

Pediatric Hearing Loss

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Educational Gap

Hearing loss is a common condition presenting in children. Speech and language outcomes for children with hearing loss are related to identification of the hearing loss and to the degree of hearing loss. Early identification and appropriate, prompt intervention yield better outcomes. An understanding of the relevant anatomy, common causes, testing strategies, and management of hearing loss can help the primary care physicians maximize communication development in children.

Objectives After reading this article, the reader will be able to:

1. Provide a basic overview of the anatomy of the ear and discuss implications for conductive and sensorineural hearing loss.
2. Discuss the importance of newborn hearing screening and provide details on how this testing is accomplished.
3. Discuss some of the causes of hearing loss in children.
4. Detail interventions that may be appropriate in children with hearing loss.

Hearing loss is a common condition in children, with 1 in 1000 live births affected with severe to profound permanent hearing loss. The prevalence increases to 6 in 1000 when all degrees of hearing loss, mild to profound, are considered. As children age, the prevalence increases, and by age 18 years, 17 in 1000 individuals are affected by some degree of permanent hearing loss. This makes hearing loss more prevalent than diabetes mellitus and all pediatric cancers. (1) These numbers, however, do not take into account all the children who are affected by long-standing chronic effusions, which, though temporary, can have a significant effect on speech and language development if not identified and appropriately treated.

Hearing loss in children can derive from many forms. It can be congenital (present at birth), genetic, syndromic, nonsyndromic, acquired, and/or progressive. It may manifest as conductive, sensorineural, or mixed hearing loss. The evaluation, diagnosis, and management of children with hearing loss will involve a multidisciplinary effort by pediatricians, otolaryngologists, audiologists, deaf educators, speech and language pathologists, early intervention specialists, and

AUTHOR DISCLOSURE Dr. Grindle has disclosed that he has a family member who works for Bristol-Myers Squibb. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

ABBREVIATIONS

ABR	auditory brainstem response
CHL	conductive hearing loss
CMV	cytomegalovirus
OAE	otoacoustic emission
SNHL	sensorineural hearing loss
UNHS	universal newborn hearing screening

many others. The main focus of care should always be on providing appropriate communication strategies to the affected individual.

A basic knowledge of the auditory system is needed to understand how to appropriately intervene in childhood hearing loss. In addition, one must understand how response to sound is measured clinically. The intensity of sound is measured in decibels. In testing paradigms, the intensity of the sound is varied, and the patient's responses are recorded. By definition, 0 dB is the threshold of human hearing, meaning that someone with normal hearing will hear a tone burst at this intensity 50% of the time. Normal hearing is defined as being within 15 dB of this threshold. Minimal hearing loss is 16 to 25 dB, mild hearing loss is 26 to 40 dB, moderate hearing loss is 41 to 55 dB, severe hearing loss is 71 to 90 dB, and profound hearing loss is greater than 90 dB. However, for any sound at any intensity to be registered by the brain, all the components of the auditory system must be working.

Essentially, there are 4 components: the outer ear, the middle ear, the inner ear, and the auditory nerve. The outer ear is a sound wave collection funnel. Its shape and configuration provide some enhancement to incoming sound, but the effect is minimal (approximately 5 dB). However, the external auditory canal must be patent to allow for the transmission of sound waves to the tympanic membrane. Numerous factors can interfere with the sound waves at this point. Cerumen impaction, foreign body, malformation of the auricle, or canal stenosis or atresia can all compromise the function of the outer ear.

The middle ear starts at the tympanic membrane and continues to the stapes footplate. The function of the middle ear is to serve as a pressure transducer and overcome the impedance mismatch that exists between sound waves in an air vs fluid medium of the sensory organ, the cochlea. Sound waves that are propagating through the air interact with the tympanic membrane. They cause a vibration of the tympanic membrane that moves through the ossicles (the malleus, then the incus, and finally the stapes). Adherent to the stapes is the stapes footplate and the oval window membrane. Primarily because of the vastly greater surface of the tympanic membrane compared with the oval window membrane, the sound waves that interact with the tympanic membrane are transduced and amplified through the middle ear. There is a small contribution from the size and orientation of the ossicles. Overall, the gain is approximately 20 dB.

For the tympanic membrane and ossicles to function properly, they must be intact and in an air-filled space. Thus, large tympanic membrane perforations, ossicular discontinuity, and stiffening of the ossicular chain or stapes footplate (otosclerosis) can cause hearing loss. In addition, if the

middle ear becomes filled with something other than air (effusion or cholesteatoma), this can disrupt its function and lead to hearing loss. This is particularly common in young children who have eustachian tube dysfunction and frequent otitis media with effusion.

Anything that disrupts sound getting to the cochlea can be considered conductive hearing loss (CHL). Loss at the point of the cochlea or proximal to the cochlea is considered sensorineural hearing loss (SNHL). A combination of the 2 is termed a mixed loss. The cochlea is the sensory organ of hearing. The cochlea itself is spiral in shape with $2\frac{1}{4}$ to $2\frac{1}{2}$ turns. Inside the cochlea are 3 distinct spaces that run its length. The scala tympani and the scala vestibuli are both filled with perilymph and surround the scala media, which contains endolymph. Perilymph and endolymph have different electrolyte components similar to extracellular and intracellular fluid, respectively. An electric potential exists between the 2 fluid compartments and is essential for generating action potentials along the cochlear nerve. The organ of Corti is within the scala media. Within the organ of Corti are the inner and outer hair cells, as well as many other supporting cells. Sound waves cause a vibration of the tympanic membrane, which is transduced through the middle ear and cause vibrations of the oval window membrane. This wave propagates through the perilymph of the cochlea and causes displacement along the basilar membrane of the scala media, effectively opening ion gated channels in the hair cells and causing an action potential. The cochlea is tonotopically organized such that low frequencies are registered at the apex and high frequencies at the base of the cochlea.

These action potentials are carried by the auditory nerve through various centers in the brainstem and to the auditory cortex for higher-level processing. Obviously, a defect anywhere along this path could cause a hearing loss. These defects can be macroscopic defects (eg, brainstem lesions or abnormally formed cochleas) but far more commonly are microscopic defects (eg, abnormal membrane gating proteins, ion channel defects, and collagen defects).

DIAGNOSIS OF HEARING LOSS

The earlier the diagnosis of hearing loss can be made, the sooner interventions can be initiated to help the child develop. Currently, all states have a universal newborn hearing screening (UNHS) program and early hearing and detection and intervention programs. In 1993, only 11 hospitals in the United States screened more than 90% of their newborns. Since this time, there has been incredible expansion of the programs such that in 2011 a total of 97% of all children

born in the United States had completed a newborn hearing evaluation. (2) Current data relating to universal newborn hearing programs and early hearing detection and intervention programs can be found at <http://www.cdc.gov/ncbddd/hearingloss/data.html>.

It is because of these early identification efforts that interventions are able to begin early in a child's life. The goal of these programs is to:

- identify all children with a permanent hearing loss by age 3 months;
- initiate appropriate interventions by age 6 months;
- help establish a medical home for infants with permanent hearing loss; and
- track data and quality metrics for public health initiatives.

Newborn hearing screening and early intervention programs rely on objective testing of hearing that can reliably be performed on newborn infants. These tests are otoacoustic emission (OAE) testing and auditory brainstem response (ABR) testing (also called brainstem auditory evoked response testing). These 2 tests are used as the key components in UNHS.

OAE testing relies on the fact that in response to certain stimuli there are responses thought to originate from the outer hair cells of the inner ear. These sounds can be recorded. To perform OAE testing, sound is presented to the cochlea through an insert speaker placed in the ear canal. This sound evokes the cochlea into making a specific response, which can be picked up and recorded by a microphone and recorder within the ear canal insert. These OAEs are reliably present in normal hearing ears and serve as a surrogate marker for the function of the cochlea. OAE testing is inexpensive, does not require sedation of the infant, and can be performed quickly. Because this testing relies on presenting sound to the cochlea at an appropriate intensity, it can be affected by external and middle ear conditions (eg, canal stenosis, cerumen, or other debris in the external canal or middle ear effusion). Results are reported as present or absent.

The other testing paradigm commonly used for UNHS is the ABR test. Electrodes placed on the scalp record the resulting electrographic activity of sound impulse as it propagates along the auditory nerve and brainstem. Presented impulses can be either high-frequency clicks or frequency-specific tones. In automated ABR testing used in UNHS, the intensity and frequency of the stimuli are preset to screen for mild hearing loss. If an infant does not pass an automated ABR test, a full ABR test can be performed. In this test, both the intensity and frequency of the stimuli can be adjusted. In this way, objective information can be attained as to the specific degree of hearing loss (mild to profound) and sound frequency of hearing loss (low to high). The ABR can also be

affected by outer ear and middle ear disease, although to a lesser extent than the OAE. However, ABR testing requires a calm, resting infant. As the child grows, the ability to perform ABR testing without sedation decreases. Usually, after approximately age 4 months, sedation is required.

