REVIEW ARTICLE

Cochlear Implants

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Abstract

The treatment of bilateral profound sensorineural hearing loss has been revolutionized over the past few decades by the development of cochlear implant technology. This review discusses the history, working of a cochlear implant, candidacy criteria for cochlear implant, surgical procedure and postoperative therapy.

Keywords: Cochlear implant, bilateral profound sensorineural hearing loss.

HISTORY

Volta, an Italian physicist, in 1790, attempted to stimulate the auditory nerve by connecting "electric" batteries to two metal rods which he inserted in his ears. When the circuit was completed he said he received a 'jolt in the head' and then a sound-'a kind of crackling, jerking or bubbling as if some dough or thick stuff was boiling'. Fifty years later Duchenne of Boulogne, France, tried using an alternating current to stimulate his hearing and heard what he described as a sound like an insect trapped between a glass pane and a curtain.[1]

However, skepticism prevented further development of any devices until the middle of the last century. In 1957, Djourno and Eyries, stimulated the cochlear nerve[2] directly in a patient with bilateral cholesteatoma. They placed the active electrode into the stump of the cochlear nerve while the induction coil was placed into the temporalis muscle. The patient apparently could distinguish between higher and lower frequency environmental sounds but could not understand speech.

In 1961, William House a neuro-otologist in Los Angeles along with another surgeon John M Doyle and an electronics engineer, James Doyle, introduced a gold wire into the round window through a postauricular approach in a patient with bilateral otosclerosis.[3] The patient reported hearing the electrical stimuli but the patient had poor loudness tolerance and hence the device had to be removed. Eight years later, House along with Jack Urban[4] developed a single electrode cochlear implant.

Simmons, in 1964,[4] introduced multiple electrodes into the cochlea. This inspired Graeme Clark an otolaryngologist in Melbourne who had realized the limited utility of the single channel implant.[5] In 1967, Clark and his multidisciplinary team started work on developing an artificial hearing device to stimulate the cochlea at multiple points. On 1st August 1978, they successfully implanted the first recipient, Rod Saunders with the first multichannel cochlear implant.[6]

Through the 80's and 90's, there was improvement in speech strategies and miniaturization of the external device with introduction of several safety features making cochlear implants safe and tremendously useful for profoundly hearing impaired adults and children. Currently four companies manufacture cochlear implants commercially: Cochlear, Advanced Bionics, MedEl and Neurelec.

WHAT IS A COCHLEAR IMPLANT?

A cochlear implant is a hearing prosthesis designed to restore or provide a level of auditory sensation to adults and children who have severe to profound bilateral sensorineural hearing impairment and who get limited benefit from hearing aids.[7]

COCHLEAR IMPLANT TEAM

Cochlear implantation is a team effort starting with the early identification of the hearing impairment by the neonatologist–pediatrician–audiologists. Subsequently the physicians, pediatricians, psychologists and radiologists evaluate the patients and the auditory–verbal therapists (habilitionists)

condition the child prior to surgery. After the surgeon has implanted the device, teachers of the hearing impaired or habilitionists provide auditory verbal therapy while the audiologists program (map) the implant. This team effort requires motivated parents/family in addition to funds to undergo the entire process. Postimplantation rigorous therapy is required to enable prelingual children to join the mainstream.

WHO IS A CANDIDATE FOR COCHLEAR IMPLANT? SELECTION OF PATIENTS

The patients are selected for cochlear implantation after thorough evaluation of their type and degree of hearing loss, lack of benefit with hearing aids, radiological determination of the temporal bone–cochlear anatomy for feasibility for implantation, adequate parental motivation for habilitation and the possibility/accessibility of pre- and postimplantation habilitation/therapy and programming centers:

- *ENT examination*: A thorough history is taken to determine the onset, duration and cause of the hearing loss. An otological examination is performed to detect congenital external ear malformations.
- *Audiological evaluation*: The aim is to determine the type and severity of hearing loss. This will include BERA (brain stem evoked response audiometry), ASSR (auditory steady state responses), otoacoustic emissions, pure tone audiometry, impedence audiometry to evaluate middle ear pressure and acoustic reflexes. Once the type and severity of hearing loss is confirmed, the patient is fitted with hearing aids and the hearing is evaluated with hearing aids (aided audiogram). The purpose of hearing aids is to stimulate the auditory cortex, amplify hearing as well as to enable to patient to get conditioned to wearing an external device. Failure to derive benefit with appropriately fitted, powerful hearing aids is an important criterion to select a patient for cochlear implantation.
- *Imaging*: Computed tomography (CT scan) and magnetic resonance imaging (MRI) of the temporal bone are essential to evaluate anatomy and identify cochlear anomalies (Figs 1 and 2), cochlear nerve and brain anomalies which could influence cochlear implantation and its outcomes. Postoperatively imaging studies (transorbital X-rays) are used to determine correct electrode placement within the cochlea (Fig. 3).
- *Speech and language evaluation*: Habilitationists or teachers of the hearing impaired evaluate patient for language and articulation disorders. This enables the patient and his family to have appropriate expectations after cochlear implantation. Postoperatively, patients require speech–language training to join mainstream life.
- *Psychological evaluation*: It is required to determine the mental function, especially in those children or adult

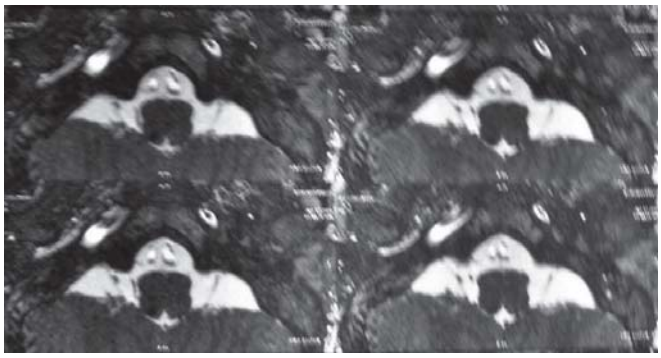

Fig. 1: MRI bilateral Michel's deformity

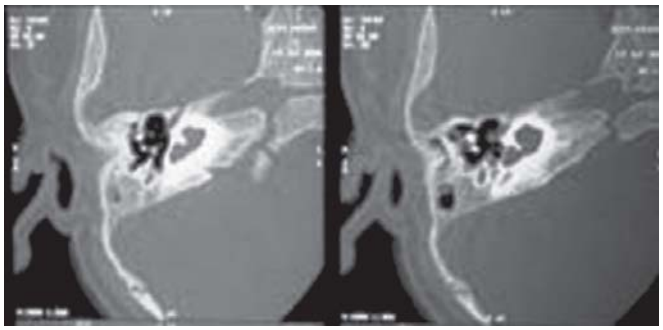

Fig. 2: HRCT temporal bone incomplete partition type 2

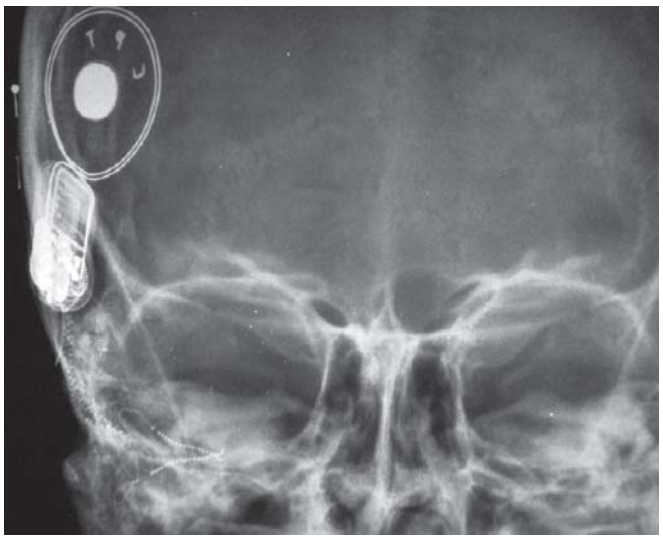

Fig. 3: Postoperative X-ray in a patient with split array for ossified cochlea

patients who have cognitive impairment, delayed milestones or brain anomalies.

