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Hearing Loss in Children

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Author Disclosure

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Objectives After completing this article, readers should be able to:

1. Determine the difference between conductive and sensorineural hearing loss.
2. Discuss the congenital and acquired causes of hearing loss.
3. Delineate the most appropriate times for screening, diagnosis, and interventions for hearing loss.
4. List the risk factors for hearing loss in all children.
5. Recognize the need for identification of hearing loss and prompt intervention in all children.

Case Studies

Case 1

During his routine health supervision visit, a healthy 2-year-old boy is noted to have an isolated speech delay. There is no family history of hearing problems. On questioning about the child's newborn hearing screen, the mother says, "They said something about repeating it, but we were pretty busy, so we never got around to it. Since no one mentioned it again, I didn't think it was important." The child is referred to audiology and diagnosed with moderate bilateral sensorineural hearing loss. A hearing aid and intensive speech therapy are recommended.

Case 2

A 4-year-old girl presents for evaluation of behavioral problems. Her parents are worried about attention-deficit disorder. They report that she frequently ignores their requests, does not seem to pay attention, and has trouble following instructions. In addition, she has not outgrown her temper tantrums like other children with whom she plays. She can understand preschool activities when the whole class is doing them, but she has the most difficulty with directions given to her individually. She has had frequent medical visits for acute otitis media. Physical examination reveals tympanic membranes that are retracted, with some scarring bilaterally. In-office tympanometry and audiometry suggest a moderate conductive hearing loss. Due to concerns that her behavioral problems are related to her hearing loss, she is referred to an otolaryngologist for consideration of tympanostomy tube placement.

Abbreviations

AAP:	American Academy of Pediatrics
ABR:	auditory brainstem response
CHL:	conductive hearing loss
EHDI:	early hearing detection and intervention
OME:	otitis media with effusion
OAE:	otoacoustic emission testing
SNHL:	sensorineural hearing loss
UNHS:	universal newborn hearing screening

Introduction

As demonstrated by these cases, childhood hearing loss can be a debilitating condition that affects 1 to 6 per 1,000 newborns to a significant degree. Such high prevalence warrants close attention because it is widely acknowledged that the first 36 months after birth represent a critical period in cognitive and linguistic development. (1) Although early identification of children who are deaf and hearing-impaired allows such children to approach their hearing peers in language skills and academic performance, those who are identified late often never can reach the same level of skill.

Targeted hearing screening used to be recommended as a method for early detection of hearing loss in neonates and

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infants. However, past studies documented that only 50% of children who had hearing loss were identified effectively by targeted screening and that the average age of hearing loss detection with this method was as late as 14 months. To initiate treatment as rapidly as possible and avoid preventable disability, every state has now mandated universal newborn hearing screening (UNHS) programs, a crucial part of early hearing detection and intervention (EHDI).

The American Academy of Pediatrics (AAP) recommends that congenital hearing loss be detected by 1 month, diagnosed definitively by 3 months, and receive intervention by 6 months of age for all infants born in the United States. Because hearing loss can be acquired at any age, it is critical for primary care practitioners to assess risk factors for hearing loss at each visit and determine which, if present, warrant a referral for full audiologic evaluation. In addition, all children ages 4 years and older who are developmentally able to cooperate with conventional audiometry should be screened regularly for hearing loss. Early intervention in all cases of hearing loss can reduce disability significantly and improve both linguistic skills and cognitive development in the hearing-impaired population.

The Mechanical Basis of Hearing

The ear is divided anatomically into outer, middle, and inner segments (Figure). The pressure waves, which form the basis for sound, are captured by the pinna and directed through the external auditory meatus. These sound waves affect the tympanic membrane, which forms the boundary between the outer and middle ear. Because effective vibration of the tympanic membrane requires that the pressure on either of its sides be equal, the middle ear is ventilated by the pharyngotympanic (eustachian) tube, which runs from the anterior wall of the middle ear to the nasopharynx.

