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Auditory Neural Prostheses – A Window to the Future

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SUMMARY

Hearing loss is one of the commonest congenital anomalies to affect children world-over. The incidence of congenital hearing loss is more pronounced in developing countries like the Indian sub-continent, especially with the problems of consanguinity. Hearing loss is a double tragedy, as it leads to not only deafness but also language deprivation. However, hearing loss is the only truly remediable handicap, due to remarkable advances in biomedical engineering and surgical techniques. Auditory neural prostheses help to augment or restore hearing by integration of an external circuitry with the peripheral hearing apparatus and the central circuitry of the brain. A cochlear implant (CI) is a surgically implantable device that helps restore hearing in patients with severe-profound hearing loss, unresponsive to amplification by conventional hearing aids. CIs 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. The only true prerequisite is an intact auditory nerve. The emphasis is on implantation as early as possible to maximize speech understanding and perception. Bilateral CI has significant benefits which include improved speech perception in noisy environments and improved sound localization. Presently, the indications for CI have widened and these expanded indications for implantation are related to age, additional handicaps, residual hearing, and special etiologies of deafness. Combined electric and acoustic stimulation (EAS / hybrid device) is designed for individuals with binaural low-frequency hearing and severe-to-profound high-frequency hearing loss. Auditory brainstem implantation (ABI) is a safe and effective means of hearing rehabilitation in patients with retrocochlear disorders, such as neurofibromatosis type 2 (NF2) or congenital cochlear nerve aplasia, wherein the cochlear nerve is damaged or absent on both sides and hence, a cochlear implant (CI) would be ineffective. In such patients, the brainstem implant bypasses the damaged / absent cochlear nerves and directly stimulates the cochlear nucleus in the brainstem. The auditory midbrain implant (AMI) has been designed for stimulation of the auditory midbrain, particularly the central nucleus of inferior colliculus (ICC). It is used especially in patients with large neurofibromatosis type 2 (NF2) wherein tumor induced damage to the brainstem/cochlear nucleus often co-exists. The efficacy and safety of auditory neural prostheses is well proven. Advancements in technology will enhance the benefit provided by these prostheses.

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Introduction:

Hearing loss is one of the commonest congenital anomalies to affect children world-over. WHO reports that nearly 2 - 3 per 1000 live births are found to have severe to profound hearing loss, making it the most common congenital abnormality to affect newborns world over. This scenario is even more pronounced in developing countries like the Indian sub-continent, especially with the problems of consanguinity. Hearing loss at birth is considered a social stigma even in present day society and ends as a double tragedy, as it leads to not only deafness but also language deprivation. However, hearing loss today, both congenital and acquired is the only truly remediable handicap, due to remarkable advances in biomedical engineering and surgical techniques. The advent of auditory neural prostheses, which are indicated for varying types and extent of hearing losses, has successfully broken the acoustic barrier, thus integrating people with hearing loss into normal society and providing them with a highly productive quality of life.

Auditory neural prostheses help to augment or restore hearing by integration of an external circuitry with the peripheral hearing apparatus and the central circuitry of the brain. They are safe and extremely effective in restoring hearing to both children and adults with severe - profound hearing loss, who do not receive benefit from conventional hearing aids. These implantable devices electronically stimulate the cochlea / auditory nerve or the higher hearing centers in the brain. The auditory system is unique in its organization because of the phenomenon of tonotopicity (place-pitch organization) which gives it the opportunity to receive and integrate external electronic circuits. This is possible because of the low potential for rejection of the device by the ear and nervous system. Thus, hearing restoration is the first successful path-breaking attempt in medical science to integrate an electronic device with the central nervous system, in order to fully restore a lost special sense organ.

COCHLEAR IMPLANTS

A cochlear implant (CI) is a surgically implantable device that helps restore hearing in patients with severe-profound hearing loss, unresponsive to amplification by conventional hearing aids. CIs 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 electrical signals are processed by an external speech processor and sent via a radiofrequency interface into an array of electrodes implanted surgically within the cochlea. The implant system preserves the tonotopic map of the cochlea and the auditory brain perceives these electrical impulses as sound(1).

Djourno and Eyries published the first description of cochlear implants in 1957. In 1961, House used a single channel CI and in 1984, Clark developed

the popular multichannel implant. FDA approval for CI was obtained in 1985. The first pediatric cochlear implantation was done in the US in 1987. Presently, a spectrum of implants is available along with improved speech processing strategies. Rapid technological advancements in bioengineering and implant manufacturing methods have led to miniaturization of the device, with refinement in sound signals, providing better "hearing in noise" and music appreciation among cochlear implantees.

