The hearing-impaired child

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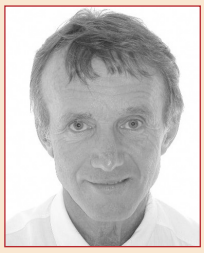

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Spoken language remains the most important means by which human beings communicate with one another. The recent explosion of communication via the electronic media is subtly changing this, bringing in a more visual component, but this will only have a major impact in years to come and will serve to complement rather than replace spoken language, which will remain central to our communication at an interpersonal level. However, the skills required for speech are not innate, and have to be acquired during the early years of life. In order for this to happen, one not only requires a brain with the necessary processing abilities, but also in particular an auditory system capable of picking up the acoustic information which is fundamental to the whole process. In a nutshell, the individual needs to be able to hear.

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Data from the recent literature indicate that the first 6 months are critical to the long-term development of language within the normal range. There is also compelling evidence that the difference between early and late identification and management of a hearing loss in children (before 6 months versus after 6 months of age) results in measurable delays of language development from 12 months of age, that do not disappear with age or intervention duration. There is thus a window of opportunity for language development with a 'use it or lose it' condition implicit, and the longer one waits, the less satisfactory the end result.

The acquisition of speech and language communication skills is also of paramount importance for children as they will use these later in order to obtain an education. It follows that children in whom this communication system has been suboptimally developed will also be disadvantaged in a school system relying heavily on the spoken word as the medium of instruction. Social and behavioural aspects of development are also affected by a hearing loss, giving added significance to early identification and intervention.

DIAGNOSIS

It has become universally accepted that early identification and intervention in children with a hearing loss is the key to a successful outcome. The ideal would naturally be universal newborn screening. Until this is a reality, the general practitioner in particular must be alert to the possibility of a hearing loss, and know how to proceed. There are some basic principles which assist in sensitising us to suspect all is not well with a child's hearing:

- Listen to the mother. Nobody is as sensitive to a child's behaviour as the mother. The most common way in which these children present is when the mother suspects a hearing loss. If a mother thinks that her child cannot hear, we must assume she is correct until we can prove that it is not so.

Age	Developmental milestones
< 3 months	Startled by loud sounds
6 months	Localises sounds
9 months	Responds to name and mimics environmental sounds
12 months	First meaningful words
18 months	Follows commands and performs simple tasks
	Able to react rapidly to sounds from any direction
24 months	Twenty words in the vocabulary of monolingual children

This is a basic responsibility of any medical practitioner.(De Lange C. Personal communication.)

- Hearing-specific developmental milestones. Knowledge of normal developmental milestones allows one to judge not only whether or not a loss is present, but also to assist in the determination of the approximate age at which the hearing loss occurred. Hayes and Northern[1] have tabled the following guidelines:
- Risk factors. A high-risk register consisting of 7 items was suggested by United States Joint Committee on Infant Hearing in 1982 as a primary means of identifying children likely to be born with hearing impairment. The acronym HEARING is useful in remembering the list:

H — Heredity: Family history of congenital sensorineural hearing loss. Consanguinity: Populations in which consanguineous marriages are common may have a prevalence of 12/1 000 births.

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E — Ear: Congenital defects of ear, nose and throat structures. Craniofacial anomalies.

A — Asphyxia: Severe neonatal asphyxia, Apgar < 5 at 1 minute and < 7 at 5 minutes.

R — Ototoxic drugs taken during pregnancy or in the neonatal period.

I — Infections: Congenital pre- or perinatal infection (e.g. Storch diseases — syphilis, toxoplasmosis, rubella, cytomegalovirus and herpes). Bacterial meningitis – all children with bacterial meningitis should have their hearing tested as soon as possible after recovering from the disease. Approximately 8% of these patients develop sensorineural hearing loss.

N — Neonatal intensive care, during the newborn period, with mechanical ventilation for > 4 days. These children are at special risk for hearing loss, resulting in 1 child in 50 from intensive care units being hearing impaired. If the child was treated in an incubator for some time, the hearing should be tested.

G — Growth: Birth weight less than 1 500 g. All premature babies should be tested for hearing loss. Also consider possible hearing loss in children with growth retardation.

