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Hearing Loss in Children
A Review

Judith E. C. Lieu, MD, MSPH; Margaret Kenna, MD, MPH; Samantha Anne, MD; Lisa Davidson, PhD

IMPORTANCE Hearing loss in children is common and by age 18 years, affects nearly 1 of every 5 children. Without hearing rehabilitation, hearing loss can cause detrimental effects on speech, language, developmental, educational, and cognitive outcomes in children.

OBSERVATIONS Consequences of hearing loss in children include worse outcomes in speech, language, education, social functioning, cognitive abilities, and quality of life. Hearing loss can be congenital, delayed onset, or acquired with possible etiologies including congenital infections, genetic causes including syndromic and nonsyndromic etiologies, and trauma, among others. Evaluation of hearing loss must be based on suspected diagnosis, type, laterality and degree of hearing loss, age of onset, and additional variables such as exposure to cranial irradiation. Hearing rehabilitation for children with hearing loss may include use of hearing aids, cochlear implants, bone anchored devices, or use of assistive devices such as frequency modulating systems.

CONCLUSIONS AND RELEVANCE Hearing loss in children is common, and there has been substantial progress in diagnosis and management of these cases. Early identification of hearing loss and understanding its etiology can assist with prognosis and counseling of families. In addition, awareness of treatment strategies including the many hearing device options, cochlear implant, and assistive devices can help direct management of the patient to optimize outcomes.

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Author Affiliations: Author affiliations are listed at the end of this article.

Corresponding Author: Judith E. C. Lieu, MD, MSPH, 660 S Euclid Ave, Campus Box 8115, St Louis, MO 63110 (lieujudithe@wustl.edu).

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Hearing loss in children is common (**Box 1**); by age 18 years, it affects nearly 1 of every 5 children in the United States. Without hearing rehabilitation, hearing loss can cause detrimental effects on speech, language, developmental, educational, and cognitive outcomes in children. Hearing rehabilitation can mitigate those detrimental effects for many children, particularly when identified soon after birth or onset.

The diagnosis and management of pediatric hearing loss have undergone significant changes in the past 30 years. In 1993, the National Institutes of Health recommended newborn hearing screening within the first 3 months of life.[1] The Joint Committee on Infant Hearing, consisting of representatives from many national organizations dedicated to ensuring early identification, intervention, and follow-up care of infants and young children with hearing loss, published statements in 1994, 2000, 2007, and 2019 to establish guidelines for newborn hearing screening and for early hearing detection and intervention programs, benchmarks for quality, tracking of outcomes, and initial management of infants with hearing loss.[2] Through the Individuals with Disabilities Act (2004), Part C provides free intervention services from birth to age 3 years for any child in the United States identified with hearing loss, and Part B provides educational assistance for children aged 3 through 21 years through individualized educational plans and programs for hearing disability.

The multichannel cochlear implant was initially approved in the United States in 1990 for children 2 years or older; the age was lowered to 18 months in 1998, 12 months in 2000, and then 9 months in March 2020.[3] The combination of newborn hearing screening programs (**Box 2**), advances in cochlear implant and hearing aid technology, and legislative policy changes have allowed more than 75% of children with hearing loss to attend public schools mainstreamed with normal-hearing students.[4] The ability of screenings to detect hearing loss in infancy, the efficacy of hearing aids and cochlear implants to mitigate consequences of hearing loss, the proliferation of genetic studies expanding the understanding of genes involved with hearing, and the knowledge about the interaction between hearing and cognition have fundamentally altered the understanding about children with hearing loss. This review will summarize what is known about the current diagnosis and management of pediatric hearing loss, with a focus on some of the current controversies in management.

Methods

PubMed was searched with the Medical Subject Heading term *hearing loss* with filters for *English language*, *child* (birth-18 years), and *humans* from 1993 through July 31, 2020. The search was

Box 1. Definitions of Hearing Loss

Hearing Loss

Any impairment in the ability to hear sounds at thresholds considered normal. For children, a pure tone threshold average of more than 15 dB at 500, 1000, 2000, and 4000 Hz is considered outside the normative range, with larger reductions in hearing levels classified by severity. Severity of hearing loss is categorized below. *Deaf* is often used as an alternative to profound hearing loss when a person cannot hear typical conversations without hearing amplification. *Hard of hearing* is a general term for anyone who has some hearing loss that ranges from mild to moderate to moderately severe and who often benefits from hearing aids.

Severity of Hearing Loss

Slight: hearing thresholds 16 to 25 dB

Mild: hearing thresholds 26 to 40 dB

Moderate: hearing thresholds 41 to 55 dB

Moderately severe: hearing thresholds 56 to 70 dB

Severe: hearing thresholds 71 to 90 dB

Profound: hearing thresholds more than 90 dB

Timing of Hearing Loss

Congenital: identified in the neonatal period

Delayed-onset: identified after the neonatal period but attributed to etiologies present at birth

Acquired: occurs after the neonatal period and is attributed to etiologies not present at birth

Sensorineural hearing loss: due to injury or defect within the cochlea, cochlear nerve, or the brainstem pathways to the auditory cortex

Conductive hearing loss: due to injury or defect within the external or middle ear, including the external auditory canal, tympanic membrane, middle ear cavity, and ossicles

Mixed hearing loss: combination of sensorineural and conductive types of hearing loss

Box 2. Newborn Hearing Screening

Technology Used to Perform Screening

Otoacoustic Emissions (OAEs)

Sounds produced by outer hair cells in the cochlea in response to acoustic signals in the ear; this noninvasive test has different forms, known as transient evoked and distortion product OAE.

