

Congenital Hearing Loss

**Jacque Cundall, R.N., B.S.N.
Newborn Hearing Screening Coordinator
Women's Health and Genetics
Tennessee Department of Health**

Outcome Without Screening:

If hearing loss is not identified until 2 or 3 years of age a child may suffer delays in speech, language, and cognitive development.

Incidence:

The incidence of congenital hearing loss is 2 to 3 infants per 1000 births with moderate to profound bilateral hearing loss. Hearing loss is the most common birth defect. 50% of infants with hearing loss do not have a risk indicator for hearing loss.

Outcome With Screening:

Hearing screening at birth can detect mild and unilateral (one ear) hearing loss as well as severe to profound losses. Infants who are screened prior to one month of age, diagnosed with hearing loss prior to three months old, and receive early intervention/habilitation services prior to 6 months old have been documented to have significantly higher levels of receptive and expressive language, personal-social development, expressive and receptive vocabulary, general development, situation comprehension, and vowel production than later identified infants with hearing loss. (Yoshinaga-Itano, C., & Apuzzo, M. (1998). The development of deaf and hard-of-hearing children identified early through the high risk registry. *American Annals of the Deaf*, 143, 416-424.). These infants function on normal or near-normal developmental levels.

Causes of Hearing Loss:

Hearing loss may be caused by abnormalities in the outer ear (pinna and ear canal), middle ear (eardrum and ossicles), and/or the inner ear (cochlea and auditory nerve).

Congenital: Genetic forms of hearing loss must be carefully distinguished from acquired (non-genetic) causes of hearing loss. 50% of permanent hearing loss in infants is due to genetic causes; 50% are to environmental causes (see risk indicators below).

Progressive, Delayed Onset or Acquired: Causes may be due to a genetic syndrome, in utero infection, postnatal infection, ototoxic medications, recurrent or persistent otitis media. (see risk indicators below)

Screening Test Confirmation:

Infants who do not pass the hearing screen at birth are referred for further hearing screening and/or diagnostic evaluation by the primary care provider for an audiology evaluation with an audiologist skilled in working with infants and young children. Infants found to have hearing loss will need further medical evaluation by an otologist/otolaryngologist/ENT and genetic provider to correctly diagnose the specific cause of hearing loss, identify other medical diagnoses/syndromes, need of surgical treatment if appropriate and provide information on communication options and prognosis. The genetic forms of hearing loss are diagnosed by otologic, audiologic, and physical examination, family history, ancillary testing (such as CT examination of the temporal bone), and DNA-based testing.

Newborn Screening Considerations:

Infants may have false positive results due debris in the ear canal, noisy test environment, level of infant activity during testing and on the screening method.

Treatment:

Treatment may be initiated immediately after diagnosis. There are a variety of communication methods such as oral, auditory-verbal, auditory-oral, sign or a combination of these. Amplification may include hearing aids, FM systems, and cochlear implants. Non-oral methods may include American Sign Language, Cued Speech, Signed Exact English and Total Communication.

Special Concerns:

The health professional should provide the family with information about the type and degree of the hearing impairment, its potential impact on speech/language and cognitive development, the treatment and communication options available, and the positive impact of early intervention. Health provider should have a clear understanding that a child's family has the final choice as to whether or not the infant should use hearing aids, cochlear implants, other assistive technology or other methods of communication.

Interdisciplinary early intervention may include vision evaluation, medical treatment, home/center based early intervention services and family support to enhance communication, thinking, and behavioral skills needed to achieve academic and social success.

A thorough assessment of hearing may require multiple sessions. Serial evaluations may be necessary to develop reliable profiles of hearing status and developmental abilities. Prolonged delays between assessments should be avoided.

Infants that pass the first hearing screening but who have a risk indicator for progressive and delayed onset hearing loss or acquired hearing loss, should be monitored every 6 months during the first three years of life. Risk indicator will be listed on the revised blood spot form.

Risk Indicators for Progressive and Delayed Onset or Acquired Hearing Loss

(Joint Committee on Infant Hearing 2000 Position Statement JCIH)

1. The JCIH risk indicators for **birth through age 28 days** where universal hearing screening is not yet available. These indicators are as follows:
 - a. An illness or condition requiring admission of 48 hours or greater to a neonatal intensive care unit.
 - b. Stigmata or other findings associated with a syndrome known to include a sensorineural and or conductive hearing loss.
 - c. Family history of permanent childhood sensorineural hearing loss.
 - d. Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal.
 - e. In utero infection such as cytomegalovirus, herpes, toxoplasmosis, or rubella.
2. The JCIH recommends the following indicators for use with neonates or infants (**29 days through 2 years**). These indicators place an infant at risk for progressive or delayed-onset sensorineural hearing loss and/or conductive hearing loss. Any infant with these risk indicators for progressive or delayed-onset hearing loss who has passed the birth screen should, nonetheless, receive audiologic monitoring every 6 months until age 3 years. These indicators are as follows:
 - a. Parental or caregiver concern regarding hearing, speech, language, and/or developmental delay.
 - b. Family history of permanent childhood hearing loss.

- c. Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or eustachian tube dysfunction.
- d. Postnatal infections associated with sensorineural hearing loss including bacterial meningitis.
- e. In utero infections such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis.
- f. Neonatal indicators—specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation (ECMO).
- g. Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher's syndrome.
- h. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich's ataxia and Charcot-Marie-Tooth syndrome.
- i. Head trauma.
- j. Recurrent or persistent otitis media with effusion for at least 3 months.