Both OAE and ABR tests are useful for testing newborns because they are objective, reliable, and repeatable and require limited cooperation. The tests, however, are surrogate markers for normal hearing. It is thought that if the cochlea is structurally intact, as reflected by present OAEs, and if there is integrity of the auditory nerve, as measured by a normal ABR test result, then hearing should be normal. It is only when the child gets older that hearing can be fully tested. Behavioral audiometry or visual reinforcement audiometry can be performed on children as young as ages 6 to 9 months. Play audiometry typically is used for children older than 2 years. At ages 4 to 5 years, children can cooperate with routine or conventional audiometry. In these tests, pure tones are presented to the ears separately or simultaneously. Behavioral responses can be observed as the intensity and frequency of the stimuli are modulated. These responses coupled with word discrimination scores in older children can give an accurate reflection of a patient's ability to hear and understand at a particular hearing level.

An adjunct to all these forms of testing is tympanometry, which can be used to measure the function of the external auditory canals and the middle ear. Normal middle ear pressures (type A) indicate normal tympanic membrane and ossicular mobility and compliance. Type B or flat tympanograms are present if there is no mobility of the tympanic membrane (eg, otitis media with effusion) or if there is complete fixation of the ossicles. This will also be present if there is a tympanic membrane perforation. Type C tympanograms are typically caused by retraction of the tympanic membrane secondary to eustachian tube dysfunction. Results of the tympanograms can be a useful adjunct when interpreting all other audiometric testing. For example, someone who fails his/her newborn screen on OAE testing and who presents with a moderate CHL on ABR testing but who has flat, type B tympanogram may have transient middle ear effusion and not permanent hearing loss. Conversely, tympanograms will appear completely normal in an individual with pure SNHL.

Newborn hearing screens have significantly decreased the age at which children are diagnosed as having hearing loss from a mean age of 2.5 years to 2 to 3 months. (3) Irrespective of newborn hearing programs, there are known high-risk criteria for hearing loss (Table 1). These high-risk criteria do not effectively delineate which children will have hearing loss and should not replace newborn hearing

screens. However, they are a useful adjunct in identifying children who have or may be at high risk for developing hearing loss. For those children with any high-risk criteria, formal audiometric evaluation with OAE and full ABR testing should be pursued if there is any concern about hearing loss. It must be remembered that children with progressive onset hearing loss will typically pass their UNHS. Also, children with auditory neuropathy spectrum disorder can present a diagnostic and management challenge. These children typically have present OAEs but absent or abnormal ABRs. Thus, they may pass screening paradigms that use only OAEs. These children can have variable audiologic presentations with hearing loss or hearing ability that do not match what would be expected from their audiometric data. These patients should be closely monitored for speech and language development and may benefit from traditional hearing aids or cochlear implantation. (4)

Children with severe to profound hearing loss will develop their prelingual language skills at a normal pace,

cooing and babbling until ages 6 to 9 months. They will, however, begin to lose these skills and will not progress. Thus, they will present with concerns for speech delay. It also may be reported that the children do not turn their heads in response to sounds and that they do not make eye contact when spoken to. Parents may also note that there may be delays in sitting, crawling, and walking. Paradoxically, some parents and caregivers of children will report that the child can hear, or they may dismiss their child's inattentive behavior as ignoring or "just kids being kids." The intensity of normal conversation is approximately 30 to 50 dB. Normal street noise is approximately 60 dB. Loud conversations (shouting) are approximately 80 dB. Chainsaws are approximately 90 dB. Thus, a child with mild to moderate hearing loss will hear and react to loud shouting and machinery. However, he or she will not be able to hear soft sounds and will often lose the high-frequency consonant sounds that give words meaning. Even children with profound hearing loss may seem to react to loud shouts and bangs because they can feel the vibrations.

Concern about hearing loss should prompt further evaluation, regardless of the age of the child. As detailed above, there are many different types of testing, both objective and subjective, that can accurately determine a child's hearing thresholds, irrespective of age.

