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REVIEW

Cochlear Implantation with Labyrinthectomy: Indications, Considerations, and Outcomes

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Abstract

Purpose of Review The purpose of this review is to examine current evidence concerning cochlear implantation in the setting of previous or simultaneous labyrinthectomy.

Recent Findings Recent data support the use of cochlear implants (CIs) in certain settings where labyrinthectomy is indicated. Clinical situations where this was examined include refractory Meniere's disease, inner ear schwannoma, and vestibular schwannoma. For Meniere's disease and inner ear schwannoma, data support outcomes that are on par with general CI recipients. Implantation in the setting of labyrinthectomy for vestibular schwannoma is less consistent; however, simultaneous cochlear implantation with labyrinthectomy provides unique options for nerve monitoring in these patients.

Summary CI is a viable strategy for hearing rehabilitation among patients with vestibular disease or vestibular schwannoma in cases where management incorporates labyrinthectomy. The use of CIs in these settings requires unique considerations given risk of cochlear ossification, potential partial cochlectomy for tumor access, manipulation of the cochlear modiolus or cochlear nerve, and the ability to use CIs for cochlear nerve monitoring during vestibular schwannoma resection.

Keywords Cochlear implant · Labyrinthectomy · Meniere's disease · Schwannoma

Introduction

Certain otologic conditions are managed preferentially or by necessity with labyrinthectomy. Violation of the otic capule in this manner almost universally yields profound sensorineural hearing loss. [1] Therefore, management of such conditions requires careful patient counselling and consideration of both the therapeutic benefit of intervention and the resulting sensorineural deafness.

Cochlear implants (CIs) offer a modality by which sound and speech perception can be "salvaged" in patients with sensorineural hearing loss in the setting of labyrinthectomy. [2] Provided an intact auditory nerve is present with sufficient health to propagate an electrical stimulus, patients can undergo labyrinthectomy and still access sound via electrical stimulation with a CI. [2] This review will consider recent literature analyzing CIs in the setting of labyrinthectomy, for management of intractable Meniere's disease, inner ear schwannoma, and vestibular schwannoma. This review will also consider clinically noteworthy factors such as timing of implantation in the setting of labyrinthectomy given attendant risk of ossification, and MRI surveillance with a CI in place.

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Historical Perspective

Historically, an appropriate CI patient would have a patent and undamaged otic capsule, as it was thought that labyrinthine trauma would potentiate spiral ganglion cell loss and portend a nonfunctional or poorly functioning implant. [3] However, in 1988 Gantz et al. [4] reported on the successful implantation of patients requiring skeletonization of the modiolus due to cochlear ossification, and, soon after, Kvetan et al. [5] reported on the successful implantation of a patient who had undergone labyrinthectomy for temporal

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bone adenocarcinoma. These reports, along with supporting histopathological studies showing survival of spiral ganglion cells after otic capsule surgery, spurred consideration of implantation in patients having undergone or requiring labryinthectomy [3]. In 1993, Zwolen et al. [6] published on simultaneous implantation with labyrinthectomy for the treatment of refractory Meniere's disease. And in 1992, Hoffman et al. [7] reported on the successful implantation after labyrinthectomy for resection of a vestibular schwannoma. As CI candidacy criteria have expanded, particularly for patients with single-sided deafness, so has the use of CIs in patients undergoing labyrinthectomy. In this review, we will discuss current literature on the three main patient populations undergoing labyrinthectomy with implantation, namely, those with refractory Meniere's disease, vestibular schwannoma, or inner ear schwannoma, and we will consider the benefits, limitations, and special considerations of each clinical scenario.

Current Use/Indications

Meniere's Disease

Meniere's disease is a disorder of the inner ear that yields fluctuating hearing loss, vertigo, tinnitus, and fullness in the involved ear. While treatment algorithms for Meniere's disease focus on hearing-sparing medical and surgical therapy, primarily for the treatment of vertigo, many patients with irretractable vertigo or significant hearing loss due to the natural course of the disease with persistent vertigo have undergone labyrinthectomy for effective vertigo management. [8] However, disruption of the otic capsule in this manner leads to profound ipsilateral sensorineural hearing loss. CIs offer a way to rehabilitate hearing in patients with Meniere's disease who have undergone such hearing-ablative therapies.

