

Cochlear implants and brain stem implants

Richard T Ramsden

Department of Otolaryngology, Head and Neck Surgery, Manchester Royal Infirmary, Manchester, UK

This chapter describes the development of two implantable prosthetic neurostimulators which, in the last 20 years, have revolutionised the management of severe-to-profound sensorineural deafness. We have witnessed their rapid evolution from the realms of esoteric laboratory abstraction, with many critics and little perceived clinical use, to a routine treatment which is safe, effective and, indeed, cost effective. It is one of the great triumphs of biomedical and surgical collaboration, and is without any doubt the greatest ever advance in the treatment of deafness.

The anatomy and physiology of the auditory pathways have been described in a previous special issue¹. To summarise, a sound wave entering the ear causes the tympanic membrane and ossicular chain to vibrate. The stapes, the smallest and innermost of the three ossicles moves in-and-out in the oval window and is the interface between the middle and inner ears. The mechanical movements of the middle-ear sound-conducting apparatus are transmitted to the inner-ear fluids and a wave, the travelling wave of von Békésy, passes up the cochlea to reach a maximum at a point determined by its frequency. For the process to continue, the physical energy of the travelling wave has to be converted into electrical energy that can be propagated through the auditory nerve to the brain stem and from there to the higher auditory centres. This process of transduction occurs in the organ of Corti. Depolarisation in the inner hair cells initiates transmission through the first order neurones, the cell bodies of which are in the spiral ganglion. The synapse with the second order neurone occurs in the cochlear nucleus which is situated in the lower pons cranial to the foramen of Luschka. The more cranial nuclear projections in the brain stem and auditory cortex are complex and a detailed understanding of their anatomy is not necessary for the understanding of this essay.

Many disease processes may lead to loss of hair cells in the organ of Corti. The commonest is the natural process of ageing in which there is a progressive loss of cells typically starting in the basal turn of the cochlea and advancing apically, accompanied by a hearing loss that initially affects the high frequencies and, with time, the middle and lower frequencies. These cells are incapable of spontaneous regeneration

*Correspondence to:
Prof. Richard T Ramsden,
Professor of
Otolaryngology,
Department of
Otolaryngology, Head and
Neck Surgery, Manchester
Royal Infirmary,
Oxford Road,
Manchester M13 9WL, UK*

(although there are hopes that some time in the next few decades neurotropic factors may be identified that might make this dream a possibility). Chronic noise exposure is another good example of progressive hair cell loss. In neither of these conditions does the hair cell loss become complete, so total hearing loss is unlikely and treatment with hearing aids may be effective. There are, however, a number of conditions in which total or near total loss of the organ of Corti may occur and the most common are listed in Table 1. It will be seen that these are all acquired conditions and may affect both adults and children. To these must be added a number of causes of total deafness that are present at birth or soon after birth, and, of these, recessively inherited non-syndromic deafness is the commonest. Whatever the cause, the absence of the organ of Corti prevents transduction and, although all other components of the auditory system may be intact, profound deafness results.

The cochlear implant is a device that takes over the role of mechano-electric transduction and delivers to the auditory nerve a processed signal that can be transmitted to the auditory cortex and interpreted as sound. There are two recognizable components of a typical cochlear implant system: (i) the implanted electrode array with associated microcircuitry; and (ii) the external component that refines the raw signal before delivering it to the implanted electrode. In the early days, the cochlea was regarded as inviolable, and fairly simple single channel analogue systems were inserted on to the surface of the cochlea. The modern electrode system is multichannel with up to 22 electrodes and is inserted into the scala tympani of the cochlea. It takes advantage of the highly developed tonotopicity of the cochlea with an orderly progression from high frequencies at the basal end of the cochlea to low frequencies at the apical, like a piano keyboard. Digitised processing is now almost universal. The external component comprises a microphone that sends an electrical signal to the 'brains' of the system – the speech processor.

