Deafness in adults: the role of the cochlear implant

PAUL FAGAN, MD, FRCS, FRACS; RONALD Y.K. CHIN, BSc(Med); JOHN P. TONKIN, FRCS, FRACS; MARCUS D. ATLAS, FRACS; SARAH PLATT-HEPWORTH, MA(Aud); CELENE McNEILL, MA(Aud)

In the past, major damage to both inner ears resulted in total or near-total sensorineural hearing loss and complete isolation of the patient. The development of the cochlear implant – the so-called bionic ear – has brought such patients back to the hearing world. It has become common in recent times for these patients to manage to hear well without any visual clues.

■ Hearing loss can be conveniently divided into that of middle ear type (conductive) and the so-called sensorineural group (inner ear hearing loss). A conductive hearing loss responds very well either to the use of a hearing aid or to surgery. In the era of the operating microscope, major advances have been made in the surgical correction of these problems. On the other hand, inner ear hearing loss, caused by either damage to the cochlear hair cells themselves or to their central connections, is mostly irreversible. In the past, major damage to both inner ears resulted in total or near-total hearing loss and complete isolation of the patient. The development of the cochlear implant has meant that these patients can now be given significant help. It is now common for cochlear implant patients to manage to hear well without any visual clues – for example, they can carry on a conversation over the telephone.

Dr Fagan, Dr Tonkin and Dr Atlas are otologists, Department of Otology/Neuro-Otology, St Vincent's Hospital; Mr Chin is a final year medical student; Miss Platt-Hepworth and Mrs McNeill are audiologists, Hearing and Balance Centre, St Vincent's Hospital, Sydney, NSW.

History

The idea that electrical stimulation of the ear could cause the brain to hear a sound is an old concept, beginning with Volta, the inventor of the battery. He is said to have used one of his primitive batteries to stimulate his own ear to see what it felt or sounded like. The battery charge must have been a large one because it caused him to fall down unconscious. However, he reported to his friends that he had heard a loud sound.

In the 1930's, Wever and Bray passed a tiny electrode directly into the cochlear nerve of a cat. When sound was directed into the ear, they were able to detect the passage of an electrical impulse along the nerve. They thus proved that the inner ear was a transducer – that is, it converted the mechanical energy of sound into electrical energy, which passes along the cochlear nerve to the brain where it is interpreted as sound.

In 1951 the French ENT surgeon D'Journo operated on a man who had a large cholesteatoma which had caused complete deafness in one ear. This operation was carried out under local anaesthesia. D'Journo found that the cochlear nerve was exposed, and during the operation was able to insert tiny electrodes directly into the nerve. When the nerve was stimulated with an electric current the patient reported hearing distinct sounds. D'Journo also demonstrated that increasing the frequency of the current increased the pitch of the sound reported by the patient. Simmons did similar work in the USA in 1966.

In California, William House read the work of D'Journo and, working with a totally deaf patient, stimulated the inner ear by means of tiny electrodes passed through the tympanic membrane onto the promontory of the middle ear. Later, House passed a short wire into the inner ear through the round window membrane bringing it much closer to the cochlear neurones and giving more precise measurements and results. House was the true pioneer of cochlear implantation and all subsequent work has followed his efforts

The first commercially available cochlear implant, a single-electrode device jointly developed by House and the 3M company, was widely

In 1951, direct electrical stimulation of the cochlear nerve enabled the patient to hear distinct sounds.

used from 1977 until it was supplanted by the multichannel device made by Cochlear in 1982. During this time, one of the authors of this article (Dr Tonkin) performed regular implantation of the House 3M single channel device.

In Australia, Clark and his associates at the University of Melbourne experimented with a longer wire on which were placed multiple platinum bands. This wire, like the one used by House, was passed into the inner ear via the round window membrane. Each platinum band acts as a separate electrode which can be stimulated independently. This multichannel implant stimulates the different parts of the cochlea that are responsible for hearing low tones, middle tones and high tones. Those workers demonstrated that the multichannel system gave better sound discrimination than did the single channel device. Most modern implants are of this type.

Clark and his associates, in collaboration with Cochlear, have continued to improve the sound processor and, most recently, the implant, thereby increasing the degree of speech discrimination available. The latest processor is called the Nucleus Twenty Four, named after the number of electrodes. In addition, the processor which was formerly only available as a body-worn instrument can now be used at ear level. Further refinement and improvement of the processor will undoubtedly take place.

