



Hearing Assessment in Infants, Children, and Adolescents: Recommendations Beyond Neonatal Screening

Charles Bower, MD, FAAP,^a Brian Kip Reilly, MD, FAAP,^b Julia Richerson, MD, FAAP,^c Julia L. Hecht, MD, FAAP,^d
COMMITTEE ON PRACTICE & AMBULATORY MEDICINE, SECTION ON OTOLARYNGOLOGY–HEAD AND NECK SURGERY

Children who are deaf or hard of hearing (D/HH) are at high risk for permanent deficits in language acquisition and downstream effects such as poor academic performance, personal–social maladjustments, and emotional difficulties. Identification of children born D/HH through newborn hearing screening and subsequent timely early intervention can prevent or reduce many of these adverse consequences. Ongoing surveillance for changes in hearing thresholds after infancy is also important and should be accomplished by subjective assessment for signs of atypical hearing and with objective screening tests. Scheduled hearing screening may take place in the primary care setting, or via referral to an audiologist according to the Bright Futures/American Academy of Pediatrics “Recommendations for Preventive Pediatric Health Care” (also known as the periodicity schedule). This report covers hearing assessment beyond the newborn period, reviews risk factors for hearing level change, and provides guidance for providers of pediatric primary care on the assessment and care of children who are D/HH.

TERMINOLOGY

The terminology used in this clinical report is the result of careful consideration. It is informed by published materials^{1,2} and engagement with Deaf and Hard of Hearing professionals and partner organizations (ie, National Association of the Deaf) working with the American Academy of Pediatrics (AAP) Early Hearing Detection and Intervention (EHDI) program. The authors wish to thank our partners for their work and dedication in advancing the AAP’s commitment to reversing ableist practices in pediatrics (<https://www.aap.org/en/about-the-aap/american-academy-of-pediatrics-equity-and-inclusion-efforts/words-matter-aap-guidance-on-inclusive-anti-biased-language/>).

abstract

^aDepartment of Otolaryngology, Arkansas Children’s Hospital, Little Rock, Arkansas; ^bDivision of Otolaryngology, Children’s National Medical Center, Washington, District of Columbia; ^cFamily Health Centers, Louisville, Kentucky; and ^dDepartment of Pediatrics, University of New Mexico School of Medicine, Albuquerque, New Mexico

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Readers may note the removal of deficit-framing terminology such as “loss,” “failed,” “impairment,” etc, to reflect that children who are deaf or hard of hearing (D/HH) are equal, healthy, and whole. The use of nonpathological terminology does not signify a change in the need for expeditious identification and care for children who are D/HH to reach their optimal potential. In this report, the capitalized term Deaf refers to the community of individuals who identify with the culture of deaf people that has been historically created and actively transmitted across generations.³ The terms deaf and hard of hearing are audiological designations. The terms Deafblind, deaf-blind, and deafblind refer to individuals who have both vision and hearing impacts. For more information, see <https://www.nad.org/resources/american-sign-language/community-and-culture-frequently-asked-questions/>.

INTRODUCTION

Providers of pediatric primary care can recognize infants, children, and adolescents who are at risk for or who have changes in hearing and be prepared to evaluate their hearing and arrange for proper referral, family support, and language access through appropriate medical, audiology, early intervention, and Deaf community resources within their communities. This clinical report replaces the 2009 clinical report, “Hearing Assessment in Infants and Children: Recommendations Beyond Neonatal Screening.” Changes include updated information on risk assessment, alignment with the Bright Futures/EHDI “Promoting EHDI” implementation tip sheet, updated terminology, and expansion of the considerations for providing a medical home for children who are D/HH. Note that the word children refers to infants, children, and adolescents for the purposes of this clinical report.

Infants and young children who have hearing thresholds greater than 20 dB are referred to as D/HH. They are naturally at risk for consequences of delayed and incomplete access to language, which is also known as language deprivation. The actions required to support timely and complete first language acquisition during the neuro-linguistic critical period can be implemented to prevent downstream impacts on cognition and socioemotional development. Delayed identification of changes in hearing status during early childhood and lack of timely or adequate support for language access often result in delayed development and permanent deficits. Impacts on sequential memory, abstract thinking, executive function, poor academic performance, personal–social maladjustments, and emotional difficulties have been well documented.^{4–6} For children born D/HH, early identification of hearing levels and measures to achieve access to language within the first 6 months of life have been demonstrated to facilitate language acquisition

with the potential to mitigate many of these adverse consequences.^{7–9}

As early as 1999, the AAP recommended universal newborn hearing screening and described the components of an effective program.¹⁰ As a result, over the next decade, the majority of the United States and some territories enacted legislation mandating such programs. During this period, federal funding became available to initiate and develop statewide universal newborn hearing screening and intervention, also known as EHDI programs. Because a small but significant portion of children will become D/HH after birth, there is still a need for surveillance of hearing status beyond the newborn period.^{11,12} Currently, the AAP policy statement “2022 Recommendations for Preventive Pediatric Health Care” (<https://doi.org/10.1542/peds.2022-058044>), which includes the periodicity schedule, promotes ongoing risk assessment with subjective evaluations based on developmental milestones and physical examination between the newborn hearing screen and age 4 years and periodic objective (ie, technology-based) hearing screening for children from age 4 years through adolescence.¹³ Consequently, during the neurocritical period of language development, from birth to age 5 years, risk assessment is essential as a first step in identifying infants and young children with hearing changes who should proceed to objective hearing screening. To assist in the identification of and care for D/HH children, providers are directed to the AAP periodicity schedule (https://downloads.aap.org/AAP/PDF/periodicity_schedule.pdf) for the current objective hearing screening schedule. Additionally, the following tables are contained in this report:

- Risk Factors for Early Childhood Hearing Loss: Guidelines for Infants who Pass the Newborn Hearing Screen (Table 1)
- Expected Language/Communication Milestones (Table 2)
- Hearing Screening for Infants and Children in the Primary Care Setting (Table 3)

RISK ASSESSMENT FOR CHANGE IN HEARING THRESHOLDS

Approximately 1 to 3 per 1000 children are born with atypical hearing thresholds.¹⁴ They are identified as D/HH as a result of newborn hearing screening and subsequent audiology evaluation. Another 1 to 3 per 1000 children will become D/HH later in childhood.¹⁴ These changes in hearing thresholds can be attributable to congenital conditions or genetics that manifest with a delayed change in hearing or a variety of acquired causes.

Recommendation 1: All children should undergo a risk assessment for changes in hearing thresholds by utilizing objective and evidence-based risk factors for delayed-onset hearing changes. All children, regardless of risk factors, should have their hearing screened in accord with the current Bright Futures/AAP “Recommendations for Preventive Pediatric Health Care.”

TABLE 1 Joint Committee on Infant Hearing Year 2019 Position Statement: Risk Factors for Early Childhood Hearing Loss: Guidelines for Infants Who Pass the Newborn Hearing Screen

	Risk Factor Classification	Recommended Diagnostic Follow-up	Monitoring Frequency
	Perinatal		
1	Family history ^a of early, progressive, or delayed-onset permanent childhood hearing loss	By 9 mo	Based on etiology of family hearing loss and caregiver concern
2	Neonatal intensive care of >5 d	By 9 mo	As per concerns of ongoing surveillance of hearing skills and speech milestones
3	Hyperbilirubinemia with exchange transfusion regardless of length of stay	By 9 mo	
4	Aminoglycoside administration for more than 5 d ^b	By 9 mo	
5	Asphyxia or hypoxic ischemic encephalopathy	By 9 mo	
6	Extracorporeal membrane oxygenation ^a	No later than 3 mo after occurrence	Every 12 mo—school age or at shorter intervals based on concerns of parent or provider
7	In utero infections, such as herpes, rubella, syphilis, and toxoplasmosis	By 9 mo	As per concerns of ongoing surveillance
	In utero infection with CMV ^a	No later than 3 mo after occurrence	Every 12 mo—age 3 or at shorter intervals based on parent/provider concerns
	Mother with Zika and infant with no laboratory evidence and no clinical findings	AABR by 1 mo	ABR by 4–6 mo or VRA by 9 mo
	Mother with Zika and infant with laboratory evidence of Zika with clinical findings	AABR by 1 mo	ABR by 4–6 mo; monitor as per AAP 2022 periodicity schedule ¹⁵
8	Certain birth conditions or findings:	By 9 mo	As per concerns of ongoing surveillance of hearing skills and speech milestones
	<ul style="list-style-type: none"> • Craniofacial malformations including microtia/atresia, ear dysplasia, oral facial clefting, white forelock, and microphthalmia 		
	<ul style="list-style-type: none"> • Congenital microcephaly, congenital or acquired hydrocephalus 		
	<ul style="list-style-type: none"> • Temporal bone abnormalities 		
9	>400 syndromes have been identified with atypical hearing thresholds. ^b	By 9 mo	According to natural history of syndrome or concerns
	Perinatal or postnatal		
10	Culture-positive infections associated with sensorineural hearing loss, ^c including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis or encephalitis	No later than 3 mo after occurrence	Every 12 mo—school age or at shorter intervals based on concerns of parent or provider
11	Events associated with hearing loss:	No later than 3 mo after occurrence	According to findings or continued concerns
	<ul style="list-style-type: none"> • Significant head trauma, especially basal skull/temporal bone fractures 		
	<ul style="list-style-type: none"> • Chemotherapy 		
12	Caregiver concern ^d regarding hearing, speech, language, developmental delay, and/or developmental regression	Immediate referral	According to findings or continued concerns

VRA, visual reinforcement audiometry.

^a Infants at increased risk of delayed onset or progressive hearing loss.

^b For more information, visit the Hereditary Hearing Loss Homepage (Van Camp G, Smith RJH. Hereditary hearing loss. Available at: <https://hereditaryhearingloss.org>).

^c Infants with toxic levels or with a known genetic susceptibility remain at risk.

^d Parental/caregiver concern should always prompt further evaluation.