A recent systematic review by Selleck et al. demonstrated improvement in word recognition by an average of 23% after cochlear implantation in a cohort of patients treated with labyrinthectomy for Meniere's disease. [9] As such, CI outcomes in this population are favorable, if somewhat inferior to the normal implant population. Much of this literature has evaluated patients with some degree of bilateral hearing loss and Meniere's disease affecting at least one side; however, in a small series of patients who had implantation simultaneous with labyrinthectomy for Meniere's disease and normal contralateral hearing, Perkins et al. demonstrated improvements in speech recognition, sound localization, tinnitus handicap, and quality of life at 6 months after activation. [10]

One may postulate that less traumatic ablative therapies may yield improved CI outcomes in patients with Meniere's disease. For example, chemical ablation can remove vestibular function while not disrupting the otic capsule or placing key auditory pathway structures at risk. As such, better CI outcomes may be expected in those undergoing implantation after instillation of gentamicin or other ototoxic medications as opposed to labyrinthectomy. However, Chien et al. demonstrated in a series of 29 patients (34 implants) that there was no significant difference between CI outcomes in patients undergoing surgical labyrinthectomy versus chemical labyrinthectomy versus endolymphatic sac decompression. [11] Of note, the authors acknowledged that the sample size for the non-labyrinthectomy cohorts was lower than anticipated for appropriate statistical power. Given this, further study is needed to analyze CI performance after various techniques used for management of refractory Meniere's disease. At this time, outcomes in patients treated with surgical labyrinthectomy and implantation do appear at least as favorable as outcomes in CI recipients with Meniere's disease treated in a nonsurgical fashion.

While modern outcomes with cochlear implantation and labyrinthectomy in Meniere's disease are favorable, certain factors must be considered, primarily the status of the contralateral ear. Should the contralateral ear have Meniere's disease or otherwise have a compromised vestibular system, a patient may be steered away from surgically ablative procedures, as complete ablation of bilateral vestibular systems would yield debilitating vestibular hypofunction. [8] To this end, in such patients where the contralateral inner ear is dysfunctional or at risk of dysfunction, medical management or non-ablative surgical management (e.g., endolymphatic sac decompression) of Meniere's disease would be favored by the authors. Cochlear implantation could still be pursued should hearing be lost in the course of the disease.

Vestibular Schwannoma

Vestibular schwannomas, also referred to as acoustic neuromas, are benign neoplasms of the eighth cranial nerve complex. Management entails serial observation, radiosurgery, or operative intervention, with a general goal of preserving acoustic and cranial nerve function as long as or to the greatest degree possible. As part of the treatment algorithm, many patients with non-serviceable hearing or with large tumors may require or prefer translabyrinthine resection of their tumor, almost universally resulting in complete ipsilateral hearing loss.

Traditionally, auditory brainstem implants (ABIs) have been offered for patients with neurofibromatosis type 2-related schwannomatosis (NF2), a condition where bilateral vestibular schwannomas usually develop. However, studies have shown that ABI function is generally poor, with only a relatively small number of patients obtaining any open-set speech recognition ability. [12] On the other hand, more recent data indicates that cochlear implantation in the setting of vestibular schwannoma has been shown to outperform ABI and thus represents an alternative modality for auditory rehabilitation in select patients with a preserved auditory nerve. [13]

While generally more successful than ABI, CI outcomes in vestibular schwannoma patients are more variable than traditional implant recipients. [14–16] This is likely because the auditory nerve is at increased risk of damage, mainly in the population with large tumors who undergo surgical resection. Case-in-point, a recent multi-institutional review of 100 cases of cochlear implantation in sporadic vestibular schwannoma demonstrated that patients treated with surgical resection have poorer CI outcomes compared to those treated with nonsurgical means, obtaining open-set speech in 57% of cases compared to 94% and 80% of cases for those treated with observation and radiosurgery, respectively. [in press] A recent systematic review of surgically treated vestibular schwannomas by Wick et al. mirrors these findings. [17] While most CI recipients in this setting are daily users of their devices, only slightly more than half obtained open-set speech discrimination. These studies highlight the importance of minimizing injury of the cochlear nerve if successful cochlear implantation be desired.

To this end, as described in several prior publications by the senior author, commercial CIs can be used to monitor cochlear nerve integrity during translabyrinthine tumor resection, via either electrically-evoked auditory brainstem responses (eABR) or observing an electrically evoked stapedial reflex (eSRT) [18, 19]. In this manner simultaneous cochlear implantation may help to avoid injury to the auditory nerve, or, at the very least, prognosticate CI function after tumor resection. [18–24] First described by Lassaletta et al. [25], other authors have used a MED-EL test electrode during tumor resection to monitor eABR signal to inform decision-making regarding CI placement at the end of tumor resection.

While cochlear implantation in vestibular schwannoma may be a valuable tool for auditory rehabilitation, we must consider the impact the CI has on further schwannoma care. In our experience, imaging artifact from the internal cochlear implant device rarely inhibits successful imaging surveillance of the ipsilateral internal auditory canal. [26, 27] However, placement of the device in more posterosuperior location under the temporoparietal scalp may effectively draw the artifact further from the tumor site. Newer MRI-conditional cochlear implant systems offer greater artifact reduction, reduced complication risk, and improved patient comfort during MRI. [26] An additional consideration is the need for radiosurgery after cochlear implantation, when targeting is more complicated secondary to image distortion and artifact. For this reason, if radiosurgery is being considered, it may be preferential to have this performed prior to cochlear implant placement.

Inner Ear Schwannoma

In addition to vestibular schwannoma and other internal auditory canal/cerebellopontine angle tumors, we must also consider inner ear schwannomas. Paralleling vestibular schwannoma, inner ear schwannoma can often be managed conservatively with observation, particularly when residual functional acoustic hearing is present. [28] However, given the location of the disease and subtle radiological findings, patients often present late in disease with non-serviceable hearing or refractory vertigo, which may prompt consideration for a CI. Successful cochlear implantation has been demonstrated in patients with inner ear schwannoma left in situ, thus allowing for continued conservative management. [29] In cases such as growing tumors, uncontrollable vestibular symptoms, or patient/surgeon preferences, surgical resection may be undertaken. While these tumors do not always necessitate a formal labyrinthectomy, surgical resection does require at least some degree of otic capsule violation, ranging from a partial cochlectomy or labyrinthotomy to near-total resection of the otic capsule depending on the location and extent of the disease.

Fortunately, schwannomas isolated to the inner ear generally show favorable CI outcomes after tumor resection. A number of small reports have demonstrated successful CI use after limited cochleostomy or cochlectomy and resection of inner ear schwannoma, with open-set speech recognition reported in a vast majority of patients. [30–34] Plontke et al. [31, 35–37] have published several manuscripts detailing CI placement after more extensive subtotal cochlectomy for tumor resection. In these series, they report rates of open-set speech recognition approaching 100%. [31, 35–37] In fact, monosyllabic word recognition was equal or even better than a matched historical cohort of normal CI recipients, a fact that the team considered may be related to reduced electrical spread after cochlectomy. [38] In these cases, extensive otic capsule resection may be undertaken, and as long as the modiolus and cochlear nerve are preserved, patients often receive excellent cochlear implant outcomes.

These findings are in contrast to vestibular schwannomas, or inner ear schwannomas with transmodiolar growth that, as detailed above, may only have ~ 60% chance of open set speech recognition if treated with gross total tumor resection and cochlear implantation. This is likely secondary to the fact that in most inner ear schwannomas, the auditory nerve is not placed in direct risk during surgical resection, excluding those with transmodiolar spread to the IAC.

