

OPEN ACCESS GUIDE TO AUDIOLOGY AND HEARING AIDS FOR OTOLARYNGOLOGISTS



COCHLEAR IMPLANTS IN DEVELOPING COUNTRIES

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Cochlear Implants (CI) are surgically implanted electronic devices used to treat severe-to-profound sensorineural hearing loss in children and adults who do not benefit from conventional hearing aids. By 2012 almost 250000 people had CIs implanted, about half of them children. CIs are available in most developed countries and the vast majority of CI recipients live in such countries.

CIs are currently too expensive and as a result virtually unavailable in developing countries. Overcoming this financial hurdle coupled with the lack of specialised CI teams is a difficult and complex challenge. Some countries have fund-raising teams, pressure groups and media campaigns to increase awareness and to raise money for CIs.

The CI device

The first CIs in the 1960s were single-channel electrodes; multiple electrodes were introduced in the 1980s in Australia (*Figure 1*). The basic components of the CI comprise are illustrated in *Figure 2*: externally worn speech processor (A); a transmitting coil and magnet (B); and surgically implantable receiver/stimulator and electrode (C). Sound is detected by the microphone; a signal is transmitted to the speech processor; the speech processor filters, analyses and digitises the sound into coded signals; coded signals are sent to the transmitter coil via a cable; the transmitter coil sends the code through the skin to the internal implant via radiofrequency signals. The internal implant receiver converts the code to electrical signals; the signals are sent to the electrodes to stimulate the auditory nerve fibres and are recognised by the brain as sounds or hearing.

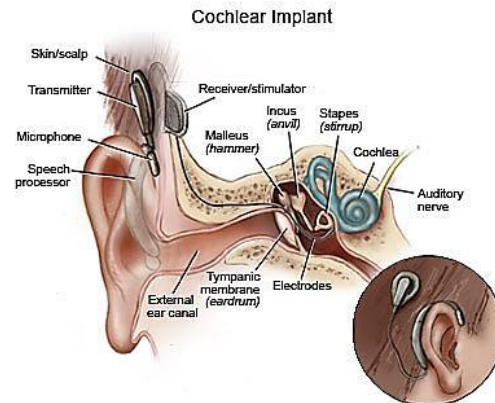


Figure 1: CI device



Figure 2: Components of CI

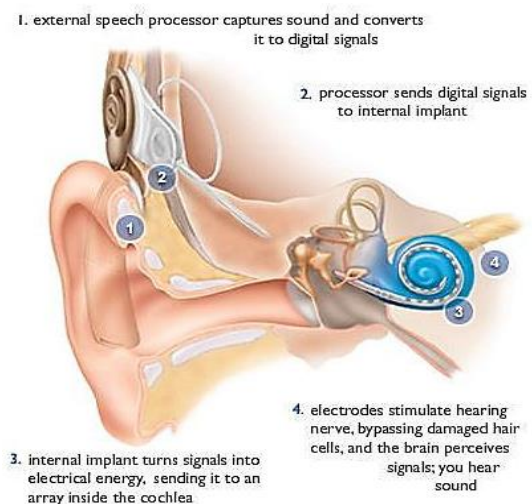


Figure 3: How a CI works

How a CI works can be viewed on U-tube:
www.youtube.com/watch?v=SmNpP2fr57A

Acoustic vs. electrical hearing

Sound stimuli normally produce patterns of electrical excitation along the nervous system pathways. When electrically stimulated, the auditory nerve generates an action potential or a “spike” which is transmitted to the auditory cortex of the brain. In severe-to-profound sensorineural hearing loss the hair cells are non-functional and the auditory system is unable to generate spikes along the auditory nerve in response to sound. The electrode of the CI is positioned in the *scala tympani* close to the spiral ganglion of the auditory nerve. CIs directly stimulate the auditory nerve electrically and bypass the middle ear and the part of the inner ear where the *Organ of Corti* is located. The brain cannot distinguish between spikes generated by hair cells or a CI and interprets it as sound. In a normal functioning cochlea, different regions of the basilar membrane in the *Organ of Corti* vibrate at different sinusoidal frequencies due to variations in thickness and width along the length of the membrane. The cell bodies of the cochlear nerve (spiral ganglion) that transmit information from different regions of the basilar membrane therefore encode frequency tonotopically. This principle of tonotopy is also applicable in electrical hearing.

