ABC of ENT

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DEAFNESS IN ADULTS

Deafness is loss of hearing, regardless of its severity. The effects depend on the severity of the loss, the rate of onset, whether one or both ears are affected, and the age of onset. A baby born deaf in both ears cannot learn to speak without special help, and normal language development will be impossible if the deafness is severe. A child with good speech will almost certainly lose it if severely deafened in the first years of life. An adult who becomes very deaf does not lose his vocabulary, but the lack of auditory feedback degrades his voice into a harsh flat monotone. Rapid total deafness in both ears is a catastrophe that affects every aspect of the victim's life, while gradually developing loss causes serious but less severe problems. By contrast, total loss of hearing in one ear is relatively trivial, regardless of age.

Deafness is the cruellest form of sensory deprivation. Unlike blindness, it often provokes ridicule rather than sympathy; unable to hear what is said and unable to control his own voice, the severely deaf person appears stupid. Isolated from family and friends and greeted by unsympathetic attitudes, he is often depressed. Tinnitus, which often accompanies deafness and is rarely found without it, can cause distress almost as great as the lack of hearing.

The prevalence of hearing loss is not accurately known: probably over 3 million adults (6 in every 100 in the UK) have impaired hearing, and over 10 000 children need special education.

The two main types of defect are conductive and sensorineural. Conductive deafness arises from any impediment to the transmission of sound waves through the external ear canal and middle ear, as far as the footplate of the stapes. Sensorineural deafness implies a defect central to the oval window: in the cochlea (sensory), the cochlear nerve (neural), or, more rarely, in central neural pathways.

Conductive deafness

In conductive deafness there are five possible mechanical defects; (a) obstruction of the external ear canal; (b) perforation of the ear drum; (c) discontinuity of the ossicular chain; (d) fixation of the ossicular chain; (e) inadequacy of the Eustachian tube.

Obstruction—The external ear canal is most commonly obstructed by wax, but also by inflammation of the meatal wall and accumulation of discharge in otitis externa. Less common causes are congenital abnormalities of development (atresia), and foreign bodies.

Perforation of the ear drum impairs the transmission of sound by offering a reduced area for incident sound waves; by allowing sound pressure to be exerted adversely on the inner side of the membrane; and by exposing the round window membrane to incident sound pressure, which counters the normal route of transmission within the cochlea. Perforations usually result from infective damage but also from trauma, especially flat-handed blows to the ear, and more rarely, after sudden pressure changes during diving.

Discontinuity of the ossicular chain is usually a sequel to infective middle ear damage. The long process of the incus is most often destroyed. Injuries to the head or ear may dislocate one ossicle from another.

Fixation of the ossicular chain is the characteristic feature of otosclerosis. This inherited disorder causes the footplate of the stapes to adhere to the oval window. No other part of the ossicular chain is affected. Otosclerosis must not be confused with tympanosclerosis, in which hyaline material is deposited under the mucosa of any part of the middle ear cleft, after repeated episodes of inflammation. These deposits may often be seen as "chalk patches" in the ear drum and they may restrict movement in any part of the chain.

Inadequacy of the Eustachian tube is common in children and is accompanied by secretory otitis media with accumulation of extremely viscous material in the middle ear—so called glue ear. Air is absorbed from the middle ear cleft; the tympanic membrane is pushed inwards by outside air pressure and its free vibration impaired; fluid then accumulates within the middle ear. "Glue" ear is the commonest cause of acquired deafness in school-age children. Apart from faults in the maturation of the normal tube opening, the nasopharyngeal orifice of the tube may be obstructed. The role of enlarged and repeatedly infected adenoids in preventing ventilation of the middle ear is controversial. Effusions in the adult middle ear are usually thin and serous. Although they often follow upper respiratory tract viral infections or barotrauma during aircraft descent, *carcinoma of the nasopharynx must be excluded* since it may present with a conductive deafness due to a middle ear effusion.

Sensorineural deafness

The three main patterns of sensorineural deafness are; bilateral and progressive, unilateral and progressive, and sudden.

The commonest reason for bilateral progressive sensorineural deafness is degeneration because of age—presbycusis. A similar progressive hearing loss is sometimes seen in middle age as "presenile" degenerative change. Bilateral progressive hearing loss may also be caused by noise and by ototoxic drugs. Excessive noise damages the hair cells of the organ of Corti; the damage may follow brief high intensity exposure (acoustic trauma) but is usually caused by high intensity exposure over long periods. Such noise-induced hearing loss is important in industry and is a hazard of noisy hobbies such as shooting and using power tools. The severity of damage depends on the intensity of the noise, duration of exposure, and individual susceptibility.

Ototoxic drugs—notably the aminoglycoside antibiotics—also destroy cochlear hair cells. Theoretically these antibiotics constitute a risk only when administered systemically, but topical applications—for example, of neomycin to large raw areas of body surface or to the bronchial tree as insufflations—are dangerous. Risks are greater in the elderly, those with impaired renal function, and after previous or concomitant ototoxic drug administration. Damage is irreversible and may continue after treatment has stopped. Some common diuretics—frusemide, when given intravenously, and ethacrynic acid—may be ototoxic. The well-known ototoxic effect of large doses of salicylates is usually reversible.

When deafness is unilateral and progressive, Menière's disease (endolymphatic hydrops) or an acoustic neuroma should be considered.

Sensorineural deafness with a sudden onset is fortunately usually unilateral. One cause is trauma to the head or ear; if there is a leak of perilymph from the oval or round window membranes this may be surgically corrected. Other causes include viral infections (particularly mumps, measles, and varicella zoster) or sudden impairment of cochlear blood flow. Sudden hearing loss may announce the presence of an acoustic neuroma.

Syphilis must always be considered with any pattern of acquired sensorineural hearing loss. In the past few years increasing numbers of cases of labyrinthine syphilis have been recognised. Serological investigations are essential whenever a reasonable explanation for the loss is lacking.

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