

Cochlear Implantation: An Overview

Nicholas L. Deep¹ Eric M. Dowling² Daniel Jethanamest¹ Matthew L. Carlson²

¹Department of Otolaryngology–Head and Neck Surgery, New York University School of Medicine, New York, New York, United States

²Department of Otorhinolaryngology, Mayo Clinic, Rochester, Minnesota, United States

Address for correspondence Matthew L. Carlson, MD, Department of Otolaryngology–Head and Neck Surgery, Mayo Clinic, 200 First Street SW, Rochester, MN, 55905, United States (e-mail: carlson.matthew@mayo.edu).

J Neurol Surg B 2019;80:169–177.

Abstract

A cochlear implant (CI) is a surgically implanted device for the treatment of severe to profound sensorineural hearing loss in children and adults. It works by transducing acoustic energy into an electrical signal, which is used to stimulate surviving spiral ganglion cells of the auditory nerve. The past 2 decades have witnessed an exponential rise in the number of CI surgeries performed. Continual developments in programming strategies, device design, and minimally traumatic surgical technique have demonstrated the safety and efficacy of CI surgery. As a result, candidacy guidelines have expanded to include both pre and postlingually deaf children as young as 1 year of age, and those with greater degrees of residual hearing. A growing proportion of patients are undergoing CI for off-label or nontraditional indications including single-sided deafness, retrocochlear hearing loss, asymmetrical sensorineural hearing loss (SNHL) in adults and children with at least 1 ear that is better than performance cut-off for age, and children less than 12 months of age. Herein, we review CI design, clinical evaluation, indications, operative technique, and outcomes. We also discuss the expanding indications for CI surgery as it relates to lateral skull base pathology, comparing CI to auditory brainstem implants, and address the concerns with obtaining magnetic resonance imaging (MRI) in CI recipients.

Keywords

- ▶ cochlear implantation
- ▶ hearing loss
- ▶ auditory prosthesis
- ▶ electric stimulation
- ▶ auditory nerve
- ▶ auditory brainstem implant

Introduction

A cochlear implant (CI) is a surgically implanted device used for hearing rehabilitation of adults and children with advanced sensorineural hearing loss (SNHL) and/or poor speech discrimination who gain limited benefit from conventional hearing aids. A CI transduces acoustic energy into an electrical signal, which is used to stimulate surviving spiral ganglion cells of the auditory nerve. The observation that electrical stimulation of the auditory pathway can create the perception of sound was discovered in 1790 by Alessandro Volta.¹ In the 1950, Lundberg was the first to be credited with the stimulation of the auditory nerve with a sinusoidal current during a neurosurgical procedure. In the 1966, Simmons was the first to demonstrate the tonotopic organization of the cochlea, whereby different regions along the

basilar membrane vibrate at different sinusoidal frequencies.¹ One of the most significant advancements came with Dr. William House in the 1960s, who along with engineer Jack Urban, created the first implantable device that could stimulate the auditory nerve, making cochlear implants a clinical reality. The original device was commercially marketed as the House/3M cochlear implant that utilized a single electrode array. During the late 1970s, Graeme Clark in Australia developed the first multichannel cochlear implant (Cochlear Nucleus Freedom), which had enhanced spectral perception and speech recognition capabilities compared with the single channel device. In the 1985, the Food and Drug Administration (FDA) approved the first multichannel CI for use in the United States (U.S.).

Over the last 3 decades, advancements in surgical technique, electrode design, and improved speech processing

received

July 14, 2018

accepted after revision

July 19, 2018

published online

September 6, 2018

© 2019 Georg Thieme Verlag KG
Stuttgart · New York

DOI <https://doi.org/10.1055/s-0038-1669411>
ISSN 2193-6331.

strategies have led to increasingly better outcomes in CI recipients. As a result, FDA candidacy criteria have gradually expanded from initially only implanting postlingual deafened adults with profound bilateral SNHL to now implanting adults and children with greater degrees of residual hearing. Furthermore, a growing proportion of patients are undergoing CI for off-label or nontraditional indications including singlesided deafness, retrocochlear hearing loss, such as with vestibular schwannoma, asymmetrical SNHL in adults and children with at least 1 ear that is better than performance cut-off for age, and children less than 12 months of age.

Cochlear Implant Design and Function

The CI system consists of separate external and internal components (►Fig. 1). The external components include the microphone, battery, speech processor, external magnet, and transmitter antenna. The internal components include the internal magnet, antenna, receiver-stimulator, and electrode array. Sound is first detected by a microphone worn on the ear and subsequently converted into an electrical signal. This signal is then sent to an external sound processor, where, according to one of the several different processing strategies, is transformed into an electronic code. This digital signal is transmitted via radiofrequency through the skin by a transmitting coil that is held externally over the receiver-stimulator by a magnet. Ultimately, this signal is translated by the receiver-stimulator into rapid electrical impulses distributed to multiple electrodes on an array implanted within the cochlea (specifically, the scala tympani). The electrodes, in turn, electrically stimulate spiral ganglion cells and auditory nerve axons, which then travel to the brain for further processing. By using these signals to systematically regulate the firing of intra-

cochlear electrodes, it is possible to convey the timing, frequency, and intensity of sound.