INITIAL EVALUATION

History

- Developmental history. The onset and progression of hearing loss is important in establishing possible aetiologies.
- Family history. Most inherited forms of hearing loss are unrelated to syndromes. However, some hereditary aetiologies non-syndromic in nature have implicated the Connexin 26 and 30 genes, and the list of genes involved with hearing is ever growing.
- Prenatal history. This is a time of complex embryogenesis and fetal development, and the possibility of chemical and biological teratogenesis should be evaluated. This includes not only the previously mentioned infections, but also other substances such as alcohol, tobacco, illicit drugs, retinoids, chemotherapeutic agents, ototoxic medications, etc.
- Perinatal history. In addition to the factors already mentioned as being high-risk, hyperbilirubinaemia has also been associated with hearing loss in infants, particularly if severe enough to require transfusions.
- Childhood history. Hearing loss that develops after the neonatal period may be congenital or acquired. Important aetiologies to consider are the following:
  - trauma
  - noise-induced loss
  - postnatal infections, e.g. mumps, measles, rubella, encephalitis and meningitis
  - ototoxic medications.

Examination

A thorough head and neck examination for craniofacial anomalies must be done.

The eyes must be examined for hypertelorism, coloboma, etc. An otological examination must be performed, as even wax can cause a meaningful hearing loss. Examination of the ears is vital before testing hearing in order for the results to be relevant.

Special investigations

These will in general be dictated by the findings after history taking and physical examination, and include serological, haematological, endocrinological, radiological investigations, etc.

HEARING TESTS

There is no reason to delay the testing of a baby's hearing because of age. In fact, the auditory brainstem response (ABR) has been detected in human neonates as early as 25 weeks' gestational age, and is not affected by sleep, sedation or attention. Ratification of any suspicion of a hearing loss is therefore always possible, but there are other ways in which one can assess hearing clinically in the young child.

Behavioural measures

The appropriate technique to be used is based largely on the developmental level of the infant or child. In children who are otherwise developing normally, the following can serve as guidelines:

From birth to 6 months — behavioural observation. Infants respond reflexively to sounds with blinking or widening of the eyes, turning towards the sound, or even with a full body startle to very loud sounds. There may also be changes in ongoing behaviour, such as increases or decreases in sucking or motor activity rate. There are however many limitations to this way of assessment, including response variability and habituation, and it is only really useful as a supplement to the observations already reported by the parents.

From 6 months to 2.5 years — visual reinforcement (VRA) or distraction audiometry. At about 6 months of age, the normally developing infant begins to localise sound on a horizontal plane. VRA uses the infant's natural tendency to turn to a sound source by rewarding head turns with an attractive visual stimulus like a nice toy. The sound is presented from behind to the child's ear while he is sitting on his mother's lap and being distracted from the front.

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This test can be quite reliably performed in the consulting room using the high-frequency rattle. Under soundproof conditions when done by an audiologist a very reliable audiogram can be obtained using these principles.

From 2.5 years on — conditioned response (play) audiometry (CPA). This entails teaching the child to perform a motor activity in response to sound. The task should be consistent with the motor development and interest level of the child, and may include activities such as putting a peg in a board or fitting a piece into a puzzle.

From 5 years on — routine pure-tone audiometry should be possible.

Physiological measures

Auditory brainstem response

As already mentioned, ABR can be measured from birth. It is a series of electrical potentials that can be recorded from the scalp during the first 10 - 20 milliseconds following the onset of a transient stimulus. It is ideal for testing auditory sensitivity in infants and children who, because of developmental stage or handicapping conditions, cannot be tested reliably using conventional behavioural techniques. The stimulus can be a click or tone bursts, and can even be presented via bone conduction. In these latter cases there is an added advantage that contralateral masking is not needed, and the test can be done on patients with effusions and congenital middle ear anomalies.

