CASE REPORT
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Restoring Hearing After Resection of Vestibular Schwannoma by Cochlear Nerve Preservation and Cochlear Implantation: Long-Term Follow-Up of Two Cases

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ABSTRACT

Hearing outcomes of two cases of growing sporadic vestibular schwannoma, resected via a translabyrinthine approach with simultaneous cochlear implantation are reported. After gross total resection and anatomical preservation of the facial and cochlear nerve, the integrity of the cochlear nerve—on an electrophysiological level—was evaluated using the intracochlear test electrode of the Auditory Nerve Test System. After confirming electrically-evoked auditory brainstem recordings, cochlear implantation and hearing rehabilitation were performed as per the single-sided deafness protocol. This report describes the audiological outcome with respect to speech understanding in quiet and noise, localization of sounds as well as phoneme discrimination up to one year after surgery.

Keywords: Vestibular schwannoma, cochlear implantation, cochlear fibrosis

Introduction

Sporadic vestibular schwannomas (VS) are benign tumors arising from the Schwann cells of the vestibulocochlear nerve and have an estimated incidence of 19-42 per million.[1,2] Most symptoms of VS tend to develop progressively and mainly include sensorineural hearing loss (SNHL), tinnitus, and vertigo. Depending on the symptomatology, location, volume, and growth pattern of the VS, different treatment options are available, including a wait-and-scan policy to observe the natural evolution of the tumor, microsurgical tumor resection, or stereotactic radiotherapy aiming to stop tumor growth.[3]

There are three main surgical approaches for VS resection, chosen based on factors such as tumor size, location, preoperative hearing level, and hearing preservation options. The translabyrinthine approach sacrifices acoustic hearing and is suitable for patients with poor preoperative hearing or limited hearing preservation options. The retrosigmoid approach offers a wide view of the cisternal tumor component and allows for preservation of inner ear structures. However, it may require cerebellar retraction and limit access to the facial and cochlear nerves in the distal internal auditory canal (IAC). The middle fossa approach targets small tumors primarily located within the IAC from a superior trajectory. Its disadvantages include placing the facial nerve between the surgeon and the tumor, along with some retraction on the temporal lobe, carrying risks of postoperative seizures and speech disturbances.[4,5]

Signal intensity in preoperative T2-weighted magnetic resonance imaging (MRI) has been reported as a prognostic marker for predicting postoperative hearing loss. More specifically, reduced T2-weighted signal in the cochlea and/or fundus already highlights reduced odds of functional hearing preservation.[6] However, even with anatomical preservation of the cochlear nerve, functional hearing preservation is often limited.

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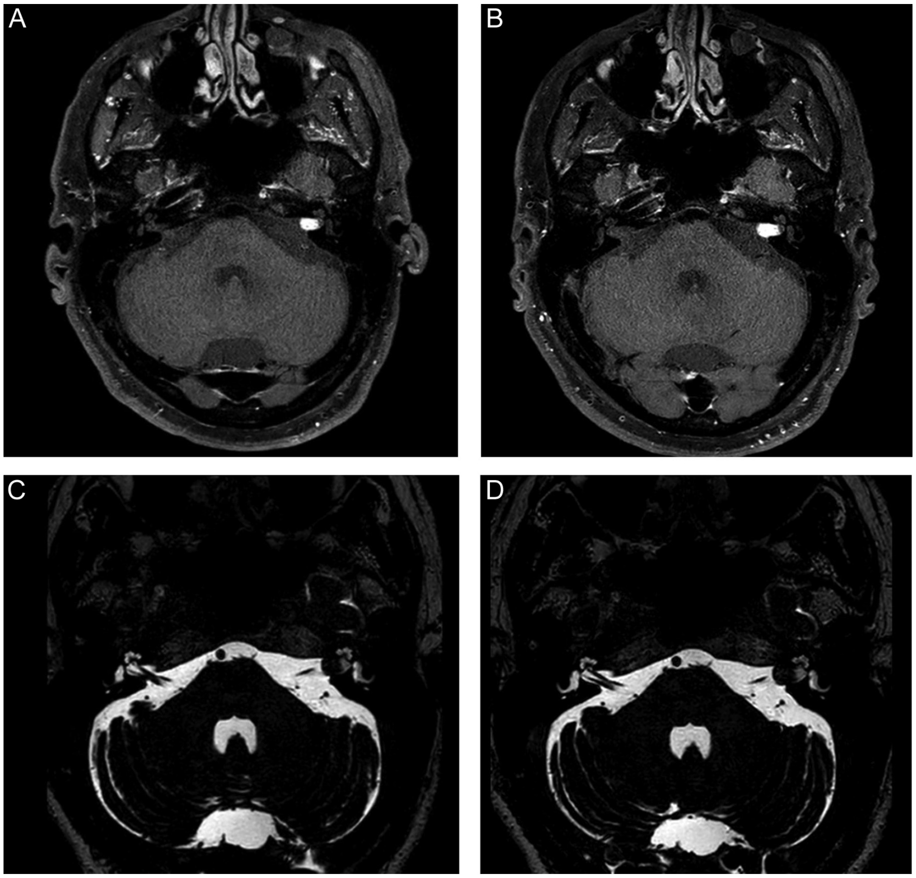