Components of a Cochlear Implant System

The implant has external components, consisting of a microphone which receives and transduces sound into an electrical waveform, a 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. The internal components include a 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. (Fig.1)

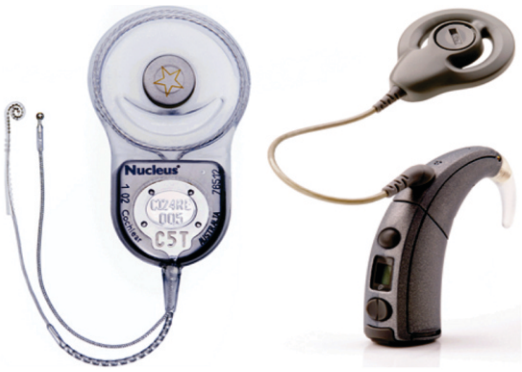

Fig. 1 : Cochlear implant-internal and external components

Speech processors are currently available as body worn and ear level speech processors. All components play an important part in converting sound to an electrical stimulus. The microphone receives and transduces sound into an electrical representation. This is done in an analog (continuous) fashion. The external speech processor and signal-transfer hardware shapes the electrical signal. This requires amplifying, compressing, filtering, and shaping. Amplification is necessary to increase some signal levels to the point that they can be used in the electrical circuits. Compression is a necessary second step of signal modulation. The normal human ear can hear gradations of sound intensity in a range of 120 dB. Persons with severe to profound hearing loss do not have this same range. In the high frequencies, their dynamic range (the difference between their absolute threshold and painful sound) can sometimes be only 5 dB. The range in the lower frequencies is often 10–25 dB. This means that significant compression of the sound energy must take place in order to render it useful. Thus, all cochlear implants employ gain control of one kind or another. These systems monitor the output voltage and adjust the ratio of compression to keep the output in a range where it provides useful, but not painful stimuli.

Filtering of the input signal is the

next step. Frequencies between 100 Hz and 4000 Hz are generally those most important for understanding speech. Sound energy is analyzed using several different types of filters. This allows the unimportant frequencies to be removed and the frequencies of interest to be separately modified. Useful sound information is filtered into frequency bands. This information can then be analyzed for speech patterns and channeled to the appropriate portion of the electrode array. The transmitter, or outer coil, is placed on the mastoid (usually held in place by magnets) and sends the processed signal to the receiver via radiofrequency. The receiver, surgically placed in a well over the mastoid, receives the signal and sends electrical energy to one or many electrodes in the array. The electrode array, which lies within the cochlea, delivers the electric signal to electrodes along its length. The electrical field generated at these locations serves to discharge the neural components of the auditory system. The eighth nerve then conveys the signal. Just as important as any of the man-made components is the individual's ability to adjust to, interpret and respond to the electrical stimulus. Length of time spent without sound stimulation of the auditory system, presence or absence of previous experience with sound, personal motivation, community or family support, and opportunities for rehabilitation have been shown to be important factors in achieving a good outcome(2). These factors likely are important in understanding significant differences in patient outcomes despite similar preoperative auditory deficits, surgical course, and CI hardware.

Types of Cochlear Implants

Cochlear implants differ in the way that they process sound and how they present electricity to the hearing nerve. Other than the speech processing strategies discussed below, there are two different ways of encoding sound information. The first form, analog coding, involves continuous coding of the sound signal with subsequent transfer to the receiver in multiple radiofrequency channels. Electrodes are continuously stimulated. The second form, digital coding, requires sampling of the sound waveform and assigning a number to these "bits" of information. These bits of information are then transferred to the receiver where they are decoded. Electrodes are stimulated in a pulse fashion. Interestingly, neither approach is 100% effective for all implant users. Recently, combining the two schemes has seen some success. Cochlear implants can also be distinguished by their use of single versus multiple channels, the number of electrodes, and their use of either monopolar or bipolar stimulation. The number of electrodes stimulated with different electrical stimuli determines the "channels" used. In other words, an implant may have multiple electrodes but if the same information is presented to all the electrodes at one time, they are essentially functioning as a single channel system. In contrast, multichannel devices

provide different information to several electrodes or groups of electrodes. Early implants had only one electrode (and one channel); recent advances have led to the development of implants with multiple electrodes (22) and multiple channels (usually 4 to 8). Having more electrodes means that multiple channels can be localized to areas of the cochlea that are most responsive, and stray current that is stimulating adjacent structures (facial nerve, vestibular nerve) can be rerouted.

Cochlear implants can employ monopolar or bipolar stimulation. In a monopolar system, there is only one ground electrode for all the others. The ground is usually located at or outside the round window. Thus, an electrical field is created from the stimulated electrode to the ground. A bipolar arrangement is such that the ground for each electrode is much closer (adjacent to, or a few electrodes away). In the highly conductive environment of the inner ear, monopolar stimulation results in some limitations. As additional electrodes are stimulated with different streams (channels) of information, the electrical fields created by stimulated electrodes may interfere with fields at other sites. This makes it difficult to stimulate more than one electrode at a time, or electrodes that are close together. The bipolar configuration was an attempt to limit this interaction by placing a ground near each electrode, such that a smaller field would be created with less interference and more discrete stimulation. Once again, one approach does not achieve satisfaction with all patients. As a result, many implants offer both grounding methods.

Indications for Cochlear Implantation

Bilateral profound cochlear hearing loss, unresponsive to amplification by the most powerful hearing aids, is the prime indication for a CI. All children below the age of 6 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 ideal candidates. The only true prerequisite is an intact auditory nerve. Postlingual candidates do extremely well with an implant and in prelingual 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 preoperative counseling regarding realistic expectations is crucial. Presently, the indications have expanded to include candidates with low frequency residual hearing and those with severe hearing loss. These expanded indications for implantation are related to age, additional handicaps, residual hearing, and special etiologies of deafness. 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. Medical and radiological criteria have expanded to include significant cochlear abnormalities including additional handicaps, as in syndromic deafness. The recent trend is towards bilateral simultaneous or sequential implantation, which provides immense benefits of binaural hearing.

