



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
- 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
- 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

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 Risk Factors for Hearing Loss and 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/>

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., Goedel, 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.