Current State-of-the-Art in Cochlear Implantation

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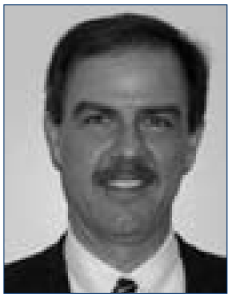

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Introduction. Cochlear implantation has been described as a miracle of modern medicine. In fact, it represents the first time in history that medicine has been able to restore one of the senses. Combining a surgically placed internal implant and an externally worn speech processor, this technology provides not only sound awareness, but now for the majority of recipients, truly useful hearing for patients who suffer from significant hearing loss. Cochlear implants have undergone tremendous change and acceptance over the past 25 years, resulting in significant benefit to thousands of patients. Advances have been achieved not only in device technology, but also in patient selection criteria and rehabilitation methods.

Historical perspective. The idea of stimulating the auditory nerve to provide sound detection has been credited to Benjamin Franklin in 1751. In 1790, a Frenchman named Volta placed an electrode in each of his ear canals and passed current between them. After awakening, he remembered hearing sound just before losing consciousness. It wasn't until 1957 that direct stimulation of the auditory nerve was performed. Djourno and Eyries were performing ear surgery on an awake patient with the auditory nerve exposed, and while stimulating the nerve, the patient reported hearing sound[1].

In 1961, William House, from the House Ear Institute in Los Angeles, performed the first single channel cochlear implant by placing a wire inside the cochlea in a deaf patient, thus giving that patient the ability to detect sound[2]. In 1978, Graeme Clark of the University of Melbourne performed the first multichannel cochlear implant, thus launching the research projects that would lead to FDA approval for adults of the first multichannel cochlear implant in the United States in 1985. In 1990, the Nucleus 22-channel cochlear implant was approved for children.

Since that time, multiple devices by 3 different manufacturers have been approved and new devices representing continued advances in this field are introduced almost every year. From first prototype to wide spread use, the cochlear implant represents an amazingly rapid technological advance.

How the device works. A cochlear implant is an electronic device that provides function to the damaged or absent hair cells of the inner ear by providing electrical stimulation to the remaining nerve fibers. All cochlear implants have 4 common features: a microphone to pick up sound; a micro-electronic processor to convert sound into electrical signals; a transmission system to deliver the signals to the implanted device; and a small electrode array that is surgically introduced into the cochlea. When the device detects a sound, it is converted into an electrical signal, which is transmitted across the skin to the internal device. The electrical signal is then presented to a specific tonotopic location in the cochlea, which corresponds to the pitch of the sound. The auditory nerve is stimulated and this is perceived as sound by the hearing center in the brain.

Patient evaluation and candidacy.

Prospective candidates for this procedure undergo extensive evaluation to insure that they meet candidacy criteria and to insure appropriate counseling regarding their potential benefit. Every patient evaluation includes medical and audiological testing, and in some patients, psychological, cognitive, and sociological evaluations are performed. In addition, children are evaluated for their rehabilitation potential and their educational system is reviewed to insure maximum benefit from this technology.

Medical history includes assessment of general health, including life expectancy and ability to undergo general anesthesia. The age of onset of hearing loss, etiology of hearing loss, use of hearing aids and oral communication, and duration of deafness are important factors in trying to predict a patient's potential for benefit. Acute and chronic otitis media must be diagnosed and managed prior to implantation and any prior ear or posterior fossae surgery must be investigated preoperatively. History of dizziness, prior meningitis, head trauma, radiation therapy and any illnesses that could affect central auditory processing are important. These factors may not exclude cochlear implantation but are part of each individual evaluation and affect patient counseling about their potential benefit.

