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

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An Office of the Department of Health and Human Services
Paul R. LePage, Governor Mary C. Mayhew, Commissioner

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-Nielson
- 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

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Monitoring Infants with Risk Factors for Hearing Loss

Level 1A Risk Factors	Level 1B Risk Factors
• Family history of permanent childhood hearing loss • In-utero infections (CMV, herpes, rubella, toxoplasmosis, syphilis) • Culture positive postnatal infection (bacterial meningitis, sepsis) • Craniofacial or temporal bone anomalies (cleft lip/palate, atresia, ear tags/pits) • Severe birth asphyxia • Mechanical ventilation • Hyperbilirubinemia *with* transfusion • Multiple risk factors from any level • ECMO • Chemotherapy	• Syndromes associated with progressive hearing loss (Neurofibromatosis, Oseteopetrosis, Usher syndrome, Waardenburg Syndrome, Pendred Syndrome, Alport Syndrome, Lange-Neilson Syndrome) • Neurodegenerative disorders or sensory motor neuropathies (Hunter Syndrome, Friedriech ataxia, Charcot-Marie-Tooth Syndrome) • Head Trauma, especially of the basal skull and temporal bone fractures • Very low Birth Weight (<1500 g) • Respiratory Distress • Bronchiopulmonary dysplasia
Level 2 Risk Factors	
• Ototoxic medication exposure (any amount) with no other risk factors • Low birth weight (1500-2500 g) with no other risk factors • Prematurity (<37 weeks) with no other risk factors • NICU stay greater than 5 days • Hyperbilirubinemia without transfusion (at risk for Auditory Neuropathy Spectrum Disorder)	

***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*

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