TABLE 1. High-Risk Criteria for Children

Birth to Age 28 Days
Family history of congenital or early-onset hearing loss
Congenital infection known to be associated with hearing loss (eg, cytomegalovirus, rubella, herpes, syphilis, toxoplasmosis, varicella)
Craniofacial abnormality
Birth weight <1500 g
Hyperbilirubinemia requiring exchange transfusion
Exposure to ototoxic medications
Bacterial meningitis
Low Apgar scores at birth (<3 at 5 minutes and <6 at 10 minutes)
Prolonged mechanical ventilation (>10 days)
Findings consistent with a syndrome with known hearing loss
Ages 29 days to 2 years
Concern about hearing, speech, language, or other developmental delay
Bacterial meningitis
Neonatal risk factors associated with hearing loss
Head trauma, especially temporal bone fracture
Findings of syndrome associated with sensorineural hearing loss
Exposure to ototoxic medications (eg, aminoglycosides, loop diuretics, cisplatin)
Neurodegenerative disorders
Infectious diseases associated with hearing loss

CAUSES OF HEARING LOSS

The causes of hearing loss can be broadly divided into genetic and acquired and further divided into congenital (present at birth) or progressive. Approximately 50% to 60% of all prelingual hearing loss is genetic. Of this, 70% are nonsyndromic forms of hearing loss. The most common type of nonsyndromic hearing loss is autosomal recessive in 75% to 80%. Furthermore, the most common type of nonsyndromic hearing loss is caused by defects in connexin 26, comprising approximately 50% of all nonsyndromic autosomal recessive hearing loss (Figure 1).

As discussed above, nonsyndromic hearing loss is the most common. Defects have been found in all inheritance patterns. To date, more than 60 different gene loci have been identified that can cause hearing loss. By convention, they are named *DFNA* for autosomal dominant, *DFNB* for autosomal recessive, and *DFNX* for X-linked. The number following the DFN (A, B, or X) identifies the gene locus. *DFNB1*, which is caused by a defect in *GJB2* (also called connexin 26), is the most common of nonsyndromic hearing loss, accounting for approximately 50% of autosomal recessive, nonsyndromic hearing loss. Defects in *GJB2* are caused by a number of different mutations, the most common of which is 35delG.

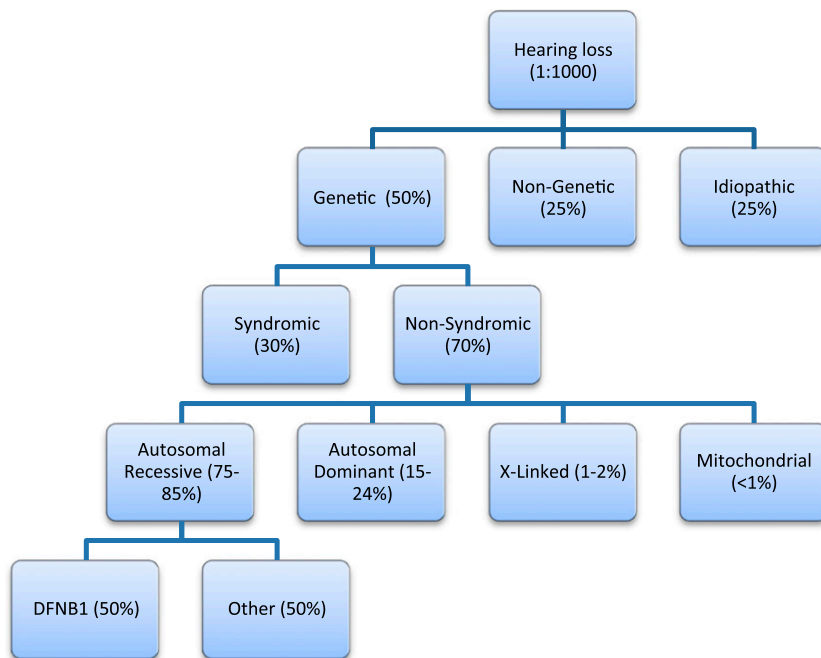


Figure 1. Summary of causes of prelingual hearing loss. Adapted from Smith RJH, Shearer AE, Hildebrand MS, et al. Deafness and hereditary hearing loss overview. In: Pagon RA, Adam MP, Ardinger HH, et al, eds. *GeneReviews*. Seattle: University of Washington; 1993–2014. Available at: <http://www.ncbi.nlm.nih.gov/books/NBK1434/>. Accessed Jan. 5, 2014

Regardless of cause, they all lead to abnormal function of the gap junction protein.

