

Pediatric Sensorineural Hearing Loss



By **Anita Jeyakumar**, MD, pediatric otolaryngologist at Children's Hospital.

This issue of *Pediatric Review* is intended for pediatricians, family physicians and all other interested medical professionals. For CME purposes, the author has no relevant financial relationships to disclose.

OBJECTIVES

At the end of this activity the participant should be able to:

1. Describe the causes of pediatric sensorineural hearing loss
2. Discuss the appropriate treatment of pediatric sensorineural hearing loss
3. Describe the clinical presentation, diagnostic evaluation, and management for infants at risk for early-onset GBS disease

INTRODUCTION

The term sensorineural hearing loss (SNHL) denotes a hearing loss (HL) attributable to an abnormality affecting the cochlea, the vestibulocochlear nerve (CN VIII) or processing centers of the central nervous system. SNHL is the most common sensory deficit in developed societies. In the United States, one child in 500 is born with bilateral SNHL of at least 40 dB. The prevalence of hearing impairment above this threshold increases to 2.7 per 1000 before the age of 5 and 3.5 per 1000 during adolescence.

DIAGNOSTICS

To understand diagnostic testing for SNHL, we need to have a brief discussion about ear anatomy and physiology.

TESTING

As of 2010, 53 out of 59 U.S. states, districts and territories have an Early Detection and Intervention (EDHI) program for HL which provides screening services and referrals for nearly 95% of all births. The EDHI goals include the 1-3-6 plan, which calls for all infants to be screened by one month of age, all children who do not pass the screening to receive diagnostic testing by three months of age, and all children with confirmed hearing loss to be enrolled in an appropriate intervention program by six months of age. Since the initiation of newborn hearing screening, the average age at which HL is confirmed has dropped from 24 – 30 months to 2 – 3 months.

The diagnosis of HL requires careful medical history, family history and physical examination. Accurate assessment of the degree of hearing loss is essential to develop an appropriate treatment plan. An evaluation for HL should always include formal audiologic evaluation.

Testing hearing in a young child with HL can be difficult and requires experience and significant expertise. Depending on the child's age and severity of HL, there are multiple ways that hearing can be tested. Physiologic tests objectively determine the functional status of the auditory system and can be performed at any age. Volitional responses are not required from the patient for physiologic tests. They include:

- Auditory brain stem response testing (ABR, also known as BAER, BSER)
- Auditory steady-state response testing (ASSR)
- Evoked otoacoustic emissions (OAEs)
- Immittance testing (tympanometry, acoustic reflex thresholds, acoustic reflex decay).

Audiometry subjectively determines how the patient processes auditory information, or hears. Volitional responses are required for audiometric testing.

- Pure-tone audiometry involves the determination of the lowest intensity at which an individual "hears" a pure tone, as a function of frequency (or pitch). Octave frequencies from 250 Hz (close to middle C) to 8000 Hz are tested using earphones. Intensity or loudness is measured in decibels (dB), defined as the ratio between two sound pressures.
- Air conduction audiometry presents sounds through earphones. Accurate assessment is dependent on the condition of the external ear canal, middle ear and inner ear.
- Bone conduction audiometry presents sounds through a vibrator placed on the mastoid bone or forehead, thus bypassing the external and middle ears. Bone condition is dependent on the condition of the inner ear and nerve pathways.
- Behavioral testing includes behavioral observation audiometry (BOA) and visual reinforcement audiometry (VRA). BOA is used in infants from birth to 6 months of age. It relies on observation of patient responses to stimuli, including startle or head turning. VRA is used in children from age 6 months – 2.5 years of age and can provide a reliable, complete audiogram. In a similar method, conditioned play audiometry (CPA) is used to test children from age 2.5 – 5 years of age. The patient is taught to respond to stimuli with a specific action, either by performing a type of "play" or interacting with the tester.

SEVERITY OF HEARING LOSS

Hearing is measured in decibels (dB). The threshold or 0 dB mark for each frequency refers to the level at which normal young adults perceive a tone burst 50% of the time. Hearing is considered normal if an individual's thresholds are within 15 dB of normal thresholds. The severity of HL is graded as shown in Figure 2.