Figure 1. A-D. Magnetic resonance images of the cerebellopontine angle. Initial MRI (A and C) vs. follow-up images at 6 months (B and D). Images A and B are 3-dimensional T1-weighted black blood sequences. Images C and D are T2 drive sequences.

Although anatomical preservation of the cochlear nerve is possible in all surgical approaches, the only approach that enables simultaneous intraoperative access to the cochlea for cochlear implantation is the translabyrinthine approach.

We have observed that patients with unilateral severe-to-profound SNHL or single-sided deafness (SSD) may suffer from reduced speech perception in noise, impaired sound localization, reduced health-related quality of life, and ipsilateral incapacitating tinnitus.[7,8] In case of deafness after VS resection, hearing rehabilitation is limited to the use of contralateral routing of signal (CROS or BiCROS) or bone conduction hearing devices to provide CROS. Neither of the CROS/BiCROS modalities will allow for binaural hearing. Moreover, there is a risk for cochlear fibrosis after microsurgical resection, which would obviate cochlear implantation at a later stage.[9]

Main Points

- Although anatomical preservation of the cochlear nerve is possible during microsurgical resection of vestibular schwannoma, functional preservation is often limited.
- The translabyrinthine approach enables access to the cochlea for the insertion of an intracochlear test electrode to perform electrically-evoked auditory brainstem recording (eABR).
- Intraoperative eABR—using the Auditory Nerve Test System—has the ability to accurately predict auditory perception with cochlear implants after vestibular schwannoma resection.
- Cochlear implantation can be performed during the same intervention and should be considered as a potential tool to restore hearing to some level.

Intraoperative electrically-evoked auditory brainstem recording (eABR), using Auditory Nerve Test System (ANTS), allows us to intraoperatively predict auditory perception with a cochlear implant (CI) quite accurately.[10] Literature supports the ability of cochlear implantation in SSD to restore binaural cues and the associated positive effect on speech perception in noise and sound localization.[11,12]

In this report, we describe the audiological outcome with respect to speech understanding in quiet and noise, localization of sounds as well as phoneme discrimination up to one year after surgery.

Case Presentation

Case 1

A 46-year-old man presented at the Ear, Nose, and Throat (ENT) clinic with subjective hearing loss, tinnitus, and a feeling of pressure in the left ear. Initial MRI of the cerebellopontine angle (CPA) demonstrated an intracanalicular VS at the left CPA (dimensions: 5 × 9 × 5 mm) (Figures 1A and C), after which the patient was referred to the ENT department of the Antwerp University Hospital. Upon first visit, tonal audiometry showed a high-frequency SNHL on the left side, and electronystagmography showed no vestibular failure. A wait-and-scan policy was proposed. At 6-month follow-up, MRI showed a significant growth of the VS (dimensions: 6 × 11 × 5 mm) (Figures 1B and D), without an increase in complaints. The indication for resection of the VS via translabyrinthine approach was made. This approach was preferred because of tumor localization, reduced speech perception scores in quiet, and significantly reduced signal intensity on preoperative T2-weighted MRI, which is correlated with poor hearing preservation rate.

Case 2

A 63-year-old female was in follow-up at our ENT department for an intracanalicular VS at the left CPA (dimensions: 9 × 5 × 4.5 mm), for which a wait-and-scan policy was initially established. She mainly experienced difficulties with speech understanding. At follow-up, repeated MRI CPA showed growth of the VS, from an intracanalicular VS to a small VS in the CPA (dimensions: 10 × 6 × 7 mm). Tonal audiometry showed moderate-severe SNHL on the left side and impaired speech perception scores in quiet, while the right side showed presbycusis according to age. Gross total resection of the VS with translabyrinthine approach was performed four months later.