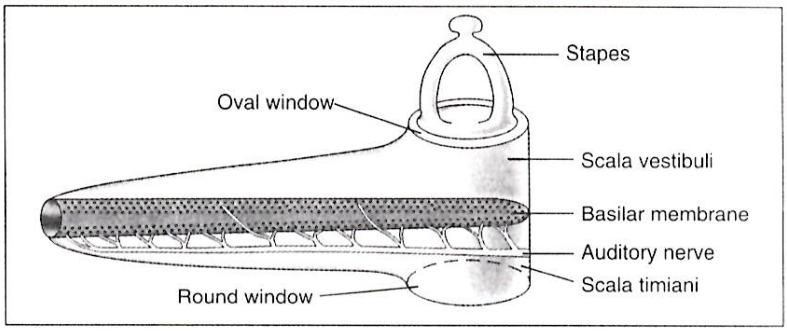


Figure 1a. The cochlea has been unrolled. Movement of the stapes footplate in the oval window causes pressure waves to travel along the cochlea. The round window moves in reciprocity with the stapes footplate. The hair cells attached to the basilar membrane are stimulated by the pressure waves.

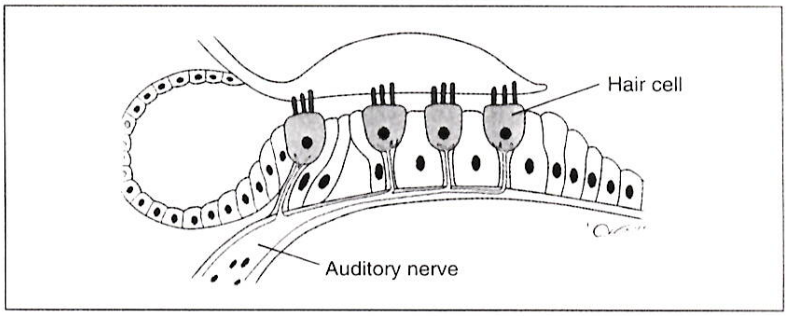


Figure 1b. The cochlear hair-cells, derived from neural tissue, have up to 20 tiny appendages. As these bend in response to a pressure wave, a pore at the free end opens, allowing a sodium–potassium interchange to take place. The resultant action potential travels over the surface of the hair cell and along the cochlear neurones to the cochlear nucleus in the brain stem.

Illustrations for *Modern Medicine* by Chris Wikoff.

How the ear works

The external, middle and inner ear combine to provide a mechanism by which an acoustic signal is transformed into a neural action potential, which is then processed in the brain. A sound wave causes a pressure change in the external ear leading to vibration of the tympanic membrane. This vibration causes movement of the ossicular chain in the middle ear (malleus, incus and stapes). This piston-like movement of the stapes in the oval window results in a fluctuation of pressure in the fluid of the middle ear.

Stapes movement is not simple. The footplate of the stapes not only vibrates, it oscillates and rocks along both axes in a subtle fashion, the details of which are beyond the scope of this discussion. Importantly though, there is a significant magnification factor here as the area of the tympanic membrane is some 20 times greater than the area of the stapes footplate.

Pressure waves generated by this movement of the stapes flow through the cochlea to impinge directly on the cochlear hair-cells (Figure 1a). As the hairs on the free surface of the hair-cell bend, a pore at the apex of each opens, leading to a sodium–potassium interchange and the development of a local potential difference, which is propagated down the outside of the hair cell to the junction with the end branches of the cochlear nerve (Figure 1b). An action potential then passes along the myelinated cochlear nerve to the cochlear nucleus in the brain stem, the first way-station in the brain, then on to the higher centres.

The first commercially available cochlear implant – a single electrode device – was widely used from 1977.

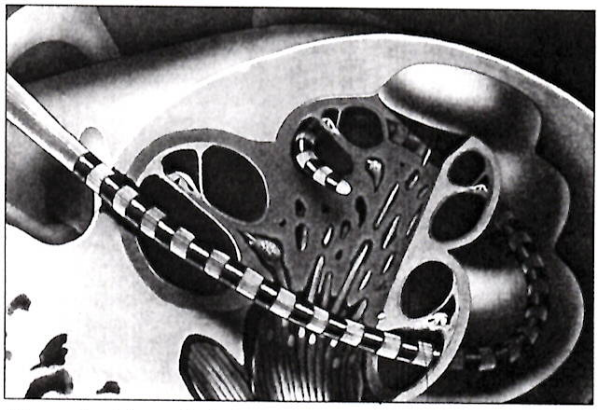


Figure 2a. The principle of the cochlear implant. An electrode array is passed into the scala tympani of the cochlea through the round window. It lies in close proximity to the ends of the cochlear neurones.