The resultant vibrations are transferred through the ossicular chain (ie, the malleus, incus, and stapes) to the oval window, a section of the cochlea that forms the beginning of the inner ear. Here the mechanical vibrations of the ossicles are converted into fluid pressure waves that impart movement to hair cells lying along the basilar membrane and within the organ of Corti. This motion subsequently is transduced into neural impulses. Sound intensity is determined by the number of hair cells firing, with louder noises triggering a greater number of neurons. Frequency is coded by both the incidence of neurons firing and the pattern of their activation along the basilar membrane. High-frequency sound causes greater vibration at its base and low frequency at its apex.

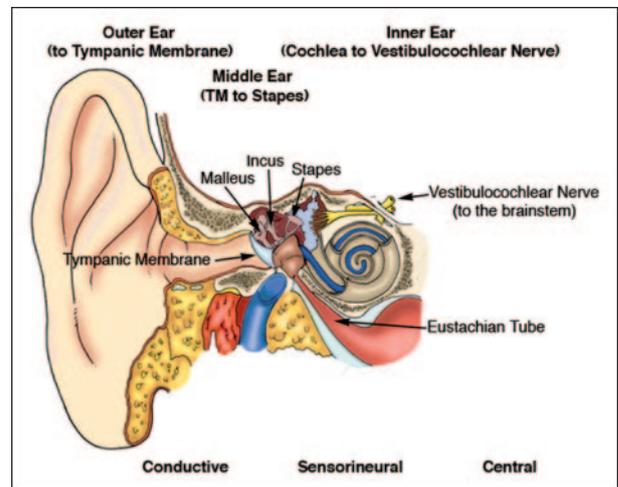


Figure. Anatomy of the ear.

The neural impulses are carried along the vestibulocochlear nerve to the brainstem and directed to various parts of the brain for additional processing. A defect in any part of this system can lead to hearing loss.

Types of Hearing Loss

Hearing loss is divided into conductive, sensorineural, mixed, and central types. Conductive hearing loss (CHL) is far more common in children and results from interference with the mechanical transmission of sound through the external and middle ear. Sensorineural hearing loss (SNHL) results from a failure to transduce vibrations to neural impulses effectively within the cochlea or transmit these impulses down the vestibulocochlear nerve. Mixed hearing loss involves a combination of these two types, usually due to damage throughout the middle ear and the inner ear. Finally, central hearing loss refers to defects in the brainstem or higher processing centers of the brain. Both CHL and SNHL may be caused by a wide variety of congenital and acquired factors (Table 1).

Congenital Hearing Loss

The capability of infants to categorize the auditory world from a very early age and the existence of a critical period for language acquisition, lasting up to approximately 36 months of age, make it vital to maximize a child's exposure to spoken language. Hearing loss early in development can be highly detrimental to the linguistic and cognitive development of an afflicted child. Yet, early intervention in the hearing-challenged child may largely offset such developmental losses.

Studies have determined that children enrolled in

Table 1. Causes of Hearing Loss

Conductive Hearing Loss	Sensorineural Hearing Loss
<p>Congenital</p> <ul style="list-style-type: none"> • Microtia/atresia • Tympanic membrane abnormalities • Ossicular malformations 	<ul style="list-style-type: none"> • Genetic disorders (syndromic, connexin 26, mitochondrial) • In utero infections (cytomegalovirus, measles, mumps, rubella, varicella, syphilis) • Anatomic abnormalities of the cochlea or temporal bone • Exposure to ototoxic drugs during pregnancy (alcohol, isotretinoin, cisplatinum) • Hyperbilirubinemia
<p>Acquired</p> <ul style="list-style-type: none"> • Infection (acute otitis media, otitis externa, ossicular erosion) • Otitis media with effusion • Foreign body (including cerumen) • Cholesteatoma • Trauma (ossicular disruption, tympanic membrane perforation) 	<ul style="list-style-type: none"> • Infections (bacterial meningitis, measles, mumps, rubella, Lyme disease) • Trauma (physical or acoustic) • Radiation therapy for head and neck tumors • Neurodegenerative or demyelinating disorders (Alport, Cogan syndromes)

