If NS, F/U every 3 months until age 3; every 6 months until age 6</td>
</tr>
<tr>
<td>Zika Virus</td>
<td>AABR</td>
<td>IP Diagnostic Eval</td>
<td>If NS, F/U @ 9 months of age for behavioral testing</td>
</tr>
<tr>
<td>ECMO</td>
<td>None / complete diagnostic at IP</td>
<td>IP Diagnostic Eval</td>
<td>F/U at 3 months of age (diagnostic ABR); then behavioral testing every 6</td>
</tr>
<tr>
<td>Condition</td>
<td>Confirmed:</td>
<td>Suspected:</td>
<td>Follow-up:</td>
</tr>
<tr>
<td>-------------------------------</td>
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<td>-----------------------------------------------</td>
</tr>
<tr>
<td><strong>Meningitis (bacterial and viral)</strong></td>
<td>None / complete diagnostic at IP</td>
<td>AABR + OAE</td>
<td>If NS, F/U at 6 months for behavioral testing</td>
</tr>
<tr>
<td><strong>Craniofacial anomalies:</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Microtia / Cleft palate</td>
<td>AABR + OAE</td>
<td>If indicated, complete diagnostic evaluation</td>
<td>If NS, F/U at 9 months for behavioral testing</td>
</tr>
<tr>
<td>Ear tags, pits</td>
<td>AABR or OAE</td>
<td>OP diagnostic, if indicated</td>
<td>Retest if parental / PCP concerns arise</td>
</tr>
<tr>
<td>Atresia well-baby</td>
<td>Screen non-affected ear (schedule OP diagnostic test)</td>
<td>Dx AC test for non-affected ear and BC for atretic ear</td>
<td>Make “Confirmed Hearing Loss” referrals</td>
</tr>
<tr>
<td>Atresia NICU</td>
<td>None / Complete IP diagnostic</td>
<td></td>
<td>If no medical or amplification management recommended, follow-up with behavioral testing at 9 months</td>
</tr>
<tr>
<td><strong>Ototoxicity:</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aminoglycosides &gt;5 days</td>
<td>AABR + OAE</td>
<td></td>
<td>If NS, F/U at 9 months for behavioral (OAEs at a minimum)</td>
</tr>
<tr>
<td>** Syndromes:**</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Syndromes assoc w/HL or Progressive / late-onset HL (Appendix 1)</td>
<td>None / IP diagnostic</td>
<td>IP Diagnostic Eval</td>
<td>If normal diagnostic, F/U @ the discretion of the audiologist or no later than 9 months for behavioral testing</td>
</tr>
<tr>
<td>Trisomy 21</td>
<td>AABR + OAE, at minimum</td>
<td>IP Diagnostic when indicated</td>
<td>If normal, F/U @ 6 months (screen) and 12 months for behavioral testing. F/U annually, if normal hearing established</td>
</tr>
</tbody>
</table>
### Neurodegenerative disorders

<table>
<thead>
<tr>
<th>Neurodegenerative disorders</th>
<th>None / IP Diagnostic</th>
<th>IP Diagnostic Eval</th>
<th>F/U at audiologist’s discretion</th>
</tr>
</thead>
</table>

### Neurologic insults:

<table>
<thead>
<tr>
<th>Hydrocephalus / Shunts</th>
<th>AABR + OAE</th>
<th>If NS, F/U at 9 months for behavioral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gestational Age / Very Low Birthweight:</td>
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</tr>
<tr>
<td>&lt;32 weeks &lt;1500 grams</td>
<td>AABR + OAE</td>
<td>If NS, F/U rescreen at 6 months (OAE at minimum)</td>
</tr>
<tr>
<td>Hypoxic Ischemic Encephalopathy (HIE) / cooling</td>
<td>AABR + OAE</td>
<td>If NS, F/U rescreen at 6 months</td>
</tr>
</tbody>
</table>