There are currently 3 FDA approved CI manufacturers: (1) Advanced Bionics Corporation (Valencia, CA, USA), (2) Cochlear Corporation (Lane Cove, Australia), and (3) MED-EL GmbH (Innsbruck, Austria). The device performance and reliability is generally comparable between all 3 implant manufactures. Each manufacturer has multiple electrode arrays to choose from, depending on the anatomy of the cochlea, the amount of residual hearing, and surgeon/patient preference. Over the recent decade, electrode arrays have been designed to be thinner, softer, and more flexible to minimize trauma during insertion and preserve the delicate neuroepithelial structures within the cochlea. Short (“hybrid”) electrode arrays have also been developed to allow the preservation of the native low-frequency hearing by not extending to the apical regions of the cochlea that contain neurons associated with low frequencies.² With atraumatic cochlear implant insertion technique, patients with residual low-frequency hearing (< 1 kHz) can preserve their natural “acoustic hearing,” while benefiting from electrical hearing in the mid to high frequencies. This type of “electroacoustic stimulation” (EAS) confers distinct advantages in speech perception in noise and music appreciation.³ Furthermore, residual hearing can be amplified with a hearing aid on the same ear as their CI, which is termed the “hybrid strategy.”

Candidacy Evaluation

The basic evaluation of CI candidates involves a medical, audiometric, and radiographic evaluation. A thorough otologic medical history should attempt to determine the etiology of the hearing loss. Prelingual versus postlingual deafness, as well as duration of deafness is valuable information to elicit since

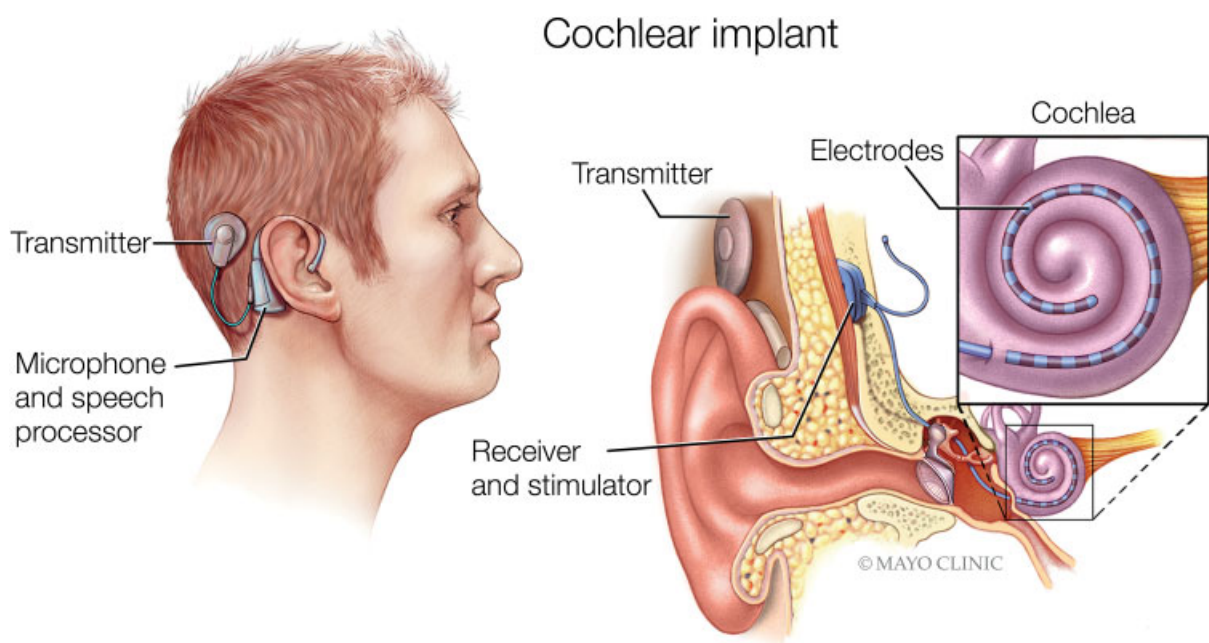


Fig. 1 Components of the cochlear implant system.

very longstanding severe or profound hearing loss may predict a poorer outcome. The majority of pediatric CI candidates are prelingual deafened children, who are born with SNHL due to genetic mutations (e.g., connexin 26), perinatal environmental exposures, or unidentified (idiopathic) causes. These patients often obtain good speech outcomes after implantation with the best results, occurring when implanted within 1 to 2 years of age.⁴

In contrast, prelingually deafened adolescents are generally not good CI candidates and have a higher risk of becoming nonusers of the device. Due to the long duration of deafness, reorganization takes place at the level of the auditory cortex, resulting in regions of the brain that would normally process auditory inputs being taken over by other somatosensory inputs, such as vision.⁵ As it turns out, once a cortical area is allocated to a different task, returning to the original task is difficult, if not impossible, so these patients often have poor outcomes and limited benefit with a CI.⁵

Preoperative discussion with patients (or the parents of children) should involve the risks, benefits, goals, and expectations of CI surgery, as well as the importance of postimplantation rehabilitation and programming. For example, patients with cognitive or developmental disorders require careful consideration prior to implantation; although these patients may not always achieve open-set-word recognition, sound awareness alone may be a reasonable goal of surgery.

Candidacy for cochlear implantation relies heavily on the audiological evaluation. The goal is to identify those patients in whom the implant is likely to provide better hearing. Patients, who endorse difficulty using the telephone, are often good candidates to refer for formal CI audiometric testing. Similarly, patients with less than 50% word recognition during standard audiometric testing should be considered for CI candidacy testing. In recent years, the accepted audiometric criteria for implantation have expanded to include patients with more residual hearing, as the safety and efficacy of CI has been more established.

