Cochlear implantation: a review of current clinical practice

Cochlear implants treat severe hearing loss by providing direct electrical stimulation to auditory nerve endings. This article reviews the clinical assessment, surgical procedure and outcomes, and looks at newer developments such as preservation of residual hearing and bilateral implantation.

Cochlear implantation has revolutionized the management of severe hearing loss and is one of the most impressive developments of medicine in recent years. It is used to treat sensori-neural hearing loss which is too severe for conventional hearing aids to be effective. Hearing aids simply amplify the acoustic signal but this cannot overcome severe inner ear hair cell loss, so cochlear implants work in a fundamentally different way. They bypass most of the structures of the ear and apply direct electrical stimulation to auditory nerve endings.

This approach is highly successful: many adults are now able to use the telephone. Congenitally deaf children may now develop virtually normal speech and language. Over 300 000 people have received cochlear implants worldwide (Sorkin, 2013).

This article explains the basic principles of cochlear implants, clinical assessment, surgery and complications, newer developments including preservation of residual hearing, bilateral implantation, treatment of unilateral deafness, and fully implantable devices.

Basic principles of cochlear implantation

Current systems include an external processor behind the ear containing a microphone, battery and sound processor. This carries power and information to the internal device through an electromagnetic induction coil. It looks much like a conventional hearing aid, with the addition of the coil (*Figure 1*). It processes the signal to reduce its complexity, optimize speech perception, and activate the appropriate electrodes.

The internal device (*Figure 2*) includes a receiver-stimulator package on the surface of the skull.

The cable for the electrode array passes through the mastoid bone and middle ear into the cochlea. A linear array of between 12 and 22 electrodes curls around the modiolus, the central column of the cochlea that contains auditory nerve endings (*Figure 3*).

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Multi-channel implants depend on the tonotopic arrangement of the cochlea: the frequencies to which different parts of the cochlea respond are arranged in a simple linear manner with high frequencies at the basal end and low frequencies at the apex. During surgery the cochlea is opened at the basal end. Each electrode covers its own range of frequencies as a result of this 'place coding' along the cochlea. In addition, 'rate coding' can convey low frequency stimulation to the apical parts of the cochlea from more basal electrodes. For speech perception, high frequencies are important so the array does not need to cover all 2.5 turns of the cochlea (O'Donoghue, 2013).

Figure 1. The external processor of a cochlear implant.

Figure 2. The internal component of a cochlear implant.

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The inner ear hair cells are bypassed, but the implant depends on functional auditory nerves and central pathways.

Clinical indications and assessment

Any patient with severe or profound bilateral hearing loss, who does not gain adequate benefit from conventional hearing aids, should be considered for cochlear implantation. In the UK, the National Institute for Health and Care Excellence (2009) has defined those clinical criteria in detail. High frequency hearing at 2 and 4 kHz should be worse than 90 dB. In children, there should be evidence of inadequate speech development, and in adults speech discrimination using BKB (Bamford-Koval-Bench) sentences should be worse than 50% at 70 dB.

In addition, cochlear implantation will not be effective if there is inadequate neural plasticity so it is essential to determine the duration of deafness affecting each ear. Most centres aim to implant congenitally deaf children before 1 year of age. Later implantation results in progressively poorer outcomes, and is of little value over the age of 5 years (Cosetti and Roland, 2010). Outcomes in adults also depend on the duration of hearing loss, but are still acceptable with severe untreated hearing loss of up to 20 years in the implanted ear. Adults who present with a history of early hearing loss may still benefit if they had useful hearing in childhood with subsequent progressive loss, but it can be very difficult to predict outcomes in such cases.

Assessment is multidisciplinary and includes detailed audiology including speech and brainstem audiometry (in part to exclude non-organic hearing loss), speech therapy and educational assessments in children by teachers of the deaf. Many patients also require vestibular and psychological assessments.

The patient's general health and cognitive function should be assessed. Many patients with severe sensorineural loss are elderly, but this does not preclude successful surgery and many patients in their 90s have good results (Wong et al, 2016). Children may have syndromes that affect fitness for surgery or the cognitive function required for good functional outcomes.

