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Guidance for the Clinician in Rendering Pediatric Care

Clinical Report—Hearing Assessment in Infants and Children: Recommendations Beyond Neonatal Screening

abstract

Congenital or acquired hearing loss in infants and children has been linked with lifelong deficits in speech and language acquisition, poor academic performance, personal-social maladjustments, and emotional difficulties. Identification of hearing loss through neonatal hearing screening, regular surveillance of developmental milestones, auditory skills, parental concerns, and middle-ear status and objective hearing screening of all infants and children at critical developmental stages can prevent or reduce many of these adverse consequences. This report promotes a proactive, consistent, and explicit process for the early identification of children with hearing loss in the medical home. An algorithm of the recommended approach has been developed to assist in the detection and documentation of, and intervention for, hearing loss. *Pediatrics* 2009;124:1252–1263

KEY POINTS

1. Every child with 1 or more risk factors on the hearing risk assessment should have ongoing developmentally appropriate hearing screening and at least 1 diagnostic audiology assessment by 24 to 30 months of age.
2. Periodic objective hearing screening of all children should be performed according to the recommendations for preventive periodic health care.[1]
3. Any parental concern about hearing loss should be taken seriously and requires objective hearing screening of the patient.
4. All providers of pediatric health care should be proficient with pneumatic otoscopy and tympanometry. However, it is important to remember that these methods do not assess hearing.
5. Developmental abnormalities, level of functioning, and behavioral problems (ie, autism/developmental delay) may preclude accurate results on routine audiometric screening and testing. In this situation, referral to an otorhinolaryngologist and a pediatric audiologist who has the necessary equipment and expertise to test infants and young children should be made.
6. The results of abnormal screening should be explained carefully to parents, and the child's medical record should be flagged to facilitate tracking and follow-up.

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KEY WORD
hearing screening, hearing loss, audiology

ABBREVIATIONS
AAP—American Academy of Pediatrics
OAE—otoacoustic emission
ABR—auditory brainstem response
VRA—visual reinforced audiometry

The guidance in this report does not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account, individual circumstances may be appropriate.

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7. Any abnormal objective screening result requires audiology referral and definitive testing.
8. A failed infant hearing screening or a failed screening in an older child should always be confirmed by further testing.
9. Abnormal hearing test results require intervention and clinically appropriate referral, including otolaryngology, audiology, speech-language pathology, genetics, and early intervention.

INTRODUCTION

Failure to detect congenital or acquired hearing loss in children may result in lifelong deficits in speech and language acquisition, poor academic performance, personal-social maladjustments, and emotional difficulties. Early identification of hearing loss and appropriate intervention within the first 6 months of life have been demonstrated to ameliorate many of these adverse consequences and facilitate language acquisition.[2] Supportive evidence is outlined in the Joint Committee on Infant Hearing's "Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs," which was endorsed by the American Academy of Pediatrics (AAP).[3] This evidence also is part of the rationale for the AAP statement "Newborn and Infant Hearing Loss: Detection and Intervention,"[4] which endorses universal hearing screening and reviews the primary objectives, important components, and recommended screening methods and parameters that characterize an effective universal hearing screening program. Furthermore, the AAP statement "Recommendations for Preventive Pediatric Health Care"[1] promotes objective newborn hearing screening as well as periodic hearing screening for every child through adolescence (Table 1).

TABLE 1 Recommendations for Preventive Pediatric Health Care[1]

Stage	Age	Sensory Screening: 6. Hearing
Infancy	Prenatal	
	Newborn	[a]
	3–5 d[1]	[b]
	By 1 mo	[b]
	2 mo	[b]
	4 mo	[b]
	6 mo	[b]
	9 mo	[b]
Early childhood	12 mo	[b]
	15 mo	[b]
	18 mo	[b]
	24 mo	[b]
	30 mo	[b]
	3 y	[b]
	4 y	[a]
Middle childhood	5 y	[a]
	6 y	[a]
	7 y	[b]
	8 y	[a]
	9 y	[b]
	10 y	[a]
Adolescence	11 y	[b]
	12 y	[b]
	13 y	[b]
	14 y	[b]
	15 y	[b]
	16 y	[b]
	17 y	[b]
	18 y	[b]
	19 y	[b]
	20 y	[b]
	21 y	[b]

[a] To be performed.

[b] Risk assessment, with appropriate action to follow if positive.

All providers of pediatric health care need to recognize children who are at risk of or who suffer from congenital or acquired hearing loss, be prepared to screen their hearing, and assist the family and arrange for proper referral and treatment by identifying available hearing resources within their communities. In addition, the pediatric health care professional can play an important role in communication with the child's schoolteacher and/or nurse and special education professionals to facilitate proper accommodation and education once a hearing deficit has been confirmed.

This clinical report replaces the previous 2003 clinical report and seeks to promote a proactive, consistent, and explicit process for the early identification of children with hearing loss in the medical home. To assist in the detection and documentation of and intervention for hearing loss, an algorithm of the recommended approach with key points has been developed (Fig 1), as have several tables.