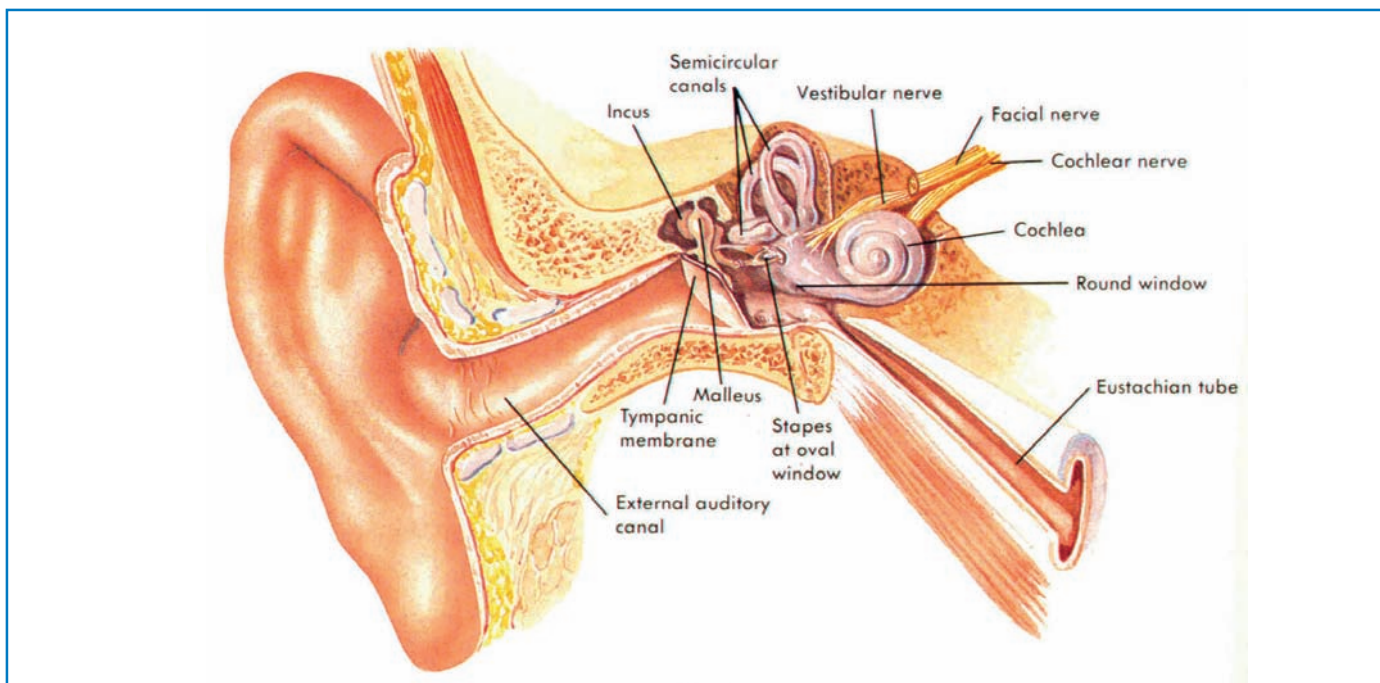


Figure 1 – Ear anatomy

DIFFERENTIAL DIAGNOSIS

HL can be described by onset: prelingual or postlingual. Most congenital HL is prelingual, but not all prelingual HL is congenital. HL can also be described by mechanism, as follows:

Conductive – In a conductive hearing loss (CHL), the sound waves are not effectively transmitted via the external ear and/or middle ear to the cochlea, thus the sensory cells receive decreased stimulation. Many forms of CHL are amenable to surgical correction. Examples of CHL include impacted wax and serous otitis media.

Sensorineural – Sensorineural hearing loss (SNHL) occurs when the sensory cells of the cochlea (inner ear) or the auditory nerve fibers are dysfunctional. The acoustic energy (sound wave) is not capable of being transformed within the cochlea to electrochemical stimuli or transmitted along the auditory pathway.

Mixed – Mixed HL is the combination of a CHL and SNHL. For example, a mixed loss could be present in a child with acute otitis media with coincident SNHL.

Central – Central HL occurs in the auditory areas of the brainstem and higher levels. Persons with central hearing loss often have normal measured hearing, but have difficulty with the processing of auditory information.

Functional – Persons with functional HL have no physiologic basis for a hearing deficit.

SENSORINEURAL HEARING LOSS

SNHL can be present at birth or progress over time. In the general population, the prevalence of hearing loss increases with age. This change reflects the impact of genetics, environment and interactions between environmental triggers and an individual's genetic predisposition. In general, it is believed that half of congenital HL is genetic in etiology and half is environmental or acquired.

PROPHYLAXIS

With the exception of acquired SNHL and noise induced HL, most hearing loss is not preventable. There is no prophylaxis for genetic HL, except for the change in counseling for the appropriate

patient. The next part of the discussion is about the significance of acquired versus genetic HL.

ACQUIRED HL

A significant portion of acquired HL in children results from maternal prenatal infections. "TORCH" organisms such as toxoplasmosis, rubella, cytomegalovirus, and herpes constitute a large percentage of the causative organisms and may cause SNHL as part of the disease process or sequelae.