Table 1 Causes of acquired severe/profound cochlear deafness

- Idiopathic (cause cannot be identified)
 - Meningitis
 - Viral infection of the inner ear (e.g. measles, mumps, rubella, CMV)
 - Bacterial labyrinthitis complicating middle ear sepsis
 - Menière's disease
 - Ototoxic drugs (e.g. aminoglycoside antibiotics)
 - Skull base fracture
 - Cochlear otosclerosis
 - Auto-immunity
 - Iatrogenic (e.g. following stapedectomy)
-

There are many different strategies employed by the various manufacturers, but the aim of speech processing is to manipulate the raw signal so that the most important features necessary for speech recognition are preserved and delivered to the ear. Underlying early speech processing strategies was the recognition that there are various bands (or formants) of spectral energy in speech. Low frequency information corresponds to the vowel sounds and contributes to the prosodic patterns of speech. The higher frequencies convey consonant information which is essential for speech recognition. Modern strategies try to convey the time-varying spectral patterns that are characteristic of speech by analyzing the signal in short time frames and signalling the most prominent spectral peaks in each frame. The other factor that influences the fidelity of the signal delivered to the ear is the rate at which speech is sampled and digitized by the speech processor. Of the current generation of cochlear implant systems, the maximum stimulus rate, summed across all electrodes and using pulsatile stimulation is approximately $96,000 \text{ s}^{-1}$. The processed signal is transmitted through the intact skin by a process of inductive coupling. An external coil is held magnetically over an internal coil that is part of the implanted component of the system. A microchip decodes the incoming wave form and directs it to the appropriate intracochlear electrode or electrodes depending on the frequency of the sound, and at a rate determined by the specification of the individual processing programme.



Fig. 1 Implanted component of the Nucleus CI24 system.

Devices implanted in the body should be reliable, safe and last for many years – in the case of a child, that means life (Fig. 1). Experience gained from pacemaker technology has proved invaluable, particularly in providing water-tight ceramic or silastic sealing systems for the electronics and cumulative failure rates of as little as 3% at 10 years are quoted by most manufacturers. Many implant programmes round the world have been operating for 15 years and most devices have continued to function without trouble. Implants that do fail can be replaced usually without difficulty and without loss in performance. Most of the important recent advances in implant design relate to speech processing strategies rather than the design of the implantable component. This means that performance with an early implant system can be improved by replacing or modifying the external component, rather than removing and replacing the intracochlear electrode. Advances in electrode design have been somewhat less spectacular. At present, the major manufacturers are pre-occupied with developing so-called ‘modiolus-hugging’ electrodes. As their name suggests, these are designed to lie close to the spiral ganglion and to the residual neural elements within the cochlea that the implant aims to stimulate. Their alleged advantages are more precise stimulus delivery and reduced power consumption. Neither of these claims can, as yet, stand up to close scrutiny, and many surgeons have fears that the new electrodes may traumatize the cochlea.

Who is suitable for a cochlear implant?

There are two main groups of candidates for implantation: (i) postlingually deafened adults; and (ii) prelingually or congenitally deaf children. To understand the issues relating to these two groups, it helps to understand the process of speech and language development in the normally hearing child. At birth, the child is not capable of speech, but by the age of a year babbling commences and by 18 months recognizable words begin to appear. From then on lexical, grammatical and semantic skills are acquired at a staggering rate, and this capacity remains immense during childhood. During this time, the auditory pathways are in a state of maximum plasticity. This facility wanes as the teens approach and is almost lost by mid-teenage. A normally hearing child, deprived of the sound of human speech during this critical period, and restored to it in adult life would be unable to acquire normal speech despite having normal peripheral auditory function. This phenomenon is similar to that observed in strabismic children who develop amblyopia². Adults who lose their hearing after the critical period already have a programmed auditory cortex. A cochlear implant rapidly re-activates dormant neural

Table 2 Factors to be considered in cochlear implant assessment

-
- Severity of hearing loss
 - Benefit from hearing aids
 - Age and duration of deafness
 - Linguistic status
 - Imaging of cochlea and auditory pathways (CT, MRI)
 - General health
 - Cognitive impairment
 - Expectations and motivation
-

networks. This is clearly observable in the many adults who can converse almost effortlessly within hours of 'switch-on'. Additional improvement occurs in many as a result of subsequent cortical re-organization. For an implant to be effective in a congenitally or prelingually deafened child, it has to be inserted while the auditory system is still plastic or programmable. There is convincing evidence that the earlier a child is implanted the better the improvement in auditory performance³. The extent of neuronal survival is clearly an important determinant of outcome, and it must also be recognized that electrical stimulation (from the implant) prevents further neural degeneration. Most cochlear implant programmes like to implant children as early as possible usually around the age of 2 years.

