COCHLEAR IMPLANTS

Current Status & Future Directions

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Abstract:
A cochlear implant is a surgically implantable electronic device that helps restore hearing in patients with severe-profound hearing loss, unresponsive to amplification by hearing aids. Cochlear implants are designed to detect mechanical sound energy and convert it into electrical signals that can be delivered to the cochlear nerve, bypassing the damaged hair cells of the cochlea. All children below the age of 10 years who have congenital or acquired profound hearing loss and who will not benefit from conventional hearing aids and all adults who have lost hearing after acquisition of language are candidates. A multidisciplinary approach is required involving the surgeon, audiologist and speech therapist, and auditory verbal habilitationist. Pre-operative counseling and post-operative rehabilitation are critical components whose importance cannot be overemphasised.

Introduction:
Cochlear implants offer safe and effective hearing habilitation and rehabilitation for profoundly deafened adults and children.1 Rather than repair diseased organs, the emphasis has changed to bypassing them using bionic prosthetic devices. Auditory neural prosthetic intervention must be done as early as possible due to the phenomenon of neural plasticity. Neural plasticity is the ability of the central nervous system to be programmed to learn a new task. This fades between 6 to 8 years of age.

In 99% of sensorineural hearing loss including congenital hearing impairment, the primary pathology is in the cochlea. In the past, patients with profound sensori-neural hearing loss unresponsive to amplification with hearing aids had to cope by lip-reading or learning sign language. Technological advances have helped restore hearing to these patients. The auditory system is unique in its organization which gives it the opportunity to receive and integrate external electronic circuits. This is due to the low rejection potential of the inner ear and nervous system.

History of implants:
Djourno and Eyries published the first description of cochlear implants in 1957. In 1961, House used a single channel cochlear implant and in 1984, Clark developed a multichannel implant. FDA approval was obtained in 1985. Initially 22 channel implants were used which have now been replaced by 24 channel straight array followed by the 24 contour implant. The first pediatric cochlear implantation was done in the U.S. in 1987.

Fig.1: Tonotopicity of the basilar membrane
COCHLEAR IMPLANTS

What is a cochlear implant?

A cochlear implant is a surgically implantable device that helps restore hearing in patients with severe-profound hearing loss, unresponsive to amplification by hearing aids. Cochlear implants are electronic devices designed to detect mechanical sound energy and convert it into electrical signals that can be delivered to the cochlear nerve, bypassing the damaged hair cells of the cochlea. These signals are then sent to an array of electrodes implanted surgically in the cochlea. The implant system preserves the tonotopic map of the cochlea. (Fig.1).

Cochlear Implant Components:

External components- a microphone which receives sound and transduces it into an electrical wave-form, speech processor which divides the signals into components for each of the electrodes and a transmitting coil which sends the signals across the scalp to the internal components.

Internal components- receiver-stimulator which receives the signals from the transmitting coil and sends it to the electrode array which is implanted in the scala tympani of the cochlea. Speech processors are available as body-worn and ear-level speech processors. (Fig. 2)

Who will benefit from a cochlear implant?

* Bilateral profound cochlear hearing loss unresponsive to amplification by the most powerful hearing aids is the indication for an implant.

* All children below the age of 10 years who have congenital or acquired profound hearing loss and all adults who have lost hearing after acquisition of language are candidates.

The only true prerequisite is an intact auditory nerve. (Fig. 7)

Postlingual candidates do extremely well with an implant. In pre and perilingual candidates, an important factor influencing candidacy is neural plasticity and the emphasis is now on implantation as early as possible to maximize speech understanding and perception. In very young children language acquisition is easier, hence the need for early implantation. Owing to the loss of neural plasticity in older prelingually deaf people, the response to implantation may not be optimal and extensive pre-op counseling regarding realistic expectations is crucial.

The minimum age for implantation in children has come down and children as young as 6 months of age have been implanted. Because the cochlea is full size at birth, there is no anatomic difficulty with electrode insertion in very young children.2

Patients with residual hearing (severe rather than profound hearing loss) are also being implanted. Medical and radiological criteria have expanded to include significant cochlear abnormalities including additional handicaps, as in syndromic deafness.

A multidisciplinary approach is required involving the surgeon, audiologist and speech therapist, and
auditory verbal habilitationist. The patient and his family must be highly motivated for the implant.

**Contraindications**

Cochlear aplasia, absence of auditory nerves, retrocochlear cause of deafness, central deafness, presence of external or middle ear infections, co-existent severe medical illness.

**How much does it cost?**
The overall cost of cochlear implantation, including surgery and post-implant habilitation (for one year) is approximately Rs.6 – 10 lakhs. Under the Tamil Nadu Chief Minister’s Comprehensive Health Insurance Scheme, cochlear implantation is provided free of cost for under-privileged children within 6 years of age.

**Pre-operative Evaluation**

Complete ENT and head & neck examination, including assessment for additional handicaps, hematological tests, TORCH serology if required, and skiagram of chest and ECG for assessing fitness for surgery.

An audiologic assessment is the primary means of determining implant candidacy. Audiological and electrophysiologic investigations include behavioral observation audiometry, puretone (Fig.3) and impedance audiometry (Fig.4), Otoacoustic Emissions (OAE), Brainstem Evoked Response Audiometry (BERA) (Fig. 5), Auditory steady state response (ASSR), aided audiometry and a hearing aid trial. Promontory stimulation testing can be done in older children and adults to assess the response of the cochlea to electrical stimulation.

**Magnetic Resonance Imaging** is the gold standard investigation for the assessment of cochlear anatomy and the vestibulocochlear bundle. (Fig.6) It reveals anomalies like Mondini deformity and Michel’s aplasia, labyrinthitis ossificans, absent eighth nerve.

