UTAH DEPARTMENT OF HEALTH
Early Hearing Detection and Intervention

WELL-BABY: RECOMMENDED NEWBORN HEARING SCREENING AND PEDIATRIC AUDIOLOGICAL ASSESSMENT PROTOCOL

1) **SCREENING BEFORE DISCHARGE** – Identification of newborns with possible hearing problems. If an infant is **born out of the hospital,** screen **before 10 days of age.**

   a) If an infant does not pass the inpatient screening, the PCP must be notified. The outpatient screening appointment should be scheduled before discharge.

   b) **Before 14 days of age**, complete outpatient screening.

      i) If the infant does not pass their 2nd hearing screening, **the PCP must be notified of the need for CMV testing.** Provide family with *Cytomegalovirus & Auditory Brainstem Response Testing Order* from the Utah Department of Health. Assist family with getting to hospital lab for CMV PCR testing.

      2nd outpatient screening is NOT recommended. It increases risk of lost-to-follow-up and delay of CMV testing past 21 days of age. See CMV Protocol

   c) Identification of newborns at risk for hearing loss (screening) falls under the ***responsibility of the birthing facility or attending midwife***. Tracking of screening remains the responsibility of the hospital to which the baby is transferred.

   d) **Readmissions**: for readmissions in the first month of life when there are conditions associated with potential hearing loss (e.g., hyperbilirubinemia that requires exchange transfusion or culture-positive sepsis), an AABR screening should be performed before discharge (JCIH, 2007).

   e) **Ototoxic medications:** Any infant who has received less than six days of Aminoglycosides only requires a newborn hearing screening, unless other risk indicators for hearing loss coincide with the ototoxic medications.

2) **DIAGNOSTIC BEFORE 3 MONTHS OF AGE** – A comprehensive audiological evaluation of infants who did not pass the hearing screening process or have other risk indicators for hearing loss.

   a) The audiological diagnostic evaluation of infants identified as at-risk for hearing loss falls under the **responsibility of audiologists** in the community who have the knowledge, capability and expertise to apply best practice infant/pediatric diagnostic procedures. Designated hospital/midwife program audiologist should see that appropriate follow-up is completed before 3 months of age, and ideally as soon as possible after the referred 2nd screening.

3) **EARLY INTERVENTION BEFORE 6 MONTHS OF AGE** – For infants and children with confirmed hearing loss, fitting of **amplification should be initiated within 1 month** of diagnosis and **referral to early intervention services be made within 2 days but no more than 7 days post diagnosis (**JCIH, 2007). Referral for early intervention services falls under the responsibility of the diagnosing audiologist.

Audiological Diagnostic Protocol

****NOTE: This document was created for the "Well-baby" population. For infants in the NICU and/or other significant risk factors for hearing loss, please refer to the *Utah EHDI NICU and High-Risk Infant Screening, Diagnostic & Follow-Up Recommended Protocol*****

1) Obtain a **detailed family, birth and medical history**. Address any parental concerns.
   a) Review JCIH (2007) indicators for infants at risk for progressive or delayed onset hearing loss. Infants who pass the newborn hearing screening, but have a risk factor should be monitored. Early and more frequent audiological monitoring is indicated for risk factors that have a higher degree of risk for hearing loss (see Appendix A). Refer to the *Utah EHDI NICU and High-Risk Infant Screening, Diagnostic & Follow-Up Recommended Protocol* for specific recommendations.

   b) Review JCIH (2000) indicators for infants at risk for auditory neuropathy (see Appendix B). They should be evaluated using OAEs, acoustic reflexes and auditory brainstem response (ABR).

2) Perform otoscopic evaluation.