- *Ophthalmologic evaluation*: Occasionally patients with hearing impairment may have associated ophthalmologic disorders, e.g. Rubella syndrome (congenital cataract with hearing impairment) or Usher's syndrome (progressive hearing impairment with visual disturbances). Hence it is essential to diagnose these prior to cochlear implantation to enable counseling and adopt appropriate training measures postoperatively.
- *Medical/pediatric evaluation*: It is performed to determine the patient's general health, fitness to undergo surgery while the surgeon attempts to identify the cause of the hearing impairment and determine the patients' candidacy for cochlear implantation. Depending on the time of onset of hearing impairment, the patient's hearing loss is classified as prelingual (hearing loss prior to development of speech and language); perilingual (hearing loss occurring around the period of language acquisition) and postlingual (hearing loss after development of speech and language).
  i. Children:
     - Six months of age or older
     - Bilateral severe to profound sensori-neural hearing loss
     - Insufficient hearing despite consistent hearing aid use to enable the development of functional speech and spoken language.
  ii. Adults:
     - Postlingual onset of profound, severe-profound, bilateral sensorineural hearing loss
     - Little or no benefit from hearing aids.

CONTRAINDICATION FOR COCHLEAR IMPLANTATION

1. Absent cochlear nerve.
2. Absent cochlea (Michel's deformity) (Figs 4 and 5).

PARTS OF A COCHLEAR IMPLANT

A. Implanted components:
- A receiver and stimulator secured in bone beneath the skin, which converts the signals into electric impulses and sends them through an internal cable to electrodes.
- An array of up to 22 electrodes (the number of electrodes varies according to the manufacturer) which are placed in the cochlea and send impulses to the nerves in the scala tympani and then directly to the brain via the auditory nerve.

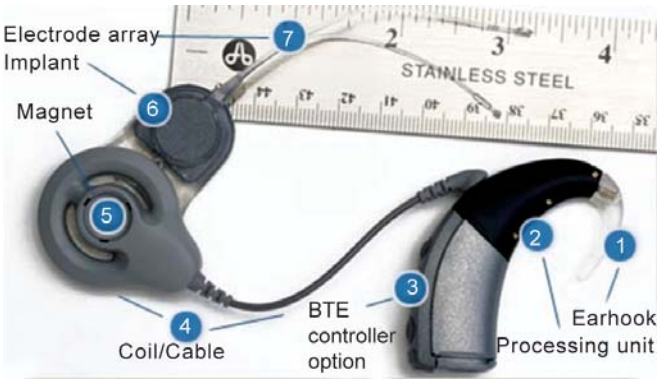

Fig. 4: Parts of cochlear implant
(*Courtesy*: Cochlear Ltd, Australia)

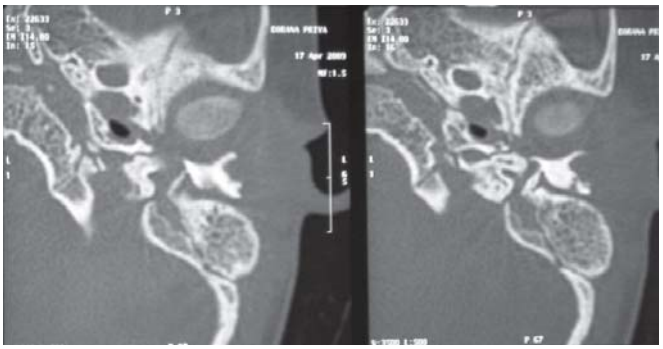

Fig. 5: HRCT temporal bone absent cochlea

B. External components:
- A microphone to pick up environmental sound.
- A speech processor which selectively filters sound to prioritize audible speech and sends the electrical sound signals through a thin cable to the transmitter.
- A transmitter coil, which is held in place by a magnet placed behind the external ear and transmits the processed sound signals to the internal device through electromagnetic induction.

