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CLINICAL REPORT

Guidance for the Clinician in Rendering Pediatric Care

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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 as well as objective hearing screening of all infants and children can prevent or reduce many of these adverse consequences. This report outlines the risk indicators for hearing loss, provides guidance for when and how to assess hearing loss, and addresses hearing referral resources for children of all ages.

ABBREVIATIONS. AAP, American Academy of Pediatrics; ABR, auditory brainstem response; OAE, otoacoustic emissions; COR, conditioned oriented response; VRA, visual reinforced audiometry.

Failure to detect children with congenital or acquired hearing loss 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 has been demonstrated to prevent many of these adverse consequences and facilitate language acquisition.¹ Supportive evidence is outlined in the Joint Committee on Infant Hearing's "Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs," which was endorsed by the American Academy of Pediatrics (AAP).² This evidence also is part of the rationale for the AAP statement, "Newborn and Infant Hearing Loss: Detection and Intervention," 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.³ Furthermore, the AAP statement, "Recommendations for Preventive Pediatric Health Care," promotes objective newborn hearing screening as well as periodic

hearing screening for every child through adolescence.⁴

Pediatricians need to recognize children who are at risk for congenital or acquired hearing loss, be prepared to evaluate their hearing, and arrange for proper referral and treatment by identifying the availability of hearing resources within their communities. This report addresses these resources and offers specific guidelines to identify hearing loss in children of all ages.

RISK INDICATORS FOR HEARING LOSS

Significant hearing loss is present in 1 to 6 per 1000 newborns.⁵ Most children with congenital hearing loss have hearing impairment at birth and are potentially identifiable by newborn and infant hearing screening. However, some congenital hearing loss may not become evident until later in childhood. Hearing impairment also can be acquired during infancy or childhood for various reasons. Infectious diseases, especially meningitis and otitis media, are leading causes 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} Certain physical findings, historical events, and developmental conditions may indicate a potential hearing problem. These 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 speech-language-auditory milestones are included in Tables 1 and 2, respectively. Every child found to have 1 or more of the high-risk indicators in Table 1 should be followed and periodically screened for late-onset congenital or acquired hearing loss.

Although questionnaires and checklists are useful in identifying a child at risk for hearing loss, studies have shown that only 50% of children with hearing loss are identified by the comprehensive use of such questionnaires.^{8,9} Therefore, periodic objective assessment of the hearing of all children should be performed.

If a parent or caregiver is concerned that a child

The recommendations in this report do 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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TABLE 1. High-Risk Indicators for Hearing Loss

Checklist of high-risk indicators for hearing loss in children from birth to 24 mo of age. These indicators are red flags and may assist physicians in referring children for audiologic testing.

Birth to 28 d

- _ Family history of sensorineural hearing loss (SNHL), presumably congenital
- _ In utero infection associated with SNHL (eg, toxoplasmosis, rubella, cytomegalovirus, herpes, syphilis)
- _ Ear and other craniofacial anomalies
- _ Hyperbilirubinemia at levels requiring exchange transfusion
- _ Birth weight less than 1500 g
- _ Bacterial meningitis
- _ Low Apgar scores: 0–3 at 5 min; 0–6 at 10 min
- _ Respiratory distress (eg, meconium aspiration)
- _ Prolonged mechanical ventilation for more than 10 d
- _ Ototoxic medication (eg, gentamicin) administered for more than 5 d or used in combination with loop diuretics
- _ Physical features or other stigmata associated with a syndrome known to include SNHL (eg, Down syndrome, Waardenburg syndrome)

29 d to 24 mo

- _ Parental or caregiver concern about hearing, speech or language, and/or developmental delay
- _ Any of the newborn risk factors listed above
- _ Recurrent or persistent OME for at least 3 mo
- _ Head trauma with fracture of temporal bone
- _ Childhood infectious diseases associated with SNHL (eg, meningitis, mumps, measles)
- _ Neurodegenerative disorders (eg, Hunter syndrome) or demyelinating diseases (eg, Friedreich ataxia, Charcot-Marie-Tooth syndrome)

Adapted with permission from Joint Committee on Infant Hearing. Year 2000 position statement: principles and guidelines for early hearing detection and intervention. *Pediatrics*. 2000;106:798–817.

cannot hear, the pediatrician needs to assume that such is true until the child has been evaluated objectively. Parental concern is of greater predictive value than the informal behavioral examination performed in the physician's office. Parents often report suspicion of hearing loss, inattention, or erratic response to sound before hearing loss is confirmed.^{10,11} One study showed parents were as much as 12 months ahead of physicians in identifying their child's hearing loss.³ Any parental concern should be taken seriously, and formal hearing evaluation should be performed.

PHYSICAL EXAMINATION

A thorough physical examination is an essential part of evaluating a child for hearing loss. Findings on head and neck examination associated with hearing impairment 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 pigmentation of the skin, hair, or eyes also may be associated with hearing loss, as in Waardenburg syndrome. Abnormalities of the eardrum should alert the physician to the possibility of hearing impairment. A leading cause of acquired hearing impairment is otitis media with effusion (OME). Temporary hearing loss has been demonstrated during episodes of acute otitis media. The child with repeated or chronic OME is at high risk of acquired hearing

TABLE 2. Expected Speech-Language-Auditory Milestones

Checklist of selected speech-language-auditory milestones achieved by infants and children who have intact cognition and hearing. Failure to achieve these milestones by expected age ranges might relate to hearing loss that necessitates audiologic testing.