EHDI programs perform significantly better than their later-detected peers on tests of vocabulary skills and intellectual development, to the point of approaching children whose auditory capacity is unimpaired. If intervention is implemented by 6 months of age, gains of as much as 20 to 40 percentile points on academic measures may be recorded. (1) This finding highlights how critically important it is that every child's hearing be screened repetitively beginning at birth, ensuring the rapid detection of all hearing problems and implementation of interventions as quickly as possible.

Causes

The causes of SNHL are much more widespread than those of CHL (Table 2). Among the most prominent are family history, in utero infections, severe hyperbilirubinemia, respiratory distress, and prolonged mechanical ventilation.

Genetic factors are a significant cause of hearing loss. Many genetic disorders represent complex syndromes, of which hearing loss is only one of multiple findings. In total, more than 500 forms of syndromic hearing loss have been recorded. Waardenburg syndrome, which is characterized by hypertelorism and pigmentary abnormalities, usually presents with hearing loss of varying degrees. Usher syndrome, a leading cause of deafness and blindness, presents with SNHL compounded by progressive retinal damage and loss of vestibular function. Pendred syndrome is another condition in which SNHL is associated with goiter and enlarged vestibular aqueducts. Alport syndrome is an inherited condition that

may present with progressive SNHL as well as progressive nephritis.

The most common nonsyndromic form of genetic

Table 2. Risk Factors for Hearing Loss

Before 28 Days of Age

- Family history
- In utero infections
- Ear and other craniofacial anomalies
- Anomalies associated with syndromic sensorineural hearing loss
- Hyperbilirubinemia requiring exchange transfusion
- Birthweight <1,500 g
- Apgar score <3 at 5 minutes or <6 at 10 minutes
- Mechanical ventilation, especially for more than 10 days
- Exposure to ototoxic medications (eg, gentamicin)

After 28 Days of Age

- Parental concern about hearing, speech, or developmental delay
- Persistent otitis media with effusion for more than 3 months
- History of head trauma
- History of bacterial meningitis
- Diagnosis of disorders that can lead to hearing loss (eg, neurodegenerative disorders, demyelinating disorders)
- Diagnosis of syndromes associated with hearing loss (eg, Alport syndrome)

hearing loss involves mutations in the gene encoding the connexin 26 (Cx26) protein. Disorders associated with Cx26 are recessive. SNHL associated with Cx26 is moderate-to-severe, congenital, and bilateral most commonly, but it also may present as mild, progressive, or asymmetric hearing loss. Mitochondrial disorders also are a significant cause of both syndromic and nonsyndromic genetic hearing loss.

Certain maternal infections can affect the immature auditory systems of the developing fetus adversely. Cytomegalovirus is the infection associated most commonly with congenital SNHL. Hearing loss from cytomegalovirus may be progressive to the point that an infant who initially passes a newborn hearing screening test may present with profound hearing loss 1 year later. Maternal measles, mumps, and rubella also are important risk factors for congenital hearing loss. Children who were exposed to these diseases in utero, as well as those who develop them during childhood, should be monitored carefully.

Anatomic disorders can cause congenital CHL. Malformations of the external ear can range from absence or blockage of the external ear canal (atresia) to only a slightly reduced pinna and external ear canal (microtia). The resulting lack of the external ear apparatus either restricts or completely arrests the transfer of sound waves through the ear. Some children may present with congenital malformations of the ossicular chain, which block the transmission of sound through the middle ear.

Universal Newborn Hearing Screening Before 1 Month of Age

Hearing loss cannot be diagnosed in an infant through simple observation because hearing-impaired infants achieve early language milestones (eg, smiling, cooing, babbling, gesturing) at the normal time. Only objective hearing tests may provide an accurate assessment of hearing loss in the newborn. Therefore, the Joint Committee on Infant Hearing of the AAP recommends UNHS for all newborns before 1 month of age.