TABLE 2 Expected Language/Communication Milestones

Developmental Age of Child	Language/Communication Milestones
Newborn, 1wk	<ul style="list-style-type: none"> • Cries with discomfort • Calms to adult voice
1 mo	<ul style="list-style-type: none"> • Alerts to unexpected sound • Makes brief short vowel sounds
2 mo	<ul style="list-style-type: none"> • Makes sounds other than crying • Reacts to loud sounds
4 mo	<ul style="list-style-type: none"> • Makes sounds like “oooo,” “aahh” (cooing) • Makes sounds back when you talk to the infant • Turns head toward the sound of your voice
6 mo	<ul style="list-style-type: none"> • Takes turns making sounds with you • Blows “raspberries” (sticks tongue out and blows) • Makes squealing noises
9 mo	<ul style="list-style-type: none"> • Makes different sounds like “mamamama” and “babababa” • Lifts arms up to be picked up
12 mo	<ul style="list-style-type: none"> • Waves “bye-bye” • Calls a parent “mama” or “dada” or another special name • Understands “no” (pauses briefly or stops when you say it)
15 mo	<ul style="list-style-type: none"> • Tries to say 1 or 2 words besides mama or dada, like “ba” for ball or “da” for dog • Looks at a familiar object when you name it • Follows directions given with both a gesture and words. For example, he gives you a toy when you hold out your hand and say, “Give me the toy.” • Points to ask for something or to get help
18 mo	<ul style="list-style-type: none"> • Tries to say 3 or more words besides mama or dada • Follows 1-step directions without any gestures, like giving you the toy when you say, “Give it to me.”
2 y	<ul style="list-style-type: none"> • Points to things in a book when you ask, like “Where is the bear?” • Says at least 2 words together, like “More milk.” • Points to at least 2 body parts when you ask the child to show you • Uses more gestures than just waving and pointing, like blowing a kiss or nodding yes
30 mo	<ul style="list-style-type: none"> • Says about 50 words • Says 2 or more words together, with 1 action word, like “Doggie run” • Names things in a book when you point and ask, “What is this?” • Says words like “I,” “me,” or “we”
3 y	<ul style="list-style-type: none"> • Talks with you in conversation using at least 2 back-and-forth exchanges • Asks “who,” “what,” “where,” or “why” questions, like “Where is mommy/daddy?” • Says what action is happening in a picture or book when asked, like “running,” “eating,” or “playing” • Says first name when asked • Talks well enough for others to understand, most of the time
4 y	<ul style="list-style-type: none"> • Says sentences with 4 or more words • Says some words from a song, story, or nursery rhyme • Talks about at least 1 thing that happened during his day, like “I played soccer” • Answers simple questions, like “What is a coat for?” or “What is a crayon for?”
5 y	<ul style="list-style-type: none"> • Tells a story the child heard or made up with at least 2 events. For example, a cat was stuck in a tree and a fire fighter saved it. • Answers simple questions about a book or story after you read or tell it to the child • Keeps a conversation going with >3 back-and-forth exchanges • Uses or recognizes simple rhymes (bat–cat, ball–tall)

Newborn to 1 month: Lipkin P, Macias M. Developmental milestones for developmental surveillance at preventive care visits. In: Hagan JF, Shaw JS, Duncan PM, eds. *Bright Futures: Guidelines for Health Supervision of Infants, Children, and Adolescents*, 4th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2017. 2 months to 5 years: Centers for Disease Control and Prevention. CDC’s Developmental Milestones. Available at: <https://www.cdc.gov/ncbddd/actearly/milestones/index.html>. Published August 17, 2022.

Many of the risk factors for a delayed onset of changes in hearing can be identified in the newborn period from the perinatal and family history and/or on

physical examination. Well-child visits during the first month of life are the opportune time to review the infant’s newborn hearing screening results and discuss

TABLE 3 Hearing Screening for Infants and Children in the Primary Care Setting⁷

Developmental Age of Child	Auditory Test/Average Time	Type of Measurement	Test Procedures	Advantages	Limitations
All ages	Evoked otoacoustic emissions, 10-min test	Physiologic test specifically measuring cochlear (outer hair cell) response to presentation of a sound stimulus	Small probe containing a sensitive microphone is placed in the ear canal for stimulus delivery and response detection.	Low cost, ease of use; patient may be asleep or awake; quick test time; responses not dependent on patient cooperation	Infant or child must be relatively inactive during the test; does not assess eighth cranial nerve or auditory cortex; not a true test of hearing; very sensitive to middle ear effusions, cerumen, or vernix in the ear canal
Birth–6 mo	AABR, 15-min test	Electrophysiologic measurement of activity in auditory nerve and brainstem pathways	Placement of electrodes on child's head detects auditory stimuli presented through earphones or probe	Not dependent on patient cooperation; eighth cranial nerve and auditory cortex assessed	Infant must be asleep; not a true test of hearing, because it does not assess functional hearing
4 y–adolescence	Pure tone audiometry, 30-min test	Behavioral test measuring auditory thresholds in response to speech- and frequency-specific stimuli presented through earphones	Patient is instructed to raise his or her hand when stimulus is heard.	Assesses functional hearing	Depends on the level of understanding and cooperation of the child; time intensive

risk factors for atypical hearing with caregivers. For a new patient visit at a later age, consider a review of potential risk factors and a thorough risk assessment. All children, regardless of risk factors, should have their hearing screened in accord with the current Bright Futures/AAP “Recommendations for Preventative Pediatric Health Care.”¹³ In the primary care setting, the recommended algorithm for detection of change in hearing status beyond newborn screening is shown in Fig 1.

Genetic causes that impact hearing may present beyond the newborn period. Therefore, a history of family members who are D/HH increases the probability, despite a negative newborn hearing screen.^{15,16} Table 1 includes the most common physical findings associated with being D/HH from inherited and de novo syndromic etiologies. These include, but are not limited to, atypical ear features and other craniofacial structures. Factors in the birth history may increase the risk beyond the neonatal period. Significant perinatal events such as asphyxia, hypoxic-ischemic encephalopathy, and extracorporeal membrane oxygenation are individual risk factors, as is a stay in the NICU for 5 days or longer. Hyperbilirubinemia requiring an exchange transfusion is a risk factor, regardless of length of NICU stay.