RISK INDICATORS FOR HEARING LOSS

Some degree of hearing loss (Table 2) is present in 1 to 6 per 1000 newborn infants.[5] Most children with congenital hearing loss are potentially identifiable by newborn and infant hearing screening. However, some congenital hearing loss may not become evident until later in childhood. Hearing loss also can be acquired during infancy or childhood for various reasons. Infectious diseases, especially meningitis, are a leading cause of acquired hearing loss. Trauma to the nervous system, damaging noise levels, and ototoxic drugs can all place a child at risk of developing acquired hearing loss.[6,7] Otitis media is a common cause of usually reversible hearing loss. Certain physical findings, historical events, and developmental conditions may indicate a potential hearing problem. These conditions include, but are not limited to, anomalies of the ear and other craniofacial structures, significant perinatal events, and global developmental or speech-language delays. All older infants and children should be screened for risk factors involving hearing problems. A summary of high-risk indicators for hearing loss and developmental milestones are included in Tables 3 and 4, respectively. All infants with a risk indicator for hearing loss, regardless of surveillance findings, should be referred for an audiologic assessment at least once by 24 to 30 months of age, even if the child passed the newborn hearing screening. Children with risk indicators that are highly associated with delayed-onset hearing loss, such as hav-

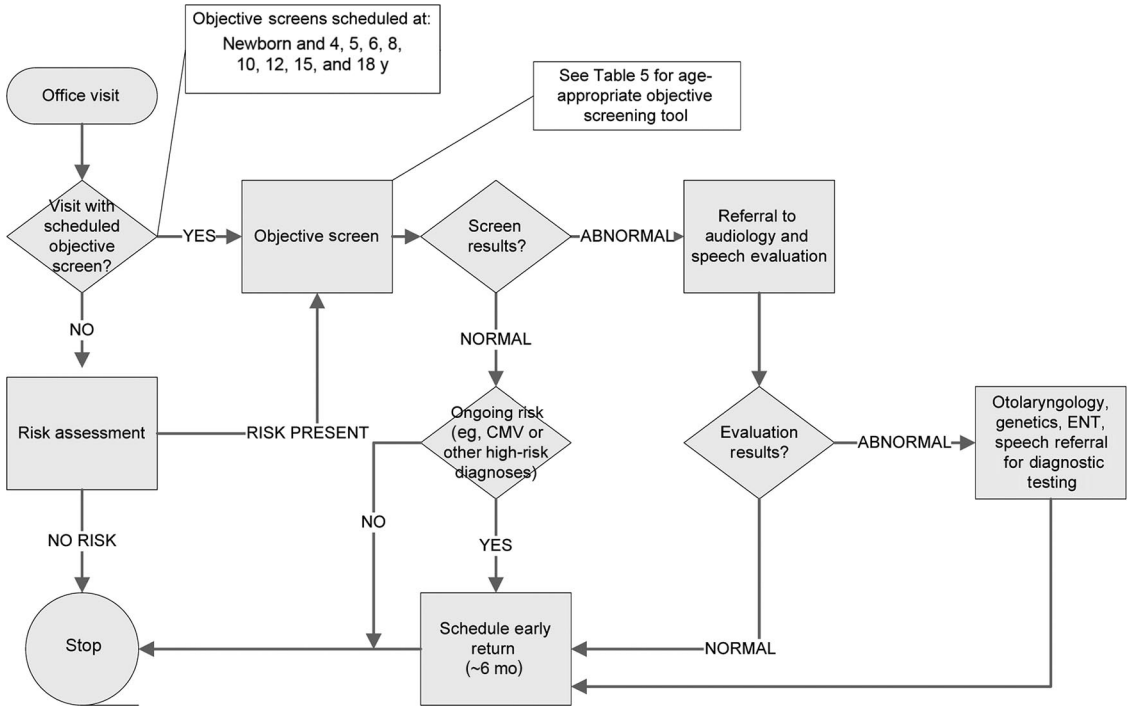

FIGURE 1
Hearing-assessment algorithm within an office visit. CMV indicates cytomegalovirus; ENT, ear, nose, and throat.

ing received extracorporeal membrane oxygenation or having cytomegalovirus infection, should have more frequent audiological assessments. Key point 1: Every child with 1 or more risk factors on the hearing risk assessment should have ongoing developmentally appropriate hearing screening and at least 1 diagnostic audiology assessment by 24 to 30 months of age (Table 1).

Although questionnaires and checklists are useful for identifying a child at risk of hearing loss, studies have shown that only 50% of children with hearing loss are identified by the comprehensive use of such questionnaires.[8,9] Key point 2: Periodic objective hearing screening of all children should be performed according to the recommendations for preventive periodic health care[1] (Table 1).

If a parent or caregiver is concerned that a child might have hearing loss, the pediatrician needs to assume that such is true until the child's hearing has been evaluated objectively. Parental concern is of greater predictive value than the informal behavioral examination performed in the physician's office.[10] Parents often report suspicion of hearing loss, inattention, or erratic response to sound before hearing loss is confirmed.[11] One study showed that parents were as much as 12 months ahead of physicians in identifying their child's hearing loss.[3] Key point 3: Any parental concern about hearing loss should be taken seriously and requires objective hearing screening of the patient.

TABLE 2 Definitions of Hearing Loss

Hearing Loss	Definition
Mild	On average, the most quiet sounds that people can hear with their better ear are between 20 and 40 dB. People who suffer from mild hearing loss have some difficulties keeping up with conversations, especially in noisy surroundings.
Moderate	On average, the most quiet sounds heard by people with their better ear are between 40 and 70 dB. People who suffer from moderate hearing loss have difficulty keeping up with conversations when not using a hearing aid.
Severe	On average, the most quiet sounds heard by people with their better ear are between 70 and 95 dB. People who suffer from severe hearing loss will benefit from powerful hearing aids, but often they rely heavily on lip reading, even when they are using hearing aids. Some also use sign language.

