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Cochlear implantation: a biomechanical prosthesis for hearing loss

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Abstract

Cochlear implants are a medical prosthesis used to treat sensorineural deafness, and one of the greatest advances in modern medicine. The following article is an overview of cochlear implant technology. The history of cochlear implantation and the development of modern implant technology will be discussed, as well as current surgical techniques. Research regarding expansion of candidacy, hearing preservation cochlear implantation, and implantation for unilateral deafness are described. Lastly, innovative technology is discussed, including the hybrid cochlear implant and the totally implantable cochlear implant.

Introduction

Cochlear implants are one of the most profound advances in modern medicine. As a therapeutic option for sensorineural hearing loss, they are fundamentally distinct from hearing aids, and work by converting sound into an electrical stimulus that bypasses the hair cells of the human cochlea and directly stimulates the cochlear nerve. As of December 2012, over 300,000 people worldwide have received cochlear implants, with approximately 60,000 adults and 40,000 children being implanted in the US [1].

As mentioned, cochlear implants are different from hearing aids in that they do not simply amplify sound. They bypass the ear's normal sound conducting mechanism, stimulating the acoustic nerve directly. An external microphone captures sound and relays it to a processor, which then sends it to a transmitter that communicates wirelessly with a receiver under the skin. The receiver then converts the sound into electrical impulses that are carried by an electrode array that is surgically placed adjacent to auditory nerve fibers within the cochlea. The signal is transmitted within the central auditory system, which generally remains intact in deaf patients, to the auditory cortex, allowing patients to hear. We present a summary review of cochlear implants, including their historical underpinnings, current indications, procedural description, including complications, as well as current topics of discussion within the field of cochlear implantation.

History of cochlear implantation

Electrical stimulation of the auditory system is not a new concept. In 1748, Benjamin Wilson used a Leyden jar to stimulate hearing in a deaf woman. Alessandro Volta, an Italian physicist, undertook similar experiments in the 1800s, but stopped because of the discomfort from the electrical stimulation [2]. In 1957, two French physician-scientists, Djourno and Eyries, implanted a device that directly stimulated the auditory nerve; the patient had sound awareness for several weeks until the device stopped working. Shortly thereafter, in 1961, Dr. William House of Los Angeles placed the first cochlear implant in a patient [3].

Initially, cochlear implants were single channel electrodes with minimal capabilities. Graeme Clark of Australia and Ingeborg and Erwin Hochmair of Austria pioneered the development of multichannel implants in the late 1970s [4,5]. Additionally, Michael Merzenich was influential in understanding the effects of electrical stimulation of the cochlear nerve and in developing multichannel implants at the University of California, San Francisco [6,7]. Using a multichannel electrode, stimulation at multiple locations

along the cochlea became possible, allowing for a more complex and realistic signal to be created. In the US, Blake Wilson further improved the quality of speech recognition with cochlear implants by programming electrodes to minimize cross-stimulation and electrode interference within the cochlea [8].

Cochlear implants were officially approved by the United States Food and Drug Administration (FDA) in 1984 [9]. The first marketed cochlear implant in the US was the single-channel implant created through a partnership between Dr. House and 3M. Soon thereafter, Nucleus Limited partnered with the Australian Department of Productivity and the University of Melbourne to produce and market one of the first commercially available multichannel implants. Currently, five companies manufacture cochlear implants: Advanced Bionics, Cochlear, MED-EL, Neurelec of France, and Nurotron out of China, the first three of which are currently available in the US.

Early on in the development and advancement of cochlear implantation, advocates and opponents of the technology were vocal with their opposing viewpoints. Supporters championed the possibility of auditory rehabilitation in patients who previously would have had no access to hearing or who would have been limited to poorly performing hearing aids. Opponents regarded the new technology with hostility and skepticism, especially regarding the proposed use in children who were born deaf [2].

Implant candidacy and evaluation

Cochlear implant candidacy is assessed through a comprehensive audiologic and medical workup. Patients are considered to be conventional cochlear implant candidates if they have bilateral moderate-to-profound sensorineural hearing loss and receive little to no benefit from hearing aids. However, recently the criteria for implant candidacy has expanded, including FDA approval in children as young as one year of age, as well as patients who may have residual low-frequency hearing [10]. In addition to preoperative auditory testing, imaging is obtained, such as a computed tomography (CT) scan of the temporal bone or magnetic resonance imaging (MRI) of the brain to define the anatomy of the temporal bone and to rule out retrocochlear causes of hearing loss, specifically abnormalities of the cochlear nerve that may preclude cochlear implantation [8]. Once the patient is cleared for surgery, pneumococcal vaccines are administered for meningitis prophylaxis per FDA guidelines.

Surgical technique

The procedure is regularly performed under general anesthesia, with facial nerve monitoring. A post-auricular incision is made, followed by dissection of soft tissue to expose the mastoid, as well as establishing a subperiosteal pocket for the implant magnet to rest in. A cortical mastoidectomy is performed, identifying key temporal bone landmarks, including the incus, the lateral semicircular canal, the tegmen tympani, and the sigmoid sinus. The facial recess is opened, the boundaries of which are the facial nerve, chorda tympani, and a shelf of bone left superiorly to protect the incus, known as the incus buttress. The round window niche is identified through the recess.