CI vs. hearing aids

CIs are very different to hearing aids for the reasons outlined below.

Hearing aids amplify sound so it may be detected by the inner ear in the presence of a damaged middle and/or inner ear; ***CIs bypass the inner ear*** and directly stimulate the auditory nerve.

Hearing with a CI is different from normal hearing and takes time to learn and accept. However, a CI almost always

allows a person to recognize warning signals, understand other environmental sounds, and to improve lip-reading capacity. With time and intensive training a patient can enjoy a face-to-face or even a telephone conversation.

The quality of sound perceived with a CI is different from natural acoustic hearing; less sound information is received and processed by the brain. Expectations to regain normal hearing should be downplayed at pre-operative counselling and post-implant training. Newer devices and processing strategies however enable CI recipients to hear better in noise, permit conversation over cell-phones, to learn foreign languages and enjoy a variety of music.

Selection criteria for CIs

Numerous factors have to be considered relating to CI candidacy *inter alia* degree of hearing loss, benefit from hearing aids, duration of deafness, modes of communication, brain plasticity, radiological and medical issues, parental commitment, educational support and financial resources.

Every effort must be made to ensure optimal utilisation of CIs and to avoid non-usage ¹. Only patients with a realistic probability of being successfully rehabilitated should be implanted so that resources are not squandered. Patients must be accessible and audiological facilities and appropriate educational facilities (for children) must be available. As a general rule a CI should not be implanted unless there is a lifetime maintenance guarantee of the CI, and of sociological and educational support.

Table 1 summarises typical referral criteria for cochlear implantation for children and adults. ***The younger the infant at the time***

of diagnosis and cochlear implantation, the better the outcome. CIs may be used in prelingually deaf children of <3yrs (preferably younger) or in children with progressive hearing loss causing post-lingual deafness.

Children	Adults
Bilateral severe-to-profound or moderate-to-profound SNHL	Bilateral severe-to-profound or moderate-to-profound SNHL
No minimum age	No upper age limit
Restricted/no useful benefit from hearing aids: <ul style="list-style-type: none"> • Birth - 2yrs: restricted access to speech sounds with hearing aids • 2-5yrs: failure to develop acceptable level of auditory skills (e.g. speech, language delay) • >5yrs: \leq50% open-set sentence discrimination 	Restricted/no useful benefit from hearing aids: <ul style="list-style-type: none"> \leq50% open-set sentence discrimination scores in ear to be implanted Pre- or post-linguistic hearing loss with oral/aural communication skills Desire to be part of hearing world

Table 1: Referral criteria for cochlear implantation for children and adults ¹

Sensitive periods for neural development exist during the 1st 3yrs of life and are critical for the establishment of auditory mechanisms including understanding speech and language. There are also critical periods for developing brain plasticity. The period of optimal brain plasticity is up to 3½-5yrs of age. From 3.6-7yrs some plasticity still persists; after 7yrs there is very little plasticity and recruitment is often irreversible ². Early auditory deprivation can lead to reallocation of perceptual resources in the auditory cortex. However electrical stimulation by a CI can result in neural survival and development of the central auditory system ³.

Age at implantation is a prognostic variable for *congenitally deaf children*; the earliest implantation is associated with the most normal developmental patterns of auditory and communication skills for congenitally and prelingually deaf

children. The following prerequisites for candidacy for CI should be taken into consideration with older congenitally deaf children and adolescents:

- Early auditory experience (early diagnosis and hearing aid fitting)
- Consistent hearing aid use (all waking hours)
- Spoken language as mode of communication

CI surgery

The CI is ideally done in an anatomically normal ear so as to simplify the mastoidectomy and middle ear surgery. Surgery is done under general anaesthesia. A 5-7cm incision is made behind and above the ear, and a subperiosteal pocket is created under the temporalis muscle. A depression is drilled in the skull to house the receiver so that it does not move around. A partial mastoidectomy and posterior tympanotomy are done, creating an opening between the 3rd segment of the facial nerve and *chorda tympani* nerve. The round window niche with its membrane facing the *scala tympani* is exposed. The CI electrode is passed through the posterior tympanotomy and inserted through an incision made in the round window membrane or through a separate opening drilled into the cochlea (cochleostomy) just inferior and anterior to the round window niche; full insertion of the electrode is preferred. The round window niche is sealed with connective tissue or muscle to reduce the risk of meningitis. Antibiotics are given perioperatively. Patients leave hospital on the 1st or 2nd day, but may also be implanted as day-case surgery (M. Bunne, Oslo, Norway - personal communication).