Adapted from: European Group on Genetics of Hearing Impairment. Martini A, ed. European Commission Directorate, Biomedical and Health Research Programme (HEAR) Infoletter 2, November 1996;8.

PHYSICAL EXAMINATION

Thorough physical examination is an essential part of evaluating a child for hearing loss. Findings on head and neck examination associated with potential hearing loss 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.[12] Hypertelorism and abnormal pig-

TABLE 3 American Academy of Pediatrics Joint Committee on Infant Hearing Year 2007 Position Statement[3]: Risk Indicators Associated With Permanent Congenital, Delayed-Onset, and/or Progressive Hearing Loss in Childhood

1	Caregiver concern[a] regarding hearing, speech, language, or developmental delay.
2	Family history[a] of permanent childhood hearing loss.
3	Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO[a], assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia[19] that requires exchange transfusion.
4	In utero infections such as CMV[a], herpes, rubella, syphilis, and toxoplasmosis.
5	Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
6	Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7	Syndromes associated with hearing loss or progressive or late-onset hearing loss[a], such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
8	Neurodegenerative disorders[a], such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
9	Culture-positive postnatal infections associated with sensorineural hearing loss[a], including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
10	Head trauma, especially basal skull/temporal bone fracture[a] that requires hospitalization.
11	Chemotherapy[a].
12	Recurrent or persistent otitis media for at least 3 months.

Risk indicators that are marked with [a] are of greater concern for delayed onset hearing loss. ECMO indicates extracorporeal membrane oxygenation; CMV, cytomegalovirus.

Reproduced with permission from: American Academy of Pediatrics, Joint Committee on Infant Hearing. *Pediatrics*. 2007; 120(4):898–921.

mentation of the skin, hair, or eyes also may be associated with hearing loss, as in Waardenburg syndrome. The presence of renal abnormalities (Alport syndrome), cardiac anomalies (prolonged QT interval in Jervell and Lange-Nielsen syndrome), and other syndromes should also prompt evaluation of hearing. Abnormalities of the eardrum should alert the physician to the possibility of hearing loss. Cerumen impactions can obscure the tympanic membrane, preventing an accurate examination, and may cause hearing loss. Dense cerumen impactions should be removed before diagnostic testing. A leading cause of acquired hearing loss is otitis media with effusion. Temporary hearing loss has been demonstrated during episodes of acute otitis media. The child with repeated or chronic otitis media with effusion is at high risk of acquired hearing loss and should undergo comprehensive hearing evaluation.[13,14] Key point 4: All providers of pediatric health care should be proficient with pneumatic otoscopy and tympanometry. However, it is important to remember that these methods do not assess hearing.

TOOLS FOR OBJECTIVE HEARING SCREENING

In addition to universal newborn hearing screening and regular surveillance of developmental milestones, auditory skills, parental concerns, and middle-ear status, objective screenings for hearing loss should be performed periodically on all infants and children in accordance with the schedule outlined in the AAP statement "Recommendations for Preventive Pediatric Health Care"[1] (Table 1). The technology used for hearing screening should be age appropriate. The child also should be comfortable with the testing situation; young children may need preparation. A variety of objective tools have been developed for screening tests. The choice of tool to use in screening depends on the child's age, degree of cooperation, and available resources. Screenings should be conducted in a quiet area where visual and auditory distractions are minimal. For children for whom screening is not possible because of developmental level, referral to a pediatric audiologist should be initiated for appropriate physiologic and/or behavioral audiological assessment. Various tests performed by audiologists are outlined in Table 5.

Tympanograms

Conductive hearing loss may be the most common cause of infant hearing screening failures.[15] Objective middle-ear assessment can best be performed by tympanometry. Tympanometry measures relative changes in tympanic membrane movement as air pressure is varied in the external auditory canal. Tympanograms (Fig 2) can most simply be classified as types A, B, and C depending on the curve shape relative to 0 as the pressure is changed (www.audiologyonline.com/askexpert/display_question.asp?question_id=451).

The presence of a type A, high-peaked tympanogram significantly decreases the probability that middle-ear effusion is the cause of hearing loss. A type B, flat tympanogram has the highest probability of the presence of middle-ear effusion or tympanic membrane perforation, which are both likely to cause some degree of hearing loss. A type C tympanogram, with a peak shifted toward negative pressure, has a low probability of middle-ear fluid and associated hearing loss. Type B and C tympanograms require clinical correlation and possibly further evaluation and treatment. Traditionally, tympanograms have been obtained by using low-frequency probe tones. These tones have been historically inaccurate for infants younger than 6 months. The use of a high-frequency probe tone (1000 Hz) was recently shown to be a better measure of middle-ear status in infants and young