Radiographic assessment is important to ensure that there are no contraindications to implantation, particularly complete labyrinthine aplasia, cochlear aplasia, cochlear nerve aplasia, and complete cochlear ossification. In addition, a history of meningitis, temporal bone fracture, or otosclerosis warrants close review of temporal bone imaging

to look for cochlear obliteration or scarring, which would complicate electrode insertion and/or require a more sophisticated drillout procedure. Magnetic resonance imaging (MRI) is the best modality for confirming a fluid-filled cochlear duct to receive the electrode, as well as the presence of a cochlear nerve to carry the signal to the brainstem and auditory cortex.⁶ Importantly, in older patients, obtaining an MRI prior to CI provides them with their last opportunity to obtain a high-quality brain image without artifact or the need to remove a magnet. High resolution computed tomography (HRCT) also has utility, particularly for surgical planning in cochlear malformations, and can also be done in a faster, more cost-effective manner compared with MRI.⁷

Conventional Indications

The FDA criteria for implantation are not consistent across devices or companies. To complicate matters, insurance companies also have varied criteria, but are generally similar to FDA guidelines. ▶ **Table 1** provides a broad overview of conventional CI indications. The cochlear implant audiometric assessment is performed by audiologists and is more comprehensive than a typical audiogram. For adults in the U.S., candidacy is based on sentence recognition test scores, most commonly Arizona Biomedical Sentences (AzBio), with properly fitted hearing aids. Scores $\leq 50\%$ in the ear to be implanted and $\leq 60\%$ in the best-aided condition are generally needed to establish candidacy. However, Centers for Medicare and Medicaid Services (CMS) has stricter criteria, requiring binaural aided sentence scores $\leq 40\%$.

In children undergoing an evaluation for cochlear implantation, it is first necessary to establish auditory thresholds. This assessment may include otoacoustic emissions, auditory brainstem response testing, auditory steady-state responses, and behavioral testing. A hearing aid trial of at least 3 months should be instituted before considering implantation regardless of the estimated level of hearing loss. Input is elicited from audiologists, parents, teachers, speech and language pathologists and the cochlear implant surgeon. Implantation is indicated in children, who fail a trial of amplification, and who have bilateral severe-to-profound SNHL and aided open-set recognition scores $\leq 30\%$ (in children capable of testing). Currently, the FDA requires children

Table 1 Cochlear implant candidacy guidelines

	Adult	Children (2–17 y)	Children (12–24 mo)
Hearing threshold	Moderate to profound SNHL in both ears (> 40 dB)	Severe to profound SNHL (> 70 dB)	Profound SNHL (> 90 dB)
Word recognition	Limited benefit from binaural amplification defined by $\leq 50\%$ sentence recognition in the ear to be implanted (or $\leq 40\%$ by CMS criteria) and $\leq 60\%$ in the contralateral ear or binaurally.	Limited benefit from binaural amplification defined by $\leq 20\text{--}30\%$ word recognition scores.	Limited benefit from binaural amplification trial based on the MAIS.

Abbreviations: CMS, centers for medicare and medicaid services; dB, decibels; SNHL, sensorineural hearing loss; MAIS, meaningful auditory integration scale.

to be ≥ 12 months of age; however, the age limit is lower in several European countries and some centers in the U.S. are implanting children as young as 6 months old.⁸ There has been a trend for early, bilateral pediatric cochlear implantation for prelingual deafened children since bilateral CIs permit optimal auditory development, language acquisition, and faster integration into society. Bilateral implantation can be done under a single surgery (simultaneous) or in staged fashion (sequential).

Expanded Indications

Over the past several decades, improvements in CI technology have translated to better performance outcomes, and as a result, there has been an expansion in CI candidacy criteria. For example, children with auditory neuropathy spectrum disorder (ANSD) have shown to achieve reliable open-set speech recognition and the majority of patients with cochlear malformations (e.g., Mondini deformity), who were previously not implant candidates are now being implanted safely.⁹

In addition, there is growing interest in implanting patients with single-sided deafness (SSD).¹⁰ CI surgery for SSD has been shown to reduce or alleviate tinnitus and improve sound localization due to the binaural input in many patients. Patients with end-stage Meniere's disease with SSD have found resolution of vertigo spells and restoration of auditory function to the deafened ear after simultaneous labyrinthectomy and cochlear implantation.¹¹ At present time, implantation for SSD is not currently FDA approved, but increasing evidence suggests that this may be a viable option in the future.¹²

Cochlear implantation after lateral skull base surgery is another area of interest. Patients with neurofibromatosis type 2 (NF2), with bilateral vestibular schwannomas or sporadic vestibular schwannomas in an only hearing ear, often pose challenging clinical dilemmas. Historically, this patient population was not considered for cochlear implantation; however, more recent experience demonstrates that open-set speech recognition can be achieved after implanting patients with a vestibular schwannoma, despite the presence of a tumor on the adjacent vestibular nerve.^{13,14} Even patients with intracochlear schwannomas, being conservatively managed, have had good outcomes after CI by leaving the tumor in situ, preserving cochlear architecture, and passing a styled electrode array through the tumor.¹⁵ Furthermore, patients who have undergone resection of a vestibular schwannoma may be considered for CI, if the cochlear nerve has been preserved. When an intact cochlear nerve exists, outcomes are generally superior to auditory brainstem implants (ABIs).¹⁶

Other retrocochlear and central nervous system disorders have shown good outcomes after CI, including properly selected patients with superficial siderosis, pachymeningitis, sarcoidosis, history of CNS radiation, and other brainstem lesions.¹⁷ The decision to proceed or not is difficult, particularly because the results are unpredictable. However, successful rehabilitation of SNHL is possible and the risks

of surgery are relatively low, thus making the attempt reasonable.