Imaging by computed tomography or magnetic resonance imaging is required to assess surgical access, cochlear anatomy, the cochlear nerves and central neural structures required for successful implantation (Mackeith et al, 2012).

Surgery

Surgery is usually performed under general anaesthetic and takes 1–2 hours per ear, often as a day case.

An incision is made behind the ear and a subperiosteal pocket dissected to house the device. Traditionally a well is drilled into the skull. A simple mastoidectomy is performed followed by a posterior tympanotomy, a window into the posterior aspect of the middle ear deep to the tympanic membrane and superficial to the facial nerve. The electrode is inserted through the round window or an adjacent cochleostomy into the basal turn (Mangus et al, 2012).

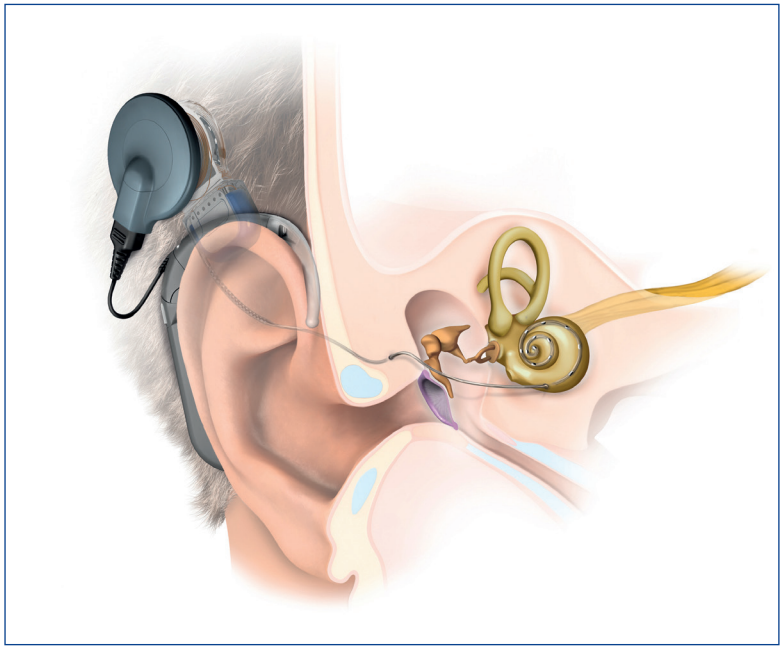

Figure 3. A cochlear implant system showing the electrode array within the cochlea.

Complications

Implant surgery has become a routine procedure with few significant complications. Major complications requiring intervention include facial paralysis, infections and device failures. Facial paralysis is rare (under 0.7%) and very unlikely with normal anatomy (Fayad et al, 2003). Device failures requiring re-implantation occur in 3–6% (Farinetti et al, 2014). Most infections are minor skin infections or acute otitis media, respond well to antibiotics, do not require explantation, and occur in 1–12% (Venail et al, 2008). The risk of meningitis is about thirty times that in the general population, although the absolute risk remains very low (0.1%). The risk can be minimized by preoperative pneumococcal immunization, perioperative antibiotics, and sealing of the cochleostomy at surgery (Reefhuis et al, 2003). Because of the risks of meningitis or a chronically infected prosthesis, it is important to treat infection in the vicinity of an implant early and aggressively.

Minor complications include transient taste disturbance from chorda tympani injury, and vertigo, which is almost always temporary (Venail et al, 2008).

Preservation of residual hearing

Traditionally, cochlear implantation has only been used in those with no or minimal hearing. It was assumed that surgery to open the inner ear would result in complete loss of remaining hearing. However, many patients have low frequency hearing that is near normal or easily aidable along with high frequency hearing that is extremely poor and unaidable. This results in good detection of sound but

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great difficulty understanding speech. Newer techniques now allow preservation of low frequency hearing in about 80% of cases: shorter electrodes provide high frequency hearing without disturbing the apical parts that provide low frequency hearing. Slimmer flexible electrodes and 'soft surgery' techniques result in less intra-cochlear trauma. Steroids given systemically and intratympanically may help preserve hearing (Huarte and Roland, 2014).

