Building a Multidisciplinary Cochlear Implant Team

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ABSTRACT

Cochlear implantation is evolving into a common modality of auditory rehabilitation for the patient with severe to profound sensorineural hearing loss. In order to provide the patient considering cochlear implantation with an understanding of how best to integrate the device into his daily life, adequate evaluation, counseling, and instruction are required. Using the multidisciplinary model, our initial experience has shown that evaluation by specialists in audiology, otolaryngology, speech pathology, pediatric genetics, social work, and child life may lead to better patient and family understanding of cochlear implantation.

INTRODUCTION

About 60,000 people worldwide have received cochlear implants to ameliorate the effects of severe to profound hearing loss. About one-third of these recipients are children (Balkany et al., 2002; Callanan and Poje, 2004). In the normally functioning ear, the tympanic membrane vibrates in response to sound waves collected and transmitted through the outer ear and external auditory canal. Vibrations of the tympanic membrane stimulate movements of the ossicles in the middle ear (incus, malleus and stapes), which in turn cause movements of the inner ear fluid. Cochlear hair cells bend in response to the fluid motion, in the process converting the mechanical signal of the fluid motion into the electrical signals of nerve impulses. The signal, transmitted by the vestibulocochlear (auditory or eighth cranial nerve [CN VIII]), is sent to the auditory centers of the brain, which interpret the sound.

Any link missing in this chain disrupts the normal sensation of and response to sound. Central hearing loss occurs with malfunction of neuronal tissues. Conductive hearing loss is due to any of a number of mechanical-related problems occurring in the outer and middle ear. Sensorineural hearing loss occurs secondary to the inability of the inner ear to transform mechanical energy into electrical stimulation of the vestibulocochlear nerve. Hearing aids are used to ameliorate conductive or sensorineural hearing loss via amplification of sound.

Cochlear implants ameliorate the effects of sensorineural dysfunction by bypassing damaged cochlear hair cells. The external components of the cochlear implant consist of a microphone, speech processor, and transmitting coil (Figure 1). The implanted internal components are the receiver-stimulator and electrode array (Figure 2). A small microphone cradling the ear gathers sound, which is then digitized by the speech processor and sent to the transmitting coil. The receiver accepts the electrical signal from the transmitting coil and then stimulates the fibers of the vestibulocochlear nerve directly via the frequency-specific electrode array.

The decision-making process involved in pursuing a cochlear implant can be an emotional and lengthy one. It involves the individual, the family, and the cochlear implant (CI) team communicating at all stages about the patients diagnosis, candidacy, and rehabilitation potential. Because of the many facets involved in successful outcomes, we have determined that keeping open lines of communication among the disciplines best meet that goal. Therefore, we have established a cochlear implantation team composed of members from audiology, otolaryngology, social work, genetics, speech therapy, and child life to provide this type of multi-disciplinary care.

TEAM MEMBERS

Audiology

The audiologist has an active role in all three of the major stages of cochlear implantation: pre-implant, surgery, and post-implant. During all three stages, the audiologist works with implant candidates/recipients and their fami-