Auditory Brainstem Response (ABR)

A noninvasive test of the integrity of the auditory pathway from middle ear, to cochlea, to the vestibulocochlear nerve, and to brainstem, where the response is measured; the ABR can be used as a pass/fail test for screening, or to identify the threshold (softest sound) at which sounds are heard.

Automated Auditory Brainstem Response (AABR)

The hearing screening version of ABR for infants in the neonatal intensive care unit (NICU).

Protocols

Normal Newborn Nursery

Hearing screening of full-term newborns usually involves the OAE. If the infant does not pass the screen in one or both ears (termed as *refer* for diagnostic testing), then they may undergo a second screening at their primary care provider visit or the birth hospital before the age of 1 month; a second screening refer should then cause the infant to undergo diagnostic ABR testing before the age of 3 months.

Neonatal Intensive Care Unit

Hearing screening of infants from the NICU usually involves the AABR; a refer usually results in a transitory evoked otoacoustic emission test to rule out auditory neuropathy spectrum disorder and a diagnostic ABR before the age of 3 months (corrected for gestational prematurity).

supplemented by literature and policy statements that were known to the authors.

Epidemiology

The prevalence of permanent bilateral severe to profound hearing loss in newborns is 1.1 per 1000 newborns and has not changed significantly over time.[5] In addition, another 1 to 2 per 1000 newborns have bilateral mild to moderate hearing loss or unilateral hearing loss of any degree.[6] However, the age at which hearing loss is detected has decreased substantially due to successful screening programs.[2] In a study from the United Kingdom, the median age of hearing loss identification for screened children using objective tests of transient-evoked otoacoustic emissions and automated auditory brainstem response with bilateral hearing loss was 10 weeks of age (n = 151), compared with 12 to 20 months with a health visitor distraction test (behavioral observation for hearing) performed between ages 7 and 8 months in homes or community centers (n = 495).[7] Because children continue to lose hearing from multiple etiologies as they age, such as temporal bone fractures, ototoxic exposures, and delayed onset of genetic hearing loss, the prevalence of hearing loss in children by age 18 years has been estimated to be as high as 18%.[8] Early identification allows for early interventions with parent-child programs, with a benchmark of no later than 3 to 6 months of age established by the Joint Committee on Infant Hearing, including hearing aids and intensive speech-language therapy, which in turn leads to better outcomes, including earlier integration into general education (ie, mainstream schooling).[2,9]

In addition to identifying infants with profound bilateral hearing loss, the newborn hearing screening programs also identify infants with bilateral mild to moderate or unilateral hearing loss. In the past, those children would have been identified much later in childhood, often when they presented with speech-language or educational delays. The past 20 years are notable for the proliferation of studies that have investigated the difficulties that children with any degree of hearing loss may encounter.

Consequences of Hearing Loss in Children

Hearing loss is a well-known prominent risk for speech and language developmental delay. The provision of hearing aids and cochlear implants early in life has been demonstrated to help many children attain near-normal speech and language trajectories, as measured by growth curves using standardized language scores.[10-12] The effectiveness of these interventions are influenced

by factors such as maternal educational level, duration of daily hearing aids use, and nonverbal intelligence.[10,13]

Despite the significant improvements in speech and language, children with hearing loss are still at risk of delays in multiple cognitive functions, such as working memory and executive functions.[14-18] These problems have long-term educational and occupational consequences. In a Danish population study involving young men appearing before a draft board, 51% with normal hearing continued education beyond age 16 years compared with 42% with mild to moderate hearing loss, and 34% with more severe hearing loss.[19] Similarly, a Norwegian cohort study found that people with hearing loss were half as likely to achieve higher education.[20]

Hearing loss has also been found to affect a child's quality of life, particularly in the school and social domains, as well as behavior and behavioral disorders.[21,22] One systematic review reported unquantified but increased associations between hearing loss and internalizing behaviors, conduct and hyperactivity disorders, and other emotional problems.[23] One study found the prevalence of psychiatric disorder in a group of deaf and hearing-impaired children to be as high as 50%.[24] In a US public health survey, hearing loss increased the likelihood of reporting child behavioral diagnoses (55% for hearing loss, adjusted odds ratio [OR] for autism, 2.9; 95% CI, 1.8-4.9), problems with behavior (95% for hearing loss, adjusted OR for attention-deficit disorder, 3.1; 95% CI, 2.5-3.9), and difficulties with socioemotional domains (90% for hearing loss, adjusted OR, 3.9; 95% CI, 3.2-4.7).[25] In addition, 17% to 48% of children with unilateral hearing loss and 50% with cochlear implant have impaired vestibular function, which can further influence their ability to participate in normal childhood activities.[26] Long-term longitudinal studies have found significant relationships between childhood hearing loss and decreased well-being and self-esteem as well as anxiety and depression among women.[27]

Etiology of Childhood Hearing Loss

Congenital

The most common causes of permanent congenital sensorineural and mixed hearing loss are congenital cytomegalovirus (CMV; 5%-20%), structural abnormalities of the temporal bones (30%-40%), and genetic causes (50%)[28-30] (See Box 1 for definitions). Additionally, many of the anatomical abnormalities are associated with genetic causes, including branchiootorenal syndrome and CHARGE syndrome. Branchiootorenal syndrome is associated with abnormalities of the second branchial arch derivatives, external ear malformations, hearing loss, and kidney malformations. CHARGE syndrome consists of coloboma, heart defects, atresia of choanae, retardation of growth, genital abnormalities, and ear abnormalities. Ear abnormalities can include malformed external, middle, or inner ears.