After identifying the round window, several methods exist of accessing the scala tympani within the cochlea. Methods including drilling a separate hole, known as a cochleostomy, as well as removing any overhanging bone, and gently opening the round window with a straight pick. Lastly, an extended cochleostomy can be performed, simply drilling on the anterior limit of the round window. Once the cochlea is opened, the implant is slowly inserted into the cochlea.

An audiologist performs an integrity test at the conclusion of the procedure, ensuring proper functioning of the implant. The soft tissue and skin are then closed in layers. Some surgeons utilize plain radiographic analysis to ensure proper location of the implant. Once awake, the patients are usually discharged home the same day of surgery, and the implant is activated 2-4 weeks postoperatively.

Complications

Cochlear implantation is a safe surgical procedure performed around the world. Complications are generally classified as major complications, requiring additional surgery or cochlear explantation, and minor complications, requiring conservative medical management. Global complication rates are generally held to be approximately 16% [11]. Complication rates have decreased with smaller surgical incisions, increased experience, and improvements in device design, and are now generally held to be approximately 11.8% for minor complications and 3.2% for major complications [12].

Major complications include infections, facial paralysis, and device failure. Infection rates range from 1-12% in the literature and the majority of these are skin infections or acute otitis media. Soft tissue infections and acute otitis media do not necessarily lead to implant removal, but do increase the risk of removal if the receiver stimulator appears to become infected. There is also a reported increased risk of bacterial meningitis after implantation that is held to be thirty-fold greater in implant recipients than in the general population, but thankfully these cases are quite sporadic with the advent of vaccination [13]. Currently the FDA recommends routine pneumococcal

vaccination with the pneumococcal conjugate vaccine (PCV13) and the polysaccharide vaccine (PPSV23) prior to undergoing implantation [14]. The rate of transient facial nerve palsy is estimated to be approximately 0.7% [15]. This is possibly from heat transferred by the drill in opening the facial recess or performing a cochleostomy or extended round window approach. Another theory is reactivation of herpes simplex virus infection from the stress of surgery. In minor injuries, complete resolution of paresis is observed. Lastly, device failure requiring reimplantation is estimated to occur in 2.5-6% of cases [11,12].

Postoperative vestibular symptoms are not uncommon. Severity varies, but it is believed that up to one-third of patients experience disequilibrium, vertigo, or vestibular weakness lasting more than 1 week postoperatively [16]. The majority of these cases resolve over weeks to months. Patients over 70 are most likely to have permanent vestibular weakness after cochlear implantation.

Unilateral *versus* bilateral implantation

Initially, cochlear implantation was only offered unilaterally. Surgeons began to question whether patients would perform better with unilateral or bilateral implants. Bilateral implantation patients demonstrated improved speech perception, and hearing in noise [11]. In addition, bilateral implant patients show significant improvement in sound localization, compared to their performance with a single implant [17].

Regarding surgical timing, bilateral implantation can be placed sequentially or simultaneously. In adults, studies have not shown a major difference in audiologic outcomes when timing between sequential implantation is minimized, and that the second ear matches the first ear's performance at 6 months [18]. However, children with simultaneous bilateral implantation demonstrated improved performance with speech recognition and language, when compared to children who were implanted sequentially [19].

While there are clear advantages to binaural stimulation, the cost-effectiveness of bilateral cochlear implantation is unclear. One study (out of Ontario, Canada) found the cost for unilateral implantation, including preoperative evaluation, surgery, and postoperative follow-up, was about $64,000, with an incremental cost of the second implant being about $48,000 (USD).The study found that cochlear implantation in adults is cost-effective, when compared to no implantation, though the benefit of sequential implantation is borderline, when compared to unilateral implantation [20]. Other studies have concluded that bilateral simultaneous pediatric implantation and unilateral adult cochlear implantation are cost-effective, but bilateral sequential pediatric implantation and bilateral (sequential or simultaneous) adult implantation are not cost-effective [21].

Cochlear implantation for single-sided sensorineural hearing loss

Implantation in the setting of single-sided deafness has recently emerged as an area of interest [22]. Historically, patients with single-sided deafness have the options of hearing aids, contralateral routing of signal (CROS) devices, or bone-anchored implants. But while CROS devices and bone-anchored implants route sound to the better hearing ear, cochlear implantation has the ability to restore sound to the deaf ear.

A particular challenge for patients with unilateral hearing loss is sound localization. Binaural stimulation and interaural time differences of stimulation allow for complex processing of sounds that facilitates localization. Studies in patients with unilateral hearing loss have demonstrated improvements in localization with cochlear implants as compared to bone anchored implants [23]. In addition to providing improved localization, a recent study analyzed the use of cochlear implantation for tinnitus suppression in patients with single-sided deafness [24]. Implantation led to increased speech perception in noise, improved sound localization, as well as decreased tinnitus. In some cases, complete tinnitus resolution is possible [25].