A thorough neurotologic examination is performed, certain laboratory studies may be helpful in select patients, and all potential candidates undergo radiological imaging prior to surgery. CT scanning remains the gold standard and is done to insure normal cochlear anatomy and patency, and to assist with surgical planning and to reduce surgical risk. In some cases, MRI is also helpful as it shows the cochlear fluid signal. This signal may be lost in cases of osteoneogenesis, sclerosing labyrinthitis, otosclerosis and temporal bone fracture. MRI can also be helpful to rule out vestibular schwannoma, in cases of inner ear malformation, and in the evaluation of patients with suspected central auditory processing issues.

Table 1. Patient Selection and Evaluation

CHILDREN	ADULTS
• Severe to profound sensorineural hearing loss in both ears	• Severe to profound sensorineural hearing loss
• Age 12 months or older	• Receive little or no benefit from hearing aids
• Receive little or no benefit from hearing aids	• Hearing loss acquired before or after learning oral speech and language
• No medical contraindications to surgery	• No medical contraindications
• Congenital or post-lingual onset of hearing loss	• A desire to be part of the hearing world
• Appropriate motivation, expectations and educational environment	

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In some patients, genetic testing is helpful. Genetic factors account for at least half of all cases of profound congenital deafness. More than 50 deafness genes have been identified and sequenced, and it is likely that hundreds more await discovery. Most cases of congenital hearing loss are due to single gene defects with no traceable family history. Genetic testing has many potential advantages. Testing can help to determine the etiology of the hearing loss when the cause is unknown. It may help to predict the progression of hearing loss and may be important in family planning issues. *(Additional information available at http://webhost.ua.ac.be/hhh.)*

Genetic testing, however, can be wrong, and having the gene doesn't always imply it is the cause of the hearing loss, and doesn't mean that every future child will be affected. Any genetic test should be combined with genetic counseling to fully understand the meaning of the results. One of the most recently discovered gene defects for hearing loss is the Connexin 26 mutation[3]. This genetic defect is thought to be responsible for 50 percent of autosomal recessive, nonsyndromic hearing loss worldwide and 20 percent of childhood deafness overall. In the United States, the carrier rate for all deafness-causing Connexin 26 mutations is 3 percent. Over 60 different variations of this gene mutation can cause hearing loss, ranging from mild to profound. Patients with this genetic mutation in general do very well with cochlear implantation.

Audiological evaluation includes pure tone testing and speech discrimination in both aided and unaided conditions. Age appropriate parameters are used and for adults, the HINT test is used. Unilateral information is helpful in deciding on the best ear for implantation. Current Medicare guidelines require that post-linguistically deafened adults must demonstrate test scores of 30 percent or less on sentence recognition tests from tape recorded tests in the patient's best listening condition.

There remain several absolute contraindications to cochlear implantation. Agenesis of the cochlea (Michel's aplasia) is a clear contraindication. In addition, agenesis or absence of a viable cochlear nerve as in cases of small IAC syndrome, temporal bone fracture, posterior fossa surgery, or radiation therapy would be contraindications. Active otitis media, central nervous system disease, and medical conditions that prevent surgery are additional contraindications.

The choice of the ear to implant is complex but there are several factors to consider in deciding which ear would be most appropriate. In general, after implantation, the residual hearing is lost and patients are no longer able to use a hearing aid for sound awareness, so the worst hearing ear is usually implanted. If the residual hearing is comparable in each ear, we would generally implant the ear with the acquired loss, shortest duration of deafness, or the ear with the longest use of hearing aid.

Predicting success. One goal of the preoperative patient evaluation is to attempt to predict the patient's success with the cochlear implant. This success is dependent on the status of the surviving auditory neurons, but unfortunately, there is no definitive way to predict the status of these neurons. It is critical to appropriately counsel patients prior to cochlear implantation regarding their potential benefit. The most critical factor to consider is the age at onset of deafness. Patients who lost their hearing after the development of spoken communication (post-lingual) do much better with the cochlear implant than those with pre-lingual onset, due the importance of auditory memory. The normal hearing child at birth has a blank auditory cortex. Repetitive sound and speech stimulation causes neural auditory pathways to develop. Without this auditory stimulation, neural pathways don't develop. Early access to the linguistic code is critical to this development; thus early identification of hearing loss and habilitation with hearing aids is of the utmost importance in these children.

Another important factor in the success of cochlear implantation is the age at the time of surgery. Implantation at a younger age is better due to the plasticity of the auditory system. We know that the ability of the brain to learn new languages decreases with age, concurrent with the decreased plasticity of the auditory neural pathways. Currently, the FDA has approved cochlear implantation down to the age of 12 months. Earlier implantation is possible, but care must be taken to insure appropriate diagnosis of hearing loss, and appropriate trial of hearing aids prior to cochlear implantation.

Duration of deafness is another factor to consider in predicting success. Clearly, shorter duration is better. Care must be taken, however, in patients who have suffered sudden hearing loss. Waiting long enough to be certain of the degree of hearing loss, awaiting possible recovery, adequate trial of hearing aids, and acceptance by the patient is very important.

One would think that the etiology of the hearing loss would help to predict the success with cochlear implantation. This is not as predictive, however, as the age of onset of hearing loss. In most etiologies, there are stimulable auditory neural elements present, but we see a variety of responses to the cochlear implant with the same etiology. It is critical to attempt to determine the etiology to insure that the cause of the hearing loss is not one of the contraindications discussed previously.

Other factors to consider in predicting success include the level of residual hearing (not as predictive as one might suspect), an educational system emphasizing an oral approach, associated handicaps, access to postoperative rehabilitation, and support of family and friends.

Surgical Procedure. The surgical procedure to place the cochlear implant meets the ideals for any surgical procedure. These ideals include do no harm (least traumatic as possible with low complication rate), simple is better (what can go wrong, will go wrong), and as time efficient as possible (to reduce cost and anesthetic risk). The current procedure for cochlear implantation is similar to a routine mastoidectomy. While it does require a general anesthesia, it is usually less than 2 hours and the majority of patients stay in the hospital less than 24 hours. There is minimal surgical risk and complication rates are very low (less than 1 percent). Potential intraoperative complications include facial nerve injury, CSF leak, hemorrhage, tympanic membrane perforation, and misplaced electrodes. Postoperative complications can include facial paresis, flap necrosis, device extrusion, excessive flap thickness, hematoma, pain, dizziness, facial nerve stimulation, infection, device failure, meningitis, and taste disturbance.

Postoperative meningitis. There is now evidence that patients who undergo cochlear implantation have a higher risk of developing meningitis after surgery than that of their age matched controls[4]. Meningitis is a bacterial infection of the lining of the brain and can be a serious, life threatening condition, which needs aggressive management. The most likely potential pathway for developing meningitis after cochlear implantation is a middle ear infection leading to bacteria tracking along the electrode array. This results in spread of infection into the inner ear, and then spread of infection to the brain space through cochlear or vestibular aqueducts. There are other possible pathways for spread as well, including hematogenous spread.

Initial concerns were raised at a European meeting in July 2002. Meningitis was reported in 26 patients after cochlear implantation, including some serious infections and 9 deaths. A complex review of data was then completed by all manufacturers and the Center for Disease Control in the U.S. This review confirmed 26 cases in the U.S. with the median time of meningitis occurring 2 months postoperatively. The longest case occurred 84 months post-operatively. In 30 percent of cases, the patient had an inner ear malformation, 65 percent had a history of recurrent acute otitis media and 11 percent had a prior history of meningitis. The risk for meningitis in the general population is 3.9 cases per 100,000. By comparison, after cochlear implantation the incidence is 243 cases per 100,000 representing a 30-fold increase[5].

Current recommendations for patients with Nucleus devices (Cochlear Americas) or Med-El devices are as follows. There appears to be no additional risk of meningitis after cochlear implantation in patients with normal cochlear anatomy. Patients with abnormal cochleas should be considered at risk for meningitis. These malformations include common cavities, enlarged vestibular aqueducts, and Mondini's deformity. In addition, patients with a history of CSF gusher, prior history of meningitis, and other central nervous system implants are considered at higher risk. Patients with the Clarion device (Advanced Bionics Corporation) that includes a 2 component system (positioner) should be considered at a higher risk for meningitis.