The incidence of hearing loss increases with premature birth and decreases with increasing gestational age and birth weight (1.2%-7.5% born at 24-31 weeks and 1.4%-4.8% with birthweight 750-1500 g) and increasing numbers of comorbidities. Hearing loss occurs in 1.2% to 7.5% of infants in neonatal intensive care units (NICUs).[28] NICU-related hearing loss also increases with combinations of hyperbilirubinemia, sepsis, neonatal bacterial meningitis, necrotizing enterocolitis, prolonged ventilation, ototoxic medication exposure, and extracorporeal membrane oxygenation.[28] Although congenital CMV is usually a primary cause of hearing loss, it is common; thus, other genetic or structural temporal bone etiologies may also be present as an additional etiology.[31,32] Congenital infections, including syphilis and rubella can cause hearing loss.[33] Rubella, once the most common viral cause of congenital sensorineural hearing loss, is now rare due to maternal vaccination. Congenital syphilis, which had decreased for decades and is still very uncommon, is unfortunately on the rise, especially in urban locations and populations (eg, non-Hispanic Black, Black, and uninsured people), with an incidence of 23.3 per 100 000 live births in 2017.[34]

Delayed-Onset

Delayed-onset hearing loss should be considered if caregivers raise concerns about their child's hearing, speech, or language delay. Delayed-onset hearing loss may also occur if there were perinatal risk factors such as congenital CMV infection or extracorporeal membrane oxygenation.

More than 119 genes are associated with sensorineural or mixed hearing loss.[35] Of these, syndromes comprise 30% of all genetic causes and are often associated with delayed onset or progressive hearing loss, including Pendred, Usher, and Alport syndromes (**Table 1**).[41] Pendred syndrome, associated with recessive variants in the *SLC26A4* gene, is the most common syndromic form of hereditary sensorineural hearing loss and is associated with thyroid dysfunction, goiter, enlarged vestibular aqueduct, and incomplete partition type II cochlear anomaly (Mondini). Usher syndrome is also autosomal recessive and has 3 clinical types, associated with at least 9 genes that are differentiated by the severity of the hearing loss, vestibular dysfunction, and age of onset of vision loss.[35] Alport syndrome is an X-linked (80%) or recessive disorder (depending on the gene) resulting in kidney failure, ocular abnormalities (anterior lenticonus, retinopathy), and progressive sensorineural hearing loss detected usually in late childhood.[42] As a caveat, many of the syndromic hearing loss etiologies may initially present as nonsyndromic hearing loss in infancy or early childhood.

Several of the nonsyndromic recessive genes are also associated with progressive sensorineural hearing loss, and children may either pass a newborn hearing screening or present with much milder loss that worsens over time. These include *GJB2* (connexin 26), *MYO15A*, and *STRC*. Autosomal dominant nonsyndromic progressive hearing loss genes include *TMC1* and *KCNQ4*.

Delayed onset of hearing loss can also occur after congenital infections. Historically, prenatal exposure to the TORCHES (toxoplasmosis, other, rubella, CMV, herpes virus, syphilis) organisms were common causes of congenital hearing loss. However, epidemiology of these organisms has changed, and only congenital CMV is currently a substantial cause of delayed onset loss in many countries. The prevalence of congenital CMV infection is 0.4% to 2.3% of all newborns.[43] Of infants with confirmed congenital loss, 6% to 7% have congenital CMV. However, up to 43% of infants with congenital CVM will initially pass a newborn hearing screening but then present with sensorineural hearing loss later in infancy or childhood.[43]

More recently congenital Zika infection has been associated with hearing loss. A 2019 review of 10 articles including 266 infants and children from Brazil, Colombia, and the United States, reported a range of hearing loss from 6% to 68% among tested infants.[44] More study is needed to determine the possibility of progression and more

Table 1. Some Common Nonsyndromic and Syndromic Genetic Hearing Loss Genes[a]