Initial screening generally is conducted in the newborn nursery before discharge. In cases of home births or when the initial “screen” was missed, a hearing test must be conducted within the first postnatal month. Screening is conducted by using evoked otoacoustic emission (OAE) testing, auditory brainstem response (ABR), or a combination of the two.

OAE detects the evoked sound from the cochlea in response to clicks or tones; ABR measures the electroencephalographic waveform response from the vestibulocochlear nerve. ABR has the potential to detect hearing

loss due to auditory neuropathy, a condition to which the newborn intensive care unit population is particularly prone, in addition to hearing loss resulting from middle and inner ear disorders. Furthermore, ABR is minimally affected by external or middle ear debris. A two-stage screening, in which ABR is used to confirm abnormal OAE results, yields the lowest number of false-positive results, thereby reducing referral rates and decreasing parental anxiety. Test accuracy increases rapidly with age due to such factors as improved tympanic membrane mobility and the reduction of middle ear fluid. Pediatricians, therefore, may reassure concerned parents that an initial positive screening result can be “false-positive.” However, they also must stress the critical importance of additional evaluation to determine if hearing loss is present so that necessary interventions occur promptly.

Full Audiologic Evaluation by 3 Months of Age

Any infant who fails the initial screen should be referred to an audiologist for a full evaluation no later than by 3 months of age. This evaluation should include a wide range of diagnostic tests to confirm and determine the nature of the hearing loss. Table 3 lists available hearing tests for all age groups. Such tests include physical examination of the outer ear, OAE, diagnostic ABR, tympanometry, and behavioral observation audiometry.

One of the major objections that has been raised to UNHS is the infeasibility of effective follow-up. The Task Force on Newborn and Infant Hearing of the AAP set 95% as the minimal threshold of follow-ups for a UNHS program to be considered successful. A dearth of audiologic evaluations after failed screening represents a waste of resources and the potential for missing the opportunity to intervene early for affected children. The results of the Colorado Newborn Hearing Screening Project, which took place between 1992 and 1999, show achieving effective referral rates to be the most difficult phase of the process. Although screening rates, referral rates, and false-positive rates all achieved or closely approached AAP recommendations, only 76% of infants failing the initial screening received documented follow-up evaluation. (2)

These initial results are discouraging, but many efforts are underway to increase the follow-up rate. Most importantly, when an abnormal screening test result is detected, practitioners need to explain to parents the test results and the importance of evaluation by audiology. It also is critical for the primary care physician to ensure that this recommended evaluation takes place. One obvious difficulty is communication among birthing hospitals,