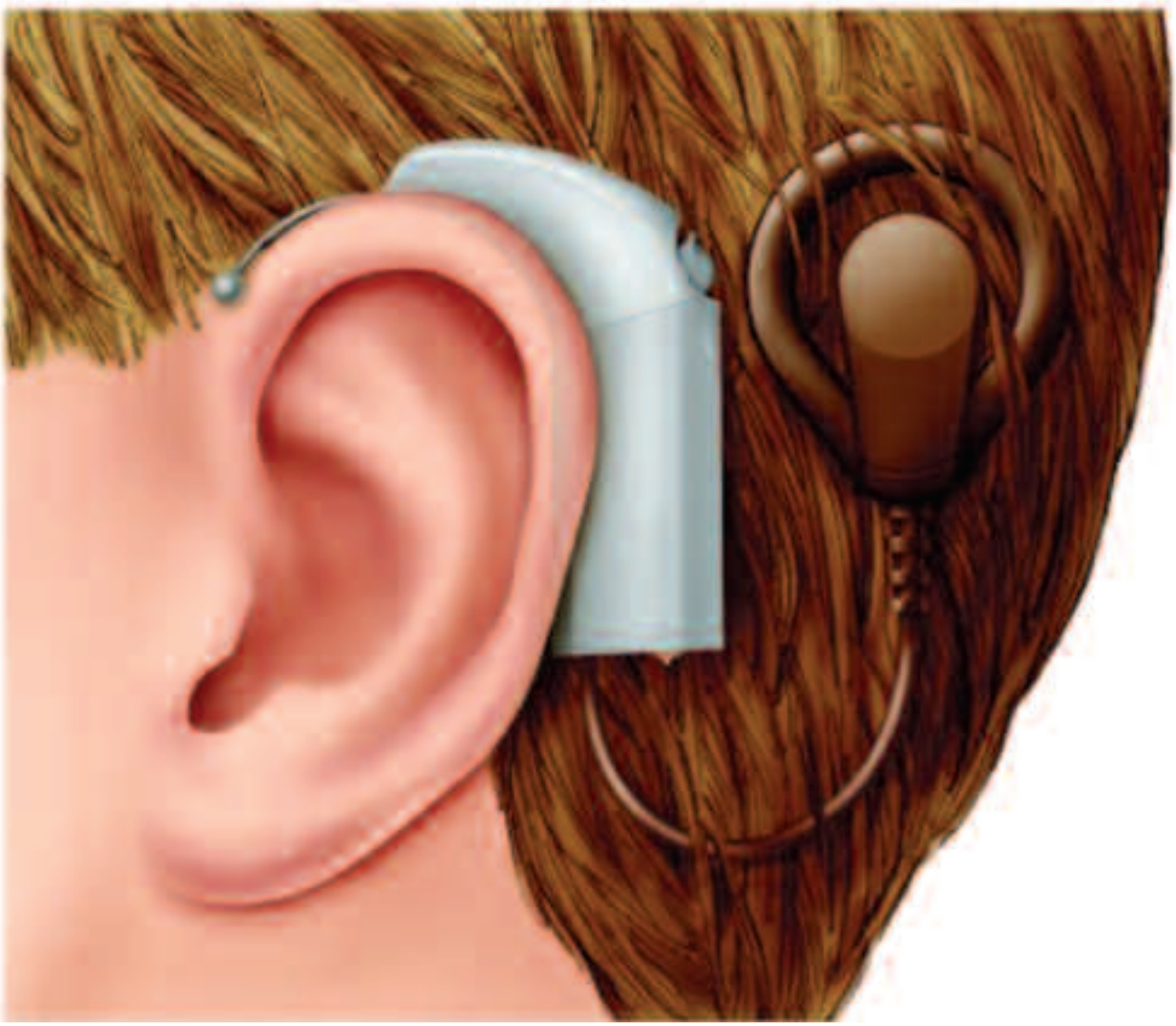

FIGURE 1 | Cochlear implant external components: microphone, behind the ear speech processor, and magnetic transmitting coil (courtesy of Cochlear Limited).

lies while liaising with other members of the CI team.

Initially, the CI candidate's hearing status and auditory function are presented to the CI team. The audiologist provides a comprehensive assessment of the candidate's hearing status delineating the nature and severity of dysfunction, such as the hearing profile with and without hearing aids, and innate auditory characteristics. Diagnostic testing includes audiometry, tympanometry, auditory brainstem testing, and oto-acoustic emissions. Hearing aid systems are evaluated, including the possibility of making a change in the candidate's existing hearing aid fit if optimal performance is not met. Auditory/listening skills are evaluated with a battery of speech perception tests, including aided benefit with current hearing aids in listening and integrating sound/speech patterns, and speech recognition words. Finally, the audiologist conducts a comprehensive communication evaluation to assess mode and communication function as well as the ability to integrate hearing into the context of everyday listening.

Audiologic rehabilitation is usually recommended preimplantation to facilitate transition to listening with CI, especially with children. Implant users benefit enormously, although not everyone with an implant will perform in the same manner. Post-implant performance is linked to auditory, listening, and communication skills at the time of implantation. Audiology provides support both educationally and emotionally to candidates and families, offering many forms of educational packets, explanations, and options to explore in regards to mode of communication,