Surgical Approach

Both surgeries were performed by the senior authors (V.V.R. and T.M.), using the translabyrinthine approach for gross total tumor resection. To enable ANTS and cochlear implantation, a facial recess approach was added to the conventional translabyrinthine approach, a bony recess with pin holes was created to hold and protect the cochlear implant receiver-stimulator and magnet in the subperiosteal tight pocket and a bony channel for the electrode lead.

Auditory Nerve Test System

The ANTS was performed before labyrinthectomy and after gross total resection of the VS. After a routine facial recess approach for cochlear implantation, the middle ear was cleaned and irrigated with a ciprofloxacin solution. Triamcinolone 40 mg/mL solution was applied to the retrotympanum, the round window, and the oval window. Finally, the round window membrane was identified by removing the niche with a low-speed diamond microdrill. The round window membrane was punctured in its anterior inferior region and enlarged with 0.2 mm microhooks. The site was prepared for intraoperative ANTS (MED-EL, Innsbruck, Austria) to evaluate the integrity of the cochlear nerve. The MAESTRO fitting software version 9.0.3 (MED-EL, Innsbruck, Austria) was used for stimulation and was connected via a trigger cable with the Synergy eABR recording system (Medelec Synergy system, VIASYS HealthCare UK, Surrey, United Kingdom). Electrode contacts 1 to 3 from the ANTS electrode array, which is 18 mm in length and has a diameter of 0.4 mm at the tip and 0.8 mm at the ring, were inserted into the cochlea, while the reference electrode with contact 4 was positioned under the temporalis muscle. Prior to the use of the ANTS and starting any eABR measurements, a pre-use check was performed successfully with the Stimulator Box. When switching the stimulator box from 1-2 to 3-4, the measured impedance (Z) changed by more than 0.4 kΩ. All measured values were between 1.5 and 9 kΩ.

Postoperative Follow-Up

Postoperatively, both cases showed normal facial function and had no cerebrospinal fluid leakage. Dizziness resolved after vestibular training. Both patients were discharged, respectively, 7 and 5 days postoperatively. The evolution of the auditory performance of both cases is presented in Tables 1 and 2, respectively.

Discussion

Simultaneous translabyrinthine resection of VS, intraoperative eABR measurement, and cochlear implantation are feasible.[15] Preoperative counseling should be performed by an experienced multidisciplinary team, with expertise in skull base surgery, cochlear implantation, and electrophysiology.

The translabyrinthine approach enables access to the cochlea for insertion of an intracochlear test electrode to perform eABR intraoperatively. More specifically, Medina et al[10] reported a diagnostic accuracy of 93% of intracochlear eABR

Table 1. Evolution of Auditory Performance Before and After Cochlear Implantation for Case 1

		1 Month After Fitting		3 Months After Fitting	6 Months After Fitting	12 Months After Fitting
	Preoperative	Unaided	CI	CI	CI	CI
Pure-tone average (PTA)[a]	27 dB HL (left)	> 117 dB HL (left)	30 dB HL (CI only, left)	35 dB HL (CI only, left)	42 dB HL (CI only, left)	32 dB HL (CI only, left)
Speech in noise (SPIN)[b]	–	−4.33 dB SNR	−5 dB SNR	−5 dB SNR	−5.67 dB SNR	−3.67 dB SNR
A§E® phoneme discrimination test[c]	–	–	–	–	–	95%
Sound localization (RMS)[d]	–	–	37.6°	11.3°	11.3°	0.0°

[a]Unaided air conduction thresholds for pure tones were determined using insert earphones. Aided thresholds for warble tones were measured postoperatively with CI in a free field with a loudspeaker at a distance of 1 m in front of the listener, with the contralateral ear masked. Both unaided and aided thresholds were determined between 125 Hz and 8 kHz according to the clinical standards (ISO 8253-1:2010) using a 2-channel Interacoustics AC-40 audiometer and in a soundproof booth. Pure-tone average thresholds were calculated as the mean of the thresholds at 500, 1000, and 2000 Hz.
[b]SRT in noise (i.e., 50% correct identification point) was determined by an adaptive procedure (fixed noise level of 65 dB SPL (decibel sound pressure level), 2 dB down −2 dB up procedure) with the Leuven Intelligibility Sentence Test in free field with both the speech and noise presented at 0°.[13]
[c]% discrimination on spectral contrasts at 70 dB using the A§E phoneme discrimination, with the contralateral ear masked.[14]
[d]Root-mean-square (RMS) was measured using the A§E azimuth localization test with narrowband noise (NBN) at 4000 Hz (except for 12 months after fitting for case 1, in which a speech noise was used), presented from 7 loudspeakers positioned at 20° intervals from −60° to 60°.Abbreviations: dB HL = decibel hearing loss, SNR = signal to noise ratio.