Contraindications for Cochlear Implantation

Not all patients with sensorineural hearing loss are good candidates for cochlear implantation. For example, patients with pure tone thresholds greater than 90 dB with residual hearing through 2000 Hz often do better with hearing aids than with implantation. The absence of the cochlea (Michel deformity) and a small internal auditory canal (associated with cochlear nerve aplasia) are contraindications to implantation on that side. Other forms of dysplasia are not necessarily contraindications. However, when implantation of a dysplastic cochlea is to be undertaken, informed consent is especially important. Cochlear implants in these patients are associated with increased risk of poor outcomes, cerebrospinal fluid (CSF) leak, and meningitis. A diagnosis of neurofibromatosis II (history of progressive hearing loss and suggestive MRI findings), mental retardation, psychosis, organic brain dysfunction, and unrealistic expectations may also be contraindications.

The presence of active middle ear disease is a contraindication to surgery. This should be treated and resolved before implantation. Patients with a history of canal wall down mastoidectomy may need surgery to reconstruct the posterior canal wall or close off the canal.

Meningitis may lead to hearing loss and ossification of the cochlea. Labyrinthitis ossificans is usually identifiable on CT scan and Magnetic resonance imaging. Adults and children with acute meningitis should be treated with steroids to avoid hearing loss. In patients with profound hearing loss, implantation must be advocated as early as possible.

Advanced otosclerosis can also cause ossification of the basal turn of the cochlea. This finding is most often noted on CT scan. This is not a contraindication as long as the surgeon is prepared to perform a drill out or pursue implantation into the scala vestibuli. Patients with otosclerosis can achieve excellent results from implantation.

Preoperative Assessment

Prior to implantation, a basic workup including hematological, chest X-ray, ECG, TORCH screen need to be performed. An audiologic assessment is the primary means of determining implant candidacy. Audiological and electrophysiologic investigations include

puretone or behavioral audiometry and impedance audiometry, otoacoustic emissions (OAE), brainstem evoked response audiometry (BERA), auditory steady state response (ASSR), aided audiometry and a hearing aid benefit evaluation. Promontory stimulation testing can be done in older children and adults to assess the response of the cochlea to electrical stimulation.

High resolution CT scans of the temporal bones are done to plan the surgical route for implantation, identify the vital structures like the facial nerve and promontory, and also to rule out any evidence of middle ear disease/ mastoiditis. MRI is the gold standard investigation for the assessment of cochlear anatomy and the vestibulocochlear bundle(3). (Fig.2 &3) CT/MRI reveal anomalies like Mondini's and Michel's aplasia, labyrinthitis ossificans, or absent eighth nerve. Rapid advances in genetics and molecular biology are revolutionizing our understanding of congenital deafness, and genetic counseling should play an important part in prevention. Hence, a genetic specialist's opinion is sought in patients with syndromic etiology of deafness. Children need to get evaluated by a child psychologist for assessment of mental functions and IQ, prior to implantation and an ophthalmologist needs to perform a fundus examination to rule out associated visual impairment as seen in Usher's syndrome. In children, pre-implant meningococcal vaccination is carried out.[4] Preoperative habilitation is important before surgery. Counseling patients and parents prior to implantation to develop realistic expectations of the likely outcome is vital. Hence, candidates and parents need to meet and interact with other cochlear implantees, to have a perspective on the procedure and its outcome.

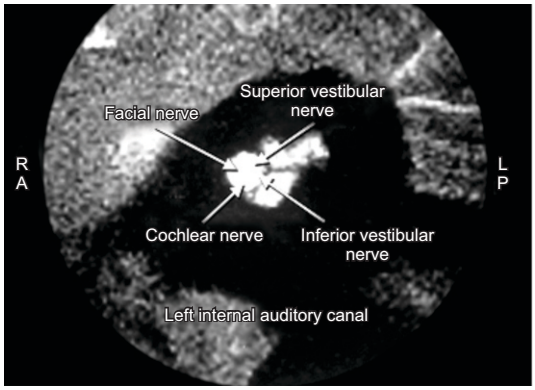

Fig. 2: MRI of the internal auditory meatus showing intact cochlear nerve

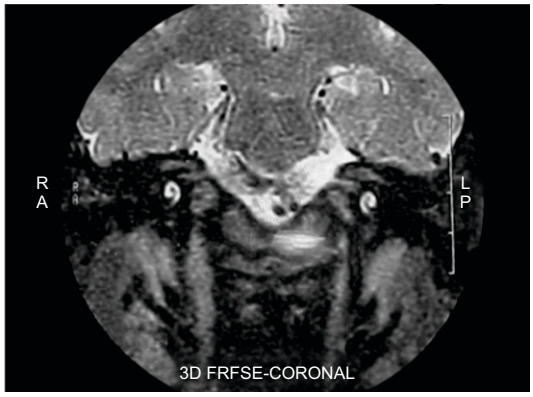

Fig. 3: MRI showing bilateral normal fluid-filled cochlea—the "Comma" sign

Cochlear Implantation Surgery

The goal of CI 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. The success of cochlear implantation depends on scrupulous attention to technique at all the various steps of the procedure. Implantation is 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. However, in very young children, there is a slightly increased risk of facial palsy and blood loss may be an issue.

