ABC of ENT

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DEAFNESS IN CHILDHOOD

Without hearing a child cannot acquire language, nor can he develop normal relationships. One to two of every 1000 liveborn infants suffer from deafness. Childhood deafness may be congenital or acquired. Congenital deafness may be (a) genetic, due to inherited defects; (b) prenatal, due to events during pregnancy such as rubella or other virus infections or the effects of drugs such as thalidomide that cross the placental barrier; or (c) perinatal, due to events at or around the time of birth. These include birth trauma, anoxia, and haemolytic disease of the newborn with kernicterus.

Deafness in the first few years of life may be due to virus infections such as measles or mumps, meningitis, ototoxic antibiotics, or other causes already discussed in relation to adults.

Screening

The aim should be to recognise every deaf child by the age of 8 months to 1 year—before the vital time for learning speech is wasted. Any medical or surgical causes need treatment, and training in acquisition of language should be started as soon as possible.

Certain high-risk groups may be recognised. They are likely to be seen in paediatric clinics, from where they will be referred, if thought to be deaf, to the audiology clinic. Those at risk are: those with a family history of deafness, those born after maternal problems in pregnancy, those with perinatal problems, those with cerebral palsy or meningitis, and those whose speech is delayed or faulty. Any child whose mother believes it to be deaf should also be suspect. Mothers are rarely wrong in this suspicion.

Screening of these high risk children will nevertheless overlook one-third of deaf children and, ideally, screening should be done by examining all infants in the population at child health clinics. Hearing *must* be assessed at 8 to 9 months and again on school entry. An additional assessment at 2-3 years is highly desirable. If deafness is suspected the infant is referred to an audiology centre for assessment by a senior clinical medical officer or a paediatric audiologist working with an educational audiologist or teacher of the deaf.

Screening methods

Screening methods depend on age. During the first 6 months there should be a "startle" or auropalpebral reflex in response to short bursts of high-intensity sound. A mother's observations are very important. The startle test produces many false-positive responses.

The second 6 months of life are the most important because hearing is easily testable and any loss discovered is early enough for help. At this age the normal child is sitting up and can be tested by distraction techniques. Appropriate sounds, presented at ear level about three feet away cause a normal child to turn towards them. The sound must be meaningful to the child. Low frequencies can be produced by the spoken voice, high frequencies by a special rattle. Crinkled tissue paper provides a broad-spectrum stimulus. These distraction techniques need two assessors—one to engage the child's attention (but not too much) and the other to make the noise. The assessor has to ensure that the response is to sound and not to movement seen or felt.

Audiometry, training, and aids

From 12 months to 2 years children become uncooperative and difficult to test. Quiet speech, with an expectation of comprehension, is an appropriate stimulus.

By the age of 3 children are once again co-operative but any severe hearing loss should have already been discovered. In addition to assessing speech comprehension pure-tone audiometry is possible using free-field sounds, then an earphone can be held to the ear, and finally earphones may be fitted. The child is conditioned to react to the stimulus—for example, by putting a brick into a box when he hears the sound.

Techniques with electric response audiometry (including electrocochleography) and other measurable responses to sound stimuli are available in special clinics when simpler tests give equivocal results and accurate information is needed. Such assessment may be necessary if suspected hearing loss is associated with other disabilities. No child is too young or too difficult to test with these techniques.

At all times the possibility of surgical correction of conductive deafness, such as that produced by glue ear, must be kept under consideration. Acquired middle ear effusions, so common in childhood, may exacerbate existing sensorineural hearing loss.

Once deafness is diagnosed the child is referred to an otologist in an ENT department for assessment of cause and possible treatment. Concurrently the teacher of the deaf plans auditory training and arranges the fitting of a hearing aid, and the child's progress will be regularly assessed in the audiology clinic. Between the ages of 2 and 3 years decisions about the child's future education are made. The goal is to achieve competence in language by the age of 5.

