**History of Cochlear Implants**

Volta, in the year 1790, became the first person to experience and publish the effects of electrical current on the auditory system. He inserted a metal rod in each ear and then subjected himself to approximately 50 volts of electricity. He reported that the sensation was that of receiving a blow to the head followed by the sound of thick soup boiling. Volta was followed by a string of scientists who continued to experiment with electricity and hearing over the next 167 years. It was Djourno and Eyries who reported the first stimulation of the acoustic nerve by direct application of an electrode in a deaf person (1957). The patient was undergoing an operation for cholesteatoma and the auditory nerve was exposed. An electrode was placed on the nerve and an induction coil and ground electrode were placed in the temporalis muscle. The coil could then be stimulated by currents produced by a second coil placed against the overlying skin. On subsequent experimentation with the patient reported hearing sounds like crickets or a roulette wheel when the second coil was applied. He was able to distinguish simple words and noted improvement of his speech reading ability. This initial implant was followed by a string of implantations performed by House, Doyle, Simmons, and others. Advances in microelectronics, biocompatible materials, and microscopic otologic surgery propelled House to produce the first single-channel implant in 1972 which stimulated the auditory nerve via the scala tympani. In 1984 the cochlear implant gained FDA approval for use in adults. This corresponded with the introduction of multichannel implants which significantly improved spectral perception and open-set speech understanding. The 1990’s saw significant improvements in speech processor designs. SPEAK and CIS speech processing strategies produced large improvements in recipient’s speech recognition. The late 1990’s and early 2000’s saw technologic improvements such as the peri-modiolar contour electrode, split electrodes, behind-the-ear processors, and implantation for children as young as 12 months.

**Components of a Cochlear Implant**

A cochlear implant consists of several components. All implants have microphones, external speech processors, signal-transfer hardware, transmitters, receivers, and electrodes.
Each plays an important part in converting sound to an electrical stimulus. The microphone simply 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 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 this stimulus to the central nervous system which decodes and interprets 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. These factors likely are important in understanding significant differences in patient outcomes despite similar preoperative auditory deficits, surgical course, and cochlear implant 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 radio-frequency 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 vs. 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, multi-channel devices provide different information to several electrodes or groups of electrodes. Early implants had only one electrode (and one channel); recent advances have lead to the development of implants with multiple electrodes (22) and multiple channels (usually 4-8). 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.

Speech Processing Strategies

There are many different ways of processing the auditory signal for presentation at the level of the cochlear ganglia. The most commonly employed are the spectral peak (SPEAK), continuous interleaved sampling (CIS), and compressed analog stimulation. The SPEAK strategy is characterized by filtering sound into 20 different bands covering the range of 200 Hz to 10,000 Hz. Each filter corresponds to an electrode on the array. The outputs for each filter are analyzed and those channels of highest amplitude that contain speech frequencies are stimulated. The stimulus rate is equal to the period of the lowest frequency of speech (F0). The dominant speech frequency between 280 and 1000 Hz (F1) is then identified and the appropriate apical electrode is stimulated. The dominant speech frequency between 800 and 4000 Hz (F2) is then identified and the appropriate basal electrode is stimulated. Three additional high frequency filters measure input in the 2000-2800 Hz, 2800-4000 Hz, and >4000 Hz ranges. Stimulus is sent to apical electrodes (in order to take advantage of the greater incidence of ganglion cell survival at the apex of the cochlea). These channels provide additional cues for consonant
perception and environmental sounds. Electrodes are stimulated sequentially, and at amplitudes specific for each frequency peak.

The continuous interleaved sampled (CIS) strategy is employed by the Clarion and MED-EL systems. This system works by filtering the speech into eight bands. The bands with the highest amplitude within the speech frequencies are subsequently compressed and their corresponding electrodes are stimulated. The CIS strategy uses high-rate pulsatile stimuli to capture the fine temporal details of speech.

The advanced combined encoder (ACE) strategy filters speech into a set number of channels and then selects the highest envelope signals for each cycle of stimulation. Stimulation is carried out in a very rapid fashion (much faster than the SPEAK strategy which stimulates at the rate of the lowest frequency of speech—180-300 cycles/second).

The simultaneous analog strategy (SAS) closely mimics the normal ear. All incoming sound is compressed and filtered into eight channels. These channels are then simultaneously and continuously presented to the appropriate tonotopic electrode. There is no effort to select for speech frequencies. Intensity is coded by either stimulus amplitude, rate or both.

The SAS strategy has met with limited success, whereas the SPEAK and CIS strategies have been relatively successful. It appears that no one system is effective for all recipients. For this reason, recent advances have made it possible for one cochlear implant to offer several speech processing strategies in the same implant. This allows the audiologist and patient to choose what strategy is best for that individual. Currently, the Nucleus systems are made to employ several processing strategies. These include spectral peak (SPEAK), advanced combined encoder (ACE), and continuous interleaved sampling (CIS). The Clarion systems use CIS to stimulate in a monopolar fashion as well as simultaneous analog stimulation (SAS). Medical Electronic (Med-El) produces a product (currently in USA clinical trials) with 12 electrode pairs suitable for deep insertion that relies on the CIS strategy with the most rapid stimulation rate of all implants. Recent advances in technologies have included the development of curved electrode arrays which are intended to more closely approximate the modiolas. Studies seem to indicate that electrodes closer to the basilar membrane need less current to stimulate the nerve and may improve spatial specificity of stimulation.

**Indications for Cochlear Implantation**

Cochlear implants are FDA approved for adults 18 years and older (no upper age limit) and children age 12 months to 17 years 11 months. Initially implants were only approved for adults who were postlingually deaf and had no improvement with high-powered hearing aids. This group of people has consistently been shown to be benefited by implantation. As more was learned about the benefits of cochlear implantation, the criteria were relaxed. Now, adult criteria include bilateral severe-to-profound sensorineural hearing loss with 70 dB pure tone average, little or no benefit from hearing aids (must attempt binaural high-powered hearing aids for at least 6 months), and psychological suitability. Audiologic examination should show word discrimination scores less than 40% in the best aided condition. The patient should have no
anatomical deformity that would preclude implantation success. Finally, the patient should have no physical condition that would preclude a general anesthetic.

Pediatric implantation is indicated in children 12 months or older with bilateral severe-to-profound sensorineural hearing loss with pure tone averages of 90 dB or greater in the better ear. The child must have had no appreciable benefit with hearing aids (evaluated with parental survey when younger than 5 and 30% or less on sentence recognition tests under best-aided conditions when 5 years old or older). Children must tolerate wearing hearing aids for a period (as all cochlear implants have external components), and show some aided communication ability. Children must be enrolled in educational programs that support aural/oral learning and have no medical contraindications. Parents must be highly motivated and have reasonable expectations.

**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. Computed tomography findings may also preclude implantation. The absence of the cochlea (Michel deformity), and a small internal auditory canal (associated with cochlear nerve atresia) 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 result, CSF leak, and meningitis.

The presence of active middle ear disease is a contraindication to surgery. This process should be treated and resolved before implantation. In a study by Luntz otitis-prone patients were treated by protocol (antibiotics, PETs, etc.) before surgery and then implanted (often with PETs in place). No delay was necessary when compared with patients who were not otitis-prone. Several were noted to have inflamed middle ear mucosa on implantation which required removal in order to identify the round window, but did very well with few postoperative episodes of otitis. Children with a history of chronic suppurative otitis media were implanted in a study by El-Kashlan without demonstrable early or late complications. Patients with a history of canal wall down mastoidectomy may need surgery to reconstruct the posterior canal wall or close off the canal before implantation.

Meningitis may lead to hearing loss and ossification of the cochlea. Labyrinthitis ossificans is usually identifiable on CT scan (brightly lit cochlea with obliteration of the basal cochlear duct) and is a relative contraindication when there is a patent contralateral basal turn. MRI is often better at delineating patency of the cochlea and should be pursued if there is any question. Very young children with hearing loss after meningitis should be followed with CT/MRI until they reach implantable age. Early implantation may be indicated if evidence of ossification is noted. Adults and children with acute meningitis should be treated with steroids to avoid hearing loss. Those that do sustain hearing loss secondary to meningitis should be observed for 6 months before implantation due to the substantial number of patients that will regain their hearing in at least one ear. 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.

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.

Work-up for a Patient Seeking Cochlear Implantation

- Audiologic examination with binaural amplification
- CT scan/MRI of temporal bones
- Trial of high-powered hearing aids
- Psychological evaluation
- Medical evaluation
- Any workup necessary to discover etiology of hearing loss

Surgical Procedure

The surgical procedure to implant the receiver and electrode array is most often a day-surgery with the patient being discharged shortly after completion of the implantation. Implanting the better or worse-hearing ear is a decision reached by the physician and the patient. The patient should understand the risk of losing all residual hearing in the implanted ear. One recent study by Chen showed no long-term advantage to implanting the patient’s better ear. Thus, many surgeons opt to implant the worse ear and have the patient continue to wear an aid in the best hearing ear.

The surgical procedure begins after the patient receives a general anesthesia. The patient’s head is shaved over the post-auricular area. The extent of hair removal depends on the incision to be used—generally four fingerbreadths above and behind the ear is sufficient. The patient is then prepped and draped in a fashion similar to other otologic procedures. A dummy receiver is placed over the skin and positioned approximately 1 cm posterior to the auricle. A postauricular incision is made 1-2 cm posterior to the implant. Several incisions have been proposed and include a large C-shaped incision, a 4-5 cm superior elliptical extension of the routine postauricular incision, a small 4-5 cm straight incision posterior and an incision posterior-superior to the auricle (minimally invasive procedure introduced by the Cochlear Company). The skin flap is elevated followed by the creation of an anteriorly-based temporoparietal fascia flap. The temporalis musculature and overlying fascia are left intact. A subperiosteal pocket is created medial to the temporalis muscle for placement of the ground electrode. A circular depression is then drilled in the temporal bone cortex superior and posterior to the area to be drilled for access to the round window. Tunnels are often drilled into the surrounding bone in order to place anchor sutures over the receiver.

A complete mastoidectomy is performed with minimal saucerization. A shelf of mastoid cortex can be helpful when securing the array and tucking the excess grounding wire. The facial recess is opened, taking care to avoid injury to the chorda tympani and facial nerves. The round
window niche is inspected. A cochleostomy is then drilled over the basal turn of the cochlea just anterior/inferior to the round window. This is carried down to endosteum of the cochlea. The endosteum is then opened using a straight pick. The electrode array is then carefully inserted through the fenestra into the scala tympani. An inserting claw or jeweler’s forceps may be used to advance the electrode array. Excess force should not be used, as the array can easily buckle and cause damage to the internal components. A deep insertion is desired in order to place the electrodes closer to the apex where the highest concentrations of surviving ganglion often are found. A small amount of connective tissue is then packed around the electrode array at the cochleostomy site in order to seal the opening. Care is taken to avoid accidental removal of the array once placed. The ground electrode is tucked into the sub-periosteal pocket and the wound is closed in several layers. No drains are placed. A bulky mastoid dressing is applied. The wound is given several weeks to heal before use of the external processor is attempted. The external processor is held in place by magnetic attraction to the magnet in the implanted receiver.

In patients who have a history of meningitis leading to hearing loss, labyrinthitis ossificans may have caused obliteration of the scala tympani. In this case, the array can be placed into the scala vestibuli. Often the ossification is incomplete and if the surgeon drills forward along the basal coil for 4-5mm the scala tympani will be identified. Care must be taken to avoid injury to the carotid artery which lies just anterior to the cochlea. In some cases of cochlear dysplasia CSF gushers have been encountered. This is managed by allowing the pressured fluid to drain off, and then proceeding with insertion as per routine.

**Postoperative Management**

The surgical complication rate after cochlear implantation is estimated to be only 5%. The most common problems are wound infection and wound breakdown. Rarely, extrusion of the device, facial nerve injury, bleeding, CSF leaks and meningitis can occur. Device-related complications include intracochlear damage, slippage of the array, breakage of the implant, and improper or inadequate insertion. Postoperative infection of the surgical site was treated by prolonged courses of postoperative antibiotics by Yu, et al with excellent results. They suggest that a long course of antibiotics and limited I&D will treat the vast majority of wound infections without the need to remove the implant. Those patients that did not respond to this treatment protocol were often found to be immunocompromised. Steenerson reported a 75% incidence of postoperative vertigo, but indicated that these patients did well after undergoing vestibular therapy. Other series do not show as high an incidence, nor the need for vestibular rehabilitation postoperatively. Stimulation of the facial or vestibular nerve by stray electrical current from electrodes outside or near the round window has also been reported. This is usually addressed by “turning off” the responsible electrodes and moving the electrical stimulation to electrodes located within the cochlea.

Recent reports of increased incidence of meningitis in cochlear implant recipients have prompted the CDC to recommend vaccination of implanted or soon to be implanted patients. Children less than 2 years old who have implants should receive pneumococcal conjugate vaccine (Prevnar). Children with implants 2 years and older who have completed the conjugate series should receive one dose of the pneumococcal polysaccharide vaccine (Pneumovax 23 or Pnu-Imune 23). Children with implants between 24 and 59 months who have never received
vaccination should receive two doses of pneumococcal conjugate vaccine two months apart and then one dose of pneumococcal polysaccharide vaccine at least two months later. Finally, persons age 5 years and older with cochlear implants should receive one dose of pneumococcal polysaccharide vaccine.

Although the incidence of device failure is very low, occasionally removal of the implant and reimplantation is necessary. These patients do surprisingly well. Alexiades, et al. showed that patients did as well or better after reimplantation (in the same ear) as with their first implant. Thus a history of implantation is not a contraindication to another cochlear implant. Long-term electrical stimulation from a cochlear implant has raised concern for damage to the auditory nerve. However, cochlear implants typically discharge less than one microcoulombs per cm2 of electrode surface and long-term studies have shown no detrimental effects. In fact, studies following patients for up to 13 years show no decline in function. This finding is still true when the study population includes those that have been implanted multiple times.

Research looking at cochlear implants under many environmental strains has shown them to be reliable and safe. Backous, et al showed them to be stable when exposed to extreme barometric pressure changes (as experienced when scuba diving). MRI exposure should be avoided generally, but may be pursued when necessary.

**Postoperative Rehabilitation**

Unless intensive postoperative rehabilitation is undertaken, cochlear implantation is likely to provide little benefit. Each patient’s need for rehabilitation is different based on pre-operative auditory experience. For the prelingually deaf patient, auditory and speech training are imperative if they are to improve their communication abilities. Postlingually deaf patients often need training in more complex listening skills. Cochlear implants in children are successful when the implantation is followed by a intensive treatment by a multidiscipline rehabilitation team. The goal of a pediatric rehabilitation team is to enable the hearing-impaired child to be able to learn passively from his environment. The rehabilitation must address both receptive language skills as well as expressive language abilities. A structured program with dedicated team members is integral to a successful cochlear implant program.

**Results of Cochlear Implantation**

Cochlear implantation is really the only effective way of treating patients with profound sensorineural hearing loss who do not benefit from hearing aids. Although the perception of a successful implantation might vary from patient to patient, the primary goal of implantation has always been improved speech perception. Since implantation began, physicians have noted a wide range of outcomes. Some patients find little benefit after implantation and may even find the stimulation annoying. Others are able to function normally even without visual cues. Still others are able to listen to and enjoy music. Years of research has given us a better understanding of what variables might influence the results of implantation.

The age of onset of deafness, as well as the length of time since the onset of hearing loss has both been shown to influence outcomes. Several studies have shown that patients who were
prelingually deafened show the poorest outcome. Prelingually deaf children implanted before age 6 appear to be able to “catch up” to implanted postlingually deaf children within 2-5 years. These children, like their postlingual counterparts, are able to achieve open-set speech discrimination. Several studies have shown that implantation at an earlier age results in earlier achievement of open-set speech discrimination. Govaerts, et al. showed that 90% of those implanted before age 2 were integrated into mainstream education whereas only 20-30% of those implanted after age 4 were ever integrated. These results are seen in children who are enrolled in aural/oral educational programs and who use oral language as their primary communication modality. The performance of implanted children is far better than those with equal hearing deficits who rely on vibrotactile devices or hearing aids. Generally, implantation of prelingually deaf adolescents and adults is significantly less successful, though results vary widely.

Most authors now believe that the shorter the period of auditory deprivation, the faster and more complete will be the achievement of open-set speech discrimination. This has been shown to be true in the adult population, as well with children. Those patients who are implanted within a short time seem to retain the plasticity of the auditory system better than those who have been deaf for a period of years. Sharma, et al. compared children implanted after different periods of deafness. He showed that children with the shortest amount of time spent without auditory stimuli regained normal cortical responses more rapidly than all others. Specifically, those with 3.5 years of deafness or less showed age-appropriate P1 latencies (a marker of plasticity) after only 6 months of stimulation with a cochlear implant. The length of time required to reach age-appropriate latencies increased with increasing length of auditory deprivation. After age 7 plasticity was greatly reduced.

Waltzman, et al. studied the long-term effects of cochlear implants in children. They followed the children after implantation for five to fifteen years and documented speech perception scores, device extrusion rates, and implant viability. He showed that implantation resulted in significant improvement of patient’s speech perception and that this benefit remained stable (often improving) over the long-term. For the vast majority of his study group this resulted in assimilation into mainstream education. There was no significant incidence of device extrusion or migration and even when device failure necessitated reimplantation, long-term performance was not decreased.

Recent studies looking at the economics of cochlear implants show cochlear implantation improves patient’s quality of life and is cost-effective even in elderly patients (>50 years old). Implantation results in significant benefit to the society as a whole, and to the individual. Unfortunately, cochlear implantation is more often a money-losing effort for everyone involved with implantation. Hospitals and physicians, as well as the other members of the rehabilitation team often find themselves without funding and support.

**Future Directions**

Partial insertion cochlear implantation has been proposed as treatment for those patients who have residual low-frequency hearing with high-frequency sensorineural hearing loss. The speech processor is coupled with a hearing aid and thus provides maximal aided hearing. The risk of such surgery is loss of the remaining hearing in that ear. Other implant strategies include
brainstem implantation for those without an intact cochlear nerve. Good results have been reported.

Nucleus products now come equipped for intraoperative testing. This allows the audiologist to map the patient’s electrode array while the patient is asleep. This is especially useful for infants and small children who are often not cooperative with conventional mapping techniques. Intraoperative repositioning is also a possibility if the mapping shows poor responses.

Bilateral implantation with the possibility of binaural hearing is currently being studied. Gantz showed that most patients with bilateral implants perform better on sound localization, but only some do better with auditory performance in noise (at one year). Despite this, most studies report increased satisfaction with two implants. At least one company is proposing a system with only one processor and receiver but implanted electrodes in both ears. Despite these results, the cost of a second implant is prohibitive. Summerfield argued that the quality of life likely to be gained (across society as a whole) by unilateral implantation is higher, per unit of expenditure, than with bilateral implants.

Implantation for patients with asymmetric sensorineural hearing loss may soon be approved. Cochlear implants are expected to help these patients with sound localization and speech comprehension in noise.

The Cochlear Company is currently testing a new “Softip” electrode array which is advanced off a stylet after traversing the straight section of the basal turn. The array then curls around the modiolus until fully inserted. The technique has been shown to cause less trauma to the basilar membrane and intracochlear structures in preliminary studies. The Cochlear Co. is also marketing its new “minimally invasive” approach which allows for implantation through a small (4-5cm) incision over the post auricular area which does not require shaving of the hair in that area. Warm response is reported by those performing this approach.

Conclusion

Cochlear implantation is no longer experimental. It is the treatment of choice for children and adults with severe-to-profound hearing loss. Significant gains in open-set speech recognition have been demonstrated by most of those who undergo implantation. Early implantation, whether in pre or postlingual patients, has shown to be effective at moving an otherwise marginalized segment of society into the mainstream. Implantation is cost-effective and results in high patient satisfaction. Although cochlear implants are still a rough and awkward imitation of our natural sense, they offer hope to thousands who must otherwise live in a silent world.
Bibliography


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