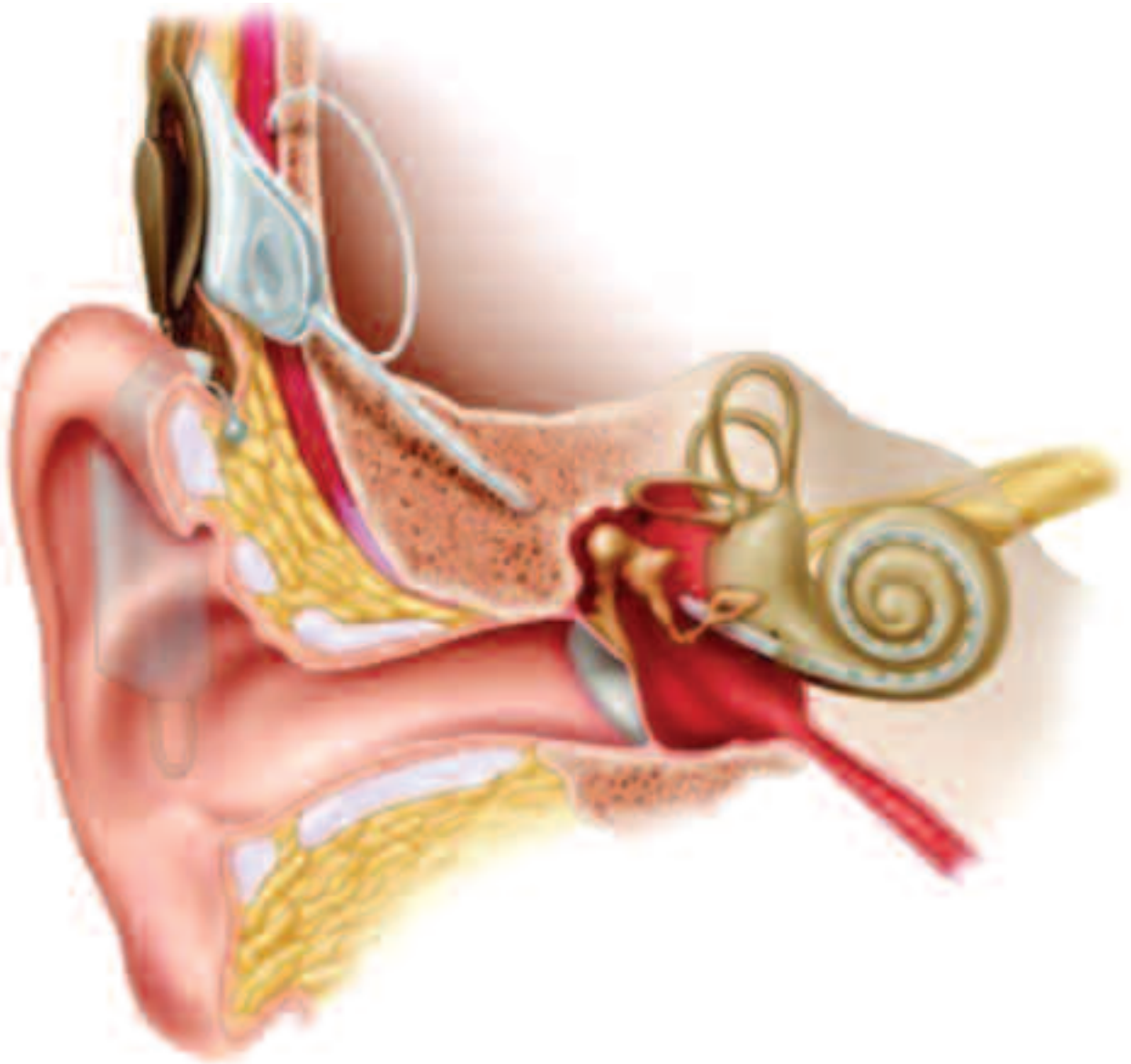

FIGURE 2 | Cochlear implant internal components: receiver stimulator under the scalp and electrode array passing through the mastoid bone into the cochlea (courtesy of Cochlear Limited).

education, and therapeutic interventions. The audiologist's goal is to empower candidates and their families to become active members in the decision process (Chute and Nevins, 2002; Schopmeyer, 2000; Watkins, 1989).

If the decision to proceed to a CI is made, the audiologist has a role during the implantation itself. During surgery, the audiologist measures the integrity of the implanted internal device and auditory nerve function via the CI electrode array. The audiologist also obtains a baseline of peripheral nerve responsiveness, which also assists programming of the CI at initial activation, helping optimize hearing performance.

Implantation marks the beginning of a commitment by the audiologist to the CI's function and to the recipient who depends on it. CI users, particularly children, need to be re-taught to "make sense" of the sounds the CI produces (Estabrooks, 1989; Nevins and Chute, 1996; Schopmeyer, 2000). In the first phase of this commitment, the most intense for recipients and their families, the external components of CI are hooked up to the implanted components and initial stimulation (mapping) of individual electrodes begins. The audiologist selects a specific speech coding strategy and stimulation parameters based on the ongoing evaluation. A series of psychophysical measures in the temporal, spectral, and intensity domains are then electrically manipulated to optimize transmission of sounds into the recipient's dynamic range. This map will be installed into the recipient's speech processor but will be changed many times in the following days, weeks, and months as the CI

patient learns to tolerate and understand more sounds. Instruction, adjustment, and practice in proper CI usage are a vital part of the long-term management, as are the continued development and enhancement of listening, speech perception, and communication skills. Finally, outcome measures and speech perception ability are evaluated on a quarterly basis, permitting optimization of the recipient's hearing, comparison of results to established norms, and collection of data for clinical research and use (Estabrooks, 1998; Moog, 2003; Rance and Dowell, 1997; Nunez, 2003). The audiologist's participation thus contributes to the "critical mass of expertise" and assures that CI recipients obtain the maximum benefit from the CI program (Clark et al., 1997).

Otolaryngology

Otolaryngologists undergo specialty training in surgery of the ear, nose, and throat. Additional fellowship training after residency in Pediatric Otolaryngology, Otology or Neurotology provides further competency in CI surgery. The otolaryngologist works with the CI team to assess whether a patient would benefit from a CI and whether he is medically fit for implantation.

Hearing data, patient motivation, family motivation, and environmental factors are assessed during initial evaluation of the CI candidate. These factors have been shown to influence CI outcome in terms of future speech development (Geers et al., 2002). Once the otolaryngologist is confident that the patient and family fully understand the process of evaluation for cochlear implantation, medical work up is instituted. Initial work up includes temporal bone computed tomography and brainstem magnetic resonance imaging, as well genetics evaluation. Diagnostic imaging is essential to ensure that the CI candidate has inner ear anatomy conducive to surgical implantation. Once all pre-operative data is accrued, the CI team meets to decide if indeed the patient is a candidate for implantation.

The surgery itself is a 3 to 5 hour operation under general anesthesia. The cochlea is approached through an incision behind the ear and through the mastoid cavity. The electrode of the CI is passed just adjacent to the facial nerve, which runs through the mastoid cavity. Postoperatively, the patient may go home that night or within one to two days after surgery. Activation of the cochlear implant occurs four to six weeks after surgery, when the incision is healed and swelling alleviated. Activation involves programming the speech processor and fitting the other external components of the CI.

Throughout all stages of evaluation for implantation, the otolaryngologist must ensure that candidates understand the risks of implantation, such as facial nerve injury or meningitis, both of which occur in less than 1% of CI patients (Kempf et al., 1997; Reefhuis et al., 2003). Candidates must also understand that profound hearing loss does not necessitate implantation. Choosing not to undergo implantation is a viable option. On top of individual counseling, the otolaryngologist must ensure that the patient will be granted access to all information and access to CI team members, so that a fully informed decision about cochlear implantation can be made.

Social Work

Families are referred for a social work assessment after they have met with the otolaryngologist and audiologist, and there is agreement about exploring the possibility of a CI. From the time of initial contact with families, the social worker obtains psychosocial information related to family dynamics and school and social issues, and assesses these factors to inform the committee's decision about patient candidacy. The social worker's interaction with the child and family involves listening to their feelings about the choice to pursue an implant and discussing their understanding of the implantation process. It includes discussing their expectations and questions for the child after surgery, processing pre-operative and post-operative habilitative demands, providing education, and offering support throughout the candidacy process. The committee's decision is further informed by other evaluations and assessments, including a psychological assessment that provides additional counseling and psychoeducation to the families during the candidacy phase.

Pediatric Genetics

All children who are being evaluated for sensorineural hearing loss should undergo pediatric genetic evaluation. Hearing loss is caused by genetic factors in 50% of cases (Jeng and Robin, 2002). Hearing loss caused by environmental factors can be caused by perinatal infection, acoustic or cerebral trauma affecting the cochlea, or ototoxic drugs such as aminoglycoside antibiotics.

Genetic causes of hearing loss can result from a mutation in a single gene (monogenic inheritance), or from a combination of mutations in different genes and environmental factors (multifactorial inheritance). Hereditary deafness may occur in combination with other somatic abnormalities, in which case it is known as syndromic deafness. Even though over 400 genetic syndromes that include hearing loss have been described, approximately 70% of genetic hearing impairment is non-syndromic and 80% of these are inherited as autosomal recessive traits (Jeng and Robins, 2002). Non-syndromic hearing loss is autosomal dominant in 20% of cases, X-linked in about 5% of cases, and mitochondrial in less than 1% of cases (Smith and Hone, 2003).

Linkage analysis has mapped many genes for non-syndromic hearing loss. The different loci that cause non-syndromic hearing loss are called DFN for deafness and are numbered in chronologic order of discovery. Autosomal dominant loci are referred to as DFNA, recessive loci

as DFNB, and X-linked recessive loci as DFN. At least 82 loci have been identified since Leon et al. mapped the first locus to 5q31 in 1992 (Leon et al., 1992).

Gene discovery in the auditory system is now progressing at the speed of sound. The cochlea is an intricate organ that contains dozens of cell types and specialized regions for the normal process of hearing. Many of the proteins encoded by genes responsible for deafness are expressed within the cochlea and can be grouped into functional categories that include hair-cell structure, extracellular matrix, compartmentalization and ion homeostasis, and transcription factors. Mutations in the gap junction subunits, connexins, are etiologic in several types of non-syndromic deafness. The connexin subunits are essential for gap junction communication between neighboring cochlear cells. Mutations in the gene for DFNB1 (the connexin-26 or GJB2 gene) are thought to be responsible for as much as 50% of profound non-syndromic deafness in American and European populations (Cohn and Kelley, 1999; Rabionet et al., 2000).

Our own routine evaluation of patients who are candidates for cochlear implants includes pedigree analysis, connexin-26 sequencing, Southern blotting for mitochondrial DNA mutations 3243, 7445, and 1555, and an electrocardiogram to rule out the dysrhythmic long QT syndrome (Jervell and Lange–Nielsen syndome). Our genetic assessment better informs CI candidates and their families of the possible etiology of their hearing loss, prognosis, and recurrence risk.

Speech Therapy

The speech pathologist holds responsibilities both before and after implantation. Before implantation, the speech pathologist plays a critical role in identifying potential candidates for cochlear implantation. Using specific selection criteria, such as age, cognitive level, current language, and communicative ability, the speech pathologist can identify those factors that will likely have an impact on outcome. With these data, future rehabilitative needs can be strategically planned. It is crucial that patients and their parents have an understanding of the lengthy rehabilitation needs after cochlear implantation. Family support has been identified as an important element in speech and language success after cochlear implantation (Geers et al., 2002). Subsequent to implantation, the speech pathologist assists in program planning, school placement, and the setting of initial goals for intervention. In some instances, the speech pathologist may also be the individual to intervene and provide ongoing therapy services and follow-up.

Child Life Specialist

Once the decision is made to undergo cochlear implantation, the child life specialist collaborates with the CI team to minimize the stress and anxiety that the child and family may experience during hospitalization perioperatively. Aside from providing children and families emotional support, advocacy, and the maintenance of a "child friendly" environment, the child life specialist provides the patient with preparation for surgery and medical procedures using a variety of tools (Gaynard et al., 1998). Prior to surgery, child and family are given a tour of the facilities, clarifying any misconceptions they may have, and making them feel more familiar and comfortable in the hospital environment. Children are encouraged to share their feelings around their hospital experience through therapeutic play techniques, art and recreational activities. The child life specialist aims to give the child and family a greater understanding of the procedure using developmentally appropriate teaching and preparation for surgery as well as other tests and procedures the child may undergo (Stanford and Thompson, 1998).

CONCLUSION

Congenital hearing loss is the most common sensory disorder in children. About one in every 1000 children born in the US is born with a loss sufficient to interfere with language development. New York State has mandatory newborn hearing screening without which the average age of diagnosis would be fourteen months. A child with bilateral profound sensorineural hearing loss who is a CI candidate requires a full biopsychosocial evaluation by a multidisciplinary team. The many health professionals involved with the patient and family including audiology, otolaryngology, speech pathology, pediatric genetics, social work, and child life each provide a unique clinical perspective. Using this team approach, the open channels of communication among all parties allow for coordinated evaluation, education, and implantation of the CI candidate.

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