Congenital cytomegalovirus (cCMV) infection is currently the most common infectious cause of childhood sensorineural hearing change (SNHC), and is identified as the etiology for 25% of deaf and hard of hearing children aged 4 years.¹⁷ Although present at birth, cCMV is usually asymptomatic (90%) and usually associated with a negative newborn hearing screen. Fifteen percent of children with asymptomatic cCMV infection develop progressive SNHC and can be identified through timely hearing screening. At this time, a definitive diagnosis of cCMV can only be made from specimens obtained by 3 weeks of age. If available, the dried blood spot obtained for the newborn genetic screen may be tested retrospectively.¹⁸ The AAP *Red Book* contains updated hearing surveillance guidelines for children with cCMV.¹⁷

Other infectious diseases, especially meningitis and otitis media, are leading causes of a change in hearing. Congenital Zika infection is a newly identified etiology for delayed-onset impact on hearing. Trauma to the nervous system, damaging noise levels, and ototoxic drugs can all place a child at risk for a permanent change in hearing thresholds.^{19,20}

Risk assessment for permanent changes in hearing thresholds goes beyond historical risk factor identification and includes real-time evaluation at the visit. This includes developmental screening of language

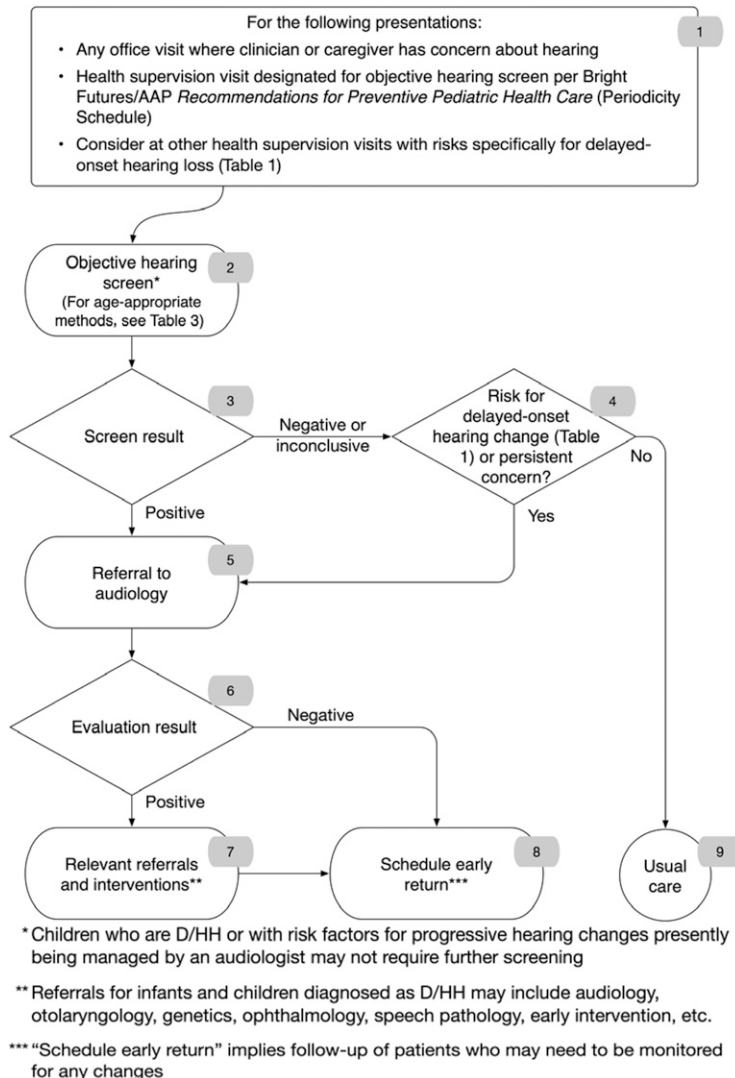


FIGURE 1
Hearing assessment algorithm within an office visit.

and auditory communication milestones, questioning caregivers regarding concern about hearing, recent medical history to assess for newly acquired risks (eg, trauma, infection, noise), and physical examination with knowledge of potential risk factors (Tables 1 and 2).

PHYSICAL EXAMINATION

A thorough physical examination is an important part of evaluating a child's hearing status. Findings on examination of the head and neck that may be associated with being D/HH include heterochromia of the irises, malformation of the auricle or ear canal, dimpling or skin tags around the auricle, cleft lip or palate, asymmetry or hypoplasia of the facial structures, and microcephaly.²¹ Middle ear structural and functional abnormalities may alert the physician to the possibility of atypical hearing.

Dense cerumen impactions should be removed before diagnostic testing because they can decrease hearing and also prevent accurate examination of the middle ear. Visualization of the eardrum and observation of mobility using pneumatic otoscopy or automated tympanometry can provide information about middle ear status. A temporary change in hearing thresholds has been demonstrated during episodes of acute otitis media. Another leading cause of acquired hearing change is otitis media with effusion (OME). The child with repeated or chronic OME may experience temporary or permanent conductive hearing change leading to impacts on language acquisition, academics, and socioemotional development, and should be referred to audiology to undergo a thorough hearing evaluation.²² Indications for referral to otolaryngology for assessment include a minimum of 3 acute infections in 6

months, 4 infections in 1 year, or persistent middle ear fluid for at least 3 months.²³

Recommendation 2: Children at all ages should have prompt objective hearing screening if there is either clinical concern or caregiver concern of permanent or ongoing hearing change.