	OMIM locus	Associated genes	Common findings	Additional diagnostic findings
Nonsyndromic hearing loss				
DFNB1	220290	*GJB2*, *GJB6*	Congenital mild to profound autosomal recessive nonsyndromic hearing loss	Usually normal temporal bone imaging[36]; rarer dominant forms are associated with skin disease; uncommon digenic inheritance with both *GJB2* and *GJB6*
DFNB16	603720	*STRC* (*CATSPER2*)	Bilateral mild to moderate congenital SNHL; deletion of both *STRC* and *CATSPER2* is associated with SNHL and infertility in males	
DFNA8/12	602574	*TECTA*	Often prelingual, often milder, and mid- or high-frequency SNHL	
DFNB21	602574	*TECTA*	Prelingual severe to profound SNHL	
DFNB3	600316	*MYO15A*	Progressive bilateral SNHL	
Mitochondrial hearing loss	561000	*MT-RNR1; 1555G>A* (this is the most common)	Maternally inherited nonsyndromic hearing loss, or hearing loss that occurs after brief exposure to aminoglycosides	There are also many mitochondrial syndromes, some of which include hearing loss
Syndromic hearing loss				
Pendred syndrome, recessive	274600	*SLC26A4*	Euthyroid (often) goiter, progressive, often asymmetric, mild to moderate sensorineural or mixed hearing loss	Intracochlear partition defect type II (Mondini) deformity in which the cochlea has less than the normal 2.5 turns and/or enlarged vestibular aqueduct on CT or MRI[3]
Usher syndrome, recessive	276900	*MYO7A*	Type I: profound hearing loss at birth, vestibular dysfunction starting at birth, vision problems early in life	Electroretinogram or dark adapted thresholds may show signs of RP earlier than routine ocular examination; there are also few variants that result in either nonsyndromic RP or nonsyndromic HL
	276904	*USH1C*		
	601067	*CDH23*		
	602083	*PCDH15*		
	606943	*SANS/USH1G*		
	276901	*USH2A*	Type II: moderate to severe hearing loss at birth, vision problems by adolescence with progression, normal balance	
	605472	*ADGRV1*		
	611383	*WHRN*		
	276902	*CLRN1*	Type III: progressive hearing loss, later onset vestibular dysfunction, and vision loss starting later in childhood or adolescence	
Alport syndrome, x-linked, recessive, dominant	301050	*COL4A5*	Progressive hearing loss, hematuria, ocular abnormalities (anterior lenticonus, retinopathy)	Kidney biopsy may reveal glomerulonephritis
	203780	*COL4A3*		
	104200	*COL4A4*		
Jervell and Lange-Nielsen syndrome, recessive	220400	*KCNQ1*	Severe to profound bilateral congenital hearing loss, syncope, sudden death	Prolongation of QT interval on electrocardiogram (ECG)
	612347	*KCNE1*		
Waardenburg syndrome, dominant or recessive	606597	*PAX3-WS1/3*	HL generally congenital, may be unilateral or bilateral and can be associated with structural inner ear anomalies, such as EVA; *WS3*, and *WS4A/B* can be autosomal dominant or recessive	Dystopia canthorum (*WS1*), synophrys, vitiligo, heterochromia iridis, white forelock; upper limb anomalies (*WS3*), Hirschsprung disease (*WS4*)
	193510	*MITF-WS2*		
	602229	*SNAI2-WS2D*		
	608890	*SOX10-WS2E*		
	277580	*4C EDNRB-WS4A*		
	613265	*EDN3-WS4B*		
Branchiooto-kidney syndrome, dominant	601653	*EYA1*	HL is generally congenital, ear anomalies may involve external, middle, and inner ear	Kidney anomalies may be structural, functional, or both
	601205	*SIX1*		
	600963	*SIX5*		

Abbreviations: CT, computed tomography; DFNB, nonsyndromic autosomal dominant deafness gene; EVA, enlarged vestibular aqueduct; HL, hearing loss; OMIM, Online Mendelian Inheritance in Man database; MRI, magnetic resonance imaging; RP, retinitis pigmentosa; SNHL, sensorineural hearing loss.

[a] Table was constructed based on Heredity Hearing Loss,[35] DiStefano et al,[37] Liming et al,[38] Shearer et al,[39] Sloan-Heggen et al,[40] and the Online Mendelian Inheritance database.[36]

clarity of the actual prevalence of hearing loss. Other congenital infections that may result in later onset sensorineural hearing loss include toxoplasmosis (1 per 10 000 in the United States) and syphilis (23.1 per 100 000 live births in 2018).[45,46]

Acquired

Postnatally acquired causes of hearing loss can be attributed to trauma, infection, ototoxic medications, or autoimmune disorders. Much of the prevalence of pediatric hearing loss is due to acquired etiologies, but specific contributions to that global prevalence have not been well studied or documented. Of the preventable causes of childhood hearing loss, the World Health Organization attributes 31% to infections, 17% to postnatal birth complications, 4% to use of ototoxic medications such as aminoglycosides by pregnant mothers and infants, and 8% to other causes.[47]

Trauma can cause conductive, mixed, or sensorineural hearing loss depending on location and type of injury to the temporal bone. Conductive hearing loss can result from tympanic membrane perforation or ossicular chain injury.[48] Temporal bone fractures can damage the cochlea, injure the cochlear nerve, or cause a perilymphatic fistula, which often result in severe to profound sensorineural hearing loss.[49] Concussive injuries to the temporal bone without fracture may also result in temporary or permanent sensorineural hearing loss.[50] Trauma to the cochlea can also be in the form of noise exposure damaging the outer hair cells resulting in permanent loss.

Infectious causes of sensorineural hearing loss include measles, mumps, varicella zoster, Lyme disease, bacterial meningitis, and rarely, otitis media. Measles and mumps disease with subsequent hearing loss is more common in unvaccinated than vaccinated children.[51] Lyme disease is an uncommon but potentially treatable cause of hearing loss. Lastly, hearing loss can often result from bacterial meningitis and can be progressive, most commonly after *Streptococcus pneumoniae* infections. Close surveillance for hearing loss is important because labyrinthine ossification can occur and implanting a cochlear device must be expedited for the patients that meet audiologic criteria for the implant.