Illustration reproduced courtesy of Cochlear.

Figure 2b. A CT scan showing an electrode array in place, following the curve of the basal coil of the cochlea.

The function of the cochlea is extremely complex and incompletely understood. However, it is known that high frequency sounds stimulate the hair cells near the base of the cochlea and low frequency sounds stimulate the apex. Intensity is probably a function of the number of hair cells that are stimulated.

How the cochlear implant works

The common causes of complete deafness in the adult – for example, viral infections, drug toxicity, meningitis and Menière's disease – damage cochlear hair-cells, leaving (for the most part) the end branches of the cochlear nerve intact. The cochlear implant delivers a current directly to these nerve endings (Figures 2a and b).

Sound enters the system though a small microphone behind the ear (Figures 3 and 4). Here it is converted into electrical impulses which are sent via a thin cable to the speech processor, which selects and codes the elements that are most useful in the understanding of speech. These electronic codes are then sent through the cable to the transmitting coil.

Magnets in the centre of both the transmitting coil and the internal coil (receiver/stimulator) hold the two in close proximity (Figure 5). The internal coil is activated by electromagnetic induction. This is based on the principle that a magnetic flux produced by passing a current through a coil (external) will induce a current in a second coil (internal). The encoded information in the receiver/stimulator is converted by an integrated circuit into electrical signals which are sent to the electrode array within the cochlea.

As each electrode has a separate wire connecting it to the receiver/stimulator, the coded electronic signals can be sent to specific electrodes, each of which has been individually programmed to deliver sounds that can vary in volume and pitch. These electrodes then stimulate the appropriate hearing nerve fibres, which send messages to the brain. The brain receives the signals and interprets them, and a sensation of hearing is experienced. This is an over simplification of the extraordinary technology that is utilised in the manufacture and design of the modern cochlear implant. Again, a detailed discussion is beyond the scope of this paper.

Who is a candidate for a cochlear implant?

Until recently, only profoundly deaf adults who acquired deafness after developing speech and language skills were considered for cochlear implantation. As technology has improved, the indications for implantation have become wider to include adults with lesser degrees of deafness. The adult patients who do best are those whose hearing loss is recent, who are clearly well motivated and who have developed speech and language. Prior to implantation it is essential that it be demonstrated that these patients gain little or no help from a hearing aid.

The audiogram of a typical cochlear implant patient demonstrates a hearing loss which is either severe (70 to 85 dB) or profound (90 dB or greater). In the past, only patients with profound hearing losses were considered

A wire with multiple platinum electrodes allows for stimulation of different parts of the cochlea responsible for different tones.

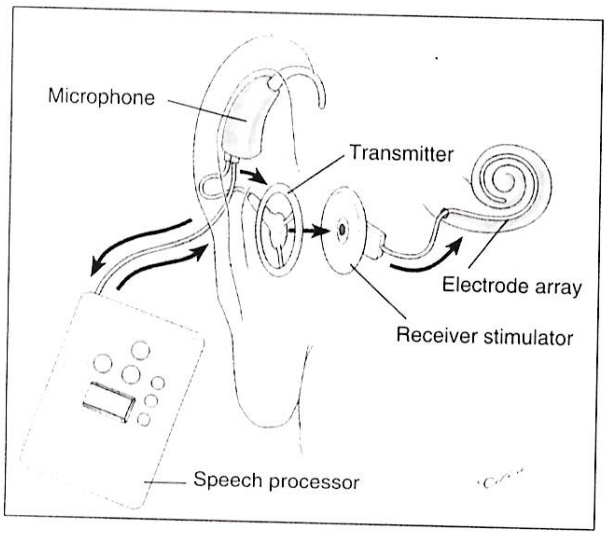
for cochlear implants. Now, patients with severe losses (Figures 6a and b) are considered, providing their speech test scores with a hearing aid are lower than 40%.