Table 3. Testing Methods for the Diagnosis and Characterization of Hearing Loss

Testing Method	Functionality	Test Length (min)	Appropriate Age	Advantages	Disadvantages
Tympanometry	External ear canal is sealed with probe tip. Ear canal pressure is varied while sound reflection by the tympanic membrane is measured. An effective assessment of middle ear pressure and function is provided.	5	All ages except newborns	<ul style="list-style-type: none"> Useful for the diagnosis of middle ear disorders such as ossicular disruption, perforated tympanic membrane, and otitis media with effusion Can help to distinguish CHL from SNHL causes (should be normal in SNHL) 	<ul style="list-style-type: none"> Not a test of hearing, only of tympanic membrane function Expected to be normal in the presence of SNHL or auditory neuropathy Does not assess inner ear function Not valuable for neonates
Auditory brainstem response (ABR)	Stimuli are delivered through earphones: <ul style="list-style-type: none"> Click stimuli for screening ABR Tone burst stimuli for diagnostic ABR The resulting electroencephalographic waveform in the auditory nerve and brainstem pathways is recorded via skin electrodes, electroencephalographic amplifiers, and computer averaging. The lowest stimulus level that results in a response gives the child's perceptual threshold.	20 for screening 60+ for diagnostic	Birth to 9 months	<ul style="list-style-type: none"> Objective test of auditory pathways; no infant participation required Ear-specific results Minimally affected by outer and middle ear debris Effectively screens for auditory neuropathy 	<ul style="list-style-type: none"> Infant must be sleeping because muscle contraction and movement create interference Children >6 months of age may require conscious sedation Does not assess cortical processing of sound Automated ABR only provides pass/fail information Diagnostic ABR requires administration and interpretation by an audiologist
Evoked otoacoustic emissions (OAE)	Click and tone bursts are presented to each ear through the OAE probe. The evoked sound generated by outer hair cell response is recorded. The presence of an OAE indicates a pass.	10	All ages	<ul style="list-style-type: none"> Objective test of auditory pathways; no infant cooperation required Ear-specific results Rapid testing time Relatively inexpensive 	<ul style="list-style-type: none"> Infant must remain quiet during test Does not assess cortical processing of sound Significantly affected by inner and middle ear debris No assessment of auditory neuropathy
Behavioral observation audiometry	The infant is seated in a soundproof room and presented with warbled pure-tones, speech, or white and narrow-band noise through loudspeakers. Responses such as the auralpalpebral reflex, startle responses, and head turning are recorded.	30	Birth to 6 months	<ul style="list-style-type: none"> Does not require infant conditioning or reinforcement Assesses auditory function and perception 	<ul style="list-style-type: none"> High-intensity stimulus required to response Requires skilled examiner; bias can be a factor Does not assess ears individually (continued)

Table 3. Testing Methods for the Diagnosis and Characterization of Hearing Loss—Continued

Testing Method	Functionality	Test Length (min)	Appropriate Age	Advantages	Disadvantages
Visual reinforcement audiometry	The child is seated between two speakers and conditioned to look toward the active speaker by means of an animated toy, which is lit when the correct head turn is given. A centering toy is used to have the infant turn the head back to a neutral position. An earplug or earphone can be used to assess each ear individually.	30	9 months to 2.5 years	<ul style="list-style-type: none"> Assesses auditory function and perception Accurate, frequency-specific thresholds can be gained from infants younger than 1 year of age 	<ul style="list-style-type: none"> Requires skilled examiner
Play audiometry	The child is conditioned to perform play activities, such as dropping a block or placing a peg in a board, in response to tones of varying frequencies delivered through headphones or a bone vibrator.	30	2.5 to 4 years	<ul style="list-style-type: none"> Assesses auditory function and perception Ear-specific results 	<ul style="list-style-type: none"> Attention span of child is a potential limiting factor
Conventional audiometry	The child is instructed to raise a hand or push a button when a tone is heard. Tones of varying frequencies are delivered through headphones or a bone vibrator.	30	4 years to adulthood	<ul style="list-style-type: none"> Assesses auditory function and perception Ear-specific results 	<ul style="list-style-type: none"> Requires cooperation from child

CHL=congenital hearing loss, SNHL=sensorineural hearing loss. Adapted from Bachmann and Arvedson. *Pediatr Res*. 1998;19:155-165.

primary care practices, and families. Some hospitals and offices designate one individual to be responsible for tracking all babies whose hearing tests are abnormal. Computer-based tracking systems also are being used to facilitate this process. By taking advantage of a systematic approach, some larger hospitals that have more experience in population-based care have reported follow-up rates approaching those in AAP guidelines. (2)

If any audiologic evaluation confirms hearing loss, other investigations are necessary. Because children who have hearing impairment rely heavily on their other senses, a vision screen and developmental screen should be performed on all children showing any degree of hearing impairment. If a syndrome is suspected, referral to a geneticist should be considered. For children who have SNHL, evaluations to determine the cause should be performed (see Table 3 for studies to consider). For all children in whom hearing loss is established by full audiologic evaluation, intervention must begin as soon as possible and no later than at 6 months of age.