Operative Technique

Cochlear implantation in the U.S. is now usually performed on an outpatient basis, under general anesthesia, and without muscle relaxation to allow for facial nerve monitoring. Selected elderly patients have also been safely implanted under conscious sedation.¹⁸ Patients should be preoperatively vaccinated according to Centers for Disease Control and Prevention (CDC) guidelines for meningitis prophylaxis.¹⁹ A 3 to 4 cm postauricular skin incision is designed to provide adequate exposure to the mastoid and place the receiver-stimulator device. A mastoidectomy with facial recess approach is performed to access the cochlea for placing the electrode (**►Fig. 2**). The mastoidectomy need not be large or exonerate every air cell, as is typical for chronic ear disease or lateral skull base surgery, rather it only needs to be large enough to permit access into the middle ear through the facial recess, which is the area bordered superiorly by the incus buttress, medially by the facial nerve, and laterally by the chorda tympani. This area is opened up very carefully and under constant irrigation as to not damage the facial nerve directly or indirectly via heat generated from the friction of drilling. Careful thinning of the posterior ear canal wall permits greater light penetration and improved visualization into the middle ear. The area of the round window can be seen once the facial recess is fully opened. The bone overhanging the round window is drilled away to provide direct visualization of the entire round window membrane and avoid confusion from hypotympanic air cells. The implant is then opened and the receiver-stimulator portion is secured either in a shallow well created in the outer cortex of the skull or in a tight subperiosteal pocket. For patients, who will require future MRI's for tumor surveillance, such as in NF2, it is advantageous to place the internal receiver-stimulator farther posterior and superior than is typical, to minimize the artifact at the internal auditory canal.

The electrode is then inserted into the scala tympani via a slit made in the round window membrane or alternatively, through a separate cochleostomy made anterior and inferior to the round window. The electrode is inserted in a slow controlled motion to prevent significant intracochlear trauma.²⁰ After insertion, a small piece of fascia is used to seal the round window opening. The electrode array is gently coiled in the mastoid cavity and the postauricular incision is closed. An intraoperative plain film radiograph may be obtained to assess electrode position. Impedance testing and neural response telemetry can also be performed by the audiologist to test the integrity of the device.

Device Programming and Outcomes

After 2 to 4 weeks of recovery, patients will follow-up with the audiologist for the "initial stimulation" and device programming, which involves setting specific parameters of

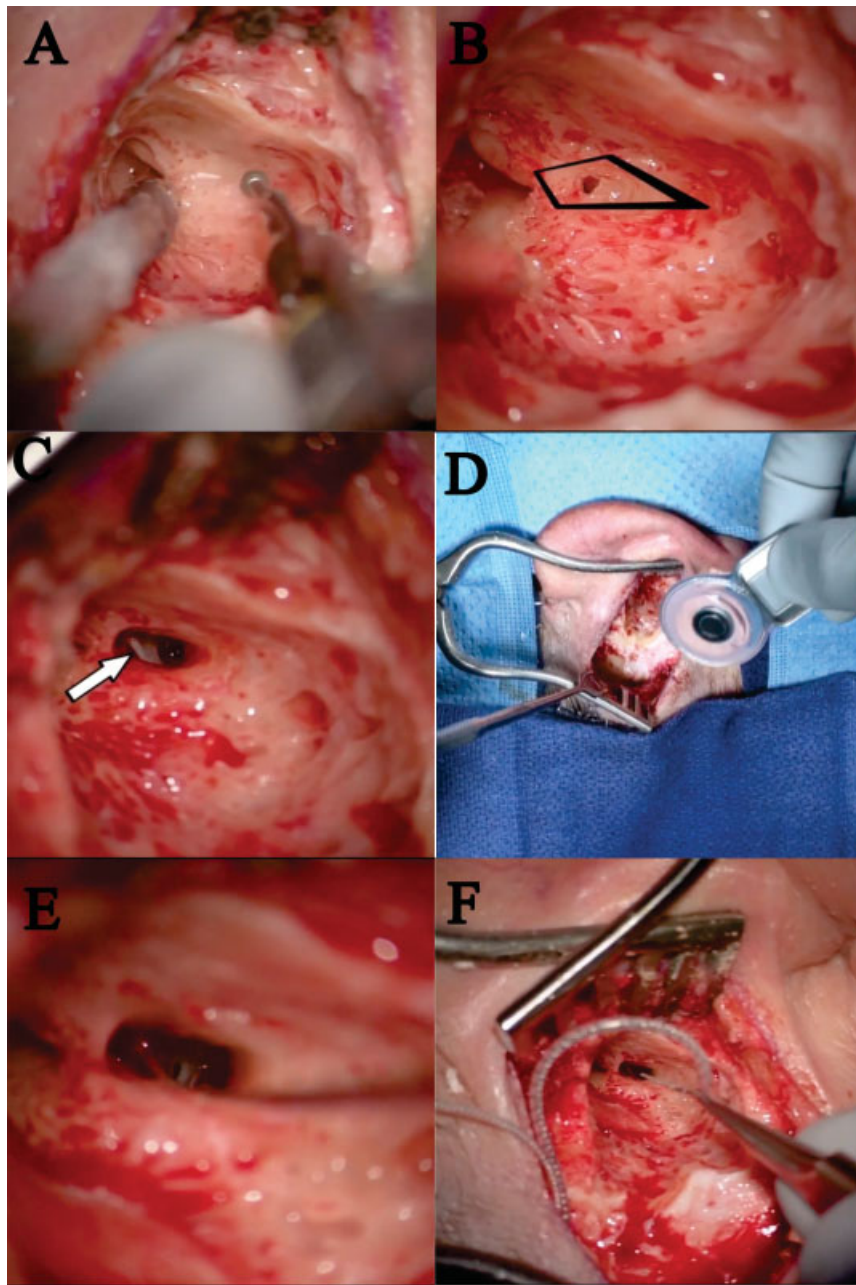


Fig. 2 (A) A mastoidectomy has been performed. (B) The short process of the incus can be seen which identifies the level of the facial recess (outlined in black). (C) The facial recess is opened and the stapedial tendon can be appreciated (arrow). (D) The receiver-stimulator is inserted within a tight subperiosteal pocket. (E) The round window membrane is fully exposed by drilling away the ledge of bone over the round window niche. A slit is then made in the round window. (F) The electrode is inserted through the round window in a slow and controlled fashion and ultimately resides in the scala tympani of the cochlea.

stimulation (e.g., loudness levels) individualized for the recipient's ear. Initially, many patients report that speech sounds distorted. Amazingly, the sound quality gradually improves as the brain adapts to the new sound over the following 3 to 6 months, depending on several factors, including age at implantation, length of deafness, previous experience with sound, and access to aural rehabilitation and therapy services.