Parental concerns and school hearing screens are the most common indicators of a change in hearing level in children after passing the newborn screen.²⁴ Caregiver concern is of even greater predictive value than the informal behavioral examination performed in the physician's office. Parents often report suspicion of decreased hearing, inattention, or erratic response to sound before confirmation that a child is D/HH.^{25,26} Parents may be as much as 12 months ahead of physicians in identifying a child's change in hearing status.¹⁰ Objective hearing screens done in the primary care setting can alleviate caregiver and clinician concern. However, all screening modalities have limitations and can produce a false-negative result. Therefore, ongoing caregiver concern after a negative screen should prompt referral to audiology for a comprehensive evaluation of hearing.

TOOLS FOR OBJECTIVE HEARING SCREENING

The appropriate technological tool or modality for objective screening depends on the child's age, ability to cooperate, and available resources. Screenings should be conducted in a quiet area where visual and auditory distractions are minimal. The child should be comfortable with the testing situation. For the procedure, infants (during the first couple of months) should be calm and preferably fed, swaddled, and asleep, and young children may need preparation with reassurance.

Billing and coding resources for screening procedures can be found at <https://www.aap.org/en/patient-care/early-hearing-detection-and-intervention/early-hearing-detection-and-intervention-coding-fact-sheet/>. Clinic staff time and attention are required for screening procedures, and provider care is required to discuss results. Therefore, hearing screening procedures should be coded, billed, and paid for by insurance companies separately from preventive care bundles.

Various hearing screening methods are outlined in Table 3 and briefly summarized here.

Middle Ear Screening

All Ages: Tympanometry

Functional assessment of the middle ear is useful in the setting of acute otitis media resolution, chronic OME, and when there is a concern of hearing change. Conductive hearing status may be the most common source of positive hearing screens in infants.²⁷ Objective middle ear assessment can best be performed by tympanometry. A flat

tympanogram indicates a high probability of the presence of middle ear effusion or tympanic membrane perforation. Both are likely to cause some impact on hearing and may require further intervention. Although low-frequency tympanograms are still used in older children, the use of a high-frequency probe tone (>660 Hz) has recently been shown to be a better measure of middle ear status in infants and young children.^{28,29}

Inner Ear Screening

Birth to 6 Months: Automated Auditory Brain Response (AABR)

The AABR is an automated and simplified screening tool based on the same technology as the auditory brain response (ABR) used by audiologists for an in-depth diagnostic evaluation. Much like an EEG, the AABR measures brain activity from sensors placed on the head. Sounds are introduced into the ear via a probe. The advantage of the AABR is that it is highly sensitive and it screens the entire hearing pathway including the eighth cranial nerve and auditory cortex. Therefore, it can detect auditory neuropathy and congenital anomalies of the eighth cranial nerve. Disadvantages of the AABR are that it requires a sleeping child and does not measure functional hearing. Without sedation, its use is generally limited to the first few months of life.

All Ages: Otoacoustic Emissions

The otoacoustic emission (OAE) is a quick, effective screening measure for inner and middle ear dysfunction. It is a practical screening tool in primary care settings because of its low cost and ease of use. The OAE screen tests the response of the cochlea's outer hair cells. When the cochlea receives sound, the hair cells vibrate. This vibration transmits neural impulses to the eighth cranial nerve to produce hearing. At the same time, the vibration echoes distally through the middle ear and this emission can be measured by a probe placed in the outer ear canal. The OAE does not assess hearing pathways proximal to the cochlea, such as the eighth cranial nerve or auditory cortex. Therefore, it cannot assess functional hearing nor detect auditory neuropathy. Use of OAE in the neonatal period will not detect an isolated congenital anomaly of the eighth cranial nerve. The OAE can be performed during sleep or while in a calm awake state.

4 Years and Older: Pure Tone Audiometry

For older children and some as young as 4 years of age, conventional screening audiometry can be used. The child is asked to raise the right or left hand when a sound is heard in the respective ear. The test should be performed in a quiet environment using earphones, and each ear should be tested at 500, 1000, 2000, and 4000 Hz. Studies show that subjective questioning of adolescents about hearing has low sensitivity to detect changes

in hearing or for whom further screening is warranted.³⁰ To detect high-frequency, noise-induced hearing changes in adolescents, office-based audiometry with the inclusion of high frequencies at 6000 and 8000 Hz³¹ is recommended once between 11 and 14 years, once between 15 and 17 years, and once between 18 and 21 years. Air conduction hearing threshold levels of >20 dB at any of these frequencies may be functionally significant. Audiometry may be a preferred screening tool for children who are able to cooperate because the entire hearing pathway is tested, as well as functional hearing.³² Screening audiometry suggesting atypical hearing thresholds in the absence of risk factors or clinical concern can be repeated. A second positive screen should be followed by prompt referral to an audiologist for a comprehensive evaluation.

Recommendation 3: A child with a positive screen for atypical hearing thresholds in one or both ears should be referred to an audiologist for diagnostic consultation and testing.

