Annals of Otology, Rhinology & Laryngology 122(2):91-99.

Vestibular Schwannoma in the Only Hearing Ear: Role of Cochlear Implants

Filippo Di Lella, MD, PhD; Paul Merkus, MD, PhD; Giuseppe Di Trapani, MD; Abdelkader Taibah, MD; Maurizio Guida, AuD; Mario Sanna, MD

Objectives: We sought to delineate the role of cochlear implantation in the management of vestibular schwannoma or other cerebellopontine angle tumors in the only hearing ear.

Methods: We performed a retrospective analysis in a quaternary referral skull base center of all patients who were affected by vestibular schwannoma (or other lesions of the cerebellopontine angle) in the only hearing ear and received a cochlear implant before or after tumor treatment (surgery or radiotherapy) or during the wait-and-scan follow-up. We also performed a systematic review of the English-language literature.

Results: The clinical and audiological results of 10 patients are reported. All patients were managed with contralateral cochlear implantation. In 7 patients, cochlear implantation was performed before tumor removal, while hearing in the ear with the tumor was still present. In 3 patients, the implant was placed after curative surgery. Nine of the 10 patients routinely use their implant with subjective benefit and fairly good auditory performance (median disyllabic word recognition, 90%; median sentence comprehension, 75%). The literature search retrieved no major series with assessment of the long-term efficacy of cochlear implantation in this rare clinical scenario.

Conclusions: Patients affected by vestibular schwannoma in their only hearing ear may significantly benefit from a cochlear implant on the contralateral side prior to tumor removal. Recent and significant hearing deterioration and tumor growth represent the main indications for cochlear implantation.

Key Words: acoustic neuroma, cochlear implant, hearing loss, only hearing ear, single-sided deafness, vestibular schwannoma.

INTRODUCTION

Vestibular schwannoma (VS) in the only hearing ear is a rare clinical setting that challenges clinicians and patients with potential bilateral total deafness, as deterioration of the hearing in