In adults, a CI can reliably restore excellent access to sound with near-normal hearing thresholds (around 25 dB HL) and greater than 75% open-set sentence recognition. Adult CI

recipients reported a restored capability to communicate on the telephone (attained by roughly 60%), the ability to converse without the necessity of lip-reading, improvement in tinnitus, and improvement in preimplantation depression.

In children, earlier implantation generally yields more favorable results. Postlingual deafened children or adolescents have excellent outcomes, achieving greater than 80% word understanding after implantation.²¹ In comparison, prelingual deafened children make slower progress toward oral communication and with more variable outcomes, but generally catch up to the postlingual deafened children by

approximately 36 to 60 months postoperatively.²¹ Similarly, among prelingual deafened children, those implanted earlier (in the first year of life) perform better on word recognition testing compared with those implanted in the second or third year of life. However, when these results are expressed as a function of “hearing age” (e.g., time following cochlear implantation) rather than chronological age, there are no significant differences among these patients as all reach an average of around 80% correct on word recognition testing, 4 years after implantation.⁴

Cochlear implantation has demonstrated one of the highest cost-effectiveness ratings of common medical interventions, particularly in children.^{22,23} The development of oral language and enrollment in mainstream schooling are common metrics for determining the effectiveness of cochlear implantation in children. Long-term studies have demonstrated that within 5 years of implantation, the rate of full-time assignment to a mainstream classroom increased from 12 to 75%, vastly reducing the utilization of support services.^{24,25}

MRI Compatibility

As candidacy criteria for CI continues to expand, there has been a greater number of CIs performed. Likewise, the use of MRI as the preferred imaging modality for many medical conditions has also steadily grown. As a result, the number of CI recipients requiring diagnostic MRI continues to increase.²⁶ This is particularly common in patients with NF2 or a history of intracranial tumors who are undergoing imaging for routine tumor surveillance.

Historically, MRI in patients with a CI and an internal magnet in place was contraindicated due to risks of device malfunction or demagnetization and patient injury from implant heating, induced electrical currents, or device torquing. Gradually, the safety concerns have been addressed, as the FDA initially approved an “MR conditional” designation to several devices, meaning that a specific protocol for a given implant model should be followed (e.g., low field strength or removal of magnet). Subsequently, centers began scanning at 1.5 T with the magnet in place with the addition of a tight headwrap to secure the implant and counteract magnet canting or dislodgement. Today, all 3 FDA approved CI manufacturers have devices that have MRI conditional labeling for 1.5 T MRI with magnet in place, as well as 3.0 T MRI with magnet removed. The newest generation device from MED-EL allows for 3.0 T MRI with magnet in place due to the innovative design that contains an internal rotatable, self-aligning removable magnet designed to effectively reduce torque, and associated discomfort.

The removable disc magnet system is advantageous, because it allows flexibility to remove the internal magnet if needed to reduce, but not resolve, MR (magnetic resonance) image degradation. The primary drawbacks of magnet removal include the wear and tear on the magnet housing, risk of device infection, the requirement for additional local or general anesthesia before and after the MRI, and the period of nonuse, while the surgical site heals.

Patients are routinely counseled regarding the potential for discomfort at the implant site, magnet migration (which may occur in up to 15% of cases, even with tight headwrap application), the very low risk of device malfunction as well as the alternative of magnet removal before MRI.^{26–28} The discomfort is usually tolerable and pretreatment with a local lidocaine block injected several centimeters posterior and inferior to the device (but not directly adjacent to it) may be offered. If the magnet becomes partially tilted on end, it can be resealed with gentle pressure over the scalp. If unsuccessful, the magnet should be surgically repositioned to prevent scalp complications. Complete magnet displacement with polarity reversal has also been reported in 6% of cases and requires surgery to revise. Device failure or soft tissue complications are rare.²⁶

Though the safety of MRI with a CI in place has steadily been established, the implant does generate significant imaging artifact, which is worse when the magnet is left in place (►Fig. 3). The severity of the artifact depends on the image acquisition technique. In general, fat suppression techniques, such as short tau inversion time inversion-recovery (STIR) or Dixon’s techniques, should be used instead of fat saturation techniques, which are known to cause significant artifact. Fast spin-echo (FSE) T2-weighted MRI can sometimes be used to better visualize structures, when contrast-enhanced T1-weighted images show significant artifact. Metal artifact reduction scan techniques are commercially available as well, which may help reducing artifact but are unlikely to fully compensate for magnetic material.²⁹ On FIESTA-C/CISS sequences, band artifacts may interfere with visualization, which is particularly problematic in NF2 patients, who are annually monitored for tumors within their cerebellopontine angle and internal auditory canal. Echo-planar imaging and fluid-attenuated inversion recovery imaging are notoriously challenging due to the gross geometric distortion and signal loss that results.²⁹

Ultimately, since different planar views and different sequences exhibit varying degrees of artifact severity, alternating between coronal and axial images, and evaluating multiple sequences is beneficial. Usually, even with the magnet in place, the ipsilateral internal auditory canal and cerebellopontine angle can be adequately examined in at least 1 plane.²⁶ NF2 patients, requiring frequent MRI for multiple, bilateral intracranial tumors may alternatively be managed by implanting the device without a magnet, and securing the receiver/stimulator with a headband instead.