It is important to remember that atypical hearing thresholds on screening tests warrant additional testing. The result is considered a “pass” or negative screen when both ears show typical thresholds on the same day. Multiple screens to achieve a negative result or repeated screening of a single ear on different days do not constitute a bilateral “pass” or negative screen. Children whose screening tests suggest a unilateral or mild hearing change should be further evaluated and receive appropriate intervention and support. Studies show that, without intervention and support, these children are also at risk for adverse effects on language, as well as difficulties with social, emotional, and academic development.³³

Recommendation 4: Behavioral audiometry may be impossible or inaccurate for some children with developmental and/or behavioral health conditions. In this situation, referral to audiology for electrophysiological hearing testing using ABR with sedation may be more appropriate.

Special consideration should be given to children who cannot cooperate with pure tone audiometric testing. It requires that a child follow directions and respond consistently to the sound stimuli to provide a reliable result. Although OAE may be a helpful screen if a child can cooperate with it, it may not be adequate for children with medical or behavioral complexity where there are ongoing concerns of hearing changes, or significant risk factors. Thirty to 40% of children who are D/HH have co-occurring developmental conditions. Foregoing the screening test and referring to audiology for an ABR may be the best option for some children. The ABR, a test that is performed in the sleeping state, can be conducted under sedation or general anesthesia. It can assess the entire hearing pathway with a degree of accuracy that allows for noninvasive therapeutic interventions, such as hearing aids.

Recommendation 5: Newborn and childhood hearing screening results should be reviewed with families and documented to facilitate tracking and follow-up. (See Bright Futures/EHDI “Promoting Early Hearing Detection and Intervention” Implementation Tip Sheet [https://downloads.aap.org/AAP/PDF/Bright%20Futures/BF_EHDI_TipSheet.pdf]). Contact state EHDI program in accord with reporting requirements.

Caregiver support in understanding the results and implications of hearing screening is essential for ensuring follow-up of risk factors, and of positive screening tests. Office practices can ensure appropriate surveillance and follow-up leading to timely intervention and support. When possible, templates may be added to electronic health records that include risk factors for delayed-onset hearing changes and prompt providers to perform and document hearing screening on schedule. Written protocols for hearing screening and follow-up can improve time to language access and support. Identification of children who are D/HH from newborn to age 4 years is reportable to all state EHDI programs for statewide public health surveillance, as well as national tracking by the Centers for Disease Control and Prevention. It is important to know state reporting requirements and document accordingly.

FOLLOW-UP OF POSITIVE SCREENING RESULTS

Recommendation 6: To prevent false-negative results, and to avoid delays in identification, access to language, and support, screening tests should not be repeated more than once before referral to audiology for consultation and further testing.

Objective screening will only result in benefit for the patient if results indicating atypical hearing thresholds are confirmed and appropriate action is undertaken. Repeated screening may delay identification and access to language and support, falsely “rule out” changes in hearing, and add unnecessary cost. Although there are no data on follow-up of office-based screening beyond the neonatal period, the most recent (2019) data from the Centers for Disease Control and Prevention on infant hearing screening programs reveal a fairly high rate of lack of follow-up and consequent failure of timely comprehensive testing.³⁴ Increasing involvement of the pediatric primary care provider in hearing screening also requires arranging and confirming appropriate follow-up testing and appropriate referrals. Optimal communication and language development, as well as optimal development in cognition, literacy, and socioemotional growth, require prompt action and support. Although availability of appropriate testing facilities is limited in some areas,³⁵ current technological advances have made tele-audiology possible for follow-up audiologic evaluations in some rural and remote communities that lack audiologists.³⁶ This is especially relevant for appropriate follow-up to infant screens, because audiologists evaluating infants and

very young children should be experienced with these age groups.³⁷ Available audiology resources can be found at the EHDI-PALS Web site (<http://www.ehdi-pals.org>).

HEARING REFERRAL RESOURCES

Recommendation 7: If comprehensive audiologic evaluation identified the child as D/HH, discuss the importance of supporting the child's communication, language development, and socioemotional needs within the family and school settings. Refer for Early Intervention specialized for children who are D/HH. Other medical specialty referrals (ie, audiology, otolaryngology, speech pathology, ophthalmology, genetics, development, etc), may also be appropriate.^{38,39}

More than 90% of children who are D/HH are born to "hearing" parents (ie, not D/HH themselves). The pediatric primary care provider who is familiar with the most common issues impacting children who are D/HH will be in a position to provide optimal care and support to families as they navigate referrals, make decisions, and address concerns regarding their child's needs. The role of the practitioner in the medical home is to provide caregivers with appropriate options so they can make well-informed decisions.

Caregivers who are also D/HH themselves, or who come from families with D/HH members, will likely have goals and preferences based on their experiences and familiarity with the issues facing D/HH children. Families who do not have such experiences, nor have knowledgeable relatives, are beginning a journey in learning about raising a D/HH child. The expression of family desires is essential for collaboration with physicians, the hearing health team (otolaryngologist, audiologist, speech-language pathologist), and developmental specialists, and should be encouraged. Family goals and expectations are influenced by a variety of factors including culture, parental education, level of income, availability of local resources, language(s) in the home, and more. Providers can enhance their roles in collaboration (ie, shared decision-making) by providing evidenced-based discussions and referring to appropriate resources and specialists.

Enrollment in Individuals with Disabilities Education Act (IDEA) Part C Early Intervention specifically for children who are D/HH is recommended for infants and children 0 through 35 months of age.³⁷ Children who are 36 months and older qualify for school-based Part B services or a 504 Plan (see Legal Rights and Advocacy, below). Practitioners in the medical home can work directly with caregivers through shared decision-making to explore the options that are presented in Early Intervention. Familiarity with the developmental risks faced by children who are D/HH can position providers to support families in ongoing mitigation of these risks.

