MAKING THE DEAF HEAR
Cochlear Implantation

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INTRODUCTION

Allowing the deaf to hear has been a goal of humanity since antiquity. The Bible reports Jesus healing a deaf man by placing his fingers in the man’s ear canals (1). Some 2000 years later modern otolaryngologists have been able to reproduce this miracle via the development of cochlear implants. These surgically-implanted devices, with externally worn hardware, convert sound energy to electrical energy which is used to stimulate the brain allowing sound perception. Since Food and Drug Administration (FDA) approval in the 1970’s and subsequent approval for children in the 1980’s, cochlear implants have been used to restore hearing in children born deaf as well as adults who lose their hearing. Approximately 40,000 individuals worldwide have cochlear implant, and approximately 8,000 implants per year are performed in the United States. It is predicted that upwards of 750,000 Americans could benefit from this technology (2, 3). This disparity between use and need is due to a number of factors including lack of awareness of health care providers to this exciting technology, cost concerns related to technology that has a large front end cost, and moral debate between the hearing and deaf cultures. In this paper, we will provide an update as to the current standard of care regarding cochlear implants. In addition, we will discuss the issues stalling their universal acceptance.

DISCUSSION

Hearing Loss

Typically, human beings perceive sound via vibration of the eardrum which initiates a chain reaction ending with higher cortical sound perception. The chain reaction is perpetuated as the vibrations are transmitted and amplified by the bones of the middle ear—the malleus, incus, and stapes (Figure 1). The last bone in the chain causes vibrations in the oval window.

Goal:

Objectives:
of the cochlea leading to fluid waves within the inner ear. These fluid waves distort cilia laden cells, hair cells, which launches a molecular cascade leading to depolarization of the auditory nerve and stimulation of Brocha’s area within the cerebral hemisphere. (Figure 1)

There are many reasons why people lose this ability to hear. Otolaryngologists categorize these into two broad categories—conductive and sensorineural hearing loss. Conductive hearing loss occurs when a mechanical problems impedes the progression of the sound wave from the outer ear to the inner ear. Plugging one’s ears with earplugs produces a temporary conductive hearing loss. Other causes of conductive hearing loss are wax impaction, middle ear infections, ear drum perforations, and diseases affecting the middle ear bones (i.e. otosclerosis). Conductive hearing loss is treated by removing or correcting the obstruction. Sensorineural hearing loss occurs when damage to the hair cells prevents transmission of the signal to the auditory nerve and brain. The most common cause of sensorineural hearing loss is presbycusis, or age-related hair cell death. This multifactorial process is most-likely genetically mediated and exacerbated by repeat exposure to loud noises. Other causes of sensorineural hearing loss are drug toxicity (i.e. gentamycin), infections (i.e meningitis), and autoimmune processes. At early stages of sensorineural hearing loss, amplifying sound with a hearing aid may overcome the depleted hair cell population and allow functional hearing. Hearing aids are typically used to treat mild to moderate levels sensorineural hearing loss. (Table I) For those individuals with advanced sensorineural hearing loss (severe to profound), cochlear implants are the treatment of choice.

Candidacy

At present, individuals with profound to severe sensorineural hearing loss are candidates for cochlear implantation (Table I). As mentioned earlier, the two largest groups of candidates are children born deaf and adults who lose their hearing.

For children born deaf, evaluation begins shortly after birth during newborn hearing screening. While not mandated in the state of Tennessee, 90% of Tennessean newborns have hearing screens before they leave the hospital (4) Such screening helps in identifying the approximately 5 per 1000 children born deaf each year. These children then undergo thorough medical evaluation in attempts to identify the etiology of hearing loss. Increasingly, genetic screening plays a role as mutations in the gene encoding a potassium channel protein, Connexin 26, may be responsible for up to 50% of autosomal recessive, non-syndromic, congenital hearing impairment (5). Following medical clearance, a 6 month trial of hearing aid use is undertaken. During

