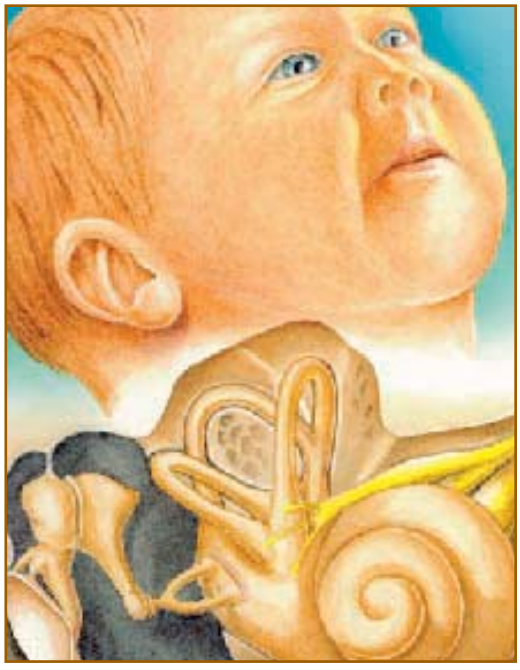


Assessing Hearing Disorders in the Young Child

An early diagnosis of hearing loss is critical in young children, as successful management interventions must be initiated early.

By Johnna A. MacCormick, MD, FRCSC

Presented at the University of Ottawa 49th Annual Refresher Course, Ottawa, Ontario, April 2000.

Statistics show six of every 1,000 newborns have a significant hearing loss.[1] Since hearing is essential for the normal development of speech and language, the consequences of a delayed diagnosis of hearing loss can be devastating. One of the most critical factors enabling successful management of a hearing-impaired child is the age at which special education is initiated.[2] Appropriate auditory stimulation prior to the age of 18 months is necessary to allow for normal language development. Delay beyond this age will likely prevent the child from attaining maximal language function.[3] In fact, in a study of 109 hearing-impaired children, when hearing loss was identified and aided before six months of age, the children scored significantly higher on tests of vocabulary, expressive language and language comprehension, as compared to those diagnosed after six months. Interestingly, these mean differences persisted beyond three years after the initial study.[4]

In the absence of hearing screening programs, severe-to-profound hearing loss may not be identi-

Dr. MacCormick is assistant professor, and director post-graduate medical education, department of otolaryngology, University of Ottawa, and chief, Division of Otolaryngology, Children's Hospital of Eastern Ontario.

fied until a child is 24 months or older, and often not until the age of five or six in milder cases. Early diagnosis of hearing loss has, until recently, depended on screening children identified by a high-risk registry (Table 1).[4] Unfortunately, this process has not been carried out uniformly in all centres. Furthermore, reports estimate approximately 50% of newborns with hearing loss of a moderate degree or greater have no known risk factors and, therefore, are missed by this screening technique.[5]

Hearing levels may be tested at any age, even in newborns, unless they are extremely premature. The Ontario Ministry of Health has allocated $7 million to establish a universal hearing program. The initial screening test will be otoacoustic emissions (OAEs), which specifically test cochlear function, primarily outer hair cell function. OAEs are low-level acoustic signals generated in the cochlea and reverse transmitted through the middle ear into the external ear, where they can be measured with a sensitive microphone. OAE responses are present in normal ears, but absent in ears with even mild hearing loss, which makes it an ideal screening tool. OAEs are of limited value, however, in quantifying the magnitude of hearing loss. Transient-evoked OAEs (TOAEs) are the response detected to a click presented to the ear. The universal screening protocol will use distortion product OAEs (DPOAEs), which enable different frequencies to be tested.

If a child does not pass the DPOAES (a "referred" outcome), he/she will have an auditory-evoked brainstem response (ABR). By using electroencephalogram (EEG) readings, repeated clicks (*i.e.*, 1,000 to 1,500) are presented to the ear of a sleeping child. The sum of synchronous neural discharges allow a wave-form to develop, which enables thresholds to be determined. The click thresholds respond to the 2,000 Hz to 4,000 Hz range. In order to test low-frequency hearing, a 500 Hz tone burst also is presented. This provides a very accurate assessment of hearing.