Birth to 3 mo

- _ Startles to loud noise
- _ Awakens to sounds
- _ Blinks or widens eyes in response (reflex) to noises

3 to 4 mo

- _ Quiets to mother's voice
- _ Stops playing, listens to new sounds
- _ Looks for source of new sounds not in sight

6 to 9 mo

- _ Enjoys musical toys
- _ Coos and gurgles with inflection
- _ Says "mama"

12 to 15 mo

- _ Responds to his or her name and "no"
- _ Follows simple requests
- _ Uses expressive vocabulary of 3 to 5 words
- _ Imitates some sounds

18 to 24 mo

- _ Knows body parts
- _ Uses expressive vocabulary 2-word phrases (minimum of 20 to 50 words)
- _ 50% of speech intelligible to strangers

By 36 mo

- _ Uses expressive vocabulary of 4- to 5-word sentences (approximately 500 words)
- _ Speech is 80% intelligible to strangers
- _ Understands some verbs

Adapted with permission from Northern J, Downs M. *Hearing in Children*. 4th ed. Baltimore, MD: Williams & Wilkins; 1991.

impairment and should undergo hearing evaluation.¹³ Pediatricians should be familiar with pneumatic otoscopy and tympanometry as useful diagnostic tools in the management of OME.

TOOLS FOR OBJECTIVE HEARING SCREENING

In addition to universal newborn hearing screening, objective screenings for hearing impairment 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."⁴ 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. Screenings should be conducted in a quiet area where visual and auditory distractions are minimal. Various audiologic tests are outlined in Table 3.

One objective means of evaluating hearing is the automated auditory brainstem response (ABR). This instrument measures ABRs at frequencies greater than 1000 Hz with a broadband click stimulus in each ear. The testing instrument incorporates a 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. Automated ABR can test each ear individually and can be performed on children of any age. Motion artifact interferes with test results. For this reason, the test is performed best in infants and young children while they are sleeping or, if neces-

TABLE 3. 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	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	Infant or child must be relatively inactive during the test; not a true test of hearing, because it does not assess cortical processing of sound
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 auditory stimuli presented through earphones 1 ear at a time	Ear-specific results; responses not dependent on patient cooperation	Infant or child must remain quiet during the test; not a true test of hearing, because it does not assess cortical processing of sound
9 mo to 2.5 y	COR or VRA, 30-min test	Behavioral tests measuring responses of the child to speech and frequency-specific stimuli presented through speakers	Both techniques condition the child to associate speech or frequency-specific sound with a reinforcement stimulus, such as a lighted toy; VRA requires a sound-treated room	Assesses auditory perception of child	Only assesses hearing of the better ear; not ear-specific; cannot rule out a unilateral hearing loss
2.5 y to 4 y	Play audiometry, 30-min test	Behavioral test measuring auditory thresholds in response to speech and frequency-specific stimuli presented through earphones and/or bone vibrator	Child is conditioned to put a peg in a peg board or drop a block in a box when stimulus tone is heard	Ear-specific results; assesses auditory perception of child	Attention span of child may limit the amount of information obtained
4 y to adolescence	Conventional audiometry, 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	Depends on the level of understanding and cooperation of the child

Adapted with permission from Bachmann KR, Arvedson JC. Early identification and intervention for children who are hearing impaired. *Pediatr Rev.* 1998;19:155-165.

sary, sedated. The ABR is currently used in many newborn screening programs.

Evoked otoacoustic emissions (OAE) 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 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, is performed quickly at any age, and is not dependent on whether the child is asleep or awake. Motion artifact does interfere with test results. The OAE is an effective screening measure for inner and middle ear abnormalities, because at hearing thresholds of 30 dB or higher, there is no OAE response. The OAE test does not further quantify hearing loss or hearing threshold level. The OAE 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 ABR test results.

The ABR and OAE are tests of auditory pathway structural integrity but are not true tests of hearing. Even if ABR or OAE test 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 impairment can be assigned. If there are distractions or the room is not soundproof, pure tone audiometry in the office should be considered solely a screening test.

Children as young as 9 to 12 months can be screened by means of conditioned oriented responses (CORs) or visual reinforced audiometry (VRA). Both of these techniques condition the child to associate speech or frequency-specific sound with a reinforcement stimulus, such as a lighted toy or dancing animal. The VRA is a more sophisticated and accurate form of COR requiring a soundproof room and is typically performed by an audiologist.

Children 2 to 4 years of age are 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.

For children 4 years and older, 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, 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 >20 dB at any of these frequencies indicate possible impairment.

Audiometric evidence of hearing loss should be substantiated by repeat screening. Earphones should be removed and repositioned, and instructions should be carefully repeated to the child to ensure

proper understanding and attention to the test. A child whose repeat test shows hearing thresholds >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. Children with unilateral or mild hearing loss also should be further evaluated; studies show such children to be similarly at risk for adverse communication skills as well as difficulties with social, emotional, and educational development.¹⁴

The results of hearing screening and ear examinations should be explained carefully to parents. The child's chart should be marked clearly to facilitate tracking of appropriate referrals, developmental skills, and school performance.

HEARING REFERRAL RESOURCES

Pediatricians should be familiar with the referral resources available in their community for hearing impaired children. Pediatric otolaryngologists and audiologists and speech and language pathologists with special training and experience in treating children should be consulted for specific diagnosis, counseling, and treatment. The primary care pediatrician and the otolaryngologist should collaborate to refer the child for comprehensive educational counseling and treatment services. Communication among professionals caring for a hearing-impaired child is essential to ensure appropriate case management.

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