Table 2. Evolution of Auditory Performance Before and After Cochlear Implantation for Case 2

		1 month After Fitting		1.5 Months After Fitting	3 Months After Fitting	6 Months After Fitting	12 Months After Fitting
	Preoperative	Unaided	CI	CI	CI	CI	CI
Pure-tone average (PTA)[a]	60 dB HL (left)	>113 dB HL (left)	35 dB HL (CI only, left)	35 dB HL (CI only, left)	32 dB HL (CI only, left)	28 dB HL (CI only, left)	33 dB HL (CI only, left)
Speech in quiet (SPIQ)[b]	–	–	–	56%	72%	83%	67%
Speech in noise (SPIN)[c]	–	–	–	−6 dB SNR	–	–	−5 dB SNR
A§E® phoneme discrimination test[d]	–	–	95%	100%	–	–	–
Sound localization (RMS)[e]	13.1°	–	–	20°	–	–	17°

[a]Unaided air conduction thresholds for pure tones were determined using supra-aural headphones. Aided thresholds for warble tones were measured postoperatively in Otocube. Both unaided and aided thresholds were determined between 125 Hz and 8 kHz according to the clinical standards (ISO 8253-1:2010) using a 2-channel Aurical audiometer and in a soundproof booth. Pure-tone average thresholds were calculated as the mean of the thresholds at 500, 1000, and 2000 Hz.
[b]Phoneme score using an open set monosyllable word list (Nederlandse Vereniging voor Audiologie (NVA) list) at 70 dB SPL in sound field.
[c]Speech recogniton threshold (SRT) in noise (i.e., 50% correct identification point) was determined by an adaptive procedure (fixed noise level of 65 dB SPL, 2 dB down −2 dB up procedure) with the Leuven Intelligibility Sentence Test in free field with both the speech and noise presented at 0°.[13]
[d]% discrimination on spectral contrasts at 70 dB using the A§E phoneme discrimination, without need for masking the contralateral ear because of the use of Otocube.[14]
[e]RMS was measured using the A§E azimuth localization test with speech noise, presented from 7 loudspeakers positioned at 20° intervals from −60° to 60°.Abbreviations: dB HL = decibel hearing loss, SNR = signal to noise ratio.

for predicting auditory perception with CIs after VS resection. On the contrary, promontory stimulation cannot be used as an intraoperative test to decide on implantation during VS resection, as it is a subjective method that lacks reliability and requires the subject to be awake.[10,16]

Long-term follow-up of auditory performance up to one year after fitting shows gradual improvement. Preoperatively, the pure-tone average (PTA) of case 1 was 27 dB HL on the left ear. Case 2 showed greater SNHL preoperatively, with a PTA of 60 dB HL on the left side. Both cases showed good evolution of auditory performance soon after fitting, as reflected by PTAs of 30 and 35 dB HL, respectively, one month after fitting. Moreover, both cases show good results on A§E® phoneme discrimination test, with scores of 95% 1 year after fitting and 100% 1.5 months after fitting, respectively. However, some degree of hearing loss remains present postoperatively. For instance, sound localization remained difficult for both cases, as reflected by the A§E azimuth localization test. However, at 1-year follow-up, the result of the localization test for case 1 was at 0°, showing a good evolution.

In a recent systematic review by Wick et al,[17] sequential and simultaneous cochlear implantations were compared. The patients in the delayed implantation group often initially underwent a surgical approach aiming to preserve hearing. The study's conclusion was that the timing of cochlear implantation did not influence CI performance. Insertion of an intracochlear spacer or depth gauge during translabyrinthine resection is an alternative to ensure a potential lumen for second-stage CI. It enables MRI control for residual or recurrent tumors before CI is offered, although current MRI compatibility of CI devices does not preclude postoperative evaluation using MRI. Simultaneous gross total tumor resection with cochlear implantation offers potential benefits, such as the ability to perform intraoperative eABR with an intracochlear electrode and to prevent secondary cochlear fibrosis to complicate optimal intracochlear positioning of the electrode array.

In conclusion, simultaneous cochlear implantation and VS resection should be considered as a potential tool to restore hearing to some level but should only be considered if intraoperative eABR demonstrates functional integrity of the cochlear nerve.

Ethics Committee Approval: N/A

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