The steps of surgery are as follows: usually an extended postauricular incision is made to expose the mastoid cortex. The incision should be made more than 1 cm away from the location of the coil of the implant. The mastoid is drilled out to expose the mastoid antrum. Saucerization of the cavity is not done. Posterior tympanotomy is performed, the promontory and round window niche are exposed, without exposing the facial nerve. A well for receiver-stimulator 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. Cochleostomy is done at the basal turn of the cochlea which is opened anterior to the round window to make the axis of introduction of the electrode array straight. The electrode array is inserted atraumatically into the scala tympani using a claw. Alternatively, a round window insertion may be performed after drilling out the anterior lip of the RW niche and adequately exposing the secondary tympanic membrane (Fig.4). Once the electrodes are inserted, diathermy should not be used. Fixation of the device and electrode array (Fig. 5) and wound closure is done.

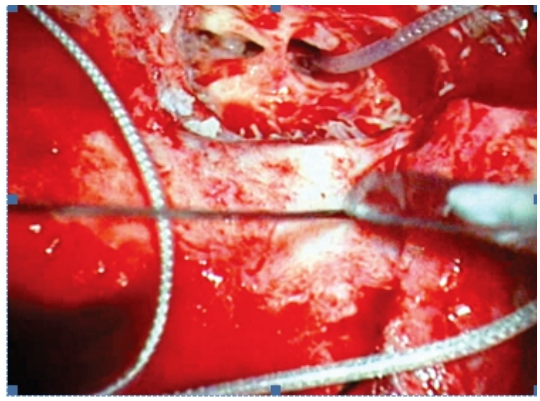

Fig. 4: Cochlear implantation electrode array in situ

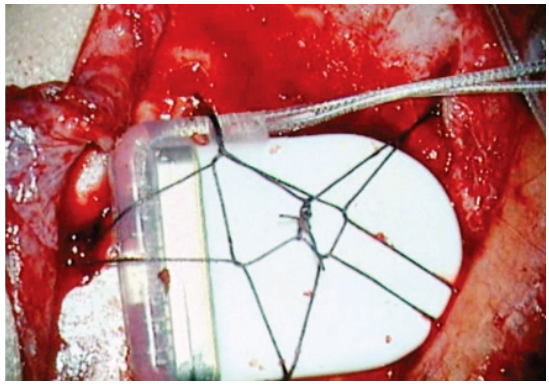

Fig. 5: Cochlear implantation receiver-stimulator coil in situ

Electrophysiological testing (impedance telemetry), neural response telemetry and electrically evoked stapedial reflex thresholds are performed intraoperatively to confirm the optimal performance of the implant in situ. This assures the implant team that the device is

functioning and that the patient is receiving an auditory stimulus and responding appropriately.

Switch-on and Mapping of Cochlear Implant

The switch on and speech processor tuning is done 3 weeks after surgery. Mapping is done at periodic intervals till a stable map is achieved. Frequent mapping sessions are required, and prolonged and intensive (re)habilitation after implantation is essential. Habilitation aims at improving receptive language skills and expressive skills. The habilitation program is started out based on baseline skills of the patient; periodical assessments of outcome need to be done in terms of environmental sound, open set, closed set speech, speech discrimination and telephonic conversation. The recommended period for auditory verbal habilitation is 1 year.

Outcomes of Cochlear Implantation

The success of a CI program is directly dependent on its ability to address the issue of patient expectations and balance it with the outcomes. A multidisciplinary approach is required involving the ENT surgeon, audiologist, speech therapist, auditory verbal habilitationist, child psychologist, and pediatrician. The patients and their family must also be highly motivated for the implant. Variables affecting the outcome of CI in children are the duration and etiology of deafness, age at onset of deafness, preimplant amplification history, communication mode, age at implantation, type of speech processor used, and duration of implant usage. In very young children, language acquisition is easier and hence the need for early implantation. Owing to the loss of neural plasticity in older prelingual deaf people, the response to implantation may not be optimal and extensive preoperative counseling regarding realistic expectations is vital. Factors influencing the overall outcomes are the transparency of the program, expertise of the team, patient motivation, family support, and facilities for habilitation.

Complications

The surgical complication rate after cochlear implantation is estimated to be less than 5%. The most common problems are wound infection, biofilms and wound breakdown. Rarely, extrusion of the device, facial nerve injury, bleeding, CSF leaks and meningitis, vertigo, tinnitus, facial nerve stimulation, numbness of scalp, loss of taste can occur. Device-related complications include intracochlear damage, slippage of the array, breakage of the implant, and improper or inadequate insertion and device failure.