Intervention for Hearing Loss by 6 Months of Age

The goal of EHDI programs is for all infants to receive targeted interventions by 6 months of age to permit them to be exposed maximally to spoken language and have the greatest likelihood of developing at the same rate as their peers who hear normally. Early intervention services can help children to make the most of their hearing devices and to develop visual communications skills to augment their hearing. Such services maximize the ability of children who have hearing loss to develop well and succeed both academically and socially. Depending on the degree and type of hearing loss, a broad range of hearing devices exists to circumvent the defective part of the hearing pathway.

To address mild-to-moderate hearing loss, external devices are available that primarily amplify sound. In general, the larger devices provide more amplification but also are more visible to others. Individuals often choose binaural hearing aids, which improve the ability to determine the direction of sound, hear soft noises, and ignore background noise. Many amplification aids can be linked to hearing aid-compatible telephones or other assistive listening devices. The fitting process for all such devices is complex and is managed by an audiologist.

A variety of implantable devices are available to address SNHL, including cochlear implants, many of which are less conspicuous than external hearing devices. Cochlear implants work by converting sound into electrical impulses that stimulate the vestibulocochlear nerve. In

this way, they bypass the entire sound conduction pathway in the external and middle ears and the sound translation that occurs in the cochlea. Early implantation allows exposure to sound during the critical period in development and leads to improved hearing, language, and speech development. Implantation occurs through surgery, which may not be covered by insurance. After the surgery, intensive training facilitated by audiologists and speech pathologists is needed to help the child learn how to understand this new type of sound input. (3)

Acquired Hearing Loss

Because children can acquire hearing loss at any age, it is critical for practitioners to be aware of its warning signs, which may be either the inciting event of hearing loss or its manifestations. Details are summarized in the tables on causes (Table 1), risk factors (Table 2), testing (Table 3), and degrees and effects of acquired hearing loss (Table 4).

Cause

The most common cause of CHL in children is otitis media with effusion (OME). This condition involves an accumulation of fluid in the middle ear due to altered eustachian tube function. Although CHL may present

after a case of acute otitis media, signs of an acute ear infection usually are absent. OME may be diagnosed with a combination of visual inspection and tympanometry. Because middle ear fluid restricts tympanic membrane mobility, pneumatic otoscopy is a helpful diagnostic tool, and tympanometry may provide a useful adjunct when diagnosis is uncertain. Cerumen impaction, otitis externa, and foreign body blockage of the external ear canal also are physical barriers that cause CHL in children.

Cholesteatoma is another possible source of CHL. This disorder is characterized by a benign tumor of skin in the middle ear that may disrupt the ossicular chain as it grows. Cholesteatomas may be congenital, but often they are caused by poorly healed perforations of the tympanic membrane or negative pressure in the middle ear. In addition to hearing loss, patients often experience drainage from the ear, pain, numbness, or dizziness. Symptoms tend to worsen gradually over time as the tumor slowly fills a greater volume of the middle ear. However, some children can be asymptomatic other than experiencing mild hearing loss, which can be undetected without screening.

Trauma may cause both acquired CHL and SNHL. The forces involved in physical blows to the head may

Table 4. Degree and Effects of Hearing Loss

Degree of Hearing Loss	Hearing Level (dB)	Effects
Normal	0 to 15	<ul style="list-style-type: none"> • Can detect all aspects of speech
Minimal	16 to 25	<ul style="list-style-type: none"> • May miss up to 10% of speech • May respond inappropriately • Peer social interaction affected
Mild	26 to 40	<ul style="list-style-type: none"> • May miss up to 50% of speech • May be labeled as "behavior problem" and "poor listener"
Moderate	41 to 55	<ul style="list-style-type: none"> • May miss 50% to 100% of speech • Speech quality likely to be poor • Vocabulary is limited • Compromised communication ability • Low self-esteem possible
Moderate/Severe	56 to 70	<ul style="list-style-type: none"> • 100% of normal volume speech lost • Delayed speech and poor intelligibility • Social isolation likely
Severe	71 to 90	<ul style="list-style-type: none"> • Loud voices only heard within 12 inches of ear • Delayed speech and language if loss is prelingual • Declining speech abilities and atonal voice if loss is postlingual
Profound	90+	<ul style="list-style-type: none"> • Sound vibrations felt rather than heard • Visual cues primary for communication • Peer group of hearing impaired children preferred