GENETIC CAUSES

Genetic causes of SNHL can be divided into syndromic or non-syndromic etiologies. Syndromic hearing impairment is the association of hearing loss with consistent clinically recognizable features involving other organ systems. Non-syndromic HL does not have associated features in other organs. As previously stated, most genetic causes of SNHL are non-syndromic.

EVALUATION OF THE SNHL PEDIATRIC PATIENT

Appropriately diagnosing the specific cause of HL in a patient can provide prognosis and counseling information. There are many tools in the physician's arsenal for formulating a correct diagnosis, including: family history, clinical examination (including an evaluation for syndromic features), audiologic findings based on the age of the patient, imaging with CT temporal bone protocols or MRI, 12-lead EKG, ophthalmologic evaluation, blood testing and urine testing for Cytomegalovirus (CMV) and genetic testing.

Computed tomography of the temporal bones is useful for detecting malformations of the inner ear (i.e., Mondini deformity, Michel aplasia, enlarged/dilated vestibular aqueduct, dilation of the internal auditory canal), which should be considered in persons with progressive HL.

CMV testing needs to be considered in infants with SNHL. The diagnosis of in utero CMV exposure requires detection of elevated CMV antibody titers or a positive urine culture in the neonatal period.

dB/Hz	250	500	1000	2000	4000	8000
-10	NORMAL HEARING					
0						
10						
20						
30	MILD HEARING LOSS					
40						
50	MODERATE HEARING LOSS					
60						
70	SEVERE HEARING LOSS					
80						
90	PROFOUND HEARING LOSS					
100						
110						
120						

Figure 2 – Severity of hearing loss

GENETIC TESTING

Genetic testing can be performed in children with SNHL. Genetic testing may involve obtaining blood from a hearing-impaired child and performing a chromosomal analysis in the hope of identifying a chromosomal defect leading responsible for HL. The caveat is that only a small portion of the chromosomal abnormalities involved in genetic hearing loss have been identified and an even smaller number are routinely tested for on a regular basis. However, many previously undiagnosed HL are identified and diagnosed with this technique.

MENINGITIS

A discussion about meningitis requires its own section because patients with profound hearing loss secondary to pneumococcal meningitis often develop cochlear ossification. Ossification within the cochlea can limit or prevent electrode insertion within the cochlea.

Despite the considerable advances in imaging modalities, CT scans of the cochlea do not reliably identify ossification or the presence of inflammatory or intra-cochlear scar tissue. In addition, the patient's medical condition may preclude evaluation due to the cognitive effects of meningitis and difficulty in proceeding with standardized behavioral measures.

However, to optimize hearing rehabilitation with a cochlear implant, the electrode placement needs to be done as soon as medically appropriate. Bilateral simultaneous cochlear implantation is also a consideration as it enables the capture of the ear with the better hearing potential and preserves the potential for binaural hearing.

MANAGEMENT OF THE SNHL PEDIATRIC PATIENT

A child with SNHL is best managed with a team approach. The team should consist of an otolaryngologist with expertise in the management of early childhood otologic disorders, an audiologist experienced in the assessment of HL in children, a speech language pathologist, a pediatric ophthalmologist and a pediatrician. The expertise of a deaf educator, neurologist, cardiologist, endocrinologist and geneticist may be required.

An important part of the evaluation is determining the appropriate habilitation option, if the parents choose to pursue hearing rehabilitation. As difficult as it might seem, it is important for the child's medical well-being that the parents are supported if they decide to pursue non-aural communication such as American Sign Language. Possibilities for hearing rehabilitation include preferential seating in school, FM system, speech and language therapy, hearing aids, vibro-tactile devices and cochlear implantation. It is important to initiate hearing rehabilitation as early as possible to improve outcomes for the child. Regardless of its etiology, uncorrected HL has consistent sequelae. Auditory deprivation through age two years is associated with poor reading performance, poor communication skills and poor speech production. Educational intervention alone is insufficient to completely remedy these deficiencies. Early auditory intervention, whether through amplification, otologic surgery or cochlear implantation, is effective in conjunction with the proper educational environment. Cochlear implantation can be considered in children over age 12 months with severe-to-profound hearing loss, and is FDA approved for these children.

SURVEILLANCE OF THE SNHL PATIENT

Sequential audiologic examinations are essential to document the stability or progression of the hearing loss, as well as to identify and treat superimposed HL, such as middle ear effusion or development of retrocochlear pathology.

PROGNOSIS

The prognosis of the SNHL patient continues to improve as technology improves. Hearing aid and cochlear implant technology continues to grow at an outstanding pace for those patients who wish to pursue hearing rehabilitation.

Noise exposure needs to be minimized by avoidance in patients with documented HL.

