**Best Practice Protocol for Pediatric Audiological Assessment:**
A guide for testing infants who refer on their newborn hearing screen OR have a risk factor for hearing loss

The following are the recommended *Minimum Standards* for a complete evaluation.
*It may take more than one appointment to obtain the complete diagnostic audiological evaluation on a pediatric patient.*

A Full Diagnostic Evaluation for **infants up to age 6 months** (to include assessment on BOTH ears, even if only one ear referred on the newborn hearing screening)
✓ Family and child history
✓ Otoscopy
✓ Frequency specific assessment at 500, 1000, 2000, and 4000 Hz via ABR with Tone Bursts;
   - If hearing loss is identified via air conduction ABR, bone conduction ABR should be completed to determine type of hearing loss
   - If neural hearing loss has been ruled out and ABR results indicate >90 dB hearing loss (no response at the limits of the equipment), ASSR testing should be completed to identify possible profound hearing loss threshold
✓ Click evoked neurodiagnostic ABR using both condensation and rarefaction stimulus, to determine if a cochlear microphonic is present, and that there is no reversal to the waveform response. A "no response" frequency specific ABR must also include a click recording with polarity reversal.
✓ Comprehensive otoacoustic emissions, DPOAE and/or TEOAE
✓ Tympanogram at 1000Hz tone for infants under 6 months of age
✓ Report results after each appointment to the Maine Newborn Hearing Program via the online reporting form

For children **>6 months of age developmentally**, and as appropriate (to include assessment on BOTH ears).
✓ Family and child history
✓ Otoscopy
✓ Conditioned Behavioral Audiometry (VRA or CPA) under insert earphones or headphones:
   - Minimal response levels for air at 500, 1000, 2000, 4000, and 8000 Hz
   - Bone conduction as needed to rule out a conductive pathology
   - Speech Awareness Thresholds/Speech Reception Thresholds
   - Word Recognition Scores when developmentally appropriate
✓ Comprehensive otoacoustic Emissions, DPOAE and/or TEOAE
✓ Immittance battery
   - 226Hz probe tone tympanometry
   - Ipsilateral and contralateral acoustic reflexes at 500, 1000, and 2000 Hz
✓ ABR testing is indicated if the responses to behavioral audiometry are unreliable or if there is suspicion of a neural hearing loss. *At least one ABR test is recommended to confirm hearing loss in children under the age of three years
✓ Report results after each appointment to the Maine Newborn Hearing Program for children up through age 3 years old, via the online reporting form

*It is recommended that all of the above information and results of each test be provided in the audiological report and sent to the child's pediatrician.*

**References**
www.asha.org/policy

Maine Newborn Hearing Program 207-287-8427
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Risk Indicators for Hearing Loss

RISK INDICATORS ASSOCIATED WITH PERMANENT CONGENITAL, DELAYED-ONSET, OR PROGRESSIVE HEARING LOSS IN CHILDHOOD

Risk indicators that are marked with an asterisk * are of 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 greater than or equal to (≥) 5 days*, exposure to ototoxic medications (gentamycin and tobramycin), loop diuretics (furosemide/Lasix), or chemotherapy, 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 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
- Severe birth asphyxia
- Hyperbilirubinemia without transfusion (at risk for Auditory Neuropathy Spectrum Disorder)

References
Monitoring Infants with Risk Factors for Hearing Loss

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<th>Level 1B Risk Factors</th>
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<td>• Family history of permanent childhood hearing loss</td>
<td>• Syndromes associated with progressive hearing loss</td>
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<tr>
<td>• In-utero infections (CMV, herpes, rubella, toxoplasmosis, syphilis)</td>
<td>(Neurofibromatosis, Osteopetrosis, Usher syndrome, Waardenburg Syndrome, Pendred Syndrome, Alport Syndrome, Lange-Neilson Syndrome)</td>
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<tr>
<td>• Culture positive postnatal infection (bacterial meningitis, sepsis)</td>
<td>• Neurodegenerative disorders or sensory motor neuropathies (Hunter Syndrome, Friedreich ataxia, Charcot-Marie-Tooth Syndrome)</td>
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<td>• Craniofacial or temporal bone anomalies (cleft lip/palate, atresia, ear tags/pits)</td>
<td>• Head Trauma, especially of the basal skull and temporal bone fractures</td>
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<td>• Severe birth asphyxia</td>
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<td>• Mechanical ventilation</td>
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<td>• Multiple risk factors from any level</td>
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<td>• NICU stay greater than 5 days</td>
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<td>• Hyperbilirubinemia without transfusion (at risk for Auditory Neuropathy Spectrum Disorder)</td>
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</table>

*Caregiver concern for hearing, speech, language, or developmental delay should indicate necessity for a diagnostic audiological evaluation at the time of concern.

**Level 1A Risk Factors:** If the infant falls within this category, and has passed the newborn screening, it is recommended the baby is referred for a full diagnostic evaluation by 3 months. Frequent follow up is recommended.

**Level 1B Risk Factors:** If the infant falls within this category, and has passed the newborn screening, it is recommended the baby is referred for a full diagnostic evaluation by 6 months. Frequent follow up is recommended.

**Level 2 Risk Factors:** If the infant falls within this category, and has passed the newborn screening, it is recommended the child be referred for a full diagnostic evaluation by 12 months, and no later than 20-24 months.

*Routine follow-up thereafter is as the discretion of the audiologist/PCP

References:


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