One major consideration in this patient population is the degree of otic capsule resection necessary for tumor removal and implant design/selection. Many studies utilized cochleostomy or limited cochlectomy with the tumor resected via various “push-through” and “pull-through” methods that utilized suture filament, absorbable foam pads, or even the cochlear implant itself to mobilize the tumor and push/pull it out through a small opening. [30–34] In this manner, trauma to the otic capsule is theoretically limited; however, such techniques may risk leaving tumor behind. A more significant otic capsule resection may help to ensure total tumor removal. However, this may make accurate placement of a CI electrode challenging as some or all of the bony lumen that normally holds the implant in place may be absent. To counter this, one may choose to use a precurved electrode so that the device remains in close approximation to the spiral ganglion for electrical stimulation. One can also use materials such as bone pate or small pieces of cartilage to secure the electrode in place. Plontke et al. demonstrated in their study the use of a perimodiolar-malleable electrode that is held in place with cartilage and perichondrium grafts, with or without bone pate and fibrin glue. [37]

Special Considerations

When considering cochlear implantation in the setting of labyrinthectomy, one must consider issues related to timing of implantation and the use of imaging surveillance after implantation. With regard to timing of implantation relative to labyrinthectomy, two issues must be considered. First is ossification secondary to otic capsule violation. Violation of the otic capsule can yield a robust inflammatory and fibrotic response that can lead to obliteration of the lumen of the cochlea. Per Feng et al., partial obliteration of the cochlear lumen can be expected in up to 70% of patients treated via labyrinthectomy for vestibular schwannoma at roughly 3 months post-surgery and up to 80% at roughly 3 years. [39] Obliteration can prohibit cochlear implantation or yield an incomplete insertion that may portend poor outcomes. To this end, simultaneous labyrinthectomy with cochlear implantation is preferred at the authors' institution. A secondary option includes placement of a dummy electrode at time of resection to facilitate subsequent electrode placement if cochlear implantation is pursued at a later date.

The second issue related to timing is the ability to use a CI as a cochlear nerve monitor. As mentioned earlier, commercial CIs and investigational test electrodes can be used to elicit eABR and eSRTs that may provide relatively real-time assessment of nerve integrity. [18–24] By performing cochlear implantation simultaneously with translabyrinthine resection of vestibular schwannomas, surgeons can theoretically use this monitoring capability to avoid injury to the cochlear nerve and prognosticate future implant outcomes. However, testing in this manner can have a false-negative result, and until better data is available, should not necessarily preclude implantation, especially if the cochlear nerve is demonstrably intact at the conclusion of tumor resection. [40]

Finally, one must consider the impact that a CI can have on imaging. MRI can be performed at low risk for patients with MRI conditional magnets. A recent publication by Johnson et al. [26] found that in 153 instances of MRI with MRI conditional magnets in place, no magnets were displaced and imaging was never terminated early due to discomfort. However, even among subjects with older models containing conventional axial magnets, imaging can be obtained under special imaging protocols [27]. One must also consider the impact the device and magnet will have on imaging resolution. Even with MRI conditional magnets, CIs can produce a large artifact in many MRI sequences. [25] That can make surveillance challenging, and any further treatment, such as salvage radiosurgery, may require magnet removal for accurate targeting. That said, modern MRI sequences, such as a multi-acquisition with variable resonance image combination (MAVRIC) sequence, have been employed successfully in reducing artifacts from CIs and other implantable devices. [25] When considering implantation in the setting of an vestibular schwannoma, collaboration with an experienced neuroradiology team is beneficial to ensure good patient care.

Conclusion

CI is a viable strategy for hearing rehabilitation among patients with vestibular disease or vestibular schwannoma in cases where management incorporates labyrinthectomy. For use in Meniere's disease and inner ear schwannoma, cochlear implantation can provide speech recognition at or close to levels anticipated in normal implant recipients while allowing for definitive disease management. The use of CIs in vestibular schwannoma treated via translabyrinthine resection may yield poorer hearing outcomes than similar disease treated by observation or radiosurgery but remains a superior alternative when feasible, compared to other available hearing restoration options such as an ABI. The use of CIs in these settings requires unique consideration to the timing of implantation with respect to labyrinthectomy and the impact the CI may have on surveillance for some disease processes.

