Vestibular schwannomas (VSs) are benign and slow-growing tumors that correspond to approximately 6% of all intracranial neoplasms. These lesions may lead to a varied clinical profile, including hearing loss, tinnitus, imbalance, vertigo, facial paralysis, sensory changes associated with the trigeminal nerve, and, not infrequently, hydrocephalus and compression of the brainstem, especially in large lesions.4,16

Hearing loss is the most common symptom of vestibular schwannomas (VSs). The management of these lesions includes observation, radiosurgery, and microsurgical resection. Hearing preservation and rehabilitation are the major challenges after the tumor treatment. A 43-year-old man with previous left-sided profound hearing loss and tinnitus presented with a 2-mm left-sided intracanalicular VS. The decision was made to perform a simultaneous cochlear implantation (CI) and microsurgical resection of the tumor. The patient did well postoperatively, with significant improvement of tinnitus, sound localization, and speech recognition in noise. Previous reports of simultaneous CI and VS resection in patients with neurofibromatosis type 2 and sporadic VS in the only hearing ear have been described. The role of CI in patients with VS and normal contralateral hearing has been recently described, showing positive outcomes due to the binaural benefits. Tinnitus also can be treated by the implantation of the cochlear device. The simultaneous microsurgical removal of VS and implantation of a cochlear device is a feasible approach in patients with unilateral hearing loss and severe tinnitus.


KEY WORDS acoustic neuroma; hearing; cochlear implantation; vestibular schwannoma

Case Report

History
This 43-year-old man presented with a 30-year-history of unilateral left-sided deafness attributed to mumps infection and tinnitus. He was treated by the otolaryngology team for tinnitus, and an attempt was made to recover his hearing. The patient did not adapt to a bone-anchored
hearing aid. In a routine MRI study we found a 2-mm intracanalicular VS (Fig. 1). Management options including observation with serial imaging, radiation, and resection were discussed. A cochlear implant was suggested, and a simultaneous tumor resection could be performed.

Examination
Audiometric testing demonstrated good hearing in the right ear. However, the left ear presented profound sensorineural hearing loss.

Operation
We routinely perform a conventional retrosigmoid approach for most VSs; however, because the patient already presented with complete hearing loss and the surgical goal was also the placement of a cochlear implant, we adopted a transmastoid translabyrinthine approach. Figure 2 illustrates the procedure stepwise. The surgical procedure was performed under electroneurophysiological monitoring. The cochlear implant was manufactured in Sydney, Australia (Freedom Implant CI24RE—Contour Advance; Cochlear Corp.).

Impedance and neuronal response telemetry was performed after the cochlear implant placement, indicating correct position of the cochlear implant and integrity of the cochlear nerve (Software Custom Sound EP—4.4; Cochlear Corp.).

Postoperative Course
The patient did well, with fitting of the sound processor 40 days after surgery with normal findings on impedance and neuronal response telemetry. The mapping strategy was to obtain progressive gain, which facilitates the patient’s adaptation. The CI mapping was conducted 30 days after activation and showed normal values for impedance and neuronal response measurements. The patient had a significant improvement in tinnitus, sound localization, and sound discrimination.

The speech recognition tests before implantation were all negative (0%). After implantation, the accuracy average in the Ling Sound Test for detection and discrimination increased to 54.2%. The identification of vocabulary extension was 50%. The patient will continue the hearing rehabilitation with speech therapy every 4 months until he achieves total adaptation. He also will receive vestibular rehabilitation, which is necessary after the translabyrinthine approach.

Discussion
The Role of CI
In the 1960s, several attempts to stimulate the cochlear nerve to treat deafness were performed. Years later, a multichannel implant formed by electrodes stimulating the cochlea allowed the perception of different frequencies. The present cochlear implant is an evolution of this device. Composed of a microphone, signal processor, transmission system, and multiple electrodes, the sound is captured, processed, and transmitted, causing electrical stimulation at the cochlear nerve.

Simultaneous CI in the management of VS was first described in 1995 by Arriaga and Marks. Since then, they have been considered in a few cases in which neurofibromatosis type 2 and sporadic VS were found in the only hearing ear. Under these conditions, the risk of complete hearing loss, either due to the treatment or from the natural course of the disease, requires more effort to provide a satisfactory result. Previous studies show a positive outcome in patients with sporadic VS in the only or better hearing ear, with similar prognosis to that in patients without VS who were treated with CI. Cochlear implants after radiosurgery in patients with neurofibromatosis type 2–associated VS have been described, with significant restoration of hearing.

However, the lack of knowledge about the benefits of CI in patients with normal contralateral hearing still exists. The first study showing the results of a series of patients with this condition who underwent simultaneous CI and tumor resection was published in 2016, with 13 patients. Sanna et al., evaluated the cochlear implant in monaural conditions, but also assessed the binaural benefit perceived by the patient. Binaural benefits are an important aspect to the patient’s quality of life that should be considered in addition to the auditory gain.

The audiological impact of sporadic VS in patients with normal contralateral hearing is mainly the loss of binaural hearing. This comprises a diminished summation effect (identical signal arriving at both ears), a reduction of the squelch effect (ability of the brain to separate noise and
speech coming from different locations), and the head shadow effect (speech discrimination when the head is between the source of the sound and the hearing ear). CI presented as a good option to recover these deficits, providing benefits in conditions like squelch effect and sound localization.\textsuperscript{11,25}

Hassepass et al. demonstrated in their 11-patient series a similar outcome regarding benefits associated with binaural hearing, and they suggested another advantage of CI after translabyrinthine VS surgery: tinnitus suppression.\textsuperscript{5,18} However, the implantation was performed 1 year after tumor resection. The late implantation may be controversial due to the induced postsurgical ossification of the cochlea in the translabyrinthine approach, recommending an intracochlear placeholder to allow later cochlear implant placement. Some studies observed total cochlear ossification, with subtotal degeneration of spiral ganglion cells and cochlear nerve.\textsuperscript{3,9} The main advantage of the simultaneous procedure is to avoid these anatomical obstacles.

Hearing preservation is the major challenge of surgical treatment because complete hearing loss and hypoacusis affect approximately 90% of patients.\textsuperscript{19} This fact justifies the concern about hearing preservation and rehabilitation. The remarkable success of cochlear implants had been a game changer in the rehabilitation of these patients and emerges as a therapeutic option in cases in which there is a low chance of hearing preservation, even with normal hearing in the contralateral ear.\textsuperscript{24} So, the preservation of the cochlear nerve can allow hearing restoration. However, we need more studies demonstrating the results of this approach.

Conclusions

The simultaneous microsurgical removal of VS and CI is a feasible approach to patients with unilateral hearing loss and severe tinnitus. In this situation, the translabyrinthine approach is preferred instead of the retrosigmoid approach because it is easier to implant the cochlear device. In patients with serviceable preoperative hearing we advocate the retrosigmoid approach, with the intention of preserving hearing. When it is not possible to achieve hearing preservation, a second-stage procedure can be done to implant the cochlear device.

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**Disclosures**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**Author Contributions**

Conception and design: Borba, dos Santos Neto. Acquisition of data: Borba, dos Santos Neto, Zamponi, Hamerschmidt, Wiemes. Analysis and interpretation of data: Borba, dos Santos Neto, Zamponi, Hamerschmidt, Wiemes. Drafting the article: dos Santos Neto. Critical revising the article: Rassi. Approved the final version of the manuscript on behalf of all authors: Borba. Study supervision: Borba.

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