Auditory Brainstem Implants

For patients with profound SNHL, who cannot benefit from a CI, the only remaining option of hearing rehabilitation of the ipsilateral ear is an ABI.¹⁶ Similar to a CI, an ABI directly stimulates the auditory pathway; however, it acts further downstream and more centrally. Whereas the CI stimulates the spiral ganglion cells of the cochlea, the ABI is implanted near the surface of the cochlear nucleus on the brainstem. Similar to CI, an ABI consists of an implanted receiver stimulator, electrode, and an external speech processor.

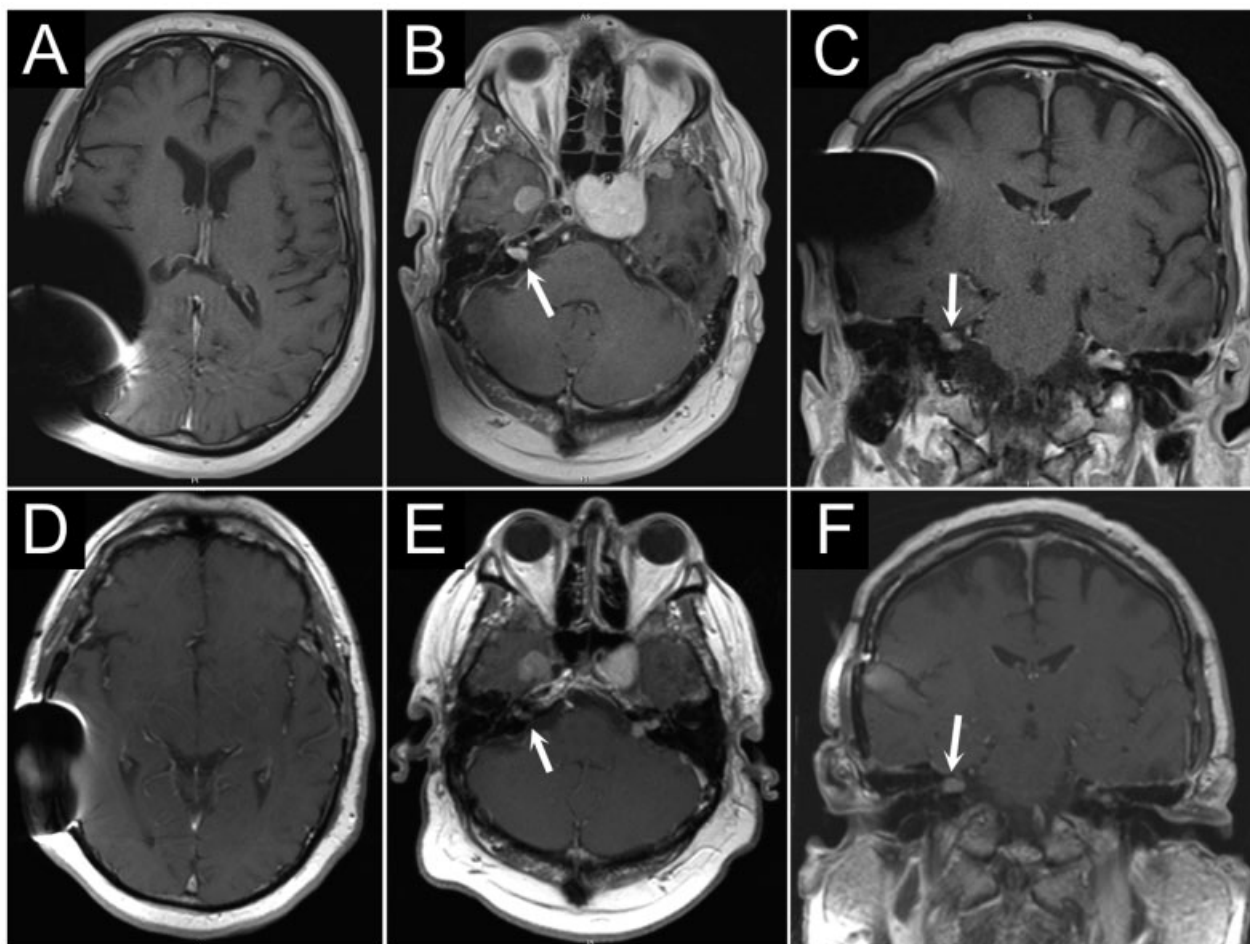


Fig. 3 Comparison of MRI artifact with (A–C) and without (D–F) the internal magnet in place. (A and D), Contrast-enhanced T1-weighted axial images at the level of maximum image distortion demonstrate less ferromagnetic artifact with the internal magnet removed. Contrast-enhanced T1-weighted axial (B and E) and coronal (C and F) images demonstrate adequate visualization of the ipsilateral internal auditory canal (arrows) in both cases despite adjacent ferromagnetic artifact from the cochlear implant. B and E are both far enough from the magnet that neither demonstrate significant artifact. (Reprinted with permission from Carlson et al.²⁶)

Indications for ABI include patients with a disrupted or absent cochlear nerve or those with a cochlea that is not suitable for implantation. The most commonly implanted patients are those with NF2, who have become deaf from disease progression or as a result of treatment. Other patients include those with deafness secondary to bilateral temporal bone fractures with cochlear nerve avulsion or labyrinthine ossification, complete ossification of the cochlea most commonly from streptococcal meningitis, and severe inner ear malformations such as labyrinthine aplasia, cochlear aplasia or cochlear nerve aplasia, whereby there is no receiving cavity to house an electrode array and/or a nerve to propagate the signal to the brainstem.¹⁶

Unlike CI, surgery for ABI requires a posterior fossa craniotomy. Placement of the electrode pad is somewhat nuanced, and results are highly variable. Furthermore, the tonotopicity of the cochlear nucleus is arranged obliquely through the pons. Therefore, to obtain frequency-discriminatory hearing, a penetrating electrode would be needed. Unfortunately, preliminary results of penetrating ABI electrodes do not demonstrate a significant advantage over

surface electrodes. This is in contrast to CI, which requires a short outpatient procedure with minimal risks. Furthermore, a CI electrode can more readily achieve reliable tonotopic stimulation of the auditory system, and the dense insulating bone of the otic capsule minimizes untoward electrical stimulation.

Outcomes: CI versus ABI

While both CI and ABI have well-established outcomes, performance is typically very different between the two.^{16,30} At present time, ABI most commonly facilitates identification of environmental sounds, and augments lip reading, but the ability to achieve high-level open-set speech recognition is limited. For this reason, situations where the cochlear nerve appears to be present and the cochlea is surgically accessible, CI should be attempted first before pursuing ABI. In fact, even though cochlear nerve aplasia is considered to be a contraindication to CI surgery, many would attempt CI prior to ABI, because there may be a nonradiographically identifiable cochlear nerve or alternative connection with the brainstem,

and therefore, if a CI works, the audiometric outcome would likely be superior to ABI.

Particularly relevant to patients with vestibular schwannoma, a CI has a high probability of affording open-set speech recognition in patients with untreated or radiated tumors, whereby the anatomical continuity of the nerve is highly likely.³¹ Single-stage CI and translabyrinthine tumor resection has also been shown to be feasible and safe.^{14,32,33} Results of CI in patients that develop nonserviceable hearing after vestibular schwannoma microsurgery are more unpredictable. If any detectable thresholds are present, this provides concrete evidence that a cochlear nerve is preserved, and these patients can develop useful hearing.³⁴ For patients with complete deafness after microsurgery, a review of the operative note may help to determine whether or not the cochlear nerve was anatomically preserved. The use of promontory stimulation to determine CI candidacy is controversial. An absent response does not exclude the possibility that the patient will derive benefit from a CI.

Future Directions

From the initial discoveries of auditory stimulation by Alessandro Volta in 1790, to reliably achieving open-set speech recognition with multichannel CI electrodes today, the field of cochlear implantation has advanced at an outstanding rate and the technology is truly nothing short of miraculous. And yet the future may witness continued improvements, from alternative stimulation strategies (e.g., radiofrequency, optical), to robotic electrode insertions with steerable arrays, minimally-invasive mastoidotomy techniques, and drug-eluting electrode arrays to deliver steroids for the prevention of intracochlear scarring or neurotrophic factors to promote neural ingrowth for improved electrode to neuron coupling. In addition to technological progress, improvements in health care delivery and awareness campaigns are needed to bring the benefits of cochlear implantation to more people, both in developed and developing countries. Despite the well-established safety and efficacy of CI surgery, and the fact that it is covered by most public and private health insurance carriers in the U.S., less than 6% of people in the U.S., who could benefit from a CI have one.³⁵ By raising awareness of the benefits of CI, educating health-care providers on the expanded indications, developing specific referral pathways, established tele-audiology services, and emphasizing the cost-effectiveness of the intervention, improved utilization, and access to this technology can be realized.

Financial Material and Support

Internal departmental funding was utilized without commercial sponsorship or support.

Conflict(s) of Interest to Declare

Matthew L. Carlson is a consultant for Advanced Bionics Corp., Cochlear Corp., and MED-EL GmbH.