TABLE 4 Developmental Milestones in the First 2 Years of Life

Milestone	Average Age of Attainment, mo	Developmental Implications
Gross motor		
Head steady in sitting	2.0	Allows more visual interaction
Pull to sit, no head lag	3.0	Muscle tone
Hands together in midline	3.0	Self-discovery
Asymmetric tonic neck reflex gone	4.0	Child can inspect hands in midline
Sits without support	6.0	Increasing exploration
Rolls back to stomach	6.5	Truncal flexion, risk of falls
Walks alone	12.0	Exploration, control of proximity to parents
Runs	16.0	Supervision more difficult
Fine motor		
Grasps rattle	3.5	Object use
Reaches for objects	4.0	Visuomotor coordination
Palmar grasp gone	4.0	Voluntary release
Transfers object hand to hand	5.5	Comparison of objects
Thumb-finger grasp	8.0	Able to explore small objects
Turns pages of book	12.0	Increasing autonomy during book time
Scribbles	13.0	Visuomotor coordination
Builds tower of 2 cubes	15.0	Uses objects in combination
Builds tower of 6 cubes	22.0	Requires visual, gross, and fine motor coordination
Communication and language		
Smiles in response to face, voice	1.5	Child more active social participant
Monosyllabic babble	6.0	Experimentation with sound, tactile sense
Inhibits to "no"	7.0	Response to tone (nonverbal)
Follows 1-step command with gesture	7.0	Nonverbal communication
Follows 1-step command without gesture (eg, "Give it to me")	10.0	Verbal receptive language
Speaks first real word	12.0	Beginning of labeling
Speaks 4–6 words	15.0	Acquisition of object and personal names
Speaks 10–15 words	18.0	Acquisition of object and personal names
Speaks 2-word sentences (eg, "Mommy shoe")	19.0	Beginning grammaticization, corresponds with vocabulary of ≥50 words
Cognitive		
Stares momentarily at spot where object disappeared (eg, yarn ball dropped)	2.0	Lack of object permanence (out of sight, out of mind)
Stares at own hand	4.0	Self-discovery, cause and effect
Bangs 2 cubes	8.0	Active comparison of objects
Uncovers toy (after seeing it hidden)	8.0	Object permanence
Egocentric pretend play (eg, pretends to drink from cup)	12.0	Beginning symbolic thought
Uses stick to reach toy	17.0	Able to link actions to solve problems
Pretend play with doll (gives doll bottle)	17.0	Symbolic thought

Modified from: Behrman RE, Jenson HB, Kliegman R, eds. *Nelson Textbook of Pediatrics*. 16th ed. Philadelphia, PA: Saunders; 2003.

children. These tympanograms are generally not classified as A, B, or C but rather as peak or no peak.

Evoked Otoacoustic Emissions

Evoked otoacoustic emissions (OAEs) are acoustic signals generated from within the cochlea that travel in a reverse direction through the middle-ear space and tympanic membrane out to the ear canal. These signals are generated in response to an auditory stimulus, either clicks or tone bursts. The signals may be detected with a very sensitive microphone/probe system placed in the external ear canal. The OAE test allows for individual ear assessment, can be performed quickly at any age, and does not depend on whether the child is asleep or awake. Mild degrees of motion artifact do not interfere with test results; however, screening results are frequently influenced by the presence of middle-ear pathologic abnormalities. The OAE test is an effective screening measure for middle-ear abnormalities and for moderate or more severe degrees of hearing loss, because normal OAE responses are not obtained if hearing thresholds are approximately 30- to 40-dB hearing levels or higher. The automated OAE screener provides a pass-fail report; no test interpretation by an audiologist is required. The OAE test does not further quantify hearing loss or hearing threshold level. The OAE test also does not assess the integrity of the neural transmission of sound from the eighth nerve to the brainstem and, therefore, will miss auditory neuropathy and other neuronal abnormalities. Infants with such abnormalities will have normal OAE test results but abnormal auditory brainstem response (ABR) test results. A "failed" OAE test only implies that a hearing loss of more than 30 to 40 dB may exist or that the middle-ear status is abnormal.

Automated ABR

One objective physiologic means of screening hearing is the automated ABR. This instrument measures cochlear response in the 1- to 4-kHz range with a broadband click stimulus in each ear. Many ABR screening instruments incorporate built-in artifact rejection for myogenic, electrical, and environmental noise interference, which ensures that data collection is halted if testing conditions are unfavorable. The automated screener provides a pass-fail report; no test interpretation by an audiologist is required. A "fail" report on an automated ABR implies a hearing level of worse than 40 dB. Automated ABR can test each ear individually and can be performed on children of any age. Motion artifacts interfere with test results. For this reason, the test