This has led to the development of hybrid 'electro-acoustic' devices that combine electrical stimulation for high frequencies and conventional acoustic amplification for the preserved low frequencies. This provides more natural sound, and improves speech scores, hearing in background noise, and appreciation of music (Gifford et al, 2013).

It is impossible to guarantee preservation of hearing, so the criteria to select patients for a hybrid device are the same as those for conventional implantation. It is worth noting that the conventional National Institute for Health and Care Excellence criteria quoted above relate only to high frequency hearing (90 dB at 2 and 4 kHz). Using those criteria, patients who lose low frequency hearing at surgery are still likely to perform substantially better after implantation than before.

The hybrid device combining an acoustic hearing aid with a cochlear implant in a single ear should be contrasted with the 'bimodal' configuration, in which the patient uses a hearing aid in one ear and a cochlear implant in the other. Funding restrictions mean that adults usually receive one implant. Where there is residual hearing in the other ear, they usually benefit from continued use of a conventional aid in that ear. This provides better localization of sound, hearing in background noise, and appreciation of music (Devocht et al, 2015).

Bilateral implantation

If both ears have very poor hearing, bilateral implantation results in better function, especially localization and hearing in background noise. It provides redundancy if one implant fails. It also eliminates the difficulty in predicting which ear will perform better after implantation (Wanna et al, 2012).

The high cost of implantation means that this is not generally available to adults in the UK. However, National Institute for Health and Care Excellence has determined that bilateral implantation is cost-effective in children (18 years and under) but only if performed simultaneously. Most children therefore now receive two implants. Studies are ongoing to determine the cost-effectiveness of bilateral implantation in adults.

Cochlear implantation in children

A number of issues specifically affect children. In congenitally deaf babies, early implantation results in better long-term outcomes in terms of numerous hearing, speech and educational measures (Cosetti and Roland, 2010). This is a simple consequence of the loss of neural plasticity over time. The results of implantation after the age of 5 years are poor. Most centres aim to implant such children between 10 and 12 months. It is not clear whether there would be any benefit to earlier implantation, which would pose several difficulties: surgical access may be significantly more difficult. Behavioural testing of hearing is less reliable, and there may be delayed maturation of hearing during the first few months, so one may not be certain that implantation is necessary.

Many children with severe hearing loss have other developmental problems, often as part of genetic syndromes, birth trauma or congenital infections. This makes assessment more complex and a multidisciplinary approach is particularly important. In cases of severe cognitive impairment, it may be impossible to predict longer term outcomes and the decision about implantation may be very difficult (Vincenti et al, 2014).

Meningitis may cause deafness, and may cause intracochlear ossification over several weeks. If this is extensive, it can make insertion of the electrode array impossible beyond the first few millimetres of the basal turn, with consequently poor function. Such cases are therefore treated as emergencies as soon as the child (or sometimes adult) is well enough to undergo assessment and surgery (Vincenti et al, 2014).

Functional outcomes of cochlear implantation

Following thorough assessment and surgery, the process of cochlear implantation has only just begun. Switch-on of the device occurs 3–4 weeks later and is followed by multiple audiological programming sessions and, especially in children, ongoing rehabilitation by speech therapists, and teachers of the deaf. In children this requires close integration with educational services. In adults who have recently had functional hearing and already have good speech and language, this process is less demanding. Nevertheless, it can take a year – or longer – to achieve optimal function: postoperative rehabilitation and consistent use of the device are essential.

The hearing provided by a cochlear implant is not normal. Recently deafened adults describe the sound as robotic, but adapt to it rapidly. The number of electrodes is far smaller than the number of distinct frequencies a normal ear can perceive, so implants do not provide a good representation of music (Limb and Roy, 2014). They are much more effective at processing speech and about three-quarters of adults are able to use a telephone. The results in adults depend mainly on the duration of deafness and on the integrity of neural structures (i.e. the auditory nerve and brain).