Centers for Disease Control and Prevention (CDC) recommend vaccination of implanted or soon to be implanted patients. Children less than 2 years of age who have implants should receive pneumococcal conjugate vaccine. Children with implants 2 years and older who have completed the conjugate series

should receive one dose of the pneumococcal polysaccharide vaccine. Children with implants between 24 months and 59 months who have never received vaccination should receive two doses of pneumococcal conjugate vaccine 2 months apart, and then one dose of pneumococcal polysaccharide vaccine at least 2 months later. Finally, persons aged 5 years and older with cochlear implants should receive one dose of pneumococcal polysaccharide vaccine.

Difficult Scenarios in Cochlear Implantation

With increasing experience in cochlear implantation, the indications for implant surgery have widened to include cochlear anomalies, syndromic associations, and multiple handicapped individuals. Implantation is beneficial in such situations. However, the surgeon must anticipate challenges during implantation and also, the subsequent habilitation may be challenging.

Cochlear Implantation in Labyrinthitis Ossificans

A common abnormality encountered is the ossified cochlea, mostly occurring as postmeningitic sequelae, 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. It is diagnosed with a CT/MRI scan. On confirmation of an obstructed basal turn, the proximal turn is drilled with a microdrill to a depth of 6–8 mm until an open lumen is discovered, and the electrode array is inserted. In total ossification, a complete drill-out of the basal turn is required and the implanted array is seated in a trough that surrounds the modiolus. A double-array implant may be used with some electrodes into the basal turn and others into the second turn.

Cochlear Implantation in Mondini's Deformity/Large Vestibular Aqueduct Syndrome

Cerebrospinal fluid leak during cochleostomy has to be sealed. 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 and intravenous mannitol drip, if necessary. Such leaks may also be encountered in cases of enlarged vestibular aqueduct (LVAS)**.** The 'pulsatile stapes sign' has been described by the author to diagnose LVAS intra-operatively.

Auditory Neuropathy/Auditory Dys-synchrony Spectrum Disorder

Normal outer hair cell (OHC) function and dys-synchronous neural responses characterize this disorder. Patients will show a normal OAE with absent BERA waveforms, which is pathognomonic of this condition. Cochlear implants are a viable management option for patients with auditory neuropathy/auditory dys-

synchrony spectrum disorder (AN/ADSD) and are beneficial in bypassing the desynchronous neural network, but the outcomes may be suboptimal or guarded, and the family needs to be counseled regarding the same.

Cochlear Implantation in Multihandicapped Individuals

Early diagnosis and rehabilitation of deafness and additional handicaps are crucial. An implant helps in the habilitation of deafness and other handicaps as well. However, patient selection criteria must be stringent. Evaluation, surgical intervention, and postimplantation management of these patients can be challenging.

Minimally Invasive Cochlear Implantation

Due to improvements in CI technology, smaller and more powerful implantable cochlear implants have evolved which has enabled smaller external incisions, smaller skin flaps, shortened surgical time, and faster healing. Current techniques in cochleostomy (Peep-hole cochleostomy) and round window electrode insertion (soft insertion) have resulted in preservation of residual hearing.

Bilateral Cochlear Implantation

Bilateral CI has significant benefits which include improved speech perception in noisy environments and improved sound localization.(5) The advantages include elimination of head-shadow effect, significant benefits from summation effects (improvement in hearing threshold from redundant information presented to each ear) and squelch effects (improvement in hearing threshold from brainstem processing of inter-aural time and intensity differences).

Cochlear Endoscopy

Cochlear endoscopy was first described by Balkany and colleagues in 1990 who used flexible fiberoptic microendoscopes (0.7–1 mm diameter)(6). Currently, the indications for cochlear endoscopy are limited and it is not recommended routinely during CI. The present indications are visualization of obstructed segments of the cochlea in labyrinthitis ossificans and the interior of the cochlea in cochlear dysplasia. Visualization of the interior of the cochlea will help in preinsertion assessment as well as to verify proper insertion of the implant.

Perimodiolar and Midscalar Cochlear Implantation

These implants are assumed to have a slightly enhanced speech perception. After the electrodes are inserted into the cochlea, the stylet is withdrawn and the electrodes come into a perimodiolar/midscalar position. The electrode-neural interface seems to be minimal in this position, and hence clarity of auditory inputs are much better.

Future Directions In Cochlear Implantation

CI surgery and technology continue to evolve. In the future, fully implanted devices (like the TIKI prototype), improved speech coding strategies, cochlear hair cell, and nerve growth factors used in conjunction with an implant may be available.

MERF Experience

Nine hundred cochlear implantations were performed over 15 years. Majority of candidates were pre-lingual, 10% candidates were postlingual and 20% were peri-lingual. Outcomes were dependent on age at implantation and duration of deafness, the best outcomes were observed when implantation was performed before 3 years of age. Children responded better with very good outcomes if implanted early. Children in 1-5 yrs age group achieved category 7 (use telephone) on CAP score and category 5 (connected speech intelligible to all listeners) on SIR score earlier than children in 6 -10 years age group.

Electroacoustic Stimulation

One of the latest applications of implantable hearing technology combines electric and acoustic stimulation (EAS) into a hybrid device designed for individuals with binaural low-frequency hearing and severe-to-profound high-frequency hearing loss.

Indications

Electro-acoustic stimulation is the latest strategy conceptualized for residual hearing preservation in the implanted ear, in order to provide combined electrical stimulation and acoustic hearing for candidates with bilateral high-frequency, severe-to-profound sensorineural hearing loss. The addition of the electrical stimulation to such patients, with existing residual low frequency hearing, can provide clear speech recognition in background noise and better appreciation of musical notes. Low-frequency thresholds generally can range from 20 dB HL to 60 dB HL through 750 Hz, and thresholds at 1000 Hz and above must generally exceed 60–70 dB HL Preoperative speech perception criteria require that aided consonant nucleus consonant (CNC) monosyllabic word recognition score in the ear to be implanted cannot exceed 50–60%. Individuals with binaural high frequency hearing loss may not gain significant benefit from traditional hearing amplification. Their relatively good low-frequency hearing may disqualify them from conventional cochlear implant (CI) candidacy. As a result, individuals with good low-frequency hearing and severe-to-profound high-frequency hearing loss can experience significant difficulty in everyday communication, particularly in noisy backgrounds, where low-frequency information alone is not sufficient to allow high levels of speech understanding.

In recent times, implant surgeons are employing soft surgical techniques

which include a smaller cochleostomy or round window insertion, performed gently with thinner electrode arrays and/or perimodiolar electrodes (atraumatic cochlear insertion) which contribute to hearing preservation with standard cochlear implants. The hybrid device uses a shortened CI electrode array that is inserted just 10-20 mm into the cochlea (versus 20-30 mm for a conventional implant), covering the basal two third of the cochlea. A successful surgical outcome allows for monaural electric stimulation of the basal cochlea for high-frequency information without damaging apical cochlear structures that transmit low-frequency acoustic information. This combination allows for the integration of electric and acoustic perception in the same ear.

Components of Hybrid Implant

The EAS system consists of two parts: a CI with a soft and flexible electrode array for preservation of residual low frequency hearing, and a speech processor which combines the CI component with conventional acoustic stimulation in one comfortable and compact device. EAS patients wear an in-the-ear (ITE) hearing aid in the implanted ear (which can amplify sound signals up to 43 dB acoustical gain) in combination with an external ear-level or body-worn speech processor or an integrated hearing aid/speech processor on the implanted side. Surgery for EAS is very similar to conventional cochlear implantation, and round window insertion is often preferred for optimal hearing preservation. The hybrid implant has a specialized microphone competent for parallel processing of sounds. The acoustic and electric digital sound processing components of the EAS processor receive sound signals from this single microphone. The parallel processing of these signals is performed separately and optimized for both acoustic stimulation (focusing on low-frequency hearing) and CI stimulation (focusing on high-frequency hearing). This microphone automatically adjusts to incoming sounds in order to capture all the vital cues necessary for understanding speech clearly without requiring special programming or the use of a switch to shuffle between the two modes of hearing.

Enhanced music perception is one of the major benefits reported by candidates who receive EAS implants.

Auditory Brainstem Implants

Auditory brainstem implantation (ABI) is a safe and effective means of hearing rehabilitation in patients with retrocochlear disorders, such as neurofibromatosis type 2 (NF2) or congenital cochlear nerve hypoplasia/aplasia, wherein the cochlear nerve is damaged or absent on both sides and hence, a cochlear implant (CI) would be ineffective. In such patients, the brainstem implant bypasses the damaged / absent cochlear nerves and directly stimulates the cochlear nucleus in the brainstem(7).

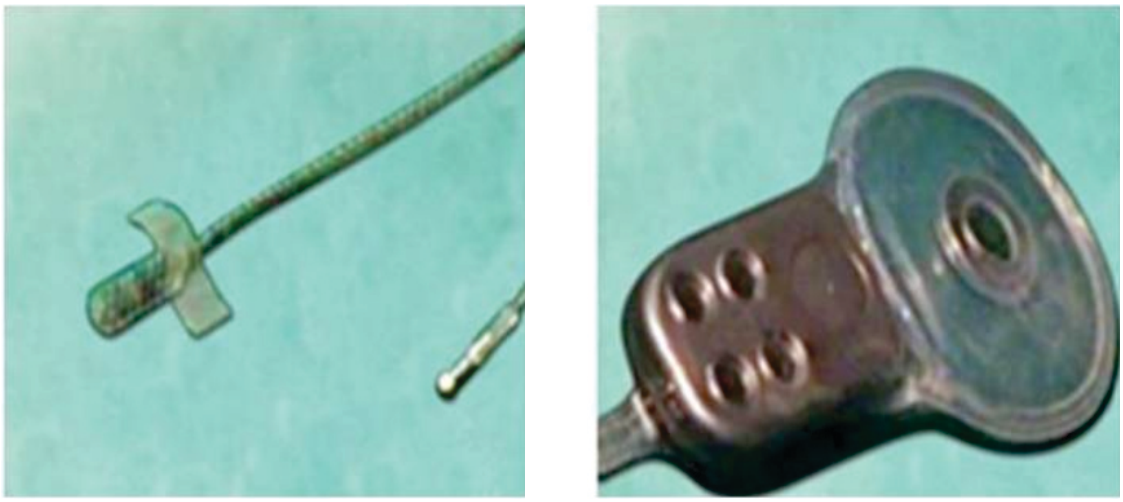

Fig. 6: Two ends of the auditory brainstem implant

Indications For Auditory Brainstem Implantation

Multichannel ABI are USFDA approved for adult patients with NF2 tumors involving both vestibule-cochlear nerves. The implant is usually placed in the lateral recess of the fourth ventricle after tumor resection(8). (Fig.6)

The indications for ABI have expanded onto non-tumoral (NT) cases, such as congenital bilateral cochlear nerve aplasia(9). In such cases, the pediatric ABI helps bypass the non-functioning hypoplastic or absent cochlear nerves and stimulates the cochlear nucleus directly, thereby restoring auditory sensation in children. Other indications for ABI include bilateral totally ossified cochleae in which a CI cannot be used, bilateral auditory neuropathy, bilateral temporal bone fractures and demyelinating diseases affecting the eight cranial nerves, but sparing at least one cochlear nucleus. Contraindications to ABI include previous stereotactic radiotherapy which has the risk of radiation necrosis of the cochlear nucleus region, and anatomic distortion and fibrosis. ABI may not be possible in very large tumors which cause distortion of the brainstem.

A multidisciplinary collaboration between neurotologist, neurosurgeon, implant audiologist and neuro-anesthetist is required in order to perform this intricate and sophisticated surgery. Most patients with the implant have good auditory awareness with appreciation of environmental sounds, but obtain more modest benefit with regard to speech perception. Majority of ABI patients use the implant, in order to facilitate lip reading while some can, in varying degrees, comprehend speech directly. It has been demonstrated that the ABI with surface electrodes may provide sufficient stimulation of the central auditory system in adults for open-set speech recognition. These favorable results motivated the clinicians to extend ABI indications onto

children with profound hearing loss who are not candidates for CI.

The incidence of cochlear nerve aplasia in the overall population world over is very low, estimated at one in every 100,000 newly born babies wherein ABI is indicated. Auditory brainstem implantation appears to be more effective in non-tumor diseases of the auditory nerve or cochlea than in patients with NF2 tumors.

Clinical Assessment for Auditory Brainstem Implantation

A meticulous work up – audiology & electrophysiology and high resolution radio-imaging with computed tomography (CT)/magnetic resonance imaging (MRI) of the brain and inner ear is mandatory.

In children, a detailed genetic study, neurological and psychomotor assessment is necessary apart from the routine work up as done for cochlear implantation.

Operative Procedure for Auditory Brainstem Implantation

For successful ABI surgery, a few important issues such as patient selection, choice of device, choice of approach, technique of tumor removal, knowledge of microanatomical variations, intraoperative identification of the cochlear nucleus and prevention of complications have to be considered. The procedure is done under intensive neuroanesthesia with intraoperative cranial nerve monitoring. Translabyrinthine approach or lateral suboccipital approach is used. Craniotomy exposes the transverse sinus superiorly and sigmoid sinus laterally. Dura needs to be opened by a vertical incision 1 cm away from the sigmoid sinus, and is reflected laterally. Cerebrospinal fluid (CSF) is let out from the basal cisterns to make the cerebellum lax. Cerebellum is retracted medially to reach the cerebellopontine (CP) angle where the VII and VIII nerve complex is identified. Inferiorly, the lower cranial nerves are seen and followed medially onto the foramen of Luschka where the choroids plexus is identified. Further dissection is done to reach the floor of the IV ventricle, where a constant vein called the straight vein is present, which leads to the site of the cochlear nucleus. In tumoral cases as in NF2, tumor excision via the same approach precedes the implantation. After tumor excision, the landmarks (VII, VIII and IX cranial nerves, choroid plexus) for the foramen of Luschka are identified. Location of the lateral recess can be confirmed by noting the egress of CSF during valsalva maneuver. The ABI electrode array is then inserted into the lateral recess and positioned once the cochlear nucleus is well delineated. Initially, temporary electrodes are placed on it and electrically evoked auditory brainstem responses (EABR), early mid-latency responses (EMLR), and device telemetry (DT) are performed to check the optimal positioning and functioning of the electrodes. Once integrity is confirmed, the permanent electrodes are then placed

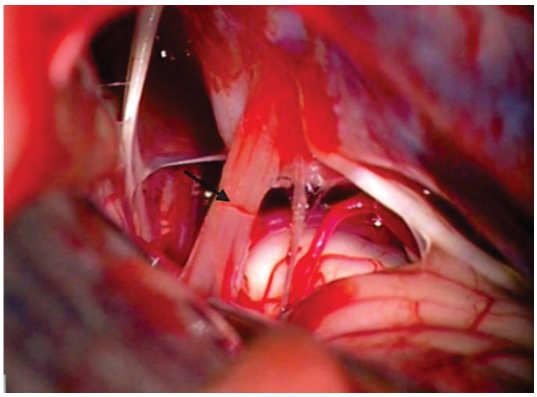

Fig. 7: Hypoplastic VIII N

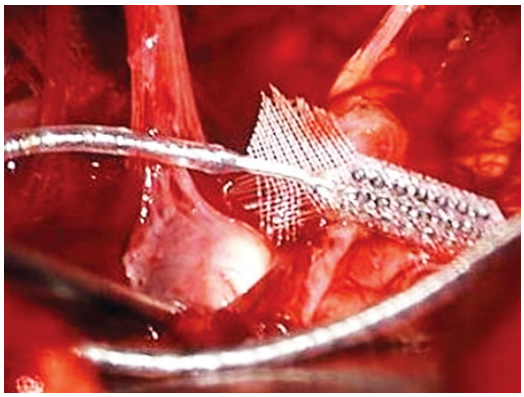

Fig. 8: Insertion of electrodes in brainstem

onto the cochlear nucleus and positioned with fibrin glue and surgicel. (Fig. 7 & 8)