TABLE 5 Audiologic Tests for Infants and Children

Developmental Age of Child	Auditory Test/ Average Time	Type of Measurement	Test Procedures	Advantages	Limitations
All ages	Evoked OAEs/10-min test	Physiologic test specifically measuring cochlear (outer hair cell) response to presentation of a stimulus; stimuli may be clicks (transient evoked OAEs) or tone pairs (distortion product OAEs)	Small probe containing a sensitive microphone is placed in the ear canal for stimulus delivery and response detection	Ear-specific results; not dependent on whether patient is asleep or awake; quick test time; screening test	Infant or child must be relatively inactive during the test; not a comprehensive test of hearing, because it does not assess cortical processing of sound; OAEs are very sensitive to middle-ear effusions and cerumen or vernix in the ear canal
Birth to 9 mo	Automated ABR/15-min test	Electrophysiologic measurement of activity in auditory nerve and brainstem pathways	Placement of electrodes on child's head detects neurologic response to auditory stimuli presented through earphones or ear inserts 1 ear at a time	Ear-specific results; responses not dependent on patient cooperation; screening test	Infant or child must remain quiet during the test (sedation is often required); not a comprehensive test of hearing, because it does not assess cortical processing of sound
9 mo to 2.5 y	VRA/15- to 30-min test	Behavioral tests measuring responses of the child to speech and frequency-specific stimuli presented through speakers or insert earphones	Technique conditions the child to associate speech or frequency-specific stimuli with a reinforcer, such as a lighted toy or video clips; VRA requires a calibrated, sound-treated room	Assesses auditory perception of child; diagnostic test.	When performed with speakers, only assesses hearing of the better ear; not ear specific; if VRA is performed with insert, earphones can rule out a unilateral hearing loss
2.5 to 4 y	Play audiometry/ 15–30 min	Behavioral test of auditory thresholds in response to speech and frequency-specific stimuli presented through earphones and/ or bone vibrator	Child is conditioned to respond when stimulus tone is heard, such as to put a peg in a pegboard or drop a block in a box	Ear-specific results; assesses auditory perception of child; screening or diagnostic test.	Attention span of child may limit the amount of information obtained
4 y to adolescence	Conventional audiometry/ 15- to 30-min test	Behavioral test measuring auditory thresholds in response to speech and frequency-specific stimuli presented through earphones and/ or bone vibrator	Patient is instructed to raise his or her hand when stimulus is heard	Ear-specific results; assesses auditory perception of patient; screening or diagnostic test	Depends on the level of understanding and cooperation of the child
All ages	Diagnostic ABR	Electrophysiologic measurement of activity in auditory nerve and brainstem pathways	Placement of electrodes on child's head detects auditory stimuli presented through insert earphones 1 ear at a time	Ear-specific results; multiple frequencies are tested, creating a map of hearing loss similar to an audiogram; responses not dependent on patient cooperation; diagnostic test	Infant or child must remain quiet during the test (sedation is often required); not a true test of hearing, because it does not assess cortical processing of sound
All ages	Tympanometry	Relative change in middle-ear compliance as air pressure is varied in the external auditory canal	Small probe placed in the ear canal and pressure varied in the ear canal	Tests for possible middle-ear pathology and pressure-equalization tube function	Not a test of hearing; depends on ear canal seal; high-frequency tone probe needed for infants younger than 6 mo

Adapted with permission from: Bachmann KR, Arvedson JC. *Pediatr Rev.* 1998;19(5):155–165.

is performed best in infants and young children while they are sleeping. If the test cannot be performed because of motion artifact, sedation may be necessary. The ABR is currently used in many newborn screening programs.

ABR and OAEs are tests of auditory pathway structural integrity but are not true tests of hearing. Even if ABR or OAE test