Congenitally deaf children who have no other morbidities and who undergo early implantation will often develop near-normal speech and language and can progress as rapidly as normal-hearing children. Many of them also enjoy music despite the limitations of current technology. Those who are implanted late or who have cognitive difficulties inevitably have poorer long term auditory, speech and educational outcomes (Ganek et al, 2012; Semenov et al, 2012).

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Cost-effectiveness

Cochlear implantation is expensive. The internal device, external processor and accessories cost around £20 000 and require upgrades every few years. The process of assessment, surgery and rehabilitation adds many thousands to that cost. Bilateral implantation adds further costs, although it does not double them. It is clear, however, that the enormous benefits of implantation make this highly cost effective (Bond et al, 2009).

Children's education is transformed (and potentially far cheaper in mainstream schools) and adults can remain in most areas of work. In the elderly for whom education and work are not considerations, their improved social function and quality of life is often dramatic. There are well-documented beneficial effects on depression and cognitive function (Wong et al, 2016).

Many agencies around the world, like the National Institute for Health and Care Excellence, have reviewed cost-effectiveness to determine funding criteria. In the UK, funding is readily available for those who meet standard audiological criteria but adults may receive funding for only one implant (National Institute for Health and Care Excellence, 2009). Ongoing studies aim to assess the cost-effectiveness of less stringent audiological criteria, and of bilateral implantation in adults.

Future developments

Implantation has been restricted to those with bilateral hearing loss, partly because of cost, but also because it had been assumed that patients would not manage to integrate normal hearing in one ear with electric hearing in the other. A number of small recent trials in patients with single-sided deafness have suggested that implantation in this group may be highly beneficial, both in improving auditory function (especially localization and hearing in background noise), and in relieving tinnitus (Cabral et al, 2016). It remains unclear whether this will ever be considered cost-effective.

There are ongoing efforts to produce a fully implantable device. This would be of great benefit in children who often lose or break external processors, it would facilitate activities like swimming, it would provide 24/7 hearing (most patients remove their external device at night), and would have cosmetic benefits. A number of test devices exist but there remain two main difficulties. One is providing sufficient power to the device without cumbersome external chargers. Possible solutions include harnessing power from the body's own heat, motion or biochemistry. The second difficulty is producing a directional microphone that does not over-amplify internal noises. One elegant approach, that has already been used with some middle ear implants, might be to attach a piezoelectric probe to the ossicular chain, thereby using the tympanic membrane as a microphone (Roche and Hansen, 2015).

In the longer term, the limitations of an array of a small number of electrodes will need to be overcome to provide more precise stimulation of auditory nerves. It is not clear how this might be achieved with current technologies: one option may be optical fibre stimulation (Jeschke and Moser, 2015).

KEY POINTS

- Cochlear implants are indicated for severe bilateral sensori-neural hearing loss outside the range of conventional hearing aids.
- They provide direct electrical stimulation of auditory nerve endings, which have a tonotopic arrangement in the cochlea.
- Outcomes depend on the duration of deafness. Congenitally deaf children should ideally be implanted around the age of 1 year. Adults with acquired deafness of up to 20 years may benefit.
- Deafness caused by meningitis requires urgent assessment because of the risk of intracochlear ossification.
- Low frequency residual hearing can be preserved allowing the use of an electro-acoustic hybrid device.
- Children in the UK are eligible for bilateral implantation.

Conclusions

Cochlear implantation has become a safe and routine procedure over the last 30 years and has revolutionized the management of severe hearing loss. The indications have progressively widened to include those with residual hearing, and to cover bilateral implantation. While outcomes are highly dependent on the duration of hearing loss, the integrity of neural structures and postoperative rehabilitation, cochlear implants are highly effective in providing useful hearing to the majority of patients with sensori-neural hearing loss that is too severe to benefit from conventional hearing aids. **BJHM**

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