Medications known to be ototoxic and can cause permanent hearing loss include aminoglycosides, antineoplastic agents (particularly cisplatin), and loop diuretics. Other medications such as salicylates and macrolides, including azithromycin, can cause hearing loss that is generally reversible. Close monitoring of dosages and serum drug levels can lessen the chance of injury to the inner ear. In addition, certain mitochondrial variants can confer increased susceptibility to aminoglycoside ototoxic effects.[52]

Autoimmune-related hearing loss can be due to primary autoimmune dysfunction localized to the inner ear or due to systemic autoimmune disorders such as Cogan syndrome (interstitial keratitis, progressive hearing loss, and vestibular dysfunction).[53] Hearing loss is often rapidly progressive and is sometimes responsive to immunosuppressants. Autoinflammatory genes, such as the *NLRP3* may also be associated with syndromic and nonsyndromic hearing loss.[54]

Evaluation of Hearing Loss Etiology

Before the availability of high-resolution temporal bone imaging, neonatal CMV screening, and genetic testing, assessment included laboratory testing for congenital syphilis or rubella, studies for the clinical aspects of syndromic genetic causes, or autoimmune etiologies; however, yield from this type of testing was low.[33] Currently, the recommendation is that testing should be based on family and medical history, patient age, age of onset of hearing loss, whether hearing loss is unilateral vs bilateral and/or progressive, and type of hearing loss. Options for tests for evaluation can include computed tomography (CT) or magnetic resonance imaging (MRI) of the temporal bones, to evaluate anatomical causes; genetic testing; ophthalmologic evaluation for coexisting abnormalities; and screening for congenital CMV in newborn infants. Tests that may yield a treatable cause of hearing loss (eg, congenital CMV, Lyme disease, autoimmune hearing loss) or diagnoses that would be important to not miss (long QT in a child with bilateral profound hearing loss, kidney failure in Alport syndrome) should be carefully considered based on individual patient presentation. Additional factors for imaging may include CT vs MRI, need for sedation, and risk of exposure to radiation. Common causes of hearing loss and possible testing options are identified in **Table 2**.

Management Options

Hearing Devices for Children With Bilateral Sensorineural Hearing Loss

Both physiological and behavioral evidence suggest bilateral input to the auditory system, as opposed to unilateral input, facilitates binaural listening skills necessary for developing spoken language skills, effective communication in daily listening and learning environments, and ultimately for academic success.[55,56] For children with bilateral sensorineural hearing loss, maximizing hearing at each ear is best for developing spoken language, ie, 2 ears each fitted with a device are better than 1 device in 1 ear. Device options for children with bilateral sensorineural hearing loss consists of 2 hearing aids, 2 cochlear implants, or a cochlear implant at one ear and a hearing aid at the opposite ear (referred to as bimodal devices). Decisions for recommending these devices are partially guided by audiometric hearing thresholds. **Table 3** illustrates the progression of hearing threshold levels as they relate to device recommendations. Hearing levels within the normal limits at each ear serve as the optimal listening condition. For children with bilateral sensorineural hearing loss, bilateral hearing aids are typically recommended for children with sufficient amounts of residual hearing. For children with severe to profound hearing loss, hearing aids may be insufficient for rehabilitating the hearing loss and cochlear implant technology should be considered. For children with intermediate levels of residual hearing or different levels of hearing at each ear, bimodal devices may be considered.

Compared with hearing aids that amplify acoustic information, cochlear implants bypass the normal transduction mechanisms of the peripheral auditory system and directly stimulate the auditory nerve using an electrical signal. Cochlear implants have an internal component that is surgically placed, consisting of an electrode array that is advanced into the cochlea and a receiver stimulator (**Figure** and **Box 3**). The external components of cochlear implants consist of a microphone, a transmitting coil with a magnet, and a processor. As noted in Table 3, device configurations progress from delivering an amplified acoustic signal (bilateral hearing aids) to an acoustic and electric signal combined (hearing aids and

Table 2. Diagnostic Studies

Diagnostic studies	Etiology	Advantages	Disadvantages
Urine, saliva or blood PCR within first 3 wks of life; DBS at birth or later	Congenital CMV	Minimally invasive	Testing not definitive for congenital CMV after 3 wks of age unless test DBS in conjunction with symptoms or imaging findings
Viral antibodies, viral DNA serological testing	Congenital TORCH infections; postnatal infections	Minimally invasive	False-positives and false-negatives can occur; work closely with ID department to order and interpret
Genetic testing[a]	Syndromic or nonsyndromic, genetic	Minimally invasive	Results may be negative or inconclusive, but HL could still be genetic because many genes still are unrecognized or tests are not available
CT temporal bones	Trauma, ear malformations	Rapid, often able to be completed without sedation	Radiation exposure
MRI of temporal bones, brain	Inner ear malformations associated with hearing loss	Painless, no radiation	May need sedation
Ophthalmologic evaluation	Concurrent disorders with eye, vision	Minimally invasive	Limited examination often based on age; may need more detailed examinations, such as ERG and DATs if suspect retinal pathology
Electrocardiogram	Long QT syndrome, generally associated with bilateral severe to profound SNHL	Minimally invasive	May identify cardiac conditions unrelated to hearing loss
Urinalysis	Alport syndrome	Minimally invasive	Results may be negative early in disease course
Thyroid function studies	Pendred Syndrome	Minimally invasive	Thyroid function may be truly normal (DFNB4) or normal early in course of Pendred syndrome

Abbreviations: CMV, cytomegalovirus; CT, computed tomography; DAT, dark adapted visual threshold; DBS, dried blood spot; DFNB, nonsyndromic autosomal dominant deafness gene; ERG, electroretinogram; HL, hearing loss; ID, infectious disease; MRI, magnetic resonance imaging; PCR, polymerase chain reaction; SNHL, sensorineural hearing loss; TORCH, toxoplasma, other (syphilis), rubella, cytomegalovirus, herpes.