In contrast to nonsyndromic hearing loss where there are typically no other associated physical findings, syndromic hearing losses have various other physical findings that are apparent. Again, syndromic hearing loss is present in all modes of inheritance. More than 500 syndromes that include hearing loss have been described. Most syndromes are rare; Table 2 summarizes some of the more common syndromic causes of hearing loss.

Of the remaining 50% of all cases of congenital hearing loss, 25% are caused by nongenetic factors. The most common acquired form of congenital hearing loss is cytomegalovirus (CMV) infection. Primary infection of the mother and fetus can result in a wide array of outcomes, ranging from no symptoms to multiple sensory deficits. If symptomatic from the CMV infection, there is 50% likelihood that the child will have SNHL to a variable degree. However, even if the infant is asymptomatic from the CMV infection, there is a 10% to 15% chance of developing SNHL. This hearing loss is often progressive and fluctuating. (5) Diagnosis of congenital CMV infection can only reliably be made during the first 3 weeks of life from a urine sample or a cheek swab. If a child does not pass his/her UNHS and there are no other obvious causes, then CMV testing should be considered. Other congenital infections, such as measles, mumps, and rubella, may also lead to hearing loss.

Another infectious origin bears particular comment. Bacterial meningitis, particularly from *Streptococcus pneumoniae*,

may lead to hearing loss. Hearing loss typically occurs early in the infection, can be mild to profound, and can often be prevented or improved with early and prompt antibiotic therapy. In addition, treatment with dexamethasone has been found to significantly reduce the incidence of hearing loss in those with *Haemophilus influenzae* type B bacterial meningitis and to some degree for those with other non-*H influenzae* type B bacterial meningitis. (6) Hearing loss in meningitis occurs in more than 30% of patients. Infections with *S pneumoniae* are associated with hearing loss with even higher frequency, 35%. (7) Patients with a diagnosis of bacterial meningitis should have hearing testing performed as part of their initial workup—once they are stable for this testing. Although hearing loss typically occurs early, it may present later and may be progressive. Therefore, audiologic follow-up is recommended. Patients should have follow-up hearing testing 4 to 8 weeks after discharge and then at 6 and 12 months after infection. (8) If there is hearing loss associated with bacterial meningitis, imaging studies should be promptly performed because there is a high association with cochlear ossification—an inflammatory process that obliterates the structures of the cochlea, particularly the scala tympani. Magnetic resonance imaging can be particularly helpful in early identification of this progressive destruction of the cochlea. If ossification is identified, early cochlear implantation may be advocated.

Other causes of acquired hearing loss include trauma, particularly temporal bone trauma. Fractures through the cochlea and vestibule can result in severe to profound

TABLE 2. **Common Syndromic Causes of Hearing Loss**

CAUSE	DESCRIPTION
Autosomal dominant	
Waardenburg syndrome	Most common type of autosomal dominant syndromic hearing loss 4 types, all with varying degrees of nonprogressive SNHL and pigmentary abnormalities Type 1: dystopia canthorum (lateral displacement of inner canthi of eyes) present Type 2: dystopia canthorum absent Type 3: limb abnormalities Type 4: Hirschsprung disease present
Stickler syndrome	Progressive SNHL Cleft palate present and spondyloepiphyseal dysplasia also may be present Often have severe myopia which increases risk for retinal detachment
Branchiootorenal syndrome	Second most common autosomal dominant hearing loss Variable degree of SNHL, CHL, or mixed hearing loss. Abnormalities in first and second branchial structures, resulting in preauricular pits, malformations of external ear, abnormalities of the ossicles, branchial cleft cysts or fistulas, and renal abnormalities. Renal involvement may be absent
Neurofibromatosis types 2	Hallmark is bilateral vestibular schwannomas Definitive diagnosis made with MRI Extremely rare in children, hearing loss often begins in third decade
Autosomal recessive	
Usher syndrome	Most common autosomal recessive syndromic hearing loss Most common cause of blindness or deafness (multisensory impairment) Blindness secondary to retinitis pigmentosa Early specialized eye examinations (electroretinography) can pick up disease as early as ages 2-4 years May also have vestibular dysfunction Type 1: congenital severe to profound SNHL and abnormal vestibular function Type 2: congenital mild to severe SNHL and normal vestibular function Type 3: progressive hearing loss and progressive loss of vestibular function
Pendred syndrome	Variable SNHL and euthyroid goiter Often have enlarged vestibular aqueduct and cochlear abnormalities on CT
Jervell and Lange-Nielsen syndrome	Congenital severe to profound SNHL Prolonged QT interval that may lead to syncopal episodes or sudden death Consider screening ECG If positive family history, consider cardiac evaluation
X-linked	
Alport syndrome	Progressive hearing loss Progressive glomerulonephritis Ophthalmologic abnormalities (anterior lenticonus)

Abbreviations: CHL=conductive hearing loss; CT=computed tomography; ECG=electrocardiography; MRI=magnetic resonance imaging; SNHL=sensorineural hearing loss.