Device programming (MAPping)

The goal of programming is to adjust the device so that it can effectively convert

acoustic input into a usable electrical dynamic range for each electrode which is stimulated.

The audiologist who does the programming/MAPping is responsible for maintenance and programming of the CI, including initial stimulation and regular follow-up programming sessions. The audiologist obtains 2 psychophysical measures *i.e.* electrical thresholds (T-levels) and comfort levels (C-levels), to create a program (MAP).

An appropriate encoding strategy as well as a stimulation mode should be selected for each patient. Encoding strategy is the means by which an implant translates the incoming acoustic signal into patterns of electrical pulses which in turn stimulate the auditory nerve fibres. Stimulation mode refers to the flow of electric current, that is, the location of the indifferent (reference) electrode relative to the active (stimulated) electrode. A combination of objective and behavioural programming techniques are utilised to establish an appropriate MAP for each CI recipient.

Bilateral CIs

In the developed world, children and growing number of post-lingual deaf adult patients are commonly implanted. Implanting bilateral CIs means that 2 separate internal and external hardware systems are implanted either concurrently or sequentially. The speech processors do not communicate with one another and programming MAPs are done independently for each processor.

Advantages of bilateral CIs include:

- Improved sound localisation
- Improved speech understanding in quiet conditions as well as with background noise
- Both auditory pathways are stimulated

which ensures that the better performing ear has been implanted (since it is difficult to determine pre-operatively which ear will perform better with a CI)

- The 2nd CI acts as a reserve should one of the implants fail
- Ensures binaural hearing and thus assists in a young child's development of central auditory processing skills

The ideal bilateral adult CI candidate has post-lingual hearing loss and a relatively short duration of profound hearing loss (<15yrs) as he/she has had the benefit of maximum binaural cortical development. Consistent and continued use of bilateral acoustic amplification prior to implantation is critical.

Older children benefit from bilateral CIs, but to a lesser degree than younger children. There is a critical time period for binaural auditory development in children. In children < 8yrs of age at the time of CI, the "gap" in speech perception between the 1st and 2nd ear is closed within the first 6-12 months following the 2nd CI ⁴. For children between the ages of 8-13yrs at the time of CI the "gap" in speech perception remains unchanged 2-3yrs after the 2nd CI.

CIs in Developing Countries

Even though the majority of deaf people live in the developing world, the availability of CIs is limited, mainly due to the high costs of the CI (30.000 - 40.000 USD), surgery and post-implantation support, but also due to the lack of specialised otolaryngology and audiology services.

The selection criteria for CIs need to be far more stringent and differ in some respects from that in developed countries due to the paucity of specialised otolaryngology and audiology services and financial constraints.

Establishing a CI Programme in a Developing Country

Audiologists are key members of a unit starting to do CIs in a developing country. Limited audiological service and insufficient rehabilitative resources are therefore the greatest challenges. Audiologists (or specially trained hearing health providers) should be well versed with assessing hearing in young children as well as in severely hearing impaired adults. Detailed knowledge of anatomy, physiology, electrical stimulation, CI technology and rehabilitation is required, and the audiologist must be educated and be familiar with the specific CI system to be used. Intensive, hands-on training with regard to programming software, intra-operative monitoring, programming (MAPping) and trouble-shooting are essential.

An ear surgeon with knowledge of audiology has to be part of the CI team from the beginning. He/she should be familiar with middle ear and mastoid surgery; surgery for CI in itself is not very complicated and in most instances is performed on ears with normal gross anatomy. Multiple CI courses for surgeons are available worldwide⁵.

The major CI brands are the predominantly used in the developing world, mainly because of their extensive experience and the support and services that the providers offer. There is an ongoing debate as to whether a more affordable CI would be more appropriate for low-income countries as it might be of lower quality and not as reliable. However, the prices of devices and accessories will have to be reduced to make CIs accessible to patients in the developing world.

Conclusions

CIs have been a major advance in the

treatment of deafness. Despite the remarkable outcomes of CIs in adult and paediatric patients, its availability as well as the required specialised services remain limited in the developing world.

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