[a] See Table 1.

Table 3. Hearing Device Configurations

Hearing level	Device configuration
Normal (all acoustic hearing)	None
Mild to moderate loss (acoustic hearing)	Bilateral hearing aids
Moderately severe to profound loss (electric + acoustic)	Bimodal cochlear implant + hearing aid
Severe or profound hearing loss (all electric hearing)	Bilateral cochlear implants

cochlear implant, bimodal) to an electrical signal only (bilateral cochlear implants).

Device Candidacy

Audiometric guidelines have been developed to determine the hearing level (unaided pure tone average), at which point children with hearing aids should be considered for a cochlear implant in the United States.[57] Initially, cochlear implants were only recommended for children with profound sensorineural hearing loss who demonstrated no benefit from conventional hearing aids; however, the guidelines have been expanded to consider cochlear implants for children with less severe loss. Recent studies have shown improved speech perception and language results with cochlear implants compared with conventional hearing aids for children with less severe loss.[58-60] Current US Food and Drug Administration (FDA) audiometric criteria for placing a cochlear implant in children with bilateral profound sensorineural hearing is from 9 through 24 months and older than 2 years for children with severe to profound sensorineural hearing loss. The documented benefits of implanting a device early for spoken language skills have supported the decrease in the age at which the FDA has approved the procedure from a minimum of 2 years to a minimum of 9 months.[13,61-63] There is evidence that early receipt of a cochlear implant is safe and is associated with a greater likelihood of improved spoken language and academic outcomes.[58,64,65]

Postsurgical complications may be minor (such as infections, skin flap break down, hematoma) or major (device failures requiring revision surgery, facial paralysis, need for explant).[66] Overall, there is a wide range of complication rates reported in the literature, ranging from 1% to 5% for major complications and 4.5% to 15% for minor complications.[67] A recent study examined outcomes within the patients who had a major complication of device failures requiring another implant (rate of 5.9%, n = 578). Even within these patients, the rate of complications from surgery was relatively low and postsurgical audiological performance was good.[68]