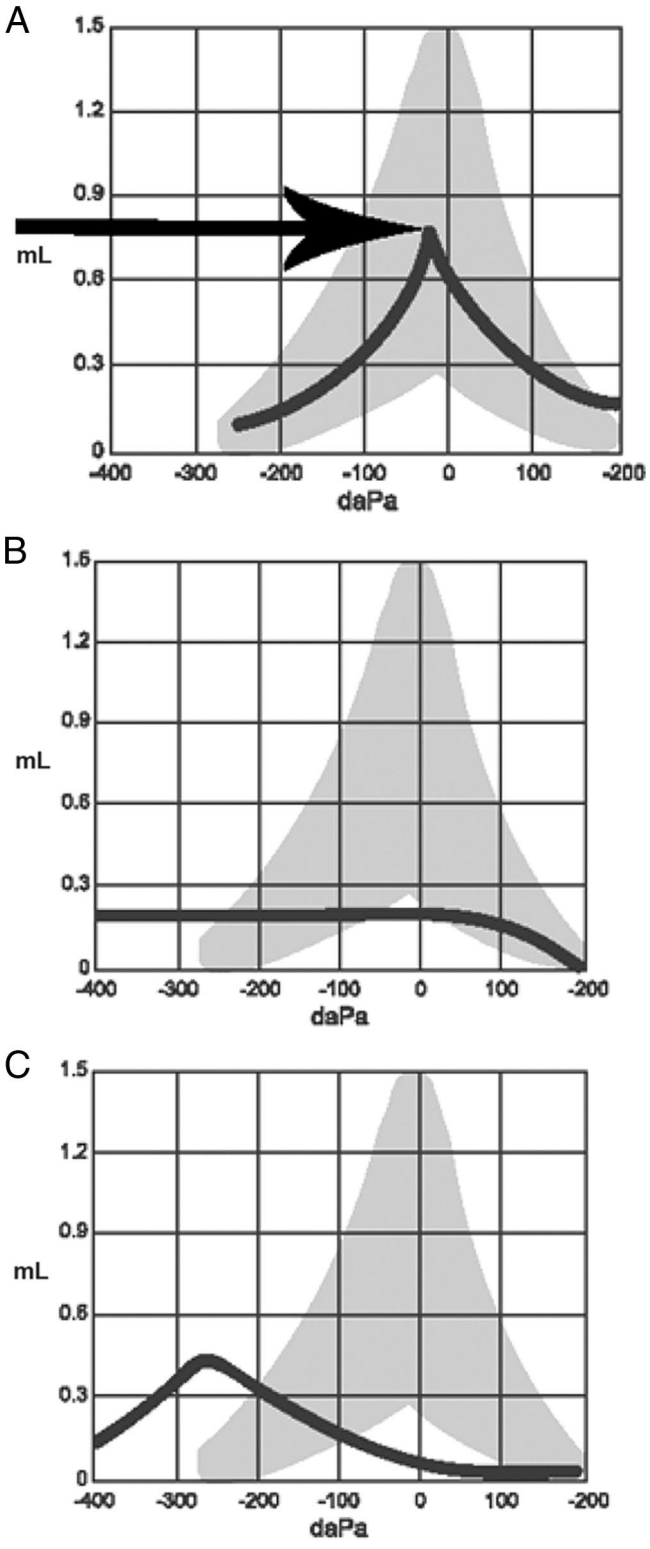

FIGURE 2
Tympanograms. Type A: normal. Type B: abnormal, needs medical attention. Type C, borderline normal; monitor; may need medical attention.

results are normal, hearing cannot be definitively considered normal until a child is mature enough for a reliable behavioral audiogram to be obtained. Behavioral pure-tone audiometry remains the standard for hearing evaluation. Hearing thresholds at specific frequencies can be determined, and the degree of hearing loss can be assigned. If there are distractions or the room is not sound treated, pure-tone audiometry in the office should be considered solely a screening test.

Play Audiometry

Children 2 to 4 years of age are screened or tested more appropriately by play audiometry. These children are conditioned to respond to an auditory stimulus through play activities, such as dropping a block when a sound is heard through earphones. Air-conduction hearing threshold levels of greater than 20 dB at any of these frequencies indicate possible hearing loss, and referral to a pediatric audiologist should be made.

Conventional Screening Audiometry

For children aged 4 years and older, conventional screening audiometry can be used. The child is asked to raise his or her hand when a sound is heard. The test should be performed in a quiet environment using earphones, because ambient noise can affect test performance significantly, especially at lower frequencies (ie, 500 and 1000 Hz). Each ear should be tested at 500, 1000, 2000, and 4000 Hz. Air-conduction hearing threshold levels of greater than 20 dB at any of these frequencies indicate possible hearing loss, and referral to a pediatric audiologist should be made.

If the child does not pass the screening, earphones should be removed and instructions carefully repeated to the child to ensure proper understanding and attention to the test and then re-screened with the earphones repositioned. A child whose repeat test shows hearing thresholds of greater than 20 dB at any of these frequencies, especially if there is no pathologic abnormality of the middle ear on physical

examination, should be referred for formal hearing testing. Key point 5: Developmental abnormalities, level of functioning, and behavioral problems (ie, autism/developmental delay) may preclude accurate results on routine audiometric screening and testing. In this situation, referral to an otorhinolaryngologist and a pediatric audiologist who has the necessary equipment and expertise to test infants and young children should be made (Table 5). Key point 6: The results of abnormal screening should be explained carefully to parents, and the child's medical record should be flagged to facilitate tracking and follow-up.

It is important to remember that a "fail" report on any 1 of a combination of tests warrants additional testing. It is also important to remember that failure of speech, language, and hearing screening assessments warrants additional testing (Tables 6–9).

TABLE 6 Ten Ways to Recognize Hearing Loss: Adolescents (11- to 21-Year Visits)

1.	Do you have a problem hearing over the telephone?
2.	Do you have trouble following the conversation when two or more people are talking at the same time?
3.	Do people complain that you turn the TV volume up too high?
4.	Do you have to strain to understand conversation?
5.	Do you have trouble hearing in a noisy background?
6.	Do you find yourself asking people to repeat themselves?
7.	Do many people you talk to seem to mumble (or not speak clearly)?
8.	Do you misunderstand what others are saying and respond inappropriately?
9.	Do you have trouble understanding the speech of women and children?
10.	Do people get annoyed because you misunderstand what they say?

Adapted from: National Institute on Deafness and Other Communication Disorders. *Ten Ways to Recognize Hearing Loss.* Bethesda, MD: National Institute of Health; 2006. NIH publication 01-4913. Available at: www.nidcd.nih.gov/health/hearing/10ways.asp.

Comprehensive Audiological Evaluation Using Physiologic and/or Behavioral Testing

The ABR test may be used as a diagnostic tool by audiologists for more definitive diagnosis of hearing loss. Usually performed in children in natural sleep up to approximately 3 to 6 months of age and then under sedation for older infants, diagnostic ABR can provide not only a general level of hearing but also frequency-specific hearing data. Diagnostic ABR is performed with different frequency tone bursts and across varying sound levels to effectively estimate an audiogram. Diagnostic ABR can also be performed with bone conduction to separate conductive from sensorineural hearing loss. Diagnostic ABR is often the definitive test used by audiologists in children and infants who are unable to cooperate with other methods of hearing testing. Audiologic evaluation using ABR or auditory steady-state response provides frequency-specific hearing thresholds by air and bone conduction in each ear separately. ABR is the gold standard for determination of hearing thresholds in infants younger than 6 months and in children who cannot be tested behaviorally.