Acute otitis media should be treated aggressively in all implant patients, and patients and primary care providers need to be informed of this potential risk and need for aggressive management of ear infections. All patients who have had a cochlear implant, no matter what device, should receive immunizations to prevent meningitis against Hemophilis Influenza type B (HIB Vaccine) and Strep pneumonia (Prevnar or Pneumovax)[6]. Most younger children are already immunized but all patients should contact their primary care physician or implant center regarding specific recommendations. *(Additional information available at http://www.cdc.gov/mnwr/preview/mmwrhtml/mm5351-Immunizationa1.htm.*

Rehabilitation. Three to 4 weeks after device placement, the patient returns to the rehabilitation center to have the device programmed for thresholds and comfortable loudness levels. Recent advances in neural response recordings have made the programming somewhat easier, particularly in the very young child. Subsequent visits are required for modification of the map, counseling about device use, aural rehabilitation, and maintenance of the external portions of the device.

For children, the most important part of the cochlear implant process is the rehabilitation following device placement. If a child is not taught how to listen with the device, and incorporate the device into the educational environment, the maximum potential benefits will not be achieved. It is critical for this rehabilitation to proceed at regular intervals with a team of audiologists and therapists experienced in cochlear implantation.

Results of cochlear implantation. Several factors appear to significantly affect an individual patient's success with the cochlear implant. These include the auditory memory, the status of surviving neurons in the cochlea, the patient's motivation and commitment to be part of the hearing world, and the rehabilitation and educational programs. The device does not restore hearing without the intense process of "learning to hear again."

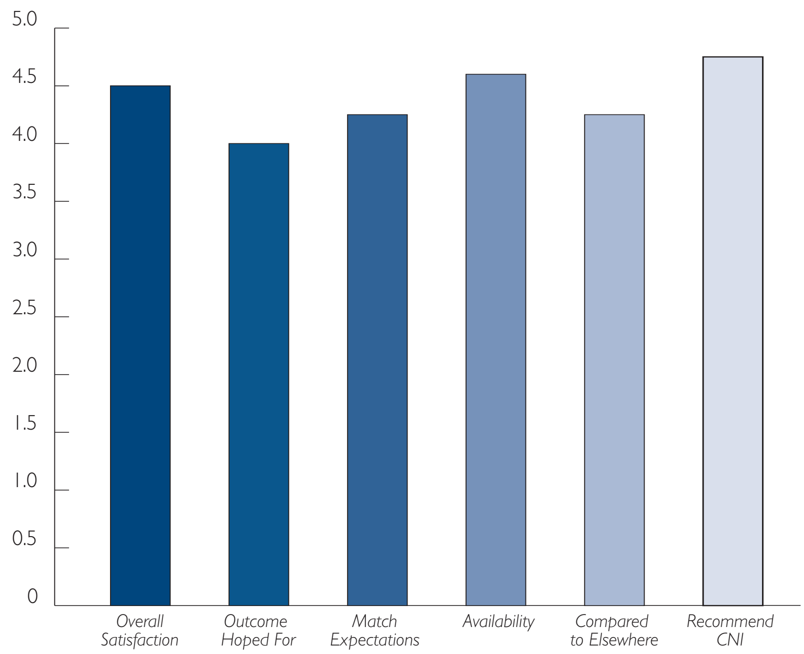
Cochlear implants do not restore "normal" hearing, and benefits vary from one individual to another. Most users find that cochlear implants help them communicate better through improved lip-reading, and over half are able to discriminate speech without the use of any visual cues. They report enjoying pleasurable sounds, feeling safer by hearing environmental sounds and feel their lives are simplified by hearing common sounds in their homes and work places. Very few patients do not receive some benefit from cochlear implants.