Middle ear infections or effusions need to be treated aggressively, especially in those children with hearing aids, to optimize the benefit they get from the hearing aids.



Children's Hospital Orthopaedics & Rehabilitation

The Orthopaedics Department provides evaluation, diagnosis and treatment for the entire spectrum of orthopaedic conditions and disorders in infants, children and adolescents. Orthopaedics offers services to patients with congenital and acquired malformations of the musculoskeletal system (clubfoot, congenital dislocation of the hip, Legg-Perthes, cerebral palsy), fractures and their late sequelae, scoliosis and other spinal abnormalities, and hand injuries. State-of-the-art sports medicine services are also available.

A specialized 13-bed, critical care spinal unit is available to all patients who undergo a spine related surgical procedure.

Management expertise includes, but is not limited to:

- Clubfoot using Ponseti Method and other foot disorders
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- Reconstruction of developmental skeletal malformations
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- Acquired and congenital hand disorders
- Musculoskeletal tumors
- Spinal disorders – Scoliosis, Kyphosis, Spondylothesis

- Limb lengthening
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Rehabilitation Center

Children's Hospital has celebrated more than 50 years of serving the medical and surgical needs of the children of our region. Today's strength resulted from the hospital's solid foundation as a rehabilitation hospital, beginning in 1955. That strong commitment to pediatric rehabilitation continues, and it is even stronger today. The comprehensive rehabilitative services and compassionate care provided by the center's dedicated staff have dramatically improved the quality of life for thousands of children. We are extremely proud of our past accomplishments, but we never lose sight of our goal — "healing one child at a time."

The goal of the center is to provide family-centered, comprehensive pediatric rehabilitative care in which the interdisciplinary team maximizes the development of independent and productive life skills through training. When a child is injured or disabled it affects the entire family — especially when rehabilitative care is necessary. Team members provide a framework to help each family adjust to the child's individual abilities and offer emotional support to preserve and strengthen the family during hospitalization. Recognizing and incorporating each family's unique qualities and individual cultural and

religious beliefs is important to the success of the program.

Family members are encouraged to attend therapy sessions with the patient to foster success in rehabilitation and reintegration into the community, school and vocation as appropriate.

Functional Pediatric Treatment

The rehabilitation goal for children and adolescents is to concentrate on restoring abilities by helping them adapt to their new skills or gaining abilities never achieved because of congenital disorders. Each patient is followed using a criteria-based system to evaluate functional skill development with respect to medical status, motor skills, cognitive and communicative abilities, self-care capabilities, and the family's needs. A comprehensive treatment plan tailored to the individual child is developed. This interdisciplinary program is reviewed and updated at regularly held staff meetings. The family is encouraged to interact with the rehabilitation team frequently, is kept informed of progress and is asked to provide input into the treatment plan. The team uses a nurse case-management approach to coordinate treatment plans with the child, family, physicians and team members.

Occupational Therapy

The Occupational Therapy Department provides evaluations and therapies to help children gain independence and promotes the development of fine motor skills, sensory motor skills, visual motor skills, self-help skills and oral motor skills that children need to function and socialize in their home, school, play and community environments. In the case of an injury or debilitating illness, Occupational Therapy will focus on rehabilitating children, allowing them to return to their daily routines at their highest level of function.

Physical Therapy

The Physical Therapy Department provides evaluation and therapy for problems with musculoskeletal and neurologic disorders in delay of developmental skills and impairments in gross motor functioning (large movements). Treatments may include range of motion, strengthening exercises, ambulation and posture control as well as performing functional activities (such as sitting, running or riding a bike). The department participates in amputee clinic working in conjunction with physicians and prosthetists to provide evaluation and training for the upper or lower extremity amputee. Orthotics evaluations are also available. Additionally the department provides burn or wound care evaluation and treatment working with surgical or plastic surgeons.

For more information or to refer a patient, please contact Lynn Kaska at (504) 896-2177 or lkaska@chnola.org.



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Please record your responses to the questions on the form below. Please circle the best possible answer. CME offer is good through September 2012.

- The term sensorineural hearing loss denotes a hearing loss attributable to an abnormality affecting :
 - the cochlea
 - the vestibulocochlear nerve
 - processing centers of the central nervous system
 - all of the above
- The average age at which hearing loss is confirmed is:
 - 2 – 3 months
 - 4 – 5 months
 - 6 – 12 months
 - 1 year
- Hearing is considered normal if an individual's thresholds are within ____ decibels of normal thresholds:
 - 5
 - 10
 - 15
 - 20
- To optimize hearing rehabilitation with a cochlear implant, the electrode placement needs to be done as soon as medically appropriate.
 - True
 - False

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