Children as young as 6 to 24 months can be tested by means of visual reinforced audiometry (VRA). This technique conditions the child to associate speech or frequency-specific sound with a reinforcement stimulus such as a lighted toy or animated toy or video clips. VRA is performed by an audiologist with experience testing young children. This testing is not readily applied in screening programs, because infants younger than 6 months' developmental age cannot perform the task, and sound-treated rooms are needed. The results of VRA can approximate those of conventional audiometry.

Children with unilateral or mild hearing loss also should be evaluated further. Studies have shown such children to be similarly at risk of adverse communication skills as well as difficulties with social, emotional, and educational development.[16]

FOLLOW-UP AND DIAGNOSTIC TESTING

Key point 7: Any abnormal objective screening result requires audiology referral and definitive testing. Screening will only result in benefit for the patient if abnormal test results are confirmed and appropriate intervention is provided. Most studies that have evaluated the success rate of infant hearing screening programs have described a fairly high rate of failure to confirm a failed screen with definitive testing. A similar problem could also occur in screening older infants and children. Improving the physician's involvement not only in screening but also in arranging and confirming appropriate follow-up testing and intervention is necessary to achieve optimal speech, language, and hearing.

Key point 8: A failed infant hearing screening or a failed screening in an older child should always be confirmed by further testing. Audiologists may repeat the audiometric test as described above in a sound booth and using a variety of other tests. ABR can also be used for definitive testing of the auditory system. A diagnostic ABR is usually performed under sedation or general anesthesia in children aged approximately 3 to 6 months and older. The test is performed with frequency-specific stimuli and presentation levels to approximate hearing threshold levels. Diagnostic ABR provides information that is accurate enough to allow for therapeutic intervention. Hearing aids can be fitted with information obtained from a diagnostic ABR. Audiologic assessment and intervention is

TABLE 7 Developmental/Behavioral Screening Tools

Resource	Description	Age Range	Where to Find
General developmental screening tools			
Ages & Stages Questionnaire (ASQ)	A series of 19 questionnaires used to screen infants and young children for developmental delays during the first 5 y of life	4–60 mo	www.brookespublishing.com/tools/asq/index.htm
Ages & Stages Questionnaire: Social-Emotional (ASQ:SE)	A series of 19 questionnaires used to screen infants and young children at risk for social or emotional difficulties, to identify behaviors of concern to caregivers, and to identify any need for further assessment	6–60 mo	www.brookespublishing.com/tools/asqse/index.htm
Parents' Evaluation of Developmental Status (PEDS)	A method for detecting developmental and behavioral-emotional problems in children	Birth to 8 y	www.pedstest.com
Parents' Evaluation of Developmental Status-Developmental Milestones (PEDS:DM)	A collection of 6–8 items per age/encounter designed to replace informal milestones checklists with highly accurate items known to predict developmental status	Birth to 11 y	www.pedstest.com/dm
Autism-specific screening tools			
Checklist for Autism in Toddlers (CHAT)	A screening tool for early detection of autism	18 to ≥24 mo	www.autismresearchcentre.com/tests/chat_test.asp
Checklist for Autism in Toddlers (CHAT), Denver Modifications	CHAT scoring modifications	18 to ≥24 mo	
Checklist for Autism in Toddlers-23 (CHAT-23)	Combination of M-CHAT and CHAT items	16–86 mo	
Childhood Asperger Syndrome Test (CAST)	A parental questionnaire to screen for autism spectrum conditions	4–11 y	www.autismresearchcentre.com/tests/cast_test.asp
Modified Checklist for Autism in Toddlers (M-CHAT)	23-item scale pointing to express interest, responsiveness to name, interest in peers, showing behavior, response to joint attention, social imitation	16–48 mo	http://depts.washington.edu/dbpeds/Screening%20Tools/MCHAT.doc
Pervasive Developmental Disorders Screening Test-II, Primary Care Screener (PDDST-II PCS)	A parental questionnaire to screen for autism spectrum conditions	18–48 mo	www.pearson-uk.com/product.aspx?n=1315&skey=2960
Autism-specific or psychosocial screening tools			
Pediatric intake form from *Bright Futures*	Questionnaire to help gather a general understanding of the history, functioning, questions and concerns of the family	Birth to 21 y	www.brightfutures.org/mentalhealth/pdf/professionals/ped_intake_form.pdf
ADHD screening tools			
Vanderbilt rating forms	Parent- and teacher-completed forms that help a clinician diagnosis ADHD and to categorize the problem into 1 of its various subtypes	6–12 y	www.brightfutures.org/mentalhealth/pdf/professionals/bridges/adhd.pdf
AAP ADHD toolkit	A comprehensive toolkit developed from evidence-based guidelines for the diagnosis and treatment of children with ADHD; this resource toolkit contains a wide array of screening, diagnosis, treatment, and support materials for clinicians and other health care professionals	6–12 y	www.aap.org

ADHD indicates attention-deficit/hyperactivity disorder.

an ongoing process. The child requires regular audiologic reevaluations to determine if there is fluctuating or progressive hearing loss. Middle-ear monitoring is also essential. Hearing aid selection, fitting, verification, and validation require ongoing and regular visits with the audiologist. Candidacy for cochlear implantation should be considered when there is limited residual hearing or when progress with amplification is insufficient. Recommendations to the family regarding cochlear implantation should be based on a team evaluation that includes audiology, otology, psychology, speech-language pathology, and other intervention personnel.