Adapted from Bachmann and Arvedson. *Pediatr Rev.* 1998;19:155–165.

disrupt the ossicular chain. Temporal bone fractures, especially transverse fractures, and penetrating trauma into the inner ear also may damage the delicate structure of the cochlea. All children presenting with signs of hearing loss after physical trauma should receive a full evaluation to determine the precise nature of the damage and plan interventions. Acoustic trauma is another major concern because it is one of the most common causes of high-frequency hearing loss in ranges greater than 4,000 Hz. Exposure to continuous noise greater than 85 dB or sudden noise greater than 140 dB is defined as the limit of damaging noise levels. Severity of hearing loss is related both to the intensity of the sound and the daily duration of exposure. Although injury can occur from a single blast injury, such as from firecrackers, more common are repeated exposure injuries from firearms, power tools, and amplified music.

Among acquired infections, bacterial meningitis is the most common cause of childhood SNHL, ranging from mild to profound, depending on the severity of illness. All children who have meningitis should have a hearing test as soon as possible. Hearing loss usually appears early in the disease but also may progress over time. Other possible infectious causes of acquired SNHL include Lyme disease, Fifth disease, and syphilis.

Testing

Tympanometry is not a test of hearing but can be helpful in augmenting findings on the clinical examination by assessing the mobility of the tympanic membrane. Depending on a child's developmental level, various hearing tests are available to assess both auditory function and perception. However, in those younger than 4 years of age, such tests require the experience of an audiologist. Table 3 has a complete list of hearing tests available for all age groups.

No simple and effective screening test is recommended for UNHS between the newborn screen and age 4 years, when conventional audiometry becomes feasible. Thus, it is important for primary care practitioners to be vigilant about assessing risk factors at every visit. Children who have any of the risk factors for hearing loss should be referred to an audiologist for specialized evaluation of their hearing. The onset of hearing loss after age 5 years has a smaller, but still significant, impact on language development. Because only 50% of children who have hearing loss are identified by the use of risk indicators, all children should have periodic objective assessments of their hearing.

Effects of Hearing Loss

Acute otitis media alone has not been associated with any long-term problems with language development. However, untreated chronic hearing loss can affect language development, leading to speech delay and difficulties with articulation. Such loss also can manifest as a child asking people to repeat themselves, not hearing instructions, ignoring a speaker, or listening to loud television or music. In extreme cases, children can present with behavioral problems or other issues that are manifestations of the effect that hearing loss has on their ability to communicate.

Because language serves as the foundation for children to learn other cognitive skills, the effects of hearing loss also can be seen in areas that require interactions for learning, such as a child's ability to understand and regulate emotions and accomplish complex motor skills. The earlier the underlying hearing loss in affected children can be identified by parents and practitioners, the earlier can the children receive appropriate interventions and accommodations to minimize the impact on their cognitive, social, and emotional development. (4)

Denouement

Case 3

A newborn girl has an abnormal ABR test and is scheduled to have an audiology appointment. The parents are busy and forget the appointment. Audiology leaves a message for the family to reschedule and sends a note to the primary care practitioner. At the 2-month health supervision visit, the primary care practitioner notices in the chart that the test has not been completed and sets up another appointment for the family, with a reminder call the day before the appointment. Diagnostic ABR reveals moderate hearing loss, and the child receives a hearing aid and speech services through the Early Intervention Program. An evaluation is initiated to determine the cause of the hearing loss. Due to early intervention, this child likely will have normal speech development, in contrast to the child in Case 1.