3) Obtain acoustic immittance measures using age-appropriate frequencies. Beware of limitations for infants less than 6 months of age (see frequencies below).
   a) Regardless of middle-ear status, **complete full audiological assessment to establish cochlear function**. See JCIH (2000) recommendations for infants with conductive hearing loss (Appendix C).
   b) 1000 Hz probe tone < 6 months (adjusted age)
   c) 226 Hz probe tone > 6 months (adjusted age)
   d) **Ipsilateral acoustic reflexes** to rule out auditory neuropathy spectrum disorder (ANSD)

4) Perform **distortion product (DPOAE) or transient evoked (TEOAE)** otoacoustic emissions.

5) Conduct an **auditory brainstem response** (ABR) evaluation:
   a) **A diagnostic evaluation is not complete without frequency-specific testing.** It is possible to miss hearing loss even with "normal" click thresholds.

b) Obtain a suprathreshold response to a click stimulus, using insert earphones. Complete rarefaction and condensation polarity click stimuli (visualize cochlear microphonic to check for presence of polarity reversal) to rule-out ANSD.
c) Obtain **frequency-specific stimuli Wave V responses** at 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz at **20 dB eHL** or better (obtain 4000 Hz and 1000 Hz at the minimum and then 500 Hz and 2000 Hz, if time permits).
d) For infants with **indication of hearing loss, obtain bone-conduction ABR** thresholds to click and toneburst stimuli, **even if middle ear involvement is suspected or comorbidities are present (**e.g. Cleft lip/palate, Down Syndrome, etc).
e) Perform ear-specific visual reinforcement audiometry (VRA) for children whose developmental age is > 6 months of age and older.

6) Discuss results and follow-up recommendations with parents.

7) Prepare a written report interpreting test results and describing the diagnostic profile.

8) If tympanograms are flat, refer to primary care provider, Medical Home or ENT specialist for medical intervention.

9) If hearing loss is confirmed, see ***Referral Protocol*** below***.***

UTAH DEPARTMENT OF HEALTH
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Permanent Hearing Loss:
Diagnostic and Early Intervention Referral Protocol

The following should be completed **for infants and children with confirmed hearing loss**:

1) Review results of the diagnostic audiologic assessment, implications of the audiologic diagnosis, and recommendations for early intervention with the parents including:
   a) Amplification and communication options
   b) Information regarding the need for medical follow-up and continuing audiological follow-up
      i) Referral to Otolaryngology (ENT) for medical clearance
      ii) Contact the Primary Care Physician
      iii) Referral to genetics, ophthalmology, developmental pediatrician, neurology, cardiology, and nephrology, as appropriate
      iv) **In order to minimize confusion, provide a hard-copy of recommendations / "next steps" for parents/caregivers prior to their leaving the appointment.**
   c) Information regarding the importance of timely enrollment in an intervention/habilitation program. **Referral should immediately be made to the Utah Schools for the Deaf and the Blind – Parent Infant Program (USDB-PIP).**

      JCIH 2007 Position Statement on early intervention enrollment:

      "…once any degree of hearing loss is diagnosed in a child, a referral should be initiated to an early intervention program ***within 2 days of confirmation of hearing loss*** *(CFR 303.321d). The initiation of early intervention services should begin as soon as possible after diagnosis of hearing loss but at* ***no later than 6 months of age****. Even when the hearing status is not determined to be the primary disability, the family and child should have access to intervention with a provider who is knowledgeable about hearing loss".*

   d) Information regarding the **Children's Hearing Aid Program (CHAP)**
      i) Website: **health.utah.gov/chap**
   e) Importance of **parent-to-parent support** (e.g., Utah EHDI Parent Consultant, Utah Parent Center, etc.)
      i) Website: **health.utah.gov/ehdi**
   f) **For infants and children < 3 years of age, see "*Infant Diagnostic & Early Intervention Referral Process*" on Page 8, for specific referral information.**