### Table 1

<table>
<thead>
<tr>
<th>Levels of Hearing Loss</th>
<th>Description</th>
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<tbody>
<tr>
<td>Mild</td>
<td>21-40 dB (whisper ~ 30dB)</td>
</tr>
<tr>
<td>Moderate</td>
<td>41-60 dB (conversation ~ 60dB)</td>
</tr>
<tr>
<td>Moderately severe</td>
<td>61-70 dB (restaurant noise ~ 70 dB)</td>
</tr>
<tr>
<td>Severe</td>
<td>71-89 dB (busy street ~ 90dB)</td>
</tr>
<tr>
<td>Profound</td>
<td>&gt; 90 dB (factory ~ 100dB, jack hammer ~ 130dB)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Device</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Borderline</td>
<td>Normal Hearing</td>
</tr>
<tr>
<td>Hearing Aid Candidate</td>
<td></td>
</tr>
<tr>
<td>Cochlear Implant Candidate</td>
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Cochlear Implants

A cochlear implant overcomes sensorineural hearing loss by bypassing damaged hair cells and directly stimulating the auditory nerve. To accomplish this, an electrode array is surgically threaded into the cochlea placing electrical contacts in close proximity to the auditory nerve (Figure 2). This electrode array is directly coupled to a subcutaneously placed internal receiver which is magnetically linked to an external receiver/processor. The external receiver processor converts sound energy to electrical energy and sends the appropriate signal across the skin and via the electrode array to the inner ear (Figure 3). A cochlear implant differs from a hearing aid in that it is not dependent upon the survival of inner hair cells within the cochlea.
the hearing aid trial, continual assessment of hearing acuity occurs via a number of age-dependent tests. If best aided thresholds remain in the severe-to-profound range, cochlear implantation is recommended. For children who qualify, FDA-guidelines allow implantation for children as young as one year of age.

Severe-to-profoundly hearing impaired adults are further stratified to either pre-lingual deafness, e.g. an individual who has never heard, and post-lingual deafness, e.g. an individual who once heard but has lost that ability. Pre-lingually-deafened adults, i.e. former Miss American Heather Whitestone, McCullum are a distinct minority. While they can undergo cochlear implantation, expected outcome is limited to acquisition of environmental sounds (i.e. hearing alarms). Ms. Whitestone’s continuing story is detailed on-line (6). More typically, adult candidates are post-lingually deafened. An example of a post-lingually deafened adult who underwent cochlear implantation is the conservative talk-show host Rush Limbaugh. Such candidates undergo a thorough assessment to identify possible causes of hearing loss. Age-related hearing loss, presbycusis, remains the leading diagnosis. While much effort is currently being expended in attempts to slow or even reverse the hair cell death characteristic of presbycusis, clinical application is years away. Adults undergo intensive hearing tests and, under best-aided conditions, must have severe-to-profound hearing loss and/or sentence identification below 40% to qualify for a cochlear implant. As a rule-of-thumb, patients who can no longer communicate over the telephone are most likely candidates for cochlear implantation. Advanced age is not a contraindication for implantation (7).

Surgery

Cochlear implantation is usually performed under general anesthesia in an outpatient surgical facility. The operation lasts 3-4 hours in duration. The technique involves a post-auricular incision (Figure 4) following which a mastoidectomy is performed. Within the mastoid, vital landmarks are identified, most notably the facial nerve which supplies innervation to the muscles of facial expression. Using the nerve as a landmark, a recess is opened anterior to the facial nerve and the middle ear is entered from behind (Figure 5). The cochlea is identified and a small hole made into it following which the electrode array is inserted. The associated internal receiver is secured in the temporoparietal skull and the incision closed. Complications are extremely rare and include facial nerve injury, pain, vertigo, CSF leak, flap necrosis, device failure, device migration out of the cochlea, cholesteroloma formation, and eardrum perforation. Because of recent concerns regarding recurrent meningitis in cochlear implant patients, Streptococcus pneumoniae vaccination to prior to surgery is recommended as per CDC guidelines (8). After implantation, care must be taken when performing magnetic resonance imaging (MRI) studies. Of the three FDA approved manufacturers, one (MED-EL Corporation; Innsbruch, Austria) is FDA approved for use in 0.2-Telsa MRI units with ongoing studies aimed at higher strength MRI units. Another (Cochlear Corporation; Melborne, Australia) recommends an in-office surgical procedure to remove the coupling magnet prior to MRI. The coupling magnet is then reimplanted after scanning is completed.