First Language Acquisition

The primary concern for a child who is newly identified as D/HH is access to language. Because the neurocritical period for language acquisition is birth to 5 years, children who become D/HH under the age of 5 years are at risk for language deprivation. Practitioners should understand that language deprivation begins in infancy and can continue during early childhood if a child does not receive fully accessible language input that is rich and immersive to stimulate typical brain development. Language deprivation results in language dysfluency and decreased literacy and impairments in higher cognitive function. Sequential memory, understanding cause and effect, mood regulation, and other features of abstract thought and executive function are affected.⁴ Ensuring full access to language, such as the family's spoken language, if possible, and/or visual language, can prevent language deprivation and its permanent negative sequelae.

It is common for hearing families to desire that their D/HH children communicate in a spoken language.⁴⁰ Hearing technologies such as hearing aids and/or cochlear implants (CIs) may be recommended by the hearing health team. CIs have become increasingly common as tools for acquiring spoken language,³⁷ with many children achieving spoken language abilities that would not have been possible with hearing aids. Younger age of implantation correlates with better spoken language outcomes, and the criteria for implantation balances this against the risks of surgery and potential adverse effects. Currently, CIs are approved by the US Food and Drug Administration (FDA) for use in children with an intact cochlea and eighth cranial nerve who are 9 months and older whose hearing thresholds are in the profound range (> 90 dB), bilaterally. Recent expansion of the FDA criteria includes children 24 months and older with severe bilateral hearing thresholds (>70 dB) and children 5 years and older with single-sided deafness or asymmetric hearing thresholds with limited word recognition. CIs are a tool for audition and, because they do not create typical hearing, caregivers and providers should be aware that children with CIs will not spontaneously acquire spoken language. Intensive therapy throughout the first 5 years is required to learn to interpret the CI's signals into receptive language and to shape articulate speech. The benefits and risks of the CI can be found on the FDA Web site at <https://www.fda.gov/medical-devices/cochlear-implants/benefits-and-risks-cochlear-implants>.

Practitioners and caregivers should be aware that these methods may not remediate hearing enough to ensure complete first language acquisition. At the same time, a natural visual language of the Deaf, such as American Sign Language (ASL), can be especially beneficial in supporting spoken language development and creating immediate communication and bonding in the home.^{41,42} In this way, ASL may provide the necessary linguistic input to prevent the risk of language

deprivation.^{43,44} Many are unaware that the historical paradigm of choosing either signed language or spoken language is a false dichotomy. Longstanding bias against ASL is well documented and has contributed to incomplete language acquisition and additional disabilities in some children who rely only on hearing aids or CIs for spoken language acquisition.⁴⁵

Evidence of the health benefits of signed and spoken language bilingualism continues to accrue.⁴⁶ Families need not be constrained into choosing a single modality for their child's communication. When hearing status changes during the period of first language acquisition (birth to age 5 years), the focus should be on the child meeting language acquisition milestones at the appropriate age to optimize brain development and prevent language deprivation. Developmental surveillance with milestones that include ASL will support families who choose bilingual language acquisition with signed and spoken language (<https://www.in.gov/health/cdhhe/files/ideal-language-milestones-english-american-sign-language.pdf>).

Postlingual Hearing Change

School-aged children with recent onset of hearing change may already have the foundation of a first language and have established communication with their family and peer group. The primary practitioner can support the family in developing a hearing health team consisting of audiologist, speech-language pathologist, and pediatric otolaryngologist, as well as referring to other medical specialists. Another area of focus should be accommodations in the school setting to support language and visual access for newly identified D/HH students to optimize their academic potential, socioemotional development, and well-being. Opportunities to socialize with the D/HH community and to learn ASL may enrich the child and family's life, and offer solutions to new challenges.⁴⁷ Providers can support the family's advocacy that their child's rights are met as determined by state and federal law.

Legal Rights and Advocacy

There are often special considerations needed when the federal IDEA is applied to children who are D/HH.^{48,49} Children whose disability does not qualify them for an Individualized Education Program under IDEA are eligible for a 504 Plan to ensure access. Many states have enacted their own Deaf Child's Bill of Rights. State EHDI programs and State Commissions for the Deaf can be a helpful source of community-based resources such as family of D/HH advocacy groups and online resources geared toward family education, peer groups for young people who are D/HH, and networking opportunities. For more information, see <https://www.infantheating.org/status/cnhs.php> and <https://www.nad.org/resources/directories/state-agencies-of-deaf-hoh/>.

Deaf Families, Culture, and Lived Experience

Culture and personal experiences will likely influence the degree of medical intervention that family members desire. Parents or caregivers who are also D/HH may consider themselves culturally Deaf and value ASL. They may have a different perspective than caregivers for D/HH children who are hearing. Culturally Deaf families will have had their own experiences with hearing care specialists, the special education system, and hearing technologies. Many D/HH adults do not identify as culturally Deaf. However, they will also be informed by lived experience. Many D/HH people report negative or traumatic experiences with the health care system.⁵⁰ Developing a rapport of trust and respect for their lived experiences, expertise, and cultural perspective is essential for providing a medical home for children from families that are D/HH. The Americans with Disabilities Act requires that interpreters be provided for caregivers and patients who prefer to communicate with medical personnel in ASL and should be arranged in advance.