Auditory training involves exposing the child to normal speech sounds so that with the help of an aid his residual hearing can be used in learning to discriminate speech differences and in monitoring his own attempts at imitation. Parents need to talk as much as possible to their child in a normal way.

Children usually need higher gain from their hearing aids than adults and the fact that the child is growing produces difficulties in mould insertion and instrument management. Although very powerful aids are often needed there is no proof that they may damage residual hearing, though investigations are continuing.

Education

Deaf children may be educated in a normal school, in a partially hearing unit in a normal school, or in a school for the deaf. About half the deaf children attend normal schools, where they are seen by visiting teachers of the deaf, who advise on use of hearing aids, help with lip reading, and assess progress. A very deaf child has great difficulty in using a hearing aid with the background noise of a normal classroom.

Partially hearing units, which are increasing in number, offer the facilities of special classrooms, like those in schools for the deaf, within a normal school. Special schools for the deaf take about half the deaf children, usually from 3 to 18 years. Their classrooms are fitted with induction loops allowing the induction coil of the hearing aid to respond to the teacher's voice transmitted over a microphone while the child moves around. Better reception is gained from a radiomicrophone system, where the teacher speaks into a light-weight microphone and the sound transmitted is received by a radioreceiver worn by the child. Various arrangements take the received signal to the child's hearing aid.

Swimming with grommets

The consensus emphasis, in education in the UK, is on oral methods, whereby the development of language and speech is encouraged with the use of residual hearing and amplification by hearing aids. Alternative (manual) forms of communication have certain disadvantages, in particular the need for long and repeated practice and the possibility of using skills only with others similarly trained. Combinations of oral and manual methods are termed "total communication." Forms of manual communication include: finger spelling—the traditional deaf and dumb alphabet; cued speech, in which the signs made near the mouth are used to resolve ambiguities in lip reading; and signing systems, such as the British and Paget-Gorman systems, in which signs indicate words, ideas, and grammatical distinctions. The majority view in Britain favours an exclusively oral approach.

Swimming is always an important issue for children. With a grommet in place the ear must be protected against water. Swimming in swimming baths should certainly be banned because the chemical irritant of chlorinated water in the postnasal space is a potent predisposing factor in secretory otitis media and acute otitis media even in ears without grommets.

A child with a grommet may probably bathe safely in the sea but parents should be warned not to allow submersion or diving. The drum can be protected during bathing with a large piece of cotton wool, moistened with petroleum jelly in the external meatus. Fitted plugs to seal the meatus can be made by hearing aid dispensing firms and provide better protection for an ear with a grommet in place. A child with a grommet who develops otitis media will suffer less pain than one with an intact drum but discharge may be profuse. The usual treatment for otitis media with systemic antibiotics should be given and the ear mopped dry followed by instillation of antibiotic and steroid drops.

Acquired conductive deafness

The commonest cause of acquired deafness in childhood is secretory otitis media. When first discovered it is reasonable to wait four to six weeks in the hope that the effusion may resolve. During this time decongestion of the nasopharyngeal mucosa may be useful and this may be encouraged by oral antihistamines together with decongestant nosedrops—1% ephedrine in normal saline or xylometazoline hydrochloride (Otrivine)—provided they are not given for over two weeks continuously. If the effusion persists operation will be needed. Myringotomy is performed under general anaesthesia, the effusion aspirated, and a grommet inserted through the myringotomy incision. The grommet allows air into the middle ear, a role eventually performed by the Eustachian tube.

A grommet will become blocked by wax and surface keratin on average about 6 to 9 months after insertion. Then it will become embedded in wax and gradually be extruded along the external ear canal to the exterior, where it will fall out or be easily removed with forceps.

Sometimes the condition recurs and repeat insertion of a grommet may be needed, occasionally up to five times. Little can be done to prevent recurrence. The role of adenoidectomy is controversial but it may occasionally help.

The photographs of a distraction technique and of audiometry were reproduced by permission of Mr J A M Martin, director of the Nuffield Hearing and Speech Centre and the illustration of the hand alphabet by permission of the Royal National Institute for the Deaf.

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