The majority of adult patients feel they communicate more effectively, many can enjoy music again, and approximately 80 percent of adult patients can communicate over the phone. In terms of overall satisfaction, most adults feel their quality of life has been significantly improved because of the cochlear implant. *(Table 2)*

After 6 months of use, the majority of children respond to their names and detect environmental sounds. Speech development is significantly improved in children after cochlear implantation, and they continue to show improvement for many years after device placement. When implanted at a young age, with appropriate habilitation, and without associated handicaps, most patients

Table 2.
Patient Satisfaction 2004
(Average Score)

0: Least Satisfied
5: 100% Satisfied



can expect to attend mainstream classrooms and have normal speech development.

Cost effectiveness of cochlear implantation. When looking at advanced medical technologies, the cost-effectiveness must be considered. The average cost of cochlear implantation in the Unites States is $65,000 for the first 2 years of care. While this seems a huge expense, studies of cochlear implant cost-effectiveness show it to be the second most cost-effective procedure of all advanced medical technologies. Only keeping premature infants alive in the neonatal nursery is more cost-effective [7].

Bilateral cochlear implantation. There are clear advantages to having binaural hearing. Studies of binaural hearing aid users show clear advantage in background noise and reverberant environments, and with sound localization with bilateral hearing aid fittings. Unilateral hearing aid users do not have these advantages. Factors to consider when assessing the incremental benefit of bilateral cochlear implantation include speech perception, sound localization, hearing in noise, quality of life and cost-effectiveness.

Studies on adult patients with 2 cochlear implants show that research subjects demonstrate significant improvements in speech understanding in noise. The benefit appears to be related to the head shadow effect, especially when adding the better ear. They also demonstrated significantly better speech understanding in quiet than did unilateral patients, possibly related to the fact that the better ear is always captured, in addition to the binaural benefit. All subjects improved localization ability as well. Subjectively, patients reported significant improvements in performance and in music appreciation with 2 cochlear implants. While there is a clear benefit to 2 cochlear implants, the cost-effectiveness of the second implant is not as dramatic as the first implant and many third party payers do not yet cover the cost of a bilateral cochlear implant.

Potential disadvantages of bilateral cochlear implants include the elimination of all residual hearing, possible detrimental effect on vestibular function, possible decreased benefit from future technologies, and the need for increased services to keep 2 devices functioning optimally. At this point, the decision to have a second cochlear implant is complex and should be individualized.

Future trends. The past 25 years of research in cochlear implants has been an amazing journey and if the next 25 years are the same, we have much to look forward to. Some of the most exciting research relates to the area of hearing recovery. Projects are underway to look at the potential for recovery of damaged hearing. These projects involve regeneration using a stem cell model, genetic alteration of endogenous cells, and repair of damaged neurons.

The *CNI Rocky Mountain Cochlear Implant Center* is currently involved in a research project to place a cochlear implant without losing residual hearing. The question is asked, what does it matter if residual hearing is lost? Clearly, patients are more comfortable with the idea of cochlear implantation if there is no loss of residual hearing. In addition, we suspect that cochlear implant performance may be better if residual hearing is preserved. This preservation would also not preclude the potential use of future technologies.

Hybrid cochlear implants are another exciting area of research in cochlear implantation. A significant number of patients have some useful low frequency hearing but no useful high frequency hearing. This puts them in a difficult area where they are not candidates for traditional cochlear implantation, but do not benefit significantly from traditional hearing aids. The hybrid cochlear implant involves a short electrode array, which is partially inserted into the cochlear in order to boost high frequency hearing without loss of low frequency hearing. This allows use of the cochlear implant to boost high frequencies and use of a traditional hearing aid to boost the low frequencies. Obviously, this involves implantation without loss of residual hearing, and the device could be customized to the level of residual hearing. Initial results look very promising but additional research is needed in this area.

Other trends in cochlear implants include continued advances in speech coding strategies and electrode design, totally implantable cochlear implants, more efficient and possibly remote programming capabilities, and use of coding strategies for music appreciation. The field of cochlear implantation has been one of dynamic change and this will no doubt continue.

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