**Fig. 3:** Pure tone audiometry showing profound hearing loss in both ears

**Fig. 4:** Impedance audiometry showing bilateral ‘A’ type with absent reflexes

**Fig. 5:** BERA showing no peak at maximum intensity

Assessment by a pediatrician is essential. Specialist referral e.g. to an ophthalmologist & cardiologist is essential to exclude any additional handicap or syndrome. exclude any additional handicap or syndrome.
Surgical Procedure

The goal of cochlear implant surgery is to insert the entire electrode array into the scala tympani with as little damage as possible to the structure of the inner ear. Implantation is a 1.5-2.5 hour procedure performed with strict aseptic precautions and is done under general anesthesia. Surgery is essentially the same in children and adults because the anatomic structures are of adult configuration at birth.

Pre-op rehabilitation is important before surgery. Counseling patients and parents prior to implantation to develop realistic expectations of the likely outcome is vital.

The steps of surgery are

1. **Incision**- usually an extended post-auricular incision is made to expose the mastoid cortex. The incision should be made more than one cms from the body of the implant.

2. **Simple Mastoidectomy**- The mastoid is drilled out to expose the mastoid antrum. Saucerization of the cavity is not done. (Fig.8)

3. **Posterior Tympanotomy**- the Facial recess is opened and the promontory and round window niche are exposed, without exposing the facial nerve

4. **Well for Receiver-Stimulator**- this is fashioned in the skull behind the mastoid cavity using a template as a guide and a groove is made to connect it to the mastoid cavity. Tie-down holes are made on either side of the well for securing the implant.

5. **Cochleostomy**- the basal turn of the cochlea is opened anterior to the round window to make the axis of introduction of the electrode array straighter. In recent years, round window insertion of the electrode array is the preferred technique.

6. **Insertion of electrode array**- the electrode array is inserted atraumatically into the scala tympani using a claw (Fig.10). Once the electrodes are inserted, diathermy should not be used.

Fixation of the device and electrode array and wound closure is done. **Electrophysiologic testing or Telemetry** is performed after implanting the electrode array. This assures the team that the device is functioning on the table and that the patient is receiving an auditory stimulus and responding appropriately.

**Post-operative Care:**

A pressure dressing is applied. Antibiotics and analgesics are administered in the post-operative period. The implantee is discharged home on the 2nd post-operative day. Suture removal is done on the 10th post-operative day The patient is asked to review 3 weeks post-op for switch-on of the device. (Fig.11). Frequent mapping sessions are required and prolonged and intensive (re)habilitation after implantation is essential. Rehabilitation aims at
improving receptive language skills and expressive skills. Post implantation rehabilitation is critical for children to optimize the usefulness of an implant.  

Cochlear implantation in specific anomalies:

Cochlear implantation in Labyrinthitis Ossificans: The most common abnormality encountered is the ossified cochlea, most commonly after meningitis, although other pathologies may predispose to ossification, including otosclerosis, chronic otitis media, ototoxicity, autoimmunity, trauma and others. This remains one of the significant surgical challenges for the otologist.

Cochlear implantation in Mondini deformity: The cochlea has only 1.5 turns instead of the usual 2.5 turns. CSF leak during cochleostomy has to be sealed.

It is important to rule out an absent eighth cranial nerve. A variety of techniques may be used to help control the flow of CSF including firm plugging of the cochleostomy using soft tissue coupled with reducing the flow of CSF by lumbar drainage, if necessary. Such leaks may also be encountered in cases of enlarged vestibular aqueduct.

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Fig 8: Cortical mastoidectomy

Fig 9: Cochleostomy in progress

Fig 10: Insertion of electrode array into the scala tympani of the basal turn of the cochlea

Fig 12: Mondinin deformity with CSF fistula

Fig 13: Enlarged vestibular aqueduct
Auditory Neuropathy / Auditory dyssynchrony: Normal outer hair cell (OHC) function and dyssynchronous neural responses characterize auditory neuropathy / auditory dyssynchrony (AN / AD). Cochlear Implants are a viable management option for patients with AN / AD.

Cochlear implantation in multi-handicapped individuals:

Early diagnosis and rehabilitation of deafness and additional handicaps such as blindness is crucial. However, patient selection criteria must be stringent. The decision to pursue implantation should be as fully informed as possible.

Jervell and Lange–Nielsen syndrome represents an important cause of congenital hearing loss. Because of the potential for cardiac arrhythmias and sudden death, additional risks are involved in cochlear implantation. 9

Usher’s syndrome is an important and common cause of blindness associated with deafness. Ophthalmic evaluation and follow up is crucial. Habilitation can be challenging in these children.

Minimally Invasive Cochlear Implantation: Due to improvements in Cochlear Implant technology, smaller and more powerful implantable cochlear stimulators have evolved which has enabled smaller external incisions, smaller skin flaps, shortened surgical time and faster healing. Current techniques in cochleostomy (Peephole Cochleostomy) and electrode insertion (soft insertion) have resulted in preservation of residual hearing. 10

Complications of Cochlear Implantation: Cochlear implantation is generally a safe procedure. However, complications may occur occasionally. Major complications include facial palsy, implant exposure due to flap loss and wound infection. Other complications include bio-film formation, facial nerve stimulation, device failure, deterioration of hearing, tinnitus, temporary balance problems, numbness of scalp, loss of taste, electrode/device extrusion, CSF leak and meningitis.

The Future: Cochlear implant surgery and technology continue to evolve. In the future, fully implanted devices, improved speech coding strategies, cochlear hair cell and nerve growth factors used in conjunction with an implant may be available.

References: