



Risk Factors for Hearing Loss and Testing Recommendations Maine CDC Newborn Hearing Program

Immediate referral for Full Diagnostic Evaluation:

- Caregiver concern regarding hearing, speech, language, developmental delay, and/or developmental regression
- In-utero infection with Cytomegalovirus (CMV)
- Bacterial meningitis
- Baseline audiogram prior to platinum-based chemotherapy

Full Diagnostic Evaluation before 3 months of age OR no later than 3 months after occurrence:

- Family history of early, progressive, or delayed onset **permanent childhood** hearing loss
- Culture-positive infections associated with sensorineural hearing loss, including viral meningitis or encephalitis
- Craniofacial anomalies such as cleft/lip palate, microtia/atresia, or ear dysplasia
- Extracorporeal membrane oxygenation (ECMO)
- Mechanical Ventilation
- Chemotherapy
- Head trauma
- Syndromes associated with atypical hearing thresholds
- Certain birth conditions or findings such as white forelock, microphthalmia, congenital microcephaly, congenital or acquired hydrocephalus, or temporal bone abnormalities
- Mother and/or infant testing positive with Zika, with or without clinical findings
- Multiple risk factors from any level

Full Diagnostic Evaluation before 9 months of age:

- NICU stay for more than 5 days
- Hyperbilirubinemia with exchange transfusion regardless of length of stay
- Ototoxic medication exposure with no other risk factors
- Asphyxia or Hypoxic Ischemic Encephalopathy
- In-utero infections, such as herpes, rubella, syphilis, and toxoplasmosis
- Preauricular tags and ear pits

References for Risk Factors for Hearing Loss:

- Horn, P., Driscoll, C., Fitzgibbons, J., & Beswick, R. (2021). Detecting hearing loss in infants with a syndrome or craniofacial abnormalities following the newborn hearing screen. *Journal of Speech, Language, and Hearing Research*, 64(9), 3594–3602.
- Joint Committee on Infant Hearing (JCIH). (2019). Year 2019 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Journal of Early Hearing Detection and Intervention*, 4(2), 1-44.
- Roth, D.A., Hildesheimer, M., Bardenstein, S., Goidel, D., Reichman, B., Maayan-Metzger, A., & Kuint, J. (2008). Preauricular skin tags and ear pits are associated with permanent hearing impairment in newborns. *Pediatrics*, 122(4), 884-890.

Call the Maine CDC Newborn Hearing Program with questions (207) 287-8427



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Best Practice Protocol for Full Diagnostic Evaluation:

A guide for testing infants who refer on their newborn hearing screen OR infants/children who have a risk factor for hearing loss

It may take more than one appointment to obtain the complete diagnostic audiological evaluation on a pediatric patient.

Children up to age 6 months:

- Family and child case history, with use of appropriate language interpreters as necessary
- Assessment of BOTH ears, even if only one ear referred on the newborn hearing screening.
- Otoscopy
- Frequency specific assessment at 500, 1000, 2000, and 4000 Hz using frequency specific (tone burst, chirp) stimuli
 - If hearing loss is identified via air conduction ABR, complete bone conduction ABR to determine type of hearing loss
- Click or chirp 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
- 1000Hz probe tone tympanometry
- Report results after each appointment to the Maine Newborn Hearing Program via the online reporting form
- Provide audiological report that includes all of the above information and results of each test to the child's primary care provider and family.

Children 6 months of age or older:

- Family and child case history, with use of appropriate language interpreters as necessary
- Assessment of BOTH ears, even if only one ear referred on the newborn hearing screening.
- Otoscopy
- Behavioral Audiometry (VRA or CPA) under insert earphones or headphones
 - Minimal response levels (MRLs) for air at 250, 500, 1000, 2000, 4000 Hz for VRA; MRLs for air at octave intervals from 250 to 8000 Hz for CPA
 - 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
 - 1000 Hz probe tone tympanometry recommended through 9 months of age
 - 226Hz probe tone tympanometry above 9 months of age
 - 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 3 years of age.**
- Report results after each appointment to the Maine Newborn Hearing Program for children through age 3 years, via the online reporting form.
- Provide audiological report that includes all of the above information and results of each test to the child's primary care provider and family.

References for Testing Recommendations:

American Academy of Audiology. (2020). *Clinical Guidance Document: Assessment of Hearing in Infants and Young Children*.

<https://www.audiology.org/publications-resources/document-library/pediatric-diagnostics>

American Speech-Language-Hearing Association. (2014). *Permanent Childhood Hearing Loss*. <https://www.asha.org/Practice-Portal/Clinical-Topics/Permanent-Childhood-Hearing-Loss/>