Summary

Hearing Disorders in the Young Child

- Appropriate auditory stimulation prior to the age of 18 months is necessary to allow for normal language development. Delay beyond this age will likely prevent the child from attaining maximal language function.
- Children who have been identified as having hearing loss by either otoacoustic emissions or an auditory-evoked brainstem response require a comprehensive auditory assessment.
- Aside from universal screening, anytime there is a parental concern, head trauma with temporal bone fracture or significant recurrent or persistent otitis media with effusion for greater than three months, an audiogram is warranted.
- Regardless of a child's age, once a significant hearing loss is confirmed, amplification and auditory-verbal therapy should begin immediately.
- Occasionally, a child will present with what appears to parents and teachers as a hearing loss, yet pure tone auditory testing is normal. This may be related to an idiopathic dysfunction of the central auditory nervous system.

Table 1

High-Risk Registry

Family history of childhood SNHL
In utero infection (*i.e.,* TORCH)
Craniofacial anomalies
BW < 1500 g
Hyperbilirubinemia at serum levels requiring exchange transfusion
Ototoxic medications, in multiple courses, or in combination with loop diuretics
Bacterial meningitis
Apgar scores 0 to 4 at one minute, or 0 to 6 at five minutes
≥ 5 days on ventilator
Stigmata of a syndrome known to be associated with hearing loss

SNHL = sensorineural hearing loss; TORCH = toxoplasmosis, other [congenital syphilis and viruses], rubella, cytomegalovirus infections, herpes simplex, syphilis; BW = birth weight

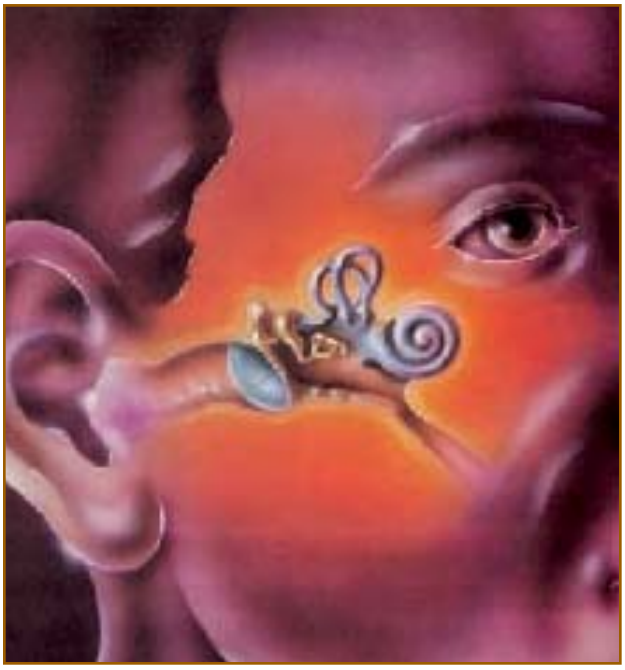
Adapted from: Joint Committee on Infant Hearing 1994 Position Statement. Pediatrics 1995; 95:152.

Children who have been identified as having hearing loss, by either OAEs or ABR, require a comprehensive auditory assessment. OAEs may be normal in certain situations, such as in cases of central auditory neuropathy. The classical example is extreme levels of bilirubin, leading to kernicterus, which may affect auditory brainstem pathways. Such patients may have robust OAEs, but may not produce an ABR. Rehabilitation with hearing aids on a normal cochlea is ineffective and potentially damaging.

As soon as a child is co-operative enough, behavioural testing should be performed by obtaining the traditional audiogram. These occasionally may be obtained after a child has reached six months of age, but tests can be performed reliably after 10 months. Visual reinforcement is used to encourage response to the presented noise. After 24 to 30 months of age, play audiometry may be used, and after three to four years of age, conventional audiometry be may be used.

Aside from universal screening, anytime there is a parental concern, head trauma with temporal bone fracture, or significant recurrent or persistent otitis media with effusion for greater than three months, an audiogram is warranted. The presence of speech delay also justifies auditory testing.