WORKING OF A COCHLEAR IMPLANT

Environmental sound is picked up by the microphone and sent to the speech processor which processes the sound and converts it into electrical signals. These signals are transcutaneously transmitted via the transmitter coil. They are received by the receiver stimulator and sent through an internal cable to the electrodes implanted in the cochlea. The electrodes stimulate the spiral ganglion cells in the scala tympani which conduct the impulses to the brain via the auditory nerve.

TYPES OF ELECTRODES

A. Electrodes for the normal cochlea:
   - *Straight/Standard electrode arrays*: The standard electrode array offers the deepest insertion (approximately 31 mm) and widest contact spacing (2.4 mm) available today.
   - *Perimodiolar electrodes*: These electrodes are designed occupy a position closer to the modiolar wall of the cochlea where the spiral ganglion cells are located. This position is expected to provide better speech understanding due to preservation of residual hearing, selective stimulation of spiral ganglion cell subpopulations and requiring less current for each stimulation.

B. Electrodes for the malformed or ossified cochlea:
   - Double array/Split array are specifically designed for severe ossification of the cochlea. It features two separate electrode branches. Each branch has electrodes and their numbers vary as per the manufacturer. The arrays are designed for insertion into different areas of the cochlea to maximize the number of channels available and optimize performance.
   - *Compressed array*: It is specifically designed for partial ossification or malformation of the cochlea. It has 12 pairs of contacts equally spaced over a shorter distance to maximize the number of electrodes available and to optimize performance.
   - *Hybrid/short array*: Combines the benefits of a hearing aid with that of a cochlear implant. It is useful for those patients who hear some/much low-frequency sounds and little or no high-frequency sounds. A cochlear implant for the high frequencies and a hearing aid for the low frequencies is theoretically an ideal solution.

COCHLEAR IMPLANT SURGERY

Typically cochlear implant surgery is performed under general anesthesia. The patient is placed in a supine position with head turned to the opposite side of implantation. An area of the scalp behind the selected ear is shaved and prepared. The position of the device is chosen on a relatively flat area of the skull. The standard incision measures 4 to 4.5 cm and is located postaurally. A cutaneous flap is raised deep to the scalp and in the avascular plane taking care to avoid perforation of the scalp. Then a large anteriorly based musculoperiosteal flap is raised to expose the areas for a mastoidectomy and for a well. The cortical skull bone is drilled to create a well to house the receiver–stimulator. In very young children the cortical skull thickness is not much and it may be necessary to drill the well almost up to the dura to prevent the receiver–stimulator from bulging through the thin scalp skin flap. Next a cortical mastoidectomy and a posterior tympanotomy are performed. Through the posterior tympanotomy, the round window is identified and the cochleostomy is made just anteroinferior to the round window membrane. The receiver–stimulator of the implant is sited in the well and tied down with sutures to prevent it from moving. The ground electrode is introduced underneath the temporalis muscle so that it is in contact with the temporal bone. Finally the electrode array is introduced into the cochleostomy and the cochleostomy is plugged with connective tissue to prevent any cerebrospinal fluid leak or implant migration. The musculoperiosteal flap is sutured followed by closure of the skin flap. A compression mastoid dressing is given for the next ten days.

POSTOPERATIVE CARE/SWITCH–ON OF IMPLANT

The patient is sent home the day after surgery with a mastoid dressing. The sutures are removed on the tenth post-operative day in adults. In children absorbable sutures are used. The implant is switched–on 10 days to 3 weeks after surgery, once the wound has healed completely.

