Cochlear Implant Outcomes in Patients with Vestibular Schwannoma

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Abstract

Objectives:
1. Describe audiologic outcomes in patients after cochlear implantation on the side with a vestibular schwannoma.
2. Describe indications for consideration of cochlear implantation in the vestibular schwannoma population.

Methods:
Study Design: Retrospective Review
Years Reviewed: 1995-2016
Disease Studied: Vestibular schwannoma
Condition Studied: Sensorineural hearing loss
Subjects studied: 5
Setting: Tertiary Academic Center
Intervention: Cochlear Implantation
Outcome Measures: Pre and Postoperative word and sentence testing scores, duration of implant benefit

Abstract

Introduction
Vestibular schwannoma (VS), whether sporadic or associated with Type 2 Neurofibromatosis (NF2), presents several unique challenges for the lateral skull base surgeon. Determination of appropriate treatment, whether surgical, systemic (bevacizumab), radiation-based, or observatory is a process wrought with complexity. One of the many factors influencing management of vestibular schwannoma patients is the concept of hearing preservation rehabilitation. When VS affects the only hearing ear, hearing rehabilitation poses several unique challenges of its own. In the past, options for hearing rehabilitation after tumor excision were limited to auditory brainstem implantation, which traditionally offers hearing improvement but limited achievement of open set speech discrimination. More recently, published data indicate that despite the retrocochlear location of VS, cochlear implants can be used to significantly improve hearing after surgical resection of VS with auditory nerve preservation, or after targeted radiation treatment.1,4

There are occasional patients who experience deafness due to VS but require no treatment for the tumor. Very little is known about the utility of cochlear implantation in the setting of ipsilateral untreated VS. In 2009, Helbig et al. (Frankfurt, Germany) first described implantation of a patient with an untreated sporadic VS. The contralateral ear (no VS but profound SNHL) was implanted first. The ear involved by tumor was sequentially implanted. Six months after surgery, the patient attained 80% speech recognition scores on monosyllabic testing, and the ear involved by tumor matched the results of the non-tumor side.2 In a 2013 multicenter (Great Britain, Australia) retrospective study, Mukherjee et al. published a series of eleven VS patients who received cochlear implants; three of these had implants placed on the side ipsilateral to an untreated tumor, and each experienced substantial hearing improvement.5

According to the above-mentioned studies, which discuss four patients in total, cochlear implants can provide hearing benefit in patients with untreated tumors. Gaps in knowledge and understanding still remain, however. One fear in this scenario is that implantation may not provide a lasting, durable benefit due to presence of untreated, slowly growing schwannoma. Another concern is that cochlear implants may not work as well in the setting of untreated VS, leading to less than ideal speech discrimination.

The present series of five NF2 patients includes two who underwent cochlear implantation after radiation of an ipsilateral schwannoma, and three who received cochlear implants ipsilaterally to untreated tumors. Regarding the three patients who received cochlear implants on the side ipsilateral to untreated tumors, this is the first published report of this practice in the United States. The information presented here calls attention to both the quality and durability of auditory rehabilitation experienced by vestibular schwannoma-bearing cochlear implant recipients these patients despite significant tumor growth after implantation.

Results

Table 1: Characteristics of patients included in series. All series patients suffered from NF2 and had bilateral vestibular schwannomas. Patient age at time of implantation ranged from 13 to 63. Maximum time since implantation was 6.5 years (Patient 3). All patients were everyday CI users at time of study end. Patient 1 and Patient 3 both experienced significant tumor growth after cochlear implantation.

Table 2: Treatment history and auditory measures for patients included in series. Two patients underwent gamma knife radiation therapy for vestibular schwannomas ipsilateral to the implanted ear. In the other three patients, the tumor involving the implanted ear was untreated at the time of implantation and remained untreated throughout the study timeframe.

Discussion
Over the last ten years, cochlear implantation has been demonstrated to be a safe and effective means of auditory rehabilitation in patients with VS. The proof of concept, and the more general acceptance of the practice that followed, came about through index cases and small case series involving patients implanted on the side ipsilateral to a previously treated (surgery or radiation) VS. Two of the patients in the present series add to the growing number of successes in this specific subset of difficult-to-manage patients.

The remaining three patients described herein underwent cochlear implantation on a side involved by an untreated VS. This practice is far less commonly cited in scientific literature than the former. In 2009, Helbig et al. (Germany) first described implantation of a patient with an untreated sporadic VS. The contralateral ear (no VS but profound SNHL) was implanted first. The ear involved by tumor was sequentially implanted. Six months after surgery, the patient attained 80% speech scores on monosyllabic testing, and the ear involved by tumor matched the results of the non-tumor side.1 In a 2013 multicenter (Great Britain and Australia) retrospective study, Mukherjee et al. published a series of eleven VS patients who received cochlear implants; three of these had implants placed on the side ipsilateral to an untreated tumor, and each experienced substantial hearing improvement.5

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Conclusions

- As previously published by several groups, deafened vestibular schwannoma patients with previously radiated tumors can experience significant hearing restoration with cochlear implants.
- Deafened VS patients whose tumors have not been treated (no radiation or surgery) can also experience significant, durable hearing restoration via cochlear implantation.
- Vestibular schwannoma patients with untreated tumors ipsilateral to the implanted ear can experience marked hearing improvement that persists for many years despite significant tumor growth.

References

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