SNHL. Fractures through the ossicles can disarticulate these bones, causing moderate CHL. Acoustic trauma (noise-induced hearing loss) can also cause permanent hearing loss. Audiograms of individuals affected by noise-induced hearing loss typically display a distinct pattern with a high-frequency dip (notch) at 4000 Hz. Certain medications are also known to be ototoxic. These medications, which include aminoglycoside antibiotics, loop diuretics, and cisplatin, should be avoided if possible, and hearing should be monitored if these medications are used.

Last, hearing loss may be caused by otitis media with effusion. This is the most common cause of childhood hearing loss, although it is often not included on lists because of its potentially transient nature. Middle ear effusion is exceedingly common in association with otitis media. In fact, 40% of all ears with resolved otitis media will have persistent fluid at 1 month, 20% at 2 months, and 10% at 3 months. (9) It is important to ask about subjective hearing when evaluating those with otitis media. If there is concern regarding hearing, objective testing should be performed.

For those individuals with persistent chronic otitis media with effusion for greater than 3 months who have an associated hearing loss, surgical management with myringotomy and tympanostomy tubes is a recommended treatment. (10) If, however, there is no hearing loss present, no surgical intervention is recommended because persistent effusion in otherwise healthy children does not cause developmental impairment. (11)

MANAGEMENT OF HEARING LOSS IN CHILDREN

Once hearing loss is identified, steps should be taken to minimize the duration of hearing loss and/or maximize the function of the residual hearing or cochlear function. In addition, efforts should be made to militate against deficits in other sensory systems. The overall goals, especially in children, are to minimize the effect that the hearing loss will have on speech and language development and to provide appropriate strategies for communication. In general terms, the more pronounced the hearing loss and the longer the duration of hearing loss without intervention, the more significant the effect on speech and language development. All the management strategies and considerations are tailored to the specific condition of the child. Children with permanent hearing loss should undergo an ophthalmology evaluation to rule out ocular disease. Children with moderate and greater bilateral SNHL should undergo screening electrocardiography to look for prolonged QT associated with hearing loss. (12) If there is a family history of sudden death, full cardiologic evaluation should be performed. Decisions regarding genetic testing and imaging should be made with the patient and family.

Medical care for hearing loss in children can include appropriately chosen antibiotic therapy for otitis media. In cases of prolonged otitis media with effusion with associated hearing loss, antibiotics are unlikely to resolve the problem. Myringotomy and tympanostomy tubes may be required for removal of the effusion and improved middle ear aeration.

For children with mild to severe hearing loss, amplification may be a suitable option. Modern hearing aids can be fit to children as early as age 3 months. The amplification on modern digital aids is adjustable to fit the patient's hearing loss curve. Early amplification, even for those with mild loss, is beneficial in that it gives them access to the full range of speech and language sounds. This and the longer duration of use that comes with early amplification lead to better speech and language development. (13)

Beyond providing hearing impaired children with equipment, it is important to make sure that the children are getting the appropriate services to help foster their growth

and development. Often, simple seating rearrangements, such as preferential seating in the front of the class and classroom FM systems, can make a significant difference.

Certain conditions that cause CHL are amenable to surgical management. As mentioned above, CHL from chronic otitis media with effusion can be treated with tympanostomy tubes. Certain ossicular abnormalities can be repaired or replaced with prosthetic ossicles. Cholesteatoma can cause erosion of the ossicles, which must be managed surgically. Bone-anchored hearing aids and middle ear implants are placed surgically.

If these options fail to provide adequate hearing for speech and language development and for communication then alternate options must be considered. Decisions need to be made concerning the desired mode of communication—manual (eg, sign language) or auditory or verbal. In appropriately selected patients who desire auditory or verbal communication, have significant hearing loss, and do not get adequate return from conventional amplification, cochlear implantation may be an option. In this procedure, a small electrode array is surgically implanted within the cochlea. This array is able to stimulate the cochlear nerve directly.