### Other Risk Factors:

<table>
<thead>
<tr>
<th>Caregiver Concern</th>
<th>AABR or OAE (at discretion of Aud)</th>
<th>At discretion of Aud</th>
<th>If NS, repeat as concerns arise</th>
</tr>
</thead>
<tbody>
<tr>
<td>Family History</td>
<td>AABR + OAE</td>
<td>At discretion of Aud</td>
<td>If NS, F/U at 9 months for behavioral</td>
</tr>
<tr>
<td>NICU admission of &gt;5 days, no risk factors</td>
<td>AABR + OAE</td>
<td>If NS, F/U at 9 months for behavioral</td>
<td></td>
</tr>
<tr>
<td>NICU admission &lt;5 days, no risk factors</td>
<td>AABR or OAEs</td>
<td>F/U as concerns arise</td>
<td></td>
</tr>
</tbody>
</table>

### Appendix 1: Common syndromes associated with hearing loss

- 22q11.2 deletion syndrome (DiGeorge / Velo-Cardio-Facial syndrome)
- Achondroplasia
- Alport
- Apert syndrome
- Branchio-oto-renal
- Charcot-Marie-Tooth disease
- CHARGE
- Cornelia de Lange syndrome
- Crouzon syndrome
- Friedreich Ataxia
- Goldenhar
- Hunter syndrome (MPS II)
- Hurler syndrome (MPS I)
- Jervell-Lange-Nielsen
- Marshall syndrome
- Neurofibromatosis
- Noonan syndrome
- Osteogenesis Imperfecta
- Pendred
- Septo-optic dysplasia (de Morsier syndrome)
- Stickler
- Treacher Collins
- Trisomy 13 (Patau syndrome)
- Trisomy 18 (Edwards syndrome)
- Turner's
- Usher
- Waardenburg

**Appendix 2: JCIH Risk Factors**

Risk indicators that are marked with a “§” are of greater concern for delayed-onset hearing loss

1. Caregiver concern§ regarding hearing, speech, language, or developmental delay

2. Family history§ of permanent childhood hearing loss

3. All infants with or without risk factors requiring neonatal intensive care for greater than 5 days, including any of the following: ECMO,§ assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix). In addition, regardless of length of stay: hyperbilirubinemia requiring exchange transfusion (Clarification of JCIH 2007)

4. In utero infections, such as CMV,§ herpes, rubella, syphilis, and toxoplasmosis

5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies

6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss

7. 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-Nielsen

8. Neurodegenerative disorders,§ such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome

9. Culture-positive postnatal infections associated with sensorineural hearing loss,§ including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis
10. Head trauma, especially basal skull/temporal bone fracture§ that requires hospitalization

11. Chemotherapy§

Appendix 3: LTFU Data

Cincinnati Children’s Hospital (CCH): The Effects of Inconclusive ABR Results on LTFU (2011)

- ABRs at 6 outpatient centers (n=764)
  - 9% didn’t achieve adequate sleep state (n=71)
    - Of those, 63% were LTFU
  - 17% had suspected fluid with incomplete ABR due to incomplete bone-conduction
    - Of those, 45% were LTFU
  - For infants needing more than one ABR, the average age of identification for permanent hearing loss was 4.3 months

CCH chart review (2013)

- 12% of infants had incomplete ABR due to poor sleep state

CCH data when infants diagnosed at the inpatient level

- Missed screening went from 30% to 0%
- Lost to documentation rates decreased from 42% to 12% (0% for children born in Ohio)
- Average age of hearing aid fitting decreased from 7 months adjusted age to 4 months adjusted age
- 95% of DHH infants had a sensory care plan while in the hospital
REFERENCES & RESOURCES


Tran, T., Ng, I. & Webb, J. et al. (2016). Late Newborn Hearing Screening, Late Follow-Up, and Multiple Follow-Ups Increase the Risk of Incomplete Audiologic Diagnosis Evaluation. The Journal of Early Hearing Detection and Intervention, 1(2), 49-55.


Utah EHDI Mandates:

Newborn Hearing Screening
Utah Code (26-10-6): Testing of Newborn Infants
https://le.utah.gov/xcode/Title26/Chapter10/26-10-S6.html
Utah Rule (R398-2): Newborn Hearing Screening

Congenital Cytomegalovirus
Utah Code (26-10-10): Cytomegalovirus Public Education and Testing
https://le.utah.gov/xcode/Title26/Chapter10/26-10-S10.html?v=C26-10-S10_1800010118000101
Utah Rule (R398-4): Cytomegalovirus Public Health Initiative