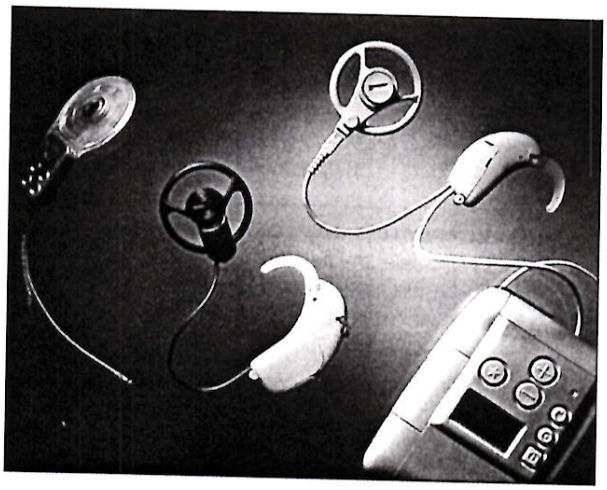
Patient assessment

There are some sophisticated audiological tests which help to decide if a hearing aid will be suitable, but the best test is an adequate trial of a properly fitted, good quality, powerful aid. The efficacy of other listening devices is also established – for example, telephone couplers, television amplifiers, hand-held microphones and loop systems via a hearing aid telecoil (T-switch).

Additional skills such as lip reading (speech reading) are very useful for any patient with hearing loss. Even people with normal hearing lip-read intuitively whenever listening conditions are not ideal. For the hearing aid or cochlear implant user, lip reading will always help to improve communication skills.

Family support and the patient's motivation are clearly important because intensive postimplant training is required to achieve good results. Occasionally, a preoperative psycho-





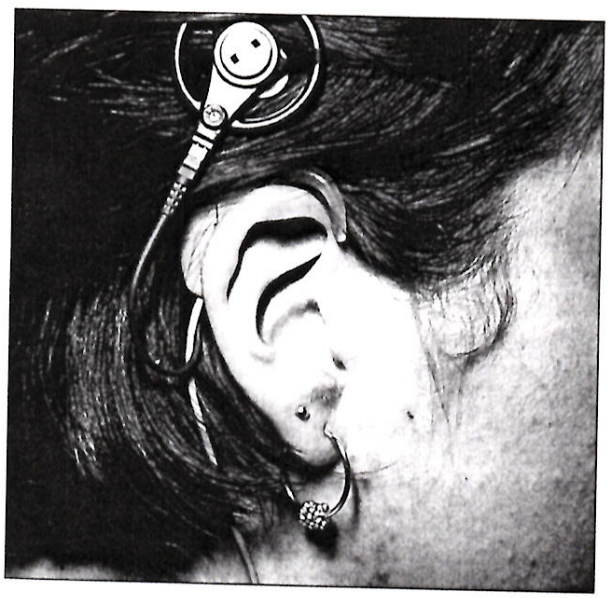


Figure 3 (above left). A basic diagram of the cochlear implant. Sound is received by the microphone and is converted to electrical impulses, which are relayed to the speech processor where they are converted to electronic codes and delivered to the transmitting coil. The transmitter sends the code across the skin to the receiver. The receiver stimulator converts the code to electrical signals which are sent to the electrodes to stimulate the nerve fibres. These signals are recognised as sounds by the brain, producing a hearing sensation.

Illustration for *Modern Medicine* by Chris Wikoff.

Figure 4 (above right). The complete device. On the left is the electrode array attached to the internal coil. On the right is a body-worn speech processor with a microphone worn at ear level. The external coil is attached by a cable to a microphone. In the centre is a more modern development in which the miniaturised speech processor, with the external (transmitting) coil attached, is worn at ear level.

Figure 5 (left). The device worn by a patient. Magnets in the centre of the transmitting coil and the internal coil hold the two in close proximity.

The latest device has 24 electrodes. The processor is small enough to be worn at ear level.

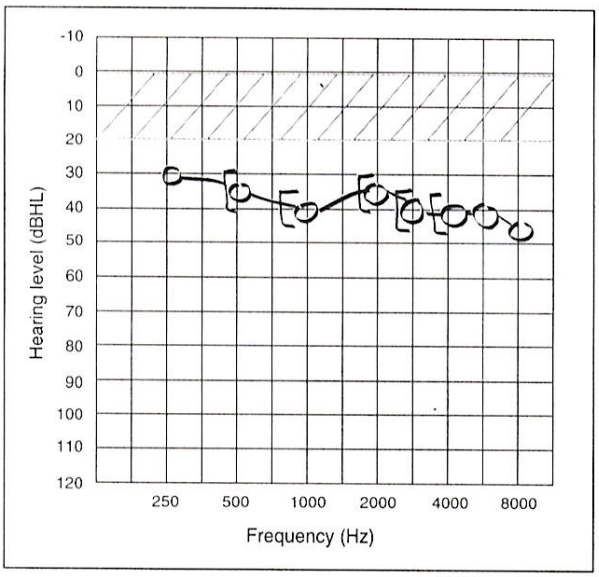


Figure 6a. Audiogram of the right ear. The vertical axis records (on a logarithmic scale) the threshold in decibels – that is, the least sound pressure that can be heard in sound-attenuated surroundings. The horizontal axis shows the frequency of the stimulus from the low (250 Hz) to the high (8,000 Hz) frequencies. Right air conduction is recorded, by convention, as a circle (O) while bone conduction is shown as a bracket ([). Where air conduction and bone conduction coincide, hearing loss is seen to be of inner ear type; that is, the middle ear is working as it should. Normal hearing falls into the shaded area (0 to 20 dB). The hearing loss shown, some 40 dB, is mild.