Implant candidates are assessed in some detail by the implant team (Table 2). Cochlear implant technology is expensive and the process of rehabilitation involves the skills of many professionals. As a result, the cost to the National Health Service of an implant with assessment, surgery and 2 years of rehabilitation is in the region of £30,000. An appropriate and rigorous selection process is, therefore, desirable. A number of criteria are considered.

Degree of deafness

This is always the first and most important consideration – is the hearing loss bad enough to warrant an implant or could as good a result be obtained with conventional hearing aids? Criteria are changing as implant technology improves. Fifteen years ago, the typical implantee was a postlingually deafened adult with a pure tone threshold of 100–110 dB and no speech discrimination (profound deafness). Now, teams are implanting patients who are still deriving some limited benefit from their hearing aids. Their maximum speech discrimination score in the best-aided condition might well be in the region of 20–40% (using material such as Bench, Kowal, Bamford sentences). In the paediatric

population, there are problems in assessing hearing thresholds and thus candidacy. Behavioural audiometry may be difficult or impossible. Objective audiometry, notably the Auditory Brain Stem Response (ABR), is widely used in assessment, but cannot as yet give accurate information about the low frequency thresholds. Speech audiometry is clearly out of the question in a prelingually deaf child with no lexical base. For these reasons, assessment of the rate of development of auditory performance over time is often necessary, particularly acquisition of language using hearing aids during a trial period which may last several months.

Age of the patient and duration of deafness

In postlingually deafened adults, age is relatively unimportant. Duration of deafness is, however, and the longer the period of auditory isolation the less good the outcome is likely to be⁴. It is impossible to set firm rules but a 55-year old, deaf for 40 years is a less favourable prospect than a 70-year-old, deaf for 5 years. The age of a congenitally deaf child is important for the reasons explained above. It is unlikely that a congenitally totally deaf child over the age of 7 years would do well with an implant, and congenitally deaf adolescents are bad candidates.

Imaging

High quality CT and MR imaging is essential to provide details of the anatomy of the cochlea and its connections, and may reveal a number of absolute or relative contra-indications to implantation. Congenital malformations (dysplasias) or acquired conditions such as cochlear obliteration, temporal bone fracture or otosclerosis should be reliably identifiable using the current generation of scanners.

General health

Common-sense rules about suitability for general anaesthesia and reasonable life expectancy apply. Of particular importance is an evaluation of central or cognitive function which may reveal potential difficulties with information processing with the implant. This is an issue after meningitis and with deaf and multiply handicapped children, more and more of whom are being referred for assessment. Each child has to be looked at on his or her merits.

Motivation, expectations and cultural issues

It is important that individuals contemplating implantation for themselves or for their children should have a realistic view of what the outcomes are likely to be. Exalted expectations are often perpetuated by the tabloid press. It is the job of the team to temper enthusiasm with realism, based on what can be predicted from knowledge of the individual subject. It is particularly important for parents to have some idea of the on-going nature of rehabilitation and that hard work is needed by them as well as the rehabilitation team and the child's teachers over a period of many years.

Many of these factors are only relative contra-indications, but taken together they allow the team to give an informed opinion to the patient or to parents of a child about likely outcome, and enlighten the discussions that precede a final decision about whether to proceed with surgery.