Key References

- Selleck AM, Dillon M, Perkins E, Brown KD. Cochlear implantation in the setting of Menière's disease after labyrinthectomy: a meta-analysis. Otol Neurotol. 2021;42(8):e973–9.

This meta-analysis evaluates audiologic outcomes in patients who undergo simultaneous cochlear implantation and ablative therapy with labyrinthectomy for unilateral Meniere's disease. The study demonstrated a statistically significant improvement in both speech recognition and sound localization, though noted that the degree of improvement may be less in this cohort compared to traditional CI recipients.

- Lassaletta L, Polak M, Huesers J, Díaz-Gómez M, Calvino M, Varela-Nieto I, Gavilán J. Usefulness of electrical auditory brainstem responses to assess the functionality of the cochlear nerve using an intracochlear test electrode. Otol Neurotol. 2017;38(10):e413–20.

This prosective cohort study assesses the viability of a test cochlear electrode that can be use to assess cochlear nerve integrity prior to cochlear implantation. In this study 10 cochlear implant candidate had a test electrode temporarily placed prior to cochlear implantation. eABR was tested using both the test and cochlear electrode. Findings were consistent between both devices, and all subjects attain useful stimulation from this implant.

- Fussell WL, Patel NS, Carlson ML, Neff BA, Watson RE, Lane JI, et al. Cochlear implants and magnetic resonance imaging: experience with over 100 studies performed with magnets in place. Otol Neurotol. 2021;42(1):51–8.

This large retrospective series evaluates the safety and feasibility of MRI in patients with cochlear implants. The study identified a low prevalence of adverse outcomes, while including a discussion of techniques that can be utilized to minimize imaging artifact in this subset of patients.

- Carlson ML, Neff BA, Sladen DP, Link MJ, Driscoll CL. Cochlear implantation in patients with intracochlear and intralabyrinthine schwannomas. Otol Neurotol. 2016;37(6):647–53.

This case series examines CI outcomes in patients with primary inner ear schwannomas who underwent implantation without tumor resection. The majority of patients achieved open-set word recognition. The study recommends use of a styleted electrode to facilitate full insertion through an obstructing tumor with a late deployment aided advancement technique.

- Plontke SK, Fröhlich L, Cozma S, Koitschev A, Reimann K, Weiß R, et al. Hearing rehabilitation after subtotal cochleoectomy using a new, perimodiolar malleable cochlear implant electrode array: a preliminary report. Eur Arch Otorhinolaryngol Off J Eur Fed Otorhinolaryngol Soc (EUFOS) Affiliated German Soc Otorhinolaryngol Head Neck Surg. 2021;278(2):353–62.

This case series evaluates the feasibility of implanting a custom malleable perimodiolar electrode after primary inner ear tumor removal via subtotal cocheloectomy. The majority of patients experienced significant improvements in sound and speech recognition.

- Feng Y, Lane JI, Lohse CM, Carlson ML. Pattern of cochlear obliteration after vestibular Schwannoma resection according to surgical approach. Laryngoscope. 2020;130(2):474–81.

This case series investigates the pattern of cochlear obliteration that results from microsurgical resection of vestibular schwannoma via translabyrinthine, retrosigmoid or middle fossa approaches. The study identified a variable time and prevalence of cochlear obliteration between surgical approaches, and found translabyrinthine approaches to have the highest risk of complete obliteration.

Author Contributions JRD, EEB, and MLC contributed to the review of the literature, writing of the manuscript, and critical review of its contents.

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Data Availability No datasets were generated or analysed during the current study.

Declarations

Conflict of Interest JRD receives research support from Cochlear Americas and Advanced Bionics.
MLC receives research support from Cochlear Americas, Advanced Bionics, and MED-EL.

Human and Animal Rights and Informed Consent All reported studies/experiments with human or animal subjects performed by the authors have been previously published and complied with all applicable ethical standards (including the Helsinki declaration and its amendments, institutional/national research committee standards, and international/national/institutional guidelines), and, where necessary, informed consent was obtained.

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