The receiver-stimulator coil is placed in a bed created in the area posterosuperior to the craniotomy. It is placed at least 10 mm behind the edge of the auricle and above the canthomeatal line, and is angled 30–45° posterosuperiorly. Tie-down holes are made on either side of the receiver-stimulator for securing the implant. Reconfirmation of implant function is done with electrophysiological tests. The stimulus to ABI is delivered by an external component comprising of a microphone, a signal processor and a transmitter coil very similar to the CI. Dura is closed primarily in a water-tight fashion. Postoperative neurointensive care is necessary with cranial nerve monitoring.

Habilitation After Auditory Brainstem Implantation

The device is switched on 2 months after implantation, providing sufficient time for wound healing and full recovery. Switch on needs to be done in the operation theater with neuro monitoring and adequate preparation for active CPR, as there may be inadvertent stimulation of other brainstem nuclei and the possibility of non-auditory stimulation of vital centers. The stimulus threshold and comfort level on each electrode is ascertained. Postoperative CT scan and X-ray of skull confirm the position of the ABI in situ. (Fig. 9) Habilitation of ABI patients requires intense dedication and skill on the part of the auditory

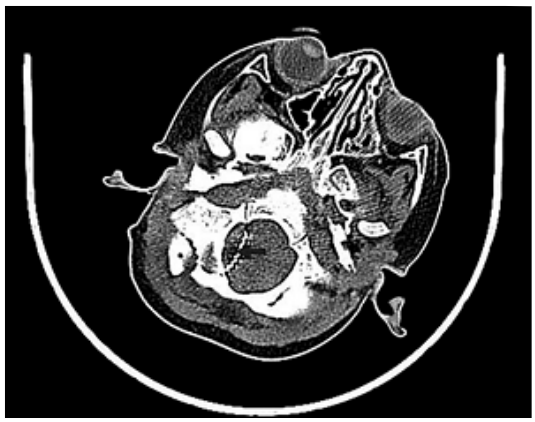

Fig. 9: CT scan image showing brainstem implant in situ

habilitationist and audiologist, along with adequate motivation and family support from the patient's side.

Outcomes of Auditory Brainstem Implantation

Intensive auditory verbal habilitation is then initiated and continues for a minimum period of 1 year (as advocated for CI) with periodical EABR and EMLR - tests done during the follow-up for confirmation of device integrity and assessment of optimal performance of the implantee. Most implantees develop very good sound awareness and good gross auditory discrimination with appropriate habilitation. Achievement of lucid environmental sound perception with pitch discrimination for closed set speech is often the culmination of the habilitation process.

MERF Experience

Of the seven candidates who underwent ABI, one patient was post-lingual and had neurofibromatosis type II and six were pre-lingual with bilateral cochlear and cochlear nerve aplasia. The outcome assessment is by habilitation scores (CAP, SIR, MUSS, MAIS), electrically evoked auditory brainstem response and cortical auditory evoked potentials.

Auditory Midbrain Implant

The auditory midbrain implant (AMI) has been designed for stimulation of the auditory midbrain, particularly the central nucleus of inferior colliculus (ICC).

Cochlear implantation (CI) is ineffective for those without an implantable cochlea or an absent, thin non-functional or tumor of the vestibulo-cochlear nerve. These patients can be implanted with the auditory brainstem implant (ABI), which directly stimulates the surface of the cochlear nucleus. Unfortunately, ABI has achieved limited success in providing environmental awareness and auditory sensations, especially in patients with large neurofibromatosis type 2 (NF2) wherein tumor induced damage to the brain-stem/cochlear nucleus often co-exists. The midbrain is a target, is more surgically accessible than the cochlear nucleus in the lateral recess and hence AMI has today emerged as a valuable alternative to the ABI. The central nucleus of ICC is tonotopically well organized like the cochlea and accesses all ascending auditory impulses from the peripheral pathways(10). AMI offers advantage over the ABI in that it can be surgically implanted under direct visual exposure of the target ICC region, with less risk of damaging critical brainstem structures and cranial nerves. Stimulation of the ICC site helps in enhancing the lip-reading capabilities and also provides environmental awareness with improvement in speech perception performance.

Conclusion

Advances in biomedical engineering have led to the development of auditory neural prostheses such as cochlear implants and auditory brainstem implants which have helped habilitate patients with severe – profound hearing loss. Their efficacy and safety are well proven. Advancements in technology will enhance the benefit provided by these prostheses.

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