Summary

- On the basis of strong research, universal newborn screening should be performed before age 1 month with repeat or follow-up testing for those who do not pass performed before age 3 months and intervention started before age 6 months.
- On the basis of strong research and consensus statement, tympanostomy tubes should be considered for individuals with bilateral persistent middle ear effusion for 3 months or greater and a documented conductive hearing loss.
- On the basis of consensus statement, all children with suspected hearing loss should have an age appropriate hearing test.
- On the basis of strong research, the most common form of congenital hearing loss is genetic. Most of this is nonsyndromic hearing loss.

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Parent Resources from the AAP at HealthyChildren.org

- <http://www.healthychildren.org/English/health-issues/conditions/ear-nose-throat/Pages/Hearing-Loss.aspx>
- Spanish: <http://www.healthychildren.org/spanish/health-issues/conditions/ear-nose-throat/paginas/hearing-loss.aspx>
- <http://www.healthychildren.org/English/health-issues/conditions/ear-nose-throat/Pages/Hearing-Loss-When-to-Call-the-Pediatrician.aspx>
- Spanish: <http://www.healthychildren.org/spanish/health-issues/conditions/ear-nose-throat/paginas/hearing-loss-when-to-call-the-pediatrician.aspx>
- <http://www.healthychildren.org/English/health-issues/conditions/ear-nose-throat/Pages/Acoustic-Trauma-Hearing-Loss-in-Teenagers.aspx>
- Spanish: <http://www.healthychildren.org/spanish/health-issues/conditions/ear-nose-throat/paginas/acoustic-trauma-hearing-loss-in-teenagers.aspx>
- English only: <http://www.healthychildren.org/English/health-issues/conditions/ear-nose-throat/Pages/Music-How-Loud-is-Too-Loud.aspx>

PIR Quiz

1. The goal of the universal newborn hearing screening (UNHS) program is which of the following:
 - A. Identify all children with moderate hearing loss by age 2 months and initiate intervention by age 4 months.
 - B. Identify all children with permanent hearing loss by age 3 months and initiate intervention by age 4 months.
 - C. Identify all children with moderate hearing loss by age 1 month and initiate intervention by age 4 months.
 - D. Identify all children with permanent hearing loss by age 3 months and initiate intervention by age 6 months.
 - E. Identify all children by age 1 month and initiate intervention by age 3 months.
2. A 3-year-old girl presents to your clinic with a 2-month history of otitis media, resistant to amoxicillin and amoxicillin clavulanate. On otoscopic examination, she has nonerythematous tympanic membranes, which are dull and do not move on insufflation. You perform a tympanogram in your office. Which of the following tympanograms would be most consistent with an ear effusion?
 - A. Type A.
 - B. Type B.
 - C. Type C.
 - D. Type D.
 - E. Type E.
3. An infant with auditory neuropathy spectrum disorder presents to your clinic. You know this disorder involves a defect in the signal transmission of sound from the inner ear to the brain. Which of the following hearing screen results would you expect in this patient?
 - A. Present otoacoustic emissions and absent auditory brainstem response.
 - B. Present otoacoustic emissions and present auditory brainstem response.
 - C. Absent otoacoustic emissions and absent auditory brainstem response.
 - D. Absent otoacoustic emissions and present auditory brainstem response.
 - E. Otoacoustic emissions and auditory brainstem response are not reliable in this disorder.
4. A 38-week-old female infant weighs 2100 g. On physical examination, she has microcephaly and a palpable liver and spleen. She exhibits petechiae on her face and trunk. You strongly suspect congenital cytomegalovirus (CMV). In counseling the parents, you inform them of the progressive sensorineural hearing loss associated with congenital CMV. Furthermore, you inform them the likelihood of sensorineural hearing loss in their infant is:
 - A. 10%.
 - B. 25%.
 - C. 50%.
 - D. 75%.
 - E. 100%.
5. A 7-year-old boy is admitted to the hospital with fever and rash and is treated for bacterial meningitis. He passes a hearing screen in the hospital before discharge. As you prepare his discharge consultations, you schedule him for follow-up audiology testing. Which of the following schedule is most appropriate?
 - A. Follow-up hearing tested at 3 months, 6 months, and 9 months after discharge.
 - B. Follow-up hearing tested at 6 months and 1 year after discharge.
 - C. Follow-up hearing tested at 1 month, 6 months, and 12 months after discharge.
 - D. Follow-up hearing tested at 1 month and 12 months after discharge.
 - E. No follow-up hearing tests indicated unless clinical symptoms of hearing loss.

