

Treating childhood hearing loss

Sensorineural hearing loss

GENERALLY, after the audiological workup is complete, a child will be referred to Australian Hearing for a hearing aid assessment and fitting, although deaf parents with a deaf baby may opt for the use of sign language.

It is critical to continue ongoing surveillance for progressive or acquired hearing loss by vigilance and frequent hearing assessments. The GP can be invaluable, monitoring the child's progress and treating intercurrent conditions such as wax impaction and OME, both of which can significantly impact on the efficient use of hearing aids.

With the coming widespread introduction of universal newborn hearing screening programs in Australia, it is expected that many more hearing-impaired babies will be diagnosed and habilitation started before the critical age of six months. As part of the diagnostic workup these babies will be seen by audiologists, otologists, geneticists and teachers of the deaf to define the degree and type of hearing loss, the possible cause, other associated conditions and the most appropriate mode of habilitation.

The expectation is that, with early habilitation of sensorineural hearing loss, more children will be able to enter mainstream schools, providing significant savings in educational terms.

The main forms of habilitation in schools for the deaf involve auditory-verbal programs or total communication. Auditory-verbal management is based on the premise that stimulation of even minimal levels of residual hearing can lead to the development of spontaneous oral speech and language. With amplification, the child processes language, supplemented with active parental help at home. Most of these children can learn to listen and talk.

Total communication allows the hearing-impaired child to communicate by whatever means is found to be most beneficial to the child. This may incorporate sign language (AUSLAN), speech, finger spelling, lip-reading, writing, cued speech or gestures. Success with total communication is seen when the parents master sign language and finger spelling.

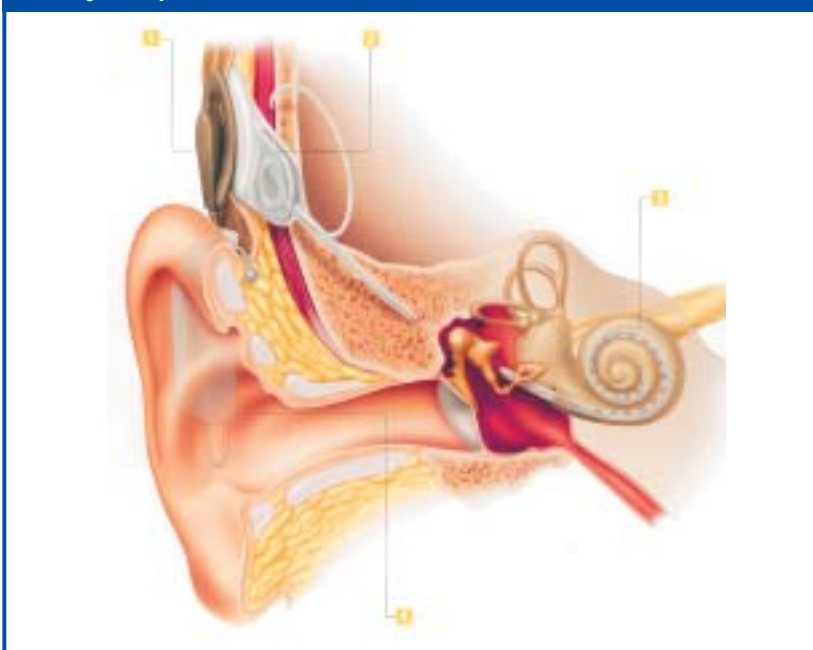
Cochlear implantation

Multi-channel cochlear implants, pioneered by Melbourne's Professor Graeme Clarke, have provided a valuable additional tool for the otologist managing children with severe to profound sensorineural hearing loss. The cochlear implant provides direct electrical stimulation to the cochlea and auditory nerve, where it is interpreted as sound by the brain (figure 7).

The device has been implanted in children as young as nine months and should be considered early for children deafened by meningitis before ossification of the inner ear occurs. It is not a panacea, and the guidelines for referral (table 7) should be adhered to strictly to allow the optimal outcome.

Generally, children with cochlear

Figure 7: How the cochlear implant works. Sound is received by the microphone behind the ear and sent to the speech processor (4), which analyses and digitises the sound into coded signals. These are sent to the transmitter coil (1), which sends the signal across the skin to the implanted receiver/stimulator (2), which in turn converts the code to electrical signals and sends them to electrodes inserted in the cochlea (3) to stimulate remaining nerve fibres. Image courtesy of Cochlear Ltd.



implants have the potential for developing good listening and spoken language. A very detailed multi-disciplinary workup is necessary, involving 10-12 sessions, as well as CT scans, steady-state evoked potentials and electrocochleography.

Postoperatively, habilitation includes device orientation, activation and tuning of the speech processor, development of listening and communication skills and long-term follow-up.

Conductive hearing loss

Children with the rare bilateral atresia syndrome are often managed with bone conductor hearing aids and, if appropriate, bone-anchored hearing aids. Those with some congenital conductive hearing loss due to stapes fixation may have an exploratory surgical procedure to ascertain if fixation can be corrected.

AOM is treated with broad-spectrum antibiotics after the diagnosis is differentiated from the other causes of red ear.

If an infant, particularly one with risk factors such as day care attendance, has three separate episodes of AOM in six months, or 5-6 episodes in 12 months (three in the Aboriginal child), insertion of ventilation tubes (grommets) should be considered. This is especially the case when there is a persistent underlying OME with hearing loss. If conventional management with antibiotics does not resolve long-standing OME with hearing loss of more than 25dB for three months, either grommets or, less commonly, hearing-aid amplification should be considered.

The child with CSOM and active otorrhoea should be treated with topical ear therapy including ear toilet with 5% Betadine followed by antibiotic or corticosteroid eardrops, preferably a non-ototoxic drop such as Ciproxin-HC from the fluoroquinolone group.

Table 7: Guidelines for referral for cochlear implantation

- Children should have severe to profound sensorineural hearing loss
- Early referral after consistent hearing aid use has demonstrated insufficient audition to facilitate the development of speech and oral language skills
- There should be no medical contraindication for undergoing cochlear implant surgery (eg, intellectual handicap, CSOM)
- Families should be motivated and have appropriate expectations
- Enrolment in an educational program with a strong auditory and verbal component

Children with CSOM may need bone conductor hearing aids or FM systems until they are old enough for repair of their tympanic membrane at age 5-6. In many schools with a high proportion of Aboriginal children with CSOM and OME, sound field systems in the classroom ensure the teacher is heard. Children with CSOM and severe conductive hearing loss may have ossicular involvement and require an ossicular chain reconstruction.

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Preventing hearing loss

PREVENTING acquired sensorineural hearing loss falls well within the province of the GP. There have been major advances in immunisation against bacteria, which can lead to meningitis and the sequela of deafness in 10-30% of cases, depending on the organism. These include pneumococcal and Hib vaccinations. Vaccination against mumps, measles and, of course, rubella will help prevent the deafness complications of these viruses.

Careful monitoring of the use and serum levels of aminoglycoside antibiotics will prevent their potential ototoxicity. Use of topical non-ototoxic antibiotic or corticosteroid eardrops in the presence of a perforated tympanic membrane or grommet is preferable to the commonly used aminoglycoside drops.

In the older child and teenager, advice regarding noise exposure to bands, concerts, Walkman devices and rifle shooting should be provided. Above all, if a parent has any doubts about their child's hearing ability, a high index of suspicion and early referral for audiological assessment will enable prompt management.

Otitis media in all its forms can be prevented, in part, by avoiding risk factors whenever possible, such as day care, passive cigarette smoking, prop feeding (which may lead to reflux into the middle ear) and early cessation of breastfeeding. Avoiding the use of dummies after 10 months of age may also help.

For the at-risk child in day care or in the indigenous community, use of the pneumococcal conjugate vaccine, Pnevna, and the Pneumovax and influenza vaccines in the older child may reduce the incidence of otitis media and help prevent progression to OME and CSOM. Public health preventive measures such as attention to hygiene, nutrition and overcrowding will also significantly help reduce the high burden of disease in the indigenous community.

Early and aggressive management of AOM in the child under six months and early referral to the otologist when there are three or more episodes of otitis media in six months may prevent the complications of the disease.

cochlear implants are being developed in Australia. The age at which cochlear implantation can be performed will decrease.

Future development of a chimera of body tissue and a biological implant, possibly on the ossicles, will allow optimal transmission of sound in the damaged middle ear.

Bacterial biofilm formation (identified by electron microscopy in the middle ear mucosa of one of my patients with CSOM) may explain in part the recalcitrant nature of the disease in the Aboriginal child with chronic infective rhinitis and otorrhoea. Topical medications designed to disperse this film may allow non-ototoxic eardrops to target the causative bacteria more readily.

What the future holds

GENETIC testing for congenital hearing loss will become more frequent as the gene loci for hereditary hearing loss are further elucidated. The possibility of gene therapy to replace a defective gene, possibly in utero, is one of the more exciting prospects of the next 10 years.

In mammalian models, researchers in the US have regrown outer hair cells, previously thought to be unable to regenerate. Micro-pumps, which can feed neurotrophic hormones into the inner ear, are being developed and various epithelial growth factors are being trialled to repair tympanic membrane perforations.

Universal newborn hearing screening will become the standard of care.

Cochlear implantation using totally implantable