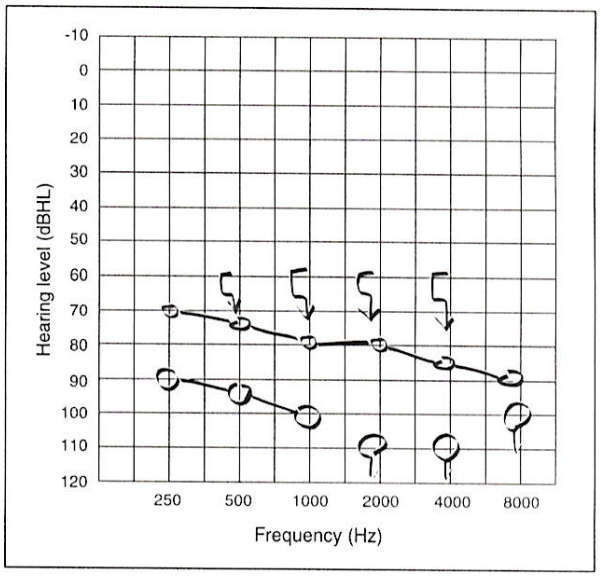


Figure 6b. An audiogram demonstrating more severe hearing loss. Bone conduction cannot be reliably recorded below 60 dB and is indicated by the symbol of a bracket with a downwards pointing arrow. The upper air conduction trace (red) at 80 dB is a severe hearing loss while the blue trace at 100 dB is profound. Formerly, only patients with a profound hearing loss underwent implantation, but advances in technology have led to successful implantation in patients with severe hearing loss.

logical assessment may be indicated.

Finally, the patient's expectations must be realistic. No matter how good the results, an implant will never restore normal hearing and this has to be well understood by the patient and the family. The prognostic factors are summarised in the Table.

Table. Prognostic factors

Poor prognosis	Good prognosis
Long term deafness	Short term deafness
Sign language is the major mode of communication	Speech is the major mode of communication
Little motivation and support	Motivated with good support
Living in a deaf community	

The surgery

The electrode array is inserted after carrying out a standard cortical mastoidectomy with a wide facial recess opening. Some bone is usually left over the exposed vertical segment of the facial nerve as instances of facial nerve stimulation by electrical current from the implanted device have been reported.

The bony lip of the round window niche is drilled off to give good exposure of the membrane. The scala tympani is opened and the electrode array is then led into the cochlea (Figure 7). This opening is sealed with soft tissue. The device itself is seated above and behind the mastoid cavity and securely fastened with heavy suture material (Figures 8a and b).

The major theoretical complication with this procedure is facial nerve injury. Occasionally, prolonged imbalance can occur if perilymph leaks around the device from the fluid-

Stapes movement is not simple. The foot plate of the stapes not only vibrates, it oscillates and rocks along both axes in a subtle fashion.

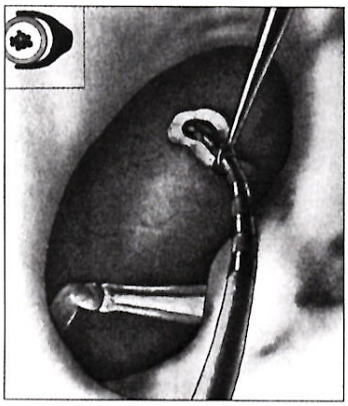


Figure 7. The electrode array is led into the cochlea through the opening in the round window.

Illustration reproduced courtesy of Cochlear.

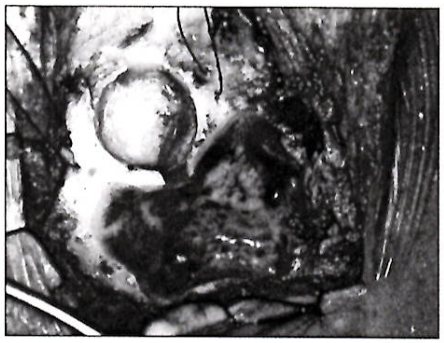


Figure 8a (above). The internal coil is seated in a circular depression drilled in the bone of the skull above and behind the pinna.