When dealing with otitis media with effusion, understanding the normal pattern of effusion clearance is important. Two weeks after an otitis



A cochlear implant may be considered for patients with profound bilateral sensorineural hearing loss who are receiving no appreciable benefit from a hearing aid.

media, 30% of effusions have cleared, 60% by four weeks, 80% by eight weeks and 90% by 12 weeks. No aggressive management is required until fluid has been present for three months. At this time, a two-week trial of second-level antibiotics is 30% effective in clearing the fluid. An otolaryngology and audiology referral would be appropriate at this time. Attention to environmental issues (*e.g.*, daycare, smoking) may be important. Temporary threshold shifts with recurrent acute otitis media, if frequent enough, also have been associated with language delay and may require intervention by an otolaryngologist.

Regardless of a child's age, once significant hearing loss is confirmed, amplification and auditory-verbal therapy should begin immediately. It is not necessary, or desirable, to wait to obtain reliable behavioural thresholds. Amplification is attempted cautiously, using ABR results as a guide, until further information becomes available. Unilateral hearing loss not amenable to amplification, or hearing loss occurring in isolated frequencies, may respond well to a frequency modulation (FM) system in the classroom.

When a child is diagnosed with sensorineural hearing loss (SNHL), the goals are to determine the type and degree of hearing loss, identify the etiology (focusing on treatable causes, such as syphilis) and provide early amplification and management. Usually, newly diagnosed children undergo a battery of tests, including blood, urine, electrocardiogram (ECG) and, sometimes, computed tomography (CT) scanning to assess for potential causes and other end-organ effects (*i.e.*, nephrotic syndrome in Alport's disease). In selected cases, genetic testing for Connexin 26 may be warranted to aid in future family planning and gain information about the potential progression of hearing loss (the genetic hearing loss affected by these alleles does not tend to deteriorate). An etiology is usually identified in less than 50% of children.

Occasionally, a child will present with what appears to parents and teachers as a hearing loss, yet pure tone auditory testing is normal. This may be related to an idiopathic dysfunction of the central auditory nervous system, referred to as central auditory processing disorder (CAPD). This can be isolated or it may co-exist with attention deficit disorders (ADHDs), learning disabilities and language disorders. Typically, these children have difficulty in perceiving spoken language, especially in the presence of background noise. They may appear to have a reduced capacity to remember verbal tasks and often have prob-

lems in the classroom understanding instructions from the teacher. Such difficulties may result in frustration and behavioural and motivational problems in the classroom. The actual diagnostic tests are time-consuming and complex, usually requiring a maturity of at least seven years of age. Few audiology centres offer this testing. If suspected or confirmed by testing, management principles should focus on optimizing the signal-to-noise ratio (*i.e.*, preferential seating). An increased reliance on visual cues and occasional FM amplification systems may be useful. Most of these children seem to develop compensatory skills as they get older. This process may be hastened by auditory training therapy.[6]

A cochlear implant may be considered for patients with profound bilateral sensorineural hearing loss who are receiving no appreciable benefit from a hearing aid. A highly motivated family is crucial to a good outcome, as a phenomenal amount of verbal therapy is required post-implant.

The possibility of hearing loss should always be considered in high-risk patients, those with language delay or concerned parents. It is never too early, nor are reasons ever too trivial, to consider audiology testing. Even after commencing universal screening, there will be a number of older children who have not been screened, and yet have a significant neurosensory hearing loss. Once the diagnosis has been made, it is critical for the primary medical caregiver to provide support during the adjustment phase and ongoing-therapy phase, as the process initially can seem very overwhelming to families. With aggressive management, language and educational outcomes can be optimized. CME

References

1. Joint Committee on Infant Hearing 1994 Position Statement. Pediatrics 1995; 95:152-6.
2. Levitt H, McGarr NS, et al: Development of language and communication skills in hearing-impaired children. Rockville, MD: American Speech-Language Hearing Association: 1987, ASHA Monograph No 26.
3. First LR, Palfrey JS: The infant or young child with developmental delay. N Engl J Med 1994; 330:478-83.
4. Yoshinaga-Itano C, Sedey M, Apuzzo A, et al: The effect of early identification on the development of deaf and hard-of-hearing infants and toddlers. Poster presented at American Academy of Audiology Conference, Salt Lake City, 1996.
5. Mauk G, White KR, Mortenson LB, et al: The effectiveness of screening programs based on high-risk characteristics in early identification of hearing impairment. Ear Hear 1991; 12:312-9.
6. Chermak GD, Musiek FE: Managing central auditory processing disorders in children and youth. Am J Audio 1992; 1(3):61-5.