COMPLICATIONS

A. Minor (Do not require device explantation):
   - *Facial paralysis*: There could be transient facial paresis or even paralysis if the facial nerve is damaged during surgery.
   - *CSF leak and meningitis*: There is risk of meningitis in cases of abnormal cochleas like common cavity and a large vestibular aqueduct, especially if the fundus of the internal auditory meatus is deficient. As a precautionary measure all children undergoing cochlear implantation have to complete a vaccination protocol as promulgated by the US FDA and Center for Disease Control, USA. In addition, the cochleostomy is packed with soft tissue during surgery to prevent any CSF leak. Occasionally a lumbar drain may have to be inserted to reduce or stop the CSF leak.[8]

- *Wound infection*: The skin flap may get infected or the edge of the flap may have poor blood supply and therefore may undergo necrosis. Hence it is advisable to keep the limbs of the incision broad.
- *Loss of taste sensation*: If the chorda tympani nerve is inadvertently damaged during posterior tympanotomy, then there could be ipsilateral loss of taste sensation.
- *Cholesteatoma*: Rarely there could be either retraction of the tympanic membrane with cholesteatoma formation or there could be iatrogenic embedding of epithelium leading to cholesteatoma later. Therefore, one has to be very careful during cochlear implant surgery to avoid epithelial implantation in the middle ear.
- Tympanic membrane perforation.
- Vestibular disturbances.
- *Hematoma/seroma*: Formation under the skin flap.

B. Major (Require device explantation):
- *Device failure*: Hard failures or implant ceasing function are usually sudden and easy to diagnose. They are caused by either internal component short circuits, seepage of fluid, etc and require the implant to be reimplanted.
- Skin flap necrosis with infection.
- Silicon allergy resulting in device explantation.

PROGRAMMING

About 2 to 4 weeks after the surgery, once the wound has healed, the implant is switched on, i.e. activated. The results are not immediate. Serial programming of the implant along with auditory verbal therapy is required before the brain can adapt to hearing and understanding sounds. The implant requires programming or mapping at frequent intervals in the first year after implantation and at regular intervals there after for up to three to five years. During this period children with prelingual hearing loss undergo intensive auditory–verbal therapy to enable them to understand sounds and their meanings thereby helping them to learn language.

CONCLUSION

Cochlear implantation is an accepted modality for the treatment of bilateral profound sensorineural hearing loss. Selection of the patient, parental motivation and intensive auditory verbal therapy are the pillars on which outcomes of cochlear implantation are based.

REFERENCES

1. Niparko, John K, Blake S Wilson. “History of Cochlear Implants”. Cochlear Implants: Principles and Practices. Lippincott Williiams and Wilkins, Philadelphia 2000;103-08.
2. S Djourno A, Eyries C, Vallancien B, De l’excitation electrique du nerf cochleaire chez l‘homme, par induction a distance, a l’aide d’un microbobinage inclus a demeure. CR Soc Biol (Paris) 1957;151:423-25.
3. House WF, Urban J. Long term results of electrode implantation and electronic stimulation of the cochlea in man. Ann Otol Rhinol Laryngol 1973;82:504-17.
4. Simmons FB, Mongeon CJ, Lewis WR. Electrical stimulation of acoustical nerve and inferior colliculus: Results in man. Arch Otolarygol 1964;79:559-67.
5. Clark G. Middle ear and neural mechanisms in hearing and in the management of deafness. Doctor of Philosophy thesis. Sydney, Australia. University of Sydney 1969.
6. Clark GM, Tong YC, Martin F. A multichannel cochlear implant: An evaluation using open-set CID sentences. Laryngoscope 1981;91:628-34.
7. Waltzmann S, Roland JT . Cochlear Implants (2nd ed), 2006 Thieme Publishers, New York, Stuttgart.
8. Migirov L, Dagan E, Kronenberg J. Surgical and medical complications in different cochlear implant devices. Acta Otolaryngol Sep 2008;1:1-4.