Indicators for Infants at Risk of Late Onset or Progressive Hearing Loss

The Joint Committee on Infant Hearing, Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs (www.jcih.org) recommended the 11 risk indicators listed below that are associated with either congenital or delayed-onset hearing loss.

All infants with a risk indicator for hearing loss should be referred for an audiological assessment at least once by 24 to 30 months of age. Children with risk indicators that are highly associated with delayed-onset hearing loss, such as having received ECMO or having CMV infection, should have more frequent audiological assessments.

There is a significant change in the definition of risk-indicator 3, which has been modified from Neonatal intensive care unit (NICU) stay more than 48 hours to a NICU stay more than 5 days.

Risk Indicators Associated with congenital, delayed-onset, or progressive hearing loss in childhood are listed below. Risk indicators that are marked with an asterisk* 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*.

Because some important indicators, such as family history of hearing loss, may not be determined during the course of Universal Newborn Hearing Screening, the presence of all risk indicators for acquired hearing loss should be determined in the medical home during early well-infant visits. Early and more frequent assessment may be indicated for children with 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 ECMO or chemotherapy, and when there is caregiver concern or a family history of hearing loss. All infants with and without risk indicators for hearing loss should be monitored during routine medical care consistent with the American Academy of Pediatrics periodicity schedule for developmental milestones, hearing skills, and parent concerns about hearing, speech, and language skills.