2) Distribute a **written report** to:
      i) Primary care provider/Medical Home and other care providers and agencies, as requested by the parents
      ii) Birthing hospital
      iii) Utah Department of Health (UDOH), Early Hearing Detection and Intervention, for entry into the State newborn hearing screening database. **The diagnostic report is**

due within 7 days. A release of information to the UDOH is not necessary (see Utah Administrative Rule R398-2-6 (3-4))

iv) USDB-PIP

UNDETERMINED Hearing Loss: Diagnostic and Early Intervention Referral Protocol

The following should be completed **for infants and children whose hearing status is "Undetermined" (i.e., hearing loss is suspected, but additional testing is needed to confirm):**

1) **Review results and recommendations** with the parents/caregivers including:
   a) Information regarding the need for medical and audiological follow-up
      i) **Schedule Follow-up ABR** prior to family leaving
      ii) Referral to Otolaryngology (ENT) for medical consultation
      iii) Contact the Primary Care Physician
      iv) Referral to EHDI Parent Consultant
   b) **In order to minimize confusion, provide a hard-copy of recommendations / "next steps" for parents/caregivers prior to their leaving the appointment.**
   c) Referral to early intervention services. **For infants and children < 3 years of age, see "*Infant Diagnostic & Early Intervention Referral Process*" on Page 7, for specific referral information.**

2) Distribute a **written report** to:
   i) Primary care provider/Medical Home and other care providers and agencies, as requested by the parents
   ii) Birthing hospital
   iii) Utah Department of Health (UDOH), Early Hearing Detection and Intervention, for entry into the State newborn hearing screening database. **The diagnostic report is due within 7 days. A release of information to the UDOH is not necessary** (see Utah Administrative Rule R398-2-6 (3-4))
   iv) USDB-PIP

FLUCTUATING CONDUCTIVE Hearing Loss: Diagnostic and Early Intervention Referral Process

The following should be completed **for infants and children whose hearing status is "Fluctuating Conductive":**

1) Review results and recommendations with the parents/caregivers including:
   a. Information regarding the need for medical and audiological follow-up
      i. **Schedule Follow-up ABR** prior to family leaving
      ii. Referral to Otolaryngology (ENT) for medical consultation

iii. Contact the Primary Care Physician

b. **In order to minimize confusion, provide a hard-copy of recommendations / "next steps" for parents/caregivers prior to their leaving the appointment.**

c. For infants and children < 3 years of age: If after two diagnostic appointments, normal hearing is not established, a referral to early intervention services is necessary. **See, "*Infant Diagnostic & Early Intervention Referral Process*" on Page 8, for specific referral information.**

A Diagnostic Written Report Should Include:

1) Case History

a) Newborn Hearing Screening results
b) Cytomegalovirus PCR testing results
   i) Specimen type (urine, saliva)
   ii) Detected or Undetected
   iii) If not completed, complete UDOH *Cytomegalovirus & Auditory Brainstem Response Testing Orders f*orm and assist family to hospital lab. NOTE: If the lab will be using a saliva swab specimen, it should not be collected until at least 2 hours after the last time the baby breastfed.
c) Risk Factors

2) Diagnostic Evaluation Results (ear specific)

a) Tympanometry (specify: 1000 Hz or 226 Hz)
b) OAEs (include both type and frequencies)
c) Acoustic Reflexes (specify: presence or absence)
d) ABR (Note: be sure to indicate eHL or nHL)
   i) Click stimuli (include rarefaction and condensation polarities to rule-out ANSD)
   ii) Toneburst stimuli (a diagnostic is **not complete** without frequency-specific testing)
   iii) Bone Conduction, where indicated (e.g., abnormal ABR responses)
e) Behavioral testing (ear-specific VRA/CPA, if developmentally appropriate)

3) Summary & Recommendations

a) If hearing loss present, include referral to:
   i) ENT*
   ii) PIP (USDB-Parent Infant Program) *
   iii) Genetics
   iv) Ophthalmology
   v) Neurology
   vi) Other specialties as needed