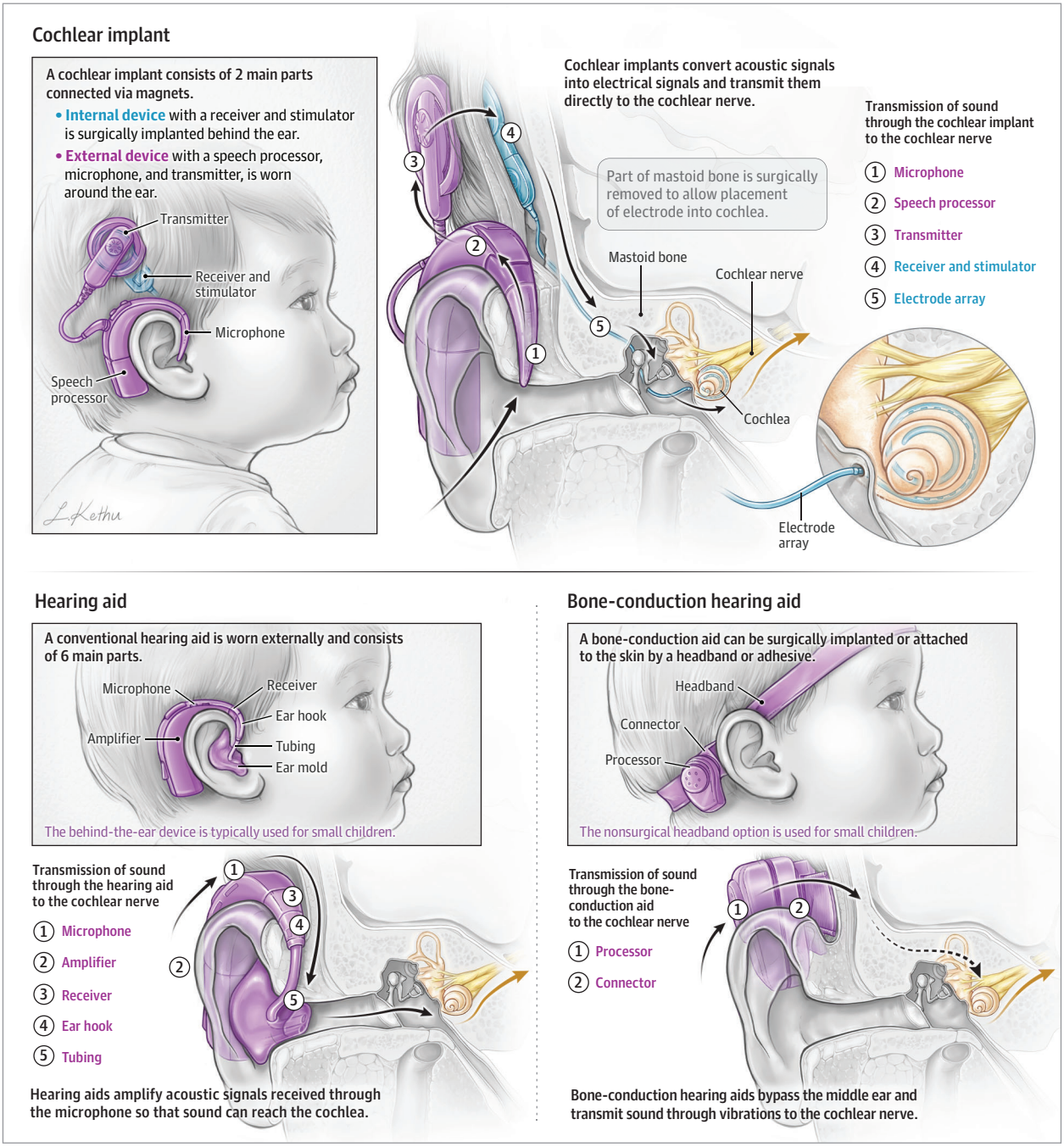
Children With Hearing Aids

Two recent studies examining a variety of spoken language outcomes for children with mild to severe hearing loss using hearing aids found that, on average, these children scored lower than their typically hearing peers.[11,69] Scores for receptive language, expressive language, speech production, and vocabulary ranged from 0.5 to 2 SDs lower than the normative mean for typically hearing peers. The degree to which children fell behind their typically hearing peers was moderated by degree of residual hearing; those with worse hearing showed greater deficits. Higher maternal educational levels and nonverbal intelligence skills coupled with earlier receipt of hearing aids, more consistent device use, and greater audibility were associated with better language outcomes.

Children With Cochlear Implants

Prior to the clinical availability of cochlear implants, children with bilateral severe to profound hearing loss using traditional hearing aids

Figure. Cochlear Implant vs Hearing Aid



acquired spoken language skills at approximately half the rate of similarly aged children with normal hearing.[70] The advent of cochlear implants has made it possible for many children with bilateral severe to profound hearing loss to attain age-appropriate speech perception, speech production, and expressive and receptive language skills by the time they enter elementary school, although a substantial proportion of children (30%-50%) fail to achieve age-appropriate spoken language skills even in the presence of factors that support successful language development.[13,61,71] Age-appropriate spoken language outcomes for pediatric recipients of cochlear implants have been associated with higher levels of nonverbal intelligence and maternal education, greater levels of preimplant residual hearing, earlier receipt of cochlear implant and early intervention services, a focus on auditory and oral instruction, and use of updated cochlear implant processor technology.[71] Improved academic attainment as well as a higher rated quality of life have been documented for children who received cochlear implants.[72,73] However, among those who received cochlear implants, long-term educational, vocational, and occupational levels achieved have continued to be significantly worse than the referenced population average.[74]

Box 3. Common Types of Hearing Devices Used by Children

Frequency Modulated (FM) System

Frequency modulated systems are often used in classroom settings, where the acoustic signal from a teacher wearing a microphone is transmitted via an FM signal to a portable speaker that sits in front of the student, allowing that student to hear the teacher better. Other terms for this technology include hearing assistive technology (HAT) or remote microphone HAT.

Hearing Aid

A personal device that is worn by the child to allow amplification of a sound; conventional hearing aids are fitted to the child's ear and use the external auditory canal to transmit the amplified sound to the cochlea; a bone-conduction hearing aid is often worn with a headband and uses vibrotactile stimulation to transmit the amplified sound to the cochlea.

Implantable Devices

Bone-Anchored Implants

Surgically placed abutments are inserted through the skin or under the skin to allow transmission of sounds through bone conduction to the cochlea.

Cochlear Implants

Cochlear implants bypass the normal transduction mechanisms of the peripheral auditory system and directly stimulate the auditory nerve using an electrical signal through a surgically placed electrode directly in the cochlea.

For children with the most profound levels of hearing loss, clinicians generally agree that bilateral cochlear implants are the most viable option for spoken language development.[75] For some children, both cochlear implants are surgically implanted simultaneously while for others, the 2 are received sequentially with varying durations (eg, few months to several years). Because some studies suggest better binaural processing skills when the interval between the first and second implant is minimal, the effect of timing for placement of the second must be discussed when the option is being considered.[76,77] Notably, as cochlear implant candidacy guidelines in the United States expand to include children with greater levels of residual hearing in at least 1 ear, many children may present with bimodal device configurations (cochlear implant combined with a hearing aids at the nonimplanted ear). For these children, clinicians must determine whether to recommend continued bimodal use or progression to bilateral implants.[75-77]

Effects of Bimodal Devices and Bilateral Cochlear Implants on Spoken Language Skills

Studies comparing the benefits of bimodal devices and bilateral cochlear implants on spoken language skills (receptive and expressive vocabulary or receptive or expressive language) have been mixed. One study found no significant group differences after accounting for various demographic variables.[78] Advantages for earlier receipt of a second cochlear implant have been found by some, yet others have noted that a period of bimodal use before the second implant was advantageous.[56,79] In many of these research studies, pediatric populations are described solely by their currently used hearing devices yet their device use prior to the first and second cochlear implant is unknown.[56] Moreover, in some cases it is unclear whether the benefits of early bilateral implants are exaggerated by deprivation of bilateral stimulation prior to the second surgery (through a lack of bimodal device use).