Most providers of pediatric health care realize the importance of referring to an otolaryngologist, an audiologist, and a speech-language pathologist. Less recognized is the potential

TABLE 8 Guidelines for Children with Abnormal Speech Development

Age, mo	Referral Guidelines for Children With "Speech" Delay
12	No differentiated babbling or vocal imitation
18	No use of single words
24	Single-word vocabulary of ≤10 words
30	Fewer than 100 words; no evidence of 2-word combinations; unintelligible
36	Fewer than 200 words; no use of telegraphic sentences; clarity <50%
48	Fewer than 600 words; no use of single sentences; clarity ≤80%

Source: Matkin ND. *Pediatr Rev.* 1984;6:151.

TABLE 9 Guidelines for Children With Suspected Hearing Loss

Age, mo	Normal Development
0–4	Should startle to loud sounds, quiet to mother's voice, momentarily cease activity when sound is presented at a conversational level
5–6	Should correctly localize to sound presented in a horizontal plane, begin to imitate sounds in own speech repertoire or at least reciprocally vocalize with an adult
7–12	Should correctly localize to sound presented in any plane, should respond to name, even when spoken quietly
13–15	Should point toward an unexpected sound or to familiar objects or persons when asked
16–18	Should follow simple directions without gesture or other visual cues; can be trained to reach toward an interesting toy at midline when a sound is presented
19–24	Should point to body parts when asked; by 21 mo, can be trained to perform play audiometry

Source: Matkin ND. *Pediatr Rev.* 1984;6:151.

benefit of genetic and ophthalmologic evaluation of children and infants with sensorineural hearing loss.[17]

Especially in young infants and children, referral to an otolaryngologist for a complete diagnostic assessment is essential. More than 90% of the children with permanent hearing loss are born to "hearing" parents, resulting in a sense of urgency for parents of newly identified children with hearing loss to seek out answers regarding etiology, risk of progression, implications, and other questions. If children are indeed identified with sensorineural hearing loss, a variety of diagnostic tests can be recommended depending on the patient's history and physical examination. Otolaryngologists may play a role in diagnosis and treating middle-ear fluid or other middle-ear disorders as well as assisting in the definitive diagnosis of the cause of sensorineural hearing loss.[18] Diagnostic testing may include imaging of the temporal bone to identify structural defects; genetic tests, such as for abnormalities of the *Connexin* gene; and, occasionally, evaluation for other metabolic defects. Evaluation by a geneticist and genetic testing can be important for diagnosis as well as for providing the family with information for future planning purposes.

The hearing health care team (comprising the audiologist, otolaryngologist, teachers of the child with hearing impairment, speech-language pathologists, and other educational and medical personnel) should assist the family with intervention for hearing loss. Interventions may include observation with increased attention to speech and language development, hearing aids, auditory-assisted systems for the school environment, or more invasive surgical hearing devices such as cochlear implants or bone-anchored hearing aids. The goal is to provide families with appropriate options so that they may make well-informed decisions. Interventions should be driven by family desires and guided by accurate and timely information from all hearing-related health care professionals. Family goals and expectations are influenced by culture, parental education, level of income, availability of local resources, language in the home, and more. The role of the hearing health care team is to assist families in identifying all the options available to them and to support them throughout the ongoing decision-making processes that will occur throughout the child's development. All members of the hearing health care team, in conjunction with parents and on the basis of informed choice, should recognize that no decision regarding intervention is "final," and periodic opportunities should be identified for discussion regarding progress, alternative interventions, and new developments.

Medical follow-up includes ongoing evaluation and management of the adequacy of hearing rehabilitation; observation for potential complications of hearing rehabilitation, such as otitis externa and cerumen impactions; and monitoring for appropriate speech and language development.

Speech and language evaluation by a speech-language pathologist with training in working with children with hearing loss is also important for documentation of baseline speech and language skills and implementing a program of intervention that reflects the family's choice regarding language development.

At least one third of children with hearing loss will have an additional coexisting condition.[3] Because many causes of hearing loss are associated with abnormal ophthalmologic findings, formal ophthalmologic evaluation is appropriate, not only to assist with the diagnosis but also to optimize vision. A diagnosis of Usher syndrome with associated progressive hearing and vision loss may influence communication choices.

Children with hearing loss should also be monitored for developmental and behavioral problems (attention-deficit/hyperactivity disorder, autism, learning disabilities) and referred for

additional evaluation when necessary. Health care professionals can use screening tools to evaluate young children periodically for such concerns (Table 7) and refer for additional evaluation when concerns arise.

A medical professional should participate as an active member of a family's hearing health care team after diagnosis and provide input to assist in the adequacy of the rehabilitative efforts to monitor the child for progression and additional disabilities.

HEARING REFERRAL RESOURCES

Key point 9: Abnormal hearing test results require intervention and clinically appropriate referral, including otolaryngology, audiology, speech-language pathology, genetics, and early intervention. Pediatric health care professionals should maintain a list of referral resources available in their community for children with hearing loss and should advocate for increasing options and choices for families. Otolaryngologists, audiologists, and speech-language pathologists with special training and experience in treating children should be consulted for specific diagnosis, counseling, and treatment. Pediatric health care professionals should collaborate to refer the child for comprehensive educational counseling and treatment services. Communication among professionals caring for a child with hearing loss is essential to ensure appropriate case management.

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