Device activation and outcome

Approximately 3 weeks following surgery, allowing time for post-operative edema to subside, the externally-worn receiver is magnetically coupled to the surgically-implanted receiver. Data is transmitted from the external receiver to the internal receiver via a coil of wire and electrical principles of inductance. At initial hook-up, adjustments are made to each electrode but stimulating each electrode individually and determining the most comfortable level of sound intensity for the patient.
This mapping process is analogous to tuning each individual instrument of an orchestra. Following this, all electrodes are activated. Analogously, the orchestra begins to play.

For children, this may be the first time recognizable sound is presented to their brain and a range of emotions is common. This represents a new birthday—a hearing birthday—following which intensive tutoring is necessary to teach children to hear. With such tutoring, speech recognition without visual clues approaches 90% and children enter mainstream education (9, 10). For adults, speech understanding exceeds 85% and many adults quickly reacclimatize to the hearing world, sometimes communicating via telephone immediately after device activation (11).

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Cost-benefit analysis

With the continued escalation of health care costs much has been written about the costs of cochlear implants. The cost (including surgery, hardware, and rehabilitation) can be upwards of $50,000 per patient. Despite this large up-front investment, cost-benefit analysis has shown that the devices are extremely economical. Using validated techniques analyzing cost per year of use, a study out of Johns Hopkins University reported that cochlear implants are much more cost-effective than other standard surgical implant procedures including implantable defibrillator and knee replacement (12). Another approach to appreciating the cost-savings is comparison of educating a deaf child versus educating a main-streamed, cochlear-implanted child. For a deaf child, the cost of primary education (K-12) approaches one million dollars—roughly 50 times the cost of educating a normal hearing child. Despite this markedly increased cost, 44% of deaf children fail to graduate from high school (compared to 19% of normal hearing individuals) and only 5% of deaf individuals graduate from college (compared to 13% of normal hearing individuals) (13).

The above cited and extensive other studies substantiate that both for children and adults, cochlear implantation is economically justifiable. Debate continues, however, as the upfront cost is being born by the health care community and the long-term savings are benefiting society at large. There are models of government support of cochlear implantation which have been successful in reducing the high cost of deaf education (14). Such programs, however, have met with resistance from the deaf community.

Deaf Culture

As cochlear implants threaten the very existence of their community, many deaf individuals and parents of deaf children oppose the use of cochlear implants. Some, most notably the National Association of the Deaf feel that hearing impairment is not a disability which needs to be fixed (15). Some also believe that their unique language—American Sign Language—assures status as a separate culture with appropriate respect for autonomy. For the interested reader, this ongoing debate between the hearing and non-hearing worlds is the subject of an Academy Award Nominated Public Broadcasting Service documentary entitled Sound and Fury (16). While the documentary does not offer any resolution to the debate, it does provide for understanding of each point-of-view.

CONCLUSION

Cochlear implants are standard of care for hearing impaired individuals who met FDA approved criteria based upon results of formal hearing tests. They are not experimental. They have a high incidence of effectiveness in restoring speech understanding. They have an extremely low incidence of complications. Most insurance carriers cover their costs. This technology, while expensive, is cost-effective. The impact of cochlear implants on deaf culture remains to be seen and is the topic of ongoing debate.
REFERENCES

1. Mark 7:31-37.