Surgery

The operation to insert the internal component of the cochlear implant system, the actual implant, has to be meticulously performed, but is well within the capability of most otologists. Through a small postauricular incision, a cortical mastoidectomy is performed and the middle ear is entered through the facial recess – the so-called posterior tympanotomy. The stapes can then usually be easily identified and 2 mm below it is the round window niche. The scala tympani of the basal turn of the cochlea is entered by drilling in front of the round window niche with a 1–1.5 mm microdrill. Usually a perilymph-filled cavity is encountered into which the electrode array can be gently introduced. As discussed previously, the latest generation of electrode arrays has a mechanism to carry it close to the modiolus and the spiral ganglion. The package comprising the receiver coil and the microchip is recessed into a bony well in the outer table of the skull, and the overlying pericranium usually provides sufficient stability without the need for anchoring ties. The operation takes about 1.5 h and, at the end of the procedure, the electrical integrity of the device can be tested by a number of measures including neural response telemetry. Using this technique, whole nerve action potentials are recorded from the auditory nerve in response to stimulation of individual electrodes within the cochlea. The information thus obtained may help the rehabilitation team in the initial estimation of threshold values of stimulation at the time of switch-on of the device, as well as confirming the integrity of each channel in the array. The incidence of postoperative complications is very low. Although the facial nerve is in the surgical field, as indeed it is with most tympanomastoid surgery, damage to it is very rare with an incidence of under 0.5%.

Special surgical problems

Surgical difficulties may be encountered in a number of well recognized conditions.

Inner ear dysplasia

The most extreme form of dysplasia, aplasia, is a contra-indication to implantation and would be picked up on pre-operative imaging. At the other end of the scale, the Mondini deformity and the large vestibular aqueduct syndrome are usually not difficult to implant. Between these extremes, the common cavity deformity in which there is little differentiation of the cochlea beyond the primitive otocyst stage is challenging, with the possibility of inadvertently inserting the implant into the posterior cranial fossa through the inner ear. Associated with this is a high risk of CSF fistula. Furthermore, in such cases of dysplasia, the facial nerve may be abnormally placed and be at increased risk. Aplasia of the auditory nerve is rare, but needs to be recognised (on MR imaging) as it is an absolute contra-indication to cochlear implantation.

Cochlear obliteration (osteoneogenesis)

In a number of conditions, the cochlear lumen may become obliterated, either by fibrous tissue or new bone, making insertion of the implant difficult, if not impossible. Meningitis, otosclerosis and skull-base fracture are associated with the deposition of new bone. Autoimmune ear disease may be associated with intracochlear fibrosis. Modified surgical techniques and electrodes have been developed to deal with these problems.

Chronic middle ear and mastoid disease

Insertion of a foreign object into the body is contra-indicated in the presence of active infection. Tympanomastoid disease, if present, including cholesteatoma must be eliminated at a first stage operation and the implant inserted at a subsequent date.

Switch-on and tuning

About 1 month after the operation, when the skin is well healed, the implant is connected to the external component and tuned up. Each

electrode in the cochlea has an electrical threshold at which the stimulus is just perceived as sound. It also has a higher level at which the stimulus just ceases to be comfortable. These are the T and C levels and the difference between them is referred to as the 'dynamic range'.

The process of establishing T and C levels for all 22 electrodes and entering them into the memory of the speech processor is called 'mapping' and requires the skills of the audiological and rehabilitation members of the cochlear implant team. Mapping is usually straightforward in adults, but can require considerable skill and patience with small prelingual children whose lack of language means that they have to be programmed using conditioning techniques. If one sets the levels too low, nothing will be heard; if too high, a non-auditory response such as pain or facial twitching may occur, which will upset the child and ensure the end of his or her co-operation in the whole mapping process. Mapping has to be repeated on a number of occasions in the first weeks and months since the psychophysical features of the auditory system change as a result of stimulation by the implant.

Outcomes

Nearly all implanted postlingually deafened adults achieve some degree of open set speech understanding using the implant alone (*i.e.* without lip reading); most achieve a high level of performance within a matter of weeks and can use the telephone with a fair degree of proficiency. Cochlear implantation in adults has been shown to be cost effective. Summerfield and Marshall calculated the cost per quality adjusted life year (QALY) for cochlear implantation and a number of other common conditions. As can be seen in Table 3, cochlear implantation compares very favourably⁵.

It is becoming increasingly clear that the majority of congenitally or prelingually deaf children implanted by the age of 2 years are able to take their place in main stream schools after 3 years of implant use and training, albeit with some degree of support. Speech perception and

Table 3 The cost per quality adjusted life year (QALY) for cochlear implantation and a number of other common conditions

Treatment	Cost/QALY (US\$)	Cost/QALY (UK£)
Cochlear implant	15,593	11,440
Coronary artery bypass graft (3 vessel disease)	11,255	2090
Heart transplantation	38,970	7840
Coronary artery bypass graft (1 vessel)	64,033	18,830
Peritoneal dialysis	83,011	19,870
Haemodialysis	86,198	21,970

production continue to improve with time and most develop the accents and modulations of their geographical regions and peer groups.