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Pediatric Hearing Loss
Christopher R. Grindle
Pediatrics in Review 2014;35;456
DOI: 10.1542/pir.35-11-456

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understood, the hormone ghrelin, the growth hormone secretagogue receptor, insulin-like growth factors, and insulin all have the potential to mediate linear growth.

In conjunction with bone age, precocious puberty can be diagnosed with Tanner staging and assessing testicular volume in males, with a postpubertal testicular volume being greater than 4 mL. Patients with late-onset congenital adrenal hyperplasia present with precocious puberty, tall stature, and an increased 17-hydroxyprogesterone level in the blood. Growth hormone excess is rare in children, yet when present is usually due to pituitary adenomas. Growth hormone excess before epiphyseal fusion results in tall stature and markedly increased height velocity. If it occurs after epiphyseal fusion, adolescents may also have signs of acromegaly, such as coarsening of facial features, enlarged jaw, and distal body overgrowth, including large hands and feet.

There are other rare genetic syndromes associated with tall stature, all of which have significant clinical stigmata that should be apparent on physical examination. Sotos syndrome should be considered if the bone age is accelerated and the child has accompanying symptoms of facial flushing, frontal bossing, and a narrow face and head. Beckwith-Wiedemann syndrome is considered if hypoglycemia is present at birth, along with anterior abdominal wall defects and macroglossia. Marshall-Smith syndrome is associated with unusually rapid physical growth, abnormal facies, and respiratory issues.

A pediatric health care professional can diagnose most causes of tall stature with a careful history and physical examination. If suspicion is high for disease or the cause is unclear, the clinician should consider radiographic bone age and focused laboratory testing. Although the social stigma behind tall stature has decreased, clinicians must still be

aware of potential pathologic causes of tall stature that would warrant intervention.

COMMENTS: This In Brief emphasizes the importance of accurate height measurements. My experience is that height measurement can often be inaccurate, especially in supine measurements in young children or very active children who have difficulty standing still. When transitioning from supine measurements to standing measurements in toddlers, there may appear to be a false perception of a decrease in height because standing height is often shorter than supine length. Accurate measurement of upper to lower body segment ratios is important in identifying several syndromes and can be performed in the following way. To measure lower body segment, measure from the symphysis pubis of the patient to the floor. The upper segment can be calculated by subtracting the lower segment from the total height. Measurement of arm span is best accomplished by measuring from the tip of the middle finger of one hand, with arms outstretched at a 90° angle to the tip of the middle finger of the other hand.

A recent study of self-reported adult height measurements in parents found that men have a tendency to overestimate their heights but women's self-report is more accurate. When looking at estimating the other parent's height, women were more likely to overestimate the men's height, whereas men were more accurate in estimating women's height. Their conclusions were that attempts should be made to use measured heights of parents when the results would have an effect on diagnostic workup or treatment of patients with tall stature.

– Janet Serwint, MD
Consulting Editor, *In Brief*

CME Quiz Correction

A revision to the order of questions in the November 2014 article “Pediatric Hearing Loss” (Grindle CR. *Pediatrics in Review*. 2014;11:456, doi: 10.1542/pir.35-11-456) led to an error in the online answers. The correct order for the article's answers should be: 1. D; 2. B; 3. A; 4. C; 5. C. The online version of the quiz and answer key have been corrected. The journal regrets the error.

ANSWER KEY FOR DECEMBER 2014 PEDIATRICS IN REVIEW:

Borrelia burgdorferi (Lyme Disease): 1. E; 2. A; 3. D; 4. D; 5. C.

Complementary and Integrative Methods in Fibromyalgia: 1. D; 2. E; 3. B; 4. C; 5. E.

Respiratory Syncytial Virus Infection and Bronchiolitis: 1. C; 2. A; 3. B; 4. E; 5. B.



Pediatrics in Review

An Official Journal of the American Academy of Pediatrics

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The online version of this article, along with updated information and services, is located on the World Wide Web at:

<http://pedsinreview.aappublications.org/content/35/11/456>

An erratum has been published regarding this article. Please see the attached page for:

<http://pedsinreview.aappublications.org/content/35/12/539.full.pdf>

Data Supplement at:

<http://pedsinreview.aappublications.org/content/suppl/2014/11/03/35.11.456.DC1>

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