Case 4

A healthy 4-year-old boy presents for a health supervision visit. His screening audiometry reveals mild hearing loss, and his physical examination demonstrates bilateral serous effusions. He does not have any symptoms of allergy or acute infection. At a follow-up appointment 3 months later, the effusions and hearing loss still are present. He likely has chronic eustachian tube dysfunction and, therefore, is referred to an ear, nose, and throat specialist for consideration of tympanostomy tube placement. This intervention could restore his hearing to normal before he has any long-

Summary

- Based on consensus statement of strong research evidence, UNHS should be performed for all newborns before 1 month of age and abnormal test results confirmed by full audiology evaluation by 3 months of age.
- Based on some research evidence, children enrolled in EHDI programs perform significantly better than their later-detected peers on tests of vocabulary skills and intellectual development, to the point of approaching children whose auditory capacity is normal.
- Based on strong research evidence, the most important risk factors for SNHL in the first 28 days after birth are low Apgar scores, positive family history, in utero infections, hyperbilirubinemia at levels requiring exchange transfusion, respiratory distress, prolonged mechanical ventilation, and symptoms indicative of syndromic hearing loss.
- Based on strong research evidence, genetic factors are a significant cause of hearing loss, accounting for 80% of congenital SNHL and 30% to 50% of all childhood SNHL. The most common form of genetic hearing loss involves mutations in the gene encoding the connexin 26 (Cx26) protein.
- Based on strong research evidence, the most common cause of CHL in children is OME. Based on consensus statement, referral to an ear, nose, and throat specialist should be considered for all children who have persistent effusion and hearing loss beyond 3 months.
- Based on strong research evidence, only 50% of children who have hearing loss can be identified by the use of risk indicators. Based on consensus statement, all children should have periodic objective assessments of their hearing.

term complications from hearing loss, similar to those reported for the child in Case 2.

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PIR Quiz

Quiz also available online at pedsinreview.aappublications.org.

6. Which of the following statements regarding hearing loss in infants and children is true?
 - A. Children born with external ear anomalies experience sensorineural hearing loss more commonly than conductive hearing loss.
 - B. Cholesteatoma is the most common cause of conductive hearing loss.
 - C. Language delay does not occur unless hearing loss is severe or profound.
 - D. Newborn hearing screens should be reserved for preterm infants and those who have a family history of deafness.
 - E. Parental concern regarding language delay or hearing loss is sufficient cause for auditory testing.

7. You are evaluating a newborn who has sensorineural hearing loss detected on her newborn hearing screen. Her mother reports an uneventful pregnancy, with normal prenatal laboratory values, and there is no family history of hearing impairment. Findings on physical examination include microcephaly and weight at the 5th percentile. The infant's liver is slightly enlarged. Of the following, the *most* likely cause of the infant's hearing loss is:
 - A. Alport syndrome.
 - B. Congenital cytomegalovirus infection.
 - C. Middle ear effusion.
 - D. Prenatal measles exposure.
 - E. Waardenburg syndrome.

8. A 3-day-old neonate has just demonstrated hearing loss on otoacoustic emission testing. The pregnancy was unremarkable, and the family history is negative for hearing loss. His physical examination findings are within normal limits. Of the following, the *most* appropriate next step in management is to:
 - A. Confirm the hearing loss with an auditory brainstem response test.
 - B. Perform renal ultrasonography to look for renal anomalies.
 - C. Refer the infant to audiology at 1 year of age if he is not saying words.
 - D. Repeat otoacoustic emission testing in 6 months.
 - E. Treat with oral antibiotics for possible middle ear effusion, then repeat otoacoustic emission testing.

9. Which of the following statements regarding hearing tests is true?
 - A. Auditory brainstem response testing requires the infant to be asleep.
 - B. Behavioral observation audiometry is the most effective type of hearing test in children older than age 4 years.
 - C. Otoacoustic emissions testing is the best method to evaluate auditory neuropathy.
 - D. Tympanometry testing typically yields abnormal results in patients who have sensorineural hearing loss.
 - E. Visual reinforcement audiometry is most effective in infants younger than 6 months of age.

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