



## Recommendations for ongoing hearing evaluations of children in the Early Intervention system

Over 99% of newborns in Massachusetts have hearing screenings soon after delivery. The system that ensures that those who do not pass the screening are seen for timely diagnostic audiology appointments and referred on to early intervention if the hearing loss is confirmed is working well. However, the newborn hearing screening is a snapshot in time. Some young children can have progressive or late onset hearing losses that are not identified by the newborn screening. Certain children are at higher risk for these types of hearing loss.

The Joint Commission on Infant Hearing 2007 Position Statement identifies risk factors associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood. The timing and number of hearing reevaluations for children with risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss. Infants who pass the neonatal screening but have a risk factor should have at least 1 diagnostic audiology assessment by 24 to 30 months of age. Early and more frequent assessment may be indicated for children with cytomegalovirus (CMV) infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, or culture-positive postnatal infections associated with sensorineural hearing loss; for children who have received extracorporeal membrane oxygenation (ECMO) or chemotherapy; and when there is caregiver concern or a family history of hearing loss.

*Risk indicators marked with a “§” are of greater concern for delayed-onset hearing loss.*

1. Caregiver concern§ regarding hearing, speech, language, or developmental delay.
2. Family history§ of permanent childhood hearing loss.
3. 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 (gentimycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.

4. In utero infections, such as CMV,§ herpes, rubella, syphilis, and toxoplasmosis.
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. 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.
8. Neurodegenerative disorders,§ such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot -Marie-Tooth syndrome.
9. Culture-positive postnatal infections associated with sensorineural hearing loss,§ including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
10. Head trauma, especially basal skull/temporal bone fracture§ that requires hospitalization.
11. Chemotherapy.§

Any infant or toddler who demonstrates delayed auditory and/or communication skills development, even if he or she passed newborn hearing screening, should receive an audiological evaluation to rule out hearing loss. Early intervention programs should refer children back to their primary health care providers with this recommendation so that necessary health plan authorization can be obtained. Be sure to obtain a copy of the audiological report and be aware of recommendations for follow up testing.