Otoacoustic emissions (OAEs)

These are low-level sounds originating within the cochlea. Through reverse propagation some of this acoustic energy leaks from the cochlea and travels through the middle ear to the external auditory meatus where it can be recorded using a sensitive microphone. OAEs are believed to be the acoustic byproducts of outer hair cell motility. They are detectable only when both the cochlea and middle-ear systems are functioning normally or near normally. Because they are recorded quickly and easily they are being used widely in paediatric audiology, particularly for infant screening. It is however important to be aware of their limitations, i.e. they are absent when there is fluid (or meconium) in the middle ear, which is very common in neonates, and they also do not measure hearing as such. Patients with some form of auditory neuropathy can be profoundly deaf, with functioning outer hair cells and hence OAEs. Therefore, they should not be done without a clinical evaluation of the middle ears, and if done in isolation they will miss a neural hearing loss.

Tympanometry

Tympanometry measures the relative change in middle-ear compliance as air pressure is varied in the external ear. It allows the objective assessment of the condition of the middle ear, i.e. whether or not fluid is present, and what the relative pressure of the air in the mid-

dle ear is. Acoustic reflexes, when they can be measured, can also give an indication as to hearing thresholds.

INTERVENTION

As emphasised earlier, our greatest responsibility is to recognise a hearing impairment in a child, and to institute treatment as soon as possible after birth. The magical age of 2 years is often given as the age before which such treatment must be instituted if a successful outcome is to be achieved as far as the development of communication skills is concerned. The danger therein, however, is that the urgency of even earlier identification becomes lost, which would be a tragedy. From the time we are born, the learning process begins immediately. Even those infants with a moderate hearing loss for the first 6 months of life will have a measurable deficit in language acquisition when compared with their normal-hearing peers at 12 months of age. The message should therefore be that we have a window of opportunity in the early years of life for acquiring these skills, which slowly but continually closes over time, and cannot be reopened.

Once recognised, treatment of a hearing impairment must be instituted immediately. For the general practitioner this would ideally be referral to an otorhinolaryngologist. A clinical (as opposed to audiometrical) assessment of the whole auditory mechanism is essential to ensure that not only potentially harmful pathology is treated, but also that normal auditory mechanisms are functioning optimally before rehabilitation processes are embarked upon. As an example, even the simple procedure of draining middle-ear fluid can improve the hearing by 20 – 30 dB, which in some cases may make hearing aids unnecessary, while in others render previously ineffective amplification very beneficial.

Once the medical and otological issues have been addressed, the further rehabilitative process should proceed under the guidance of adequately trained speech, language and hearing therapists. This involves a multidisciplinary team with the parents forming an integral part of the whole process.

An effort is made to optimise the use of what remains. Hearing-aid technology has made enormous strides over the past decades, and the quality of hearing provided and increasing social acceptability of the newer aids have resulted in far more satisfactory results. The advent of cochlear implants has also made sound accessible to those children who previously could not benefit even from the most powerful hearing aids. Numerous studies, including those from the Tygerberg Hospital Cochlear Implant Programme, have shown that young, congenitally deaf children who undergo cochlear implantation have the ability to learn language at rates comparable with those of their hearing peers. The implication is thus that even the most profound hearing loss can be overcome and permit normal speech and language development. Early diagnosis, as well as medical and acoustic intervention are however only a starting point, making the subsequent management possible.

Further optimisation of the child's learning environment will be an ongoing process, with an ultimate highly rewarding result. However, it is this process which will determine the success or failure of the child's development, and we must ensure that our patients are guided in the right direction.

There is no longer any doubt that early diagnosis and intervention in children with a hearing loss, no matter how severe, can result in a favourable outcome regarding their ability to communicate via the spoken word. The profound effect that this can have on the rest of their lives makes it imperative that we all remain ever watchful for a hearing loss in children.

FURTHER READING

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IN A NUTSHELL

Acquisition of speech and language communication skills is dependent on the ability to hear.

The earlier a hearing loss is identified and treated, the better the outcome regarding language development.

Hearing can be tested by physiological means from birth using the auditory brainstem response (ABR).

A mother's expressed suspicion that her baby cannot hear places the responsibility on the practitioner to confirm or disprove it, and to provide appropriate advice.

Children particularly at risk for hearing loss should be screened for this as a matter of routine.

The advent of newer technology (hearing aids and cochlear implants) has made access to sound possible for virtually all children.

Appropriate early intervention by a multidisciplinary team provides an excellent outcome for children with a hearing loss.