Hearing preservation after cochlear implantation

Another recent development is the emergence of evidence that acoustic hearing can be preserved despite implantation. Historically, the placement of an electrode into the fluid filled cochlea was believed to destroy the natural mechanism of hearing. However, it has been shown that hearing can be preserved in over half of patients receiving implants [26]. It is not uncommon for patients with severe-to-profound hearing loss to have some residual low-frequency hearing, and preservation of the natural hearing leads to improved speech understanding, sound localization, and hearing in complex listening environments [27]. Cochleostomy approach, electrode design, and steroids have been extensively studied for their effect on hearing preservation.

Several factors, including implant length and placement, are thought to influence hearing preservation during cochlear implantation. While the length of the electrode and its insertion depth can cause intracochlear trauma, studies have shown hearing preservation is possible in patients with full electrode insertion [28]. A recent study of 100 patients demonstrated that the best hearing

preservation outcomes were seen when the electrode was located entirely within the scala tympani [24]. Thus, studies have investigated methods to maximize atraumatic scala tympani insertion. While some report that insertion of the electrode into the cochlea through a naturally existing pathway, such as the round window, is beneficial, others believe that making a separate opening in the cochlea, also known as a cochleostomy, is preferable. However, studies demonstrate there is no significant difference between surgical approaches regarding hearing outcomes [29,30]. However, a recent study showed that round window or extended round window approaches were more likely than cochleostomy approaches to place the electrode completely within the scala tympani, and thus increase hearing preservation [24].

Electrodes have also been designed to traverse the cochlea in various orientations, again to maximize placement within the scala tympani and minimizing trauma to the native cochlear anatomy. Perimodiolar implants are designed to hug the inside of the cochlea, while lateral wall electrodes are designed to adhere to the outside wall of the cochlea during insertion. Recently, studies have shown that lateral wall implants were more likely to remain within the scala tympani than perimodiolar implants [31].

Lastly, perioperative steroids have been shown to possibly contribute to the preservation of hearing. One study found that preoperative intratympanic steroid injections led to increased rates of hearing preservation, especially at low frequencies, and increased stability of hearing preservation over the first year after implantation [27]. In addition, steroid-eluting implants have been shown to improve hearing preservation rates up to 1 year from implantation [32].

Other applications

In hopes of preserving residual hearing, Gantz and colleagues at the University of Iowa developed a hybrid cochlear implant that is only 10 mm in length [33]. This allows for shallow insertion into the cochlea, stimulating the region responsible for high frequency hearing, and avoiding the region responsible for low frequency hearing, which can be intact in some patients undergoing cochlear implantation. Early studies showed hearing preservation and increased speech perception with implant use, with benefits in speech perception in excess of 2 years [29–30,32]. Patients with the hybrid implant also demonstrated improved appreciation of music, due to the benefit of combined acoustic and electrical stimulation [32]. In addition, implant performance of the hybrid was comparable to conventional cochlear implants [33]. However, with a progressive hearing loss, the replacement of the hybrid implant with a full-length implant has been shown to improve hearing and word recognition [34,35], but adds significantly to cost.

Cochlear implantation has also been utilized in patients with profound sensorineural hearing loss and bilateral Meniere's disease. Meniere's disease consists of episodic attacks of hearing loss, tinnitus, and debilitating vertigo spells. In one study, patients that underwent cochlear implantation showed resolution of vertigo spells, improved tinnitus scores, and improved hearing performance [36]. However, their hearing outcomes are slightly worse than comparable patients implanted for other reasons [37].

The totally implantable cochlear implant

A current area of investigation is the development of a totally implantable device. Because implants require patients to wear an external microphone, processor, and transmitting coil to power the electrode, the implant use is limited to dry and relatively stable environments. The totally implantable cochlear implant is intended to bypass this challenge by having the entire system placed underneath the skin. In order for this concept to be feasible, the microphone must be sufficiently small and sensitive enough to detect sound through skin, while filtering internal noise from the body itself. In addition, the rechargeable battery must have a sufficiently long life (and be re-chargeable externally) and the entire system has to be small enough to be fully implanted. Three patients have been implanted with a totally implantable cochlear implant [38].

Conclusion

Cochlear implantation is one of the most significant advances in modern medicine. Patients also receive the benefits of relief from auditory isolation, gains in confidence, and increased ability to function in society. In addition to hearing improvement, patients note improved tinnitus and vertigo. Understanding that preservation of hearing is beneficial, soft-surgical techniques and pharmacologic interests are rapidly developing. However, cochlear implants do not restore all facets of hearing. As devices are developed with better processing, gains will be made in improving speech and noise and music appreciation, and the devices will continue to improve quality of life for patients with hearing loss for generations to come.

Abbreviations

CROS, contralateral routing of signal; FDA, Food and Drug Administration.

Disclosures

The authors declare that they have no disclosures.

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