More recently, for speech perception and ultimately language development, the benefits of bimodal use prior to a second cochlear implant were found to vary with unaided threshold hearing levels. For those with thresholds in the severely impaired range (pure tone averages of ≈ 73 dB hearing level), a period of bimodal use of 3 to 4 years was found to be advantageous for receptive vocabulary and language. For those with profound hearing loss (≈ 92 dB hearing level) the benefits were less apparent, and for those with the most profound hearing losses (≈ 111 dB hearing level) early bilateral cochlear implant was considered to be the best for speech perception and language development outcomes.[80] Recommendations for early bilateral cochlear implant and bimodal use should be made in the context that these benefits may vary depending on the hearing level. Moreover, benefits may vary across different outcome measures; early receipt of a second cochlear implant may be advantageous for certain binaural processing skills such as sound localization and listening in spatially separated noise while continued bimodal use may be advantageous for spoken language skills.[55,80]

Hearing Rehabilitation for Children With Unilateral Hearing Loss

There is increasing evidence that unilateral hearing loss leads to harmful effects on speech and language development, educational difficulties including failing a grade, and behavioral issues.[81-84] This has led to increased efforts for auditory rehabilitation for children with unilateral hearing loss with options including conventional hearing aids, frequency modulating systems, contralateral routing of signal aids, bone-conduction hearing aids, and cochlear implant. Frequency modulating systems transmit sounds from the person wearing the transmitter to the hearing aids being worn; this reduces distracting background, listening fatigue, and the distance between the speaker and listener. Contralateral routing of signal hearing aids collect sound from the ear with the hearing loss and routes it to the better hearing ear. Bone-conduction hearing aids bypass the middle ear, directly stimulating the auditory nerve on the same side if it is functional or the opposite ear with normal hearing. Cochlear implants directly stimulate the auditory nerve through an electrode placed into the cochlea (Figure).

For children with severe to profound unilateral sensorineural hearing loss, bone-conduction hearing devices have consistently shown improved hearing thresholds, speech recognition threshold (lowest level at which a person can identify spoken words), and hearing in noisy environments.[85-88] Recently, the cochlear implant procedure has been approved for profound unilateral hearing loss (also known as single-sided deafness) for children older than 5 years. Studies have shown improvement with speech outcome measures in both quiet and noisy environments; bimodal speech reception thresholds in noise; and sound localization.[89,90] Contralateral routing of signal hearing aids have had mixed outcomes reported in the literature.[88] Even when unilateral loss is less severe, frequency modulating systems and conventional hearing aids are often beneficial. With conventional hearing aids, children have been shown to experience subjective improvement at home and school, as measured with the Children's Home Inventory for Listening Difficulties (CHILD) scores, in speech recognition in noise, word recognition scores in

noise and quiet, and sound localization.[91-94] Frequency modulating systems similarly have shown improvement in objective measures of hearing such as the Bamford-Kowal-Bench sentence list and word recognition scores in noisy and quiet environments.[93,95] Although the evidence is consistent in showing improvement in audiological measures, studies are needed to evaluate if auditory rehabilitation can thwart the overall harmful effects of unilateral hearing loss on speech and language, communication, educational performance, and social functioning.

Costs Associated With Hearing Devices

Hearing devices used in hearing rehabilitation can be associated with significant cost. A pair of hearing aids can cost approximately $6000 and the cochlear implant device itself can cost nearly $20 000 or more. These costs do not include ongoing costs such as speech-language therapy, programming of the cochlear implant, and professional and surgical fees. On the contrary, costs of untreated hearing loss are substantial; one study reported more than $1 million in lifetime cost due to special education and reduced work productivity among children with prelingual severe to profound hearing loss. One cost-utility analysis showed that benefits in quality-adjusted life-years (QALYs) varied by the child's age at the time they received the implant: children younger than 18 months gained 10.7 QALYs on average over a lifetime vs 9.0 for those aged 18 through 36 months and 8.4 for those older than 36 months.[96]

Limitations

This review has several limitations. First, because of the breadth of the topic, only a few of the frontiers of pediatric hearing loss were covered. Among the aspects of hearing loss that this article did not address are (1) controversies about whether to use antiviral agents to treat infants with congenital CMV; (2) when cochlear implant ought to be considered for unilateral hearing loss; (3) the diagnosis and management of auditory neuropathy spectrum disorder; (4) the costs of rehabilitation, such as devices, surgical placement of the devices, professional fees, and therapy; (5) experimental therapies for genetic hearing loss; and (6) issues of policy, such as whether hearing aids should be covered by insurance. Second, the continued need for early detection of hearing loss throughout childhood to identify delayed-onset, progressive, or acquired hearing loss was not reviewed.

Conclusions

Hearing loss in children is common, and there has been substantial progress in diagnosis and management of these patients. Early identification of hearing loss and understanding its etiology can assist with prognosis and counseling of families. In addition, awareness of treatment strategies including the many hearing aids options, cochlear implant, and assistive devices can help direct management of the patient to optimize outcomes.

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Author Affiliations: Department of Otolaryngology-Head and Neck Surgery, Washington University in St Louis, St Louis, Missouri (Lieu, Davidson); Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Boston, Massachusetts (Kenna); Department of Otolaryngology, Head and Neck Surgery, Harvard Medical School, Boston, Massachusetts (Kenna); Head and Neck Institute, Cleveland Clinic, Cleveland, Ohio (Anne).

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Submissions: We encourage authors to submit papers for consideration as a Review. Please contact Edward Livingston, MD, at Edward.livingston@jamanetwork.org or Mary McGrae McDermott, MD, at mdm608@northwestern.edu.

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