*A referral should always be made

4) A **copy of your report** should be sent to:

a) For **normal hearing OR confirmed hearing loss**:
   i) Family**
   ii) Utah EHDI**

iii) Primary Care Provider**

b) In cases of **confirmed hearing loss**:
   i) ENT**
   ii) USDB-PIP**
   iii) Genetics
   iv) Ophthalmology
   v) Neurology
   vi) Other specialties as needed

**Should always receive a copy of your report

Reporting Results to Utah EHDI:

Email: ehdi@utah.gov or Krysta Badger at kbehring@utah.gov

Mail: **Utah Early Hearing Detection and Intervention Program**
PO Box 144620
Salt Lake City, UT 84114

Fax: **801-536-0492 (secure fax)**

Infant Diagnostic & Early Intervention Referral Process
Infant Refers IP & OP Hearing Screening
UDOH CMV & Diagnostic ABR Orders
CMV Testing*
Diagnostic ABR
Early Intervention
Normal Hearing
Results to Parents, PCP, EHDI
Undetermined
Results to Parents, PCP, EHDI
Referral to USDB-PIP & EHDI Parent Consultant
Schedule F/U Diagnostic ABR
Fluctuating Conductive
Results to Parents, PCP, EHDI
ENT referral
EHDI Parent Consultant
Schedule F/U Diagnostic ABR
If no confirmed hearing status, referral to USDB-PIP by 4 months of age
Confirmed Permanent Hearing Loss
Results to Parents, PCP, EHDI
Parent Notebook
Referral to USDB-PIP & EHDI Parent Consultant within 48 hours
deafpipreferral@usdb.org
ehdiparents@utah.gov
Referral to ENT, Genetics, Ophthalmology
Schedule F/U Diagnostic ABR
*CMV Testing also required if 1st hearing screening "refer" is > 14 days of age
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APPENDICES

APPENDIX A: JCIH Risk Factors for Hearing Loss (2000)

Indicators for progressive, delayed onset and/or conductive hearing loss; risk indicators marked with an "*" are of a greater concern for delayed-onset hearing loss

- Caregiver concern* regarding hearing, speech, language, or developmental delay
- Family history* of permanent childhood hearing loss
- Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO*, assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion
- In utero infections such as cytomegalovirus*, 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 a 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-Nielsen
- 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, especially basal skull/temporal bone fracture* that requires hospitalization
- Chemotherapy*

APPENDIX B: *Infants at increased risk for auditory neuropathy*

The population of infants cared for in the NICU may also be at increased risk for a neural condition and/or auditory brainstem dysfunction, including auditory neuropathy. Auditory neuropathy spectrum disorder (ANSD) is characterized by a unique constellation of behavioral and physiologic test results. Behaviorally, children with ANSD have been reported to exhibit normal hearing to profound hearing loss with poor speech perception. Physiologic measures of auditory function (e.g., otoacoustic emissions and auditory brainstem response) may demonstrate the finding of normal OAEs (suggesting normal outer hair cell function) and atypical or absent ABRs (suggesting neural conduction dysfunction). Reports suggest that those at increased risk for ANSD are (a) infants with a compromised neonatal course who receive intensive neonatal care, (b) children with a family history of childhood hearing loss, and (c) infants with hyperbilirubinemia. Currently, neither the prevalence of ANSD in newborns nor the

natural history of the disorder is known, and treatment options may include hearing aids or cochlear implantation. Audiological and medical monitoring of infants at risk for ANSD is recommended.

APPENDIX C: *Infants with conductive hearing loss*

The JCIH recommends ongoing audiologic and medical monitoring of infants with unilateral, mild, or chronic conductive hearing loss. Infants and children with mild or unilateral hearing loss may also experience adverse speech, language, and communication skill development, as well as difficulties with social, emotional, and educational development. Infants with unilateral hearing loss are at risk for progressive and/or bilateral hearing loss. Infants with frequent episodes of otitis media with effusion (OME) also require additional vigilance to address the persistent or recurrent OME.

For questions or comments regarding this Well-baby screening and diagnostic protocol, please contact Utah EHDI at ehdi@utah.gov.