The auditory brain stem implant (ABI)

This development from cochlear implant technology is indicated for totally deaf individuals who have no auditory nerves and who are thus not candidates for a cochlear implant. In practice, this patient group comprises almost exclusively sufferers from neurofibromatosis type 2 (NF2) who have been deafened as a result of bilateral vestibular schwannomas (acoustic neuromas) or from the surgery to remove them. Other possible indications may emerge such as cochlear nerve agenesis or the unimplantable cochlea from excessive ossification. The ABI has an electrode carrier with 20 small disc electrodes (Fig. 2) and is inserted on to the surface of the cochlear nucleus in the lateral recess of the fourth ventricle, accessed through the foramen of Luschka. Technically, this is not easy, as surgical landmarks are not always obvious. The correct position of the implant is verified by eliciting the electrically evoked auditory brain stem response (EABR). Adjacent cranial nerves (facial, glossopharyngeal, accessory and trigeminal) are monitored to minimise the risk of non-auditory stimulation. From the point of view of neuro-anatomy, there is a major problem with the frequency maps, or tonotopicity, of the cochlear nucleus compared with the cochlea. A surface electrode will function most effectively if the frequency map is distributed across the surface of the nucleus. In the cochlear nucleus, the map is disposed obliquely through the depths of the nucleus, and to take advantage of this arrangement a penetrating electrode has been developed but as yet it has not been used in clinical trials.



Fig. 2 The auditory brain stem implant with 20 disc electrodes on dacron carrier.

Multicentre trials with the surface electrode array have been carried out in North America and in Europe^{6,7}. They indicate that outcomes with the ABI are not as good as typical cochlear implant results. Nevertheless, most patients gained an awareness of environmental sounds and found the ABI enhanced their lip reading scores. A small, but important, number obtained reasonable-to-good open set speech perception using the implant alone and some obtain limited telephone use. Unwanted non-auditory side-effects such as facial twitching, pain in the throat, face or body may occur with some electrodes, but these can be programmed-out at mapping sessions.

Future developments

The near future should see the development of totally implantable devices that use either an intrinsic power source or implanted batteries that can be recharged remotely through intact skin. Similarly, remote re-programming would be possible. Research is also well advanced into mechanisms for delivering drugs or neurotrophic factors to the cochlea and auditory system through the intracochlear electrode⁸. Of course, if hair cells could be encouraged to regenerate, which again is the subject of much research, cochlear implantation would become a curiosity of medical history.

References

- 1 Haggard MP, Evans EF. Hearing. *Br Med Bull* 1987; **43**
- 2 Ryugo DK, Limb CJ, Redd EE. Brain plasticity. In: Niparko J. (ed) *Cochlear Implants: Principles and Practices*. Philadelphia, PA: Lippincott Williams and Wilkins, 2000
- 3 O'Neill CO, O'Donoghue GM, Archbold SM, Nikolopoulos TP, Sach T. Variations in gains in auditory performance from paediatric cochlear implantation. *Otol Neurotol* 2002; **23**: 44–8
- 4 Summerfield AQ, Marshall DH. *Cochlear Implantation in the UK 1990–1994*. London: HMSO, 1995; 144
- 5 Summerfield AQ, Marshall DH. *Cochlear Implantation in the UK 1990–1994*. London: HMSO, 1995; 227–8
- 6 Otto SR, Shannon RV, Brackmann DE *et al*. The multichannel auditory brainstem implant: performance in 20 patients. *Otolaryngol Head Neck Surg* 1998; **118**: 291–303
- 7 Nevison B, Laszig R, Sollmann W-P *et al*. Results from a European clinical investigation of the Nucleus multichannel auditory brainstem implant. *Ear Hear* 2002; In press
- 8 Clark GM. Cochlear implants in the third millennium. *Am J Otol* 1999; **20**: 4–8