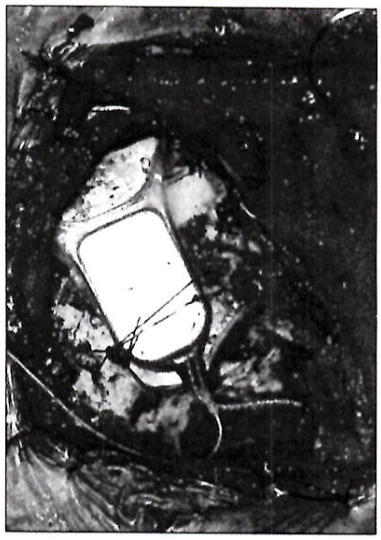


Figure 8b (right). The internal coil is fixed into place with heavy suture material.

filled inner ear into the air-filled middle ear. Wound infection in the presence of a foreign body would be a significant problem, as would device extrusion, but these problems are very rare. Perioperative antibiotic cover is usually given. Lastly, there may be a tendency for late device failure, necessitating reimplantation.

Post-surgical training

Three to six weeks after surgery the device is first 'switched on'. This means that at this time and at regular intervals through the patient's life, the implant is 'mapped'; that is, each electrode is tested for threshold comfort levels and balanced against the other electrodes (Figure 9). This is a time-consuming but essential process.

On the first day, the patient will report an understanding of a few words, but voices at this stage sound a little distorted and 'robot-like'. In the majority of patients, improvement in the quality of sound thereafter is rapid, with most or all of the electrodes stimulating. It takes 12 months of use to reach a plateau of function, but

Figure 9. 'Mapping', the process by which each electrode is individually tuned, is time-consuming but essential. It is carried out three to six weeks after surgery and at regular intervals throughout the patient's life.

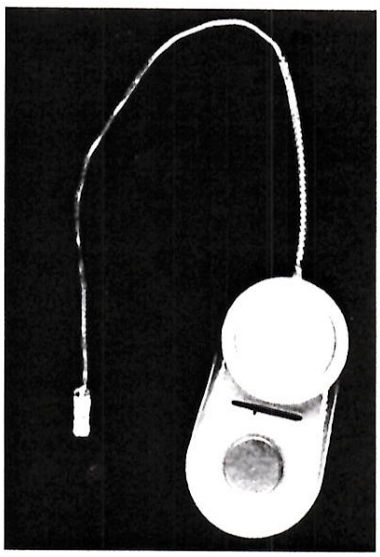


Figure 10. Auditory brainstem implant for use in deaf patients in whom the cochlear nerve has been lost (e.g. neurofibromatosis type II). The only visible change from a standard cochlear implant is that the electrode array, instead of running along a wire, is embedded in a teflon mesh which lies in contact with the cochlear nucleus in the lateral recess of the fourth ventricle.

with newer devices this period of time may be shorter. Most patients will be able to hear and understand speech with the aid of visual clues such as lip reading. With modern devices, about 70% of patients will not need these clues and will be able to converse on the telephone.

The auditory brainstem implant

Patients with neurofibromatosis type II often develop an acoustic tumour on each cochlear nerve. Hearing loss is eventually complete. The absence of a cochlear nerve rules out the standard cochlear implant. Accordingly, the electrode array has been modified by incorporating it in a teflon mesh (Figure 10) which is inserted into the lateral recess of the fourth ventricle in contact with the cochlear nucleus. Early trials of this auditory brainstem implant (ABI) have given very promising results; however, at this time the expectation is that the device will give access to environmental sound and an improvement in lip-reading ability.

The future

As improvements in electronics and computers continue, it is likely that the indications for cochlear implantation will become more liberal. These same technical advances are of course making hearing aids more efficient. It is reasonable to hope that eventually all patients with a hearing impairment will be able to be given significant help. ■

Further reading

1. Clark GM, Blamey P, Brown AM, et al. The University of Melbourne Nucleus multi-electrode cochlear implant. Switzerland: S Karger AG, 1987.
2. House WF, Berliner KI, Crary WG, et al. Cochlear implants. Ann Otol Rhinol Laryngol 1976; 85(Suppl 27).
3. Cochlear implants in adults and children. NIH Consensus Statement 1995 May 15-17; 13(2): 1-30.