Specialty Referral

The pediatric otolaryngologist has an important role in the diagnosis, workup, and treatment plan for infants and children who are D/HH. Diagnostic testing may include imaging of the temporal bone to identify structural defects and neural pathways for assessment of eighth cranial nerve integrity, assessment for middle ear dysfunction, and identification of SNHC. Pediatric otolaryngologists will address reversible conductive hearing conditions such as OME or other middle ear disorders, provide clearance and referral for hearing aids, and consider the possibility of more invasive devices, such as cochlear implants or bone-anchored hearing aids. Treatment plans should focus on the child's linguistic, cognitive, and socioemotional needs, as well as the family's desires. Otolaryngologists can encourage and support the use of school-based technologies such as frequency modulated devices, referrals to ASL educational resources in the community, and peer and parent networking.

As part of the hearing health team, the role of the otolaryngologist is similar to that of the pediatric primary care provider in the medical home: To assist families in identifying the options available to them and support them in decision-making processes that will occur throughout the child's youth and adolescence as developmental needs emerge and change. Follow-up includes ongoing evaluation and management of the adequacy of hearing habilitation with hearing technologies and observation for potential complications of device use, such as otitis externa and cerumen impactions. It is important that all providers realize that families are on a journey that can change direction. Thus, no intervention or plan is "final" and ongoing reassessment and discussion is necessary regarding progress, alternative interventions, new developments, etc.

In addition to the hearing health team, other specialists may have a role in elucidating the etiology of the change in

hearing status, probability of progression, and associated health or developmental implications. Evaluation by a medical geneticist and genetic testing⁵¹ can confirm a nonsyndromic etiology and reassure families and clinicians that other in-depth workup for associated health conditions is unwarranted. Syndromic etiologies can direct specific workups and monitoring.

All children who are D/HH should have a thorough ophthalmologic evaluation to identify associated conditions (ie, Usher syndrome, CMV chorioretinitis). For individuals who are D/HH, vision becomes a primary means of perceiving language and environment, and addressing visual needs is essential for optimal development and quality of life. Children with dual sensory involvement experience an interplay of effects and require a specialized team that works together with families to address a child's individual communication, mobility, academic, and socioemotional needs. State schools for the deaf and blind are often helpful resources regarding community-based services, and some states have federally funded resource programs for children who are deafblind.

At least one-third of children who are D/HH will have a coexisting condition affecting development and learning. These may include attention-deficit/hyperactivity disorder, autism spectrum disorder, learning disabilities, or global developmental delay.⁵² Service provision should address the whole child and the range of needs and referral to a developmental-behavioral pediatric specialist should be considered as needed.

SUMMARY OF RECOMMENDATIONS

Recommendation 1: All children should undergo a risk assessment for changes in hearing thresholds by utilizing objective and evidence-based risk factors for delayed-onset hearing changes. All children, regardless of risk factors, should have their hearing screened in accord with the current Bright Futures/AAP "Recommendations for Preventive Pediatric Health Care."

Recommendation 2: Children at all ages should have prompt objective hearing screening if there is either clinical concern or caregiver concern of permanent or ongoing hearing change.

Recommendation 3: A child with a positive screen for atypical hearing thresholds in one or both ears should be referred to an audiologist for diagnostic consultation and testing.

Recommendation 4: Behavioral audiometry may be impossible or inaccurate for some children with developmental and/or behavioral health conditions. In this situation, referral to audiology for electrophysiological hearing testing using ABR with sedation may be more appropriate.

Recommendation 5: Newborn and childhood hearing screening results should be reviewed with families and documented to facilitate tracking and follow-up (see Bright Futures/EHDI "Promoting Early Hearing Detection and

Intervention" Implementation Tip Sheet https://downloads.aap.org/AAP/PDF/Bright%20Futures/BF_EHDI_TipSheet.pdf). Contact the state EHDI program in accord with reporting requirements.

Recommendation 6: To prevent false-negative results, and to avoid delays in identification, access to language, and support, screening tests should not be repeated more than once before referral to audiology for consultation and further testing.

Recommendation 7: If comprehensive audiologic evaluation identified the child as D/HH, discuss the importance of supporting the child's communication, language development, and socioemotional needs within the family and school settings. Refer for Early Intervention specialized for children who are D/HH. Other medical specialty referrals (ie, audiology, otolaryngology, speech pathology, ophthalmology, genetics, development, etc) may also be appropriate.^{38,39}

LEAD AUTHORS

Charles Bower, MD, FAAP
Brian Kip Reilly, MD, FAAP
Julia Richerson, MD, FAAP
Julia L. Hecht, MD, FAAP

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Vivian B. Thorne

CONSULTANTS

Alexander Fiks, MD, FAAP – Partnership for Policy Implementation
Stephen (Andy) Spooner, MD, FAAP – Partnership for Policy Implementation

ABBREVIATIONS

AABR: automated auditory brainstem response
AAP: American Academy of Pediatrics
ABR: auditory brainstem response
ASL: American Sign Language
CI: cochlear implant
cCMV: congenital cytomegalovirus
D/HH: deaf or hard of hearing
EHDI: early hearing detection and intervention
FDA: US Food and Drug Administration
NICU: neonatal intensive care unit
OAE: otoacoustic emissions
OME: otitis media with effusion
SNHC: sensorineural hearing change

Address correspondence to Charles Bower, MD, FAAP. E-mail: bowercharlesm@uams.edu

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