

Sensorineural Hearing Loss

FAST FACTS

2–4

of 1,000 newborns have SNHL

50,000

children in the US annually affected by SNHL

2/3

of infants with failed newborn hearing testing is due to fluid that may resolve on its own or be treated by ear tube surgery around six months of age

1–3

of 1,000 infants develop hearing loss later in childhood

15%

of adults (18+) have some degree of hearing loss

If you have clinical questions about patients with a failed newborn hearing screen, email ENT@cchmc.org.

Sensorineural hearing loss (SNHL) is the most common congenital sensory impairment. Newborn hearing screening is performed to identify infants who have hearing loss and need further evaluation so interventions can begin right away.

NEWBORN SCREEN

The birthing hospital schedules an initial Auditory Brainstem Response (ABR) test with the Division of Audiology at Cincinnati Children's within the first month of life. If the infant fails initial ABR, a second ABR should be scheduled within a month. ABR testing should be completed by two months of age so appropriate therapy can begin by three months. If needed, hearing aids are initiated after the second ABR. Cincinnati Children's performs cytomegalovirus (CMV) testing concurrently with ABR to expedite the diagnosis, because a positive CMV test after one month may be from post-natal acquired infection.

ASSESSMENT

It is critical to perform a medical evaluation on an infant with a failed newborn hearing screen during the first month of life. Perform a standard history and physical exam, with audiologic workup. Hearing aid therapy may begin as early as three months. Cochlear implants can be performed as early as six months to allow normal access to sound.

HPE (HISTORY AND PHYSICAL EXAM) RED FLAGS

- Family history of permanent childhood hearing loss
- Hyperbilirubinemia at birth

CAUSES

SNHL may be congenital, acquired, or idiopathic. Genetics causes 50% of hearing loss, primarily through nonsyndromic recessive genes, so genetic testing is important in diagnosis. Approximately a third of SNHL cases are acquired just before or after birth, through intrauterine infections such as CMV, environmental exposure, meningitis, head trauma, ototoxic medications, prematurity, or low APGAR scores. Cases that are neither genetic nor acquired are idiopathic.

CMV causes 15% of newborn hearing loss and may have no other symptoms. Diagnostic tests should be performed while the infant is still less than one month old for antiviral medication to be effective.

MANAGEMENT/TREATMENT

Treat positive CMV infection with oral Valgancyclovir to minimize hearing loss and prevent future progression.

For patients who are CMV negative, we recommend Otoseq genetic testing and MRI of the temporal bones. Hearing aids or cochlear implants can restore hearing in nearly all pediatric patients with SNHL.

WHEN TO REFER

If your diagnosis confirms failed newborn screen results, refer to Cincinnati Children's Ear and Hearing Center via Otolaryngology, and we will coordinate the appropriate referrals to our team members, which includes audiology/aural rehabilitation, speech and language therapy, developmental pediatrics, social work, and ophthalmology.

If you would like additional copies of this tool, or would like more information, please contact the Physician Outreach and Engagement team at Cincinnati Children's.

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