References

- Eshraghi AA, Nazarian R, Telischi FF, Rajguru SM, Truy E, Gupta C. The cochlear implant: historical aspects and future prospects. *Anat Rec (Hoboken)* 2012;295(11):1967–1980
- Gifford RH, Dorman MF, Skarzynski H, et al. Cochlear implantation with hearing preservation yields significant benefit for speech recognition in complex listening environments. *Ear Hear* 2013;34(04):413–425
- Gantz BJ, Turner C, Gfeller KE, Lowder MW. Preservation of hearing in cochlear implant surgery: advantages of combined electrical and acoustical speech processing. *Laryngoscope* 2005;115(05):796–802
- Tajudeen BA, Waltzman SB, Jethanamest D, Svirsky MA. Speech perception in congenitally deaf children receiving cochlear implants in the first year of life. *Otol Neurotol* 2010;31(08):1254–1260
- Sharma A, Gilley PM, Dorman MF, Baldwin R. Deprivation-induced cortical reorganization in children with cochlear implants. *Int J Audiol* 2007;46(09):494–499
- Parry DA, Booth T, Roland PS. Advantages of magnetic resonance imaging over computed tomography in preoperative evaluation of pediatric cochlear implant candidates. *Otol Neurotol* 2005;26(05):976–982
- Ellul S, Shelton C, Davidson HC, Harnsberger HR. Preoperative cochlear implant imaging: is magnetic resonance imaging enough? *Am J Otol* 2000;21(04):528–533
- Holman MA, Carlson ML, Driscoll CL, et al. Cochlear implantation in children 12 months of age and younger. *Otol Neurotol* 2013;34(02):251–258
- Tucci DL, Telian SA, Zimmerman-Phillips S, Zwolan TA, Kileny PR. Cochlear implantation in patients with cochlear malformations. *Arch Otolaryngol Head Neck Surg* 1995;121(08):833–838
- Zeitler DM, Dorman MF, Natale SJ, Loiselle L, Yost WA, Gifford RH. Sound source localization and speech understanding in complex listening environments by single-sided deaf listeners after cochlear implantation. *Otol Neurotol* 2015;36(09):1467–1471
- Hansen MR, Gantz BJ, Dunn C. Outcomes after cochlear implantation for patients with single-sided deafness, including those with recalcitrant Ménière's disease. *Otol Neurotol* 2013;34(09):1681–1687
- Schleich P, Nopp P, D'Haese P. Head shadow, squelch, and summation effects in bilateral users of the MED-EL COMBI 40/40+ cochlear implant. *Ear Hear* 2004;25(03):197–204
- Carlson ML, Breen JT, Driscoll CL, et al. Cochlear implantation in patients with neurofibromatosis type 2: variables affecting auditory performance. *Otol Neurotol* 2012;33(05):853–862
- Ahsan S, Telischi F, Hodges A, Balkany T. Cochlear implantation concurrent with translabyrinthine acoustic neuroma resection. *Laryngoscope* 2003;113(03):472–474
- Carlson ML, Neff BA, Sladen DP, Link MJ, Driscoll CL. Cochlear implantation in patients with intracochlear and intralabyrinthine schwannomas. *Otol Neurotol* 2016;37(06):647–653
- Sanna M, Di Lella F, Guida M, Merkus P. Auditory brainstem implants in NF2 patients: results and review of the literature. *Otol Neurotol* 2012;33(02):154–164
- Modest MC, Carlson ML, Wanna GB, Driscoll CL. Cochlear implantation in patients with superficial siderosis: seven cases and systematic review of the literature. *Otol Neurotol* 2015;36(07):1191–1196
- Shabashev S, Fouad Y, Huncke TK, Roland JT. Cochlear implantation under conscious sedation with local anesthesia; safety, efficacy, costs, and satisfaction. *Cochlear Implants Int* 2017;18(06):297–303
- Reefhuis J, Honein MA, Whitney CG, et al. Risk of bacterial meningitis in children with cochlear implants. *N Engl J Med* 2003;349(05):435–445
- Carlson ML, Driscoll CL, Gifford RH, et al. Implications of minimizing trauma during conventional cochlear implantation. *Otol Neurotol* 2011;32(06):962–968

- 21 Ahmad FI, Demason CE, Teagle HF, Henderson L, Adunka OF, Buchman CA. Cochlear implantation in children with postlingual hearing loss. *Laryngoscope* 2012;122(08):1852–1857
- 22 Palmer CS, Niparko JK, Wyatt JR, Rothman M, de Lissovoy G. A prospective study of the cost-utility of the multichannel cochlear implant. *Arch Otolaryngol Head Neck Surg* 1999;125(11):1221–1228
- 23 Cheng AK, Rubin HR, Powe NR, Mellon NK, Francis HW, Niparko JK. Cost-utility analysis of the cochlear implant in children. *JAMA* 2000;284(07):850–856
- 24 Francis HW, Koch ME, Wyatt JR, Niparko JK. Trends in educational placement and cost-benefit considerations in children with cochlear implants. *Arch Otolaryngol Head Neck Surg* 1999;125(05):499–505
- 25 Koch ME, Wyatt JR, Francis HW, Niparko JK. A model of educational resource use by children with cochlear implants. *Otolaryngol Head Neck Surg* 1997;117(3, Pt 1):174–179
- 26 Carlson ML, Neff BA, Link MJ, et al. Magnetic resonance imaging with cochlear implant magnet in place: safety and imaging quality. *Otol Neurotol* 2015;36(06):965–971
- 27 Gubbels SP, McMenomey SO. Safety study of the Cochlear Nucleus 24 device with internal magnet in the 1.5 Tesla magnetic resonance imaging scanner. *Laryngoscope* 2006;116(06):865–871
- 28 Crane BT, Gottschalk B, Kraut M, Aygun N, Niparko JK. Magnetic resonance imaging at 1.5 T after cochlear implantation. *Otol Neurotol* 2010;31(08):1215–1220
- 29 Edmonson HA, Carlson ML, Patton AC, Watson RE. MR imaging and cochlear implants with retained internal magnets: reducing artifacts near highly inhomogeneous magnetic fields. *RadioGraphics* 2018;38(01):94–106
- 30 Peng KA, Lorenz MB, Otto SR, Brackmann DE, Wilkinson EP. Cochlear implantation and auditory brainstem implantation in neurofibromatosis type 2. *Laryngoscope* 2018
- 31 Pai I, Dhar V, Kelleher C, et al. Cochlear implantation in patients with vestibular schwannoma: a single United Kingdom center experience. *Laryngoscope* 2013;123(08):2019–2023
- 32 DeHart AN, Broaddus WC, Coelho DH. Translabyrinthine vestibular schwannoma resection with simultaneous cochlear implantation. *Cochlear Implants Int* 2017;18(05):278–284
- 33 Rooth MA, Dillon MT, Brown KD. Prospective evaluation of patients undergoing translabyrinthine excision of vestibular schwannoma with concurrent cochlear implantation. *Otol Neurotol* 2017;38(10):1512–1516
- 34 Lloyd SK, Glynn FJ, Rutherford SA, et al. Ipsilateral cochlear implantation after cochlear nerve preserving vestibular schwannoma surgery in patients with neurofibromatosis type 2. *Otol Neurotol* 2014;35(01):43–51
- 35 Sorkin DL. Cochlear implantation in the world's largest medical device market: utilization and awareness of cochlear implants in the United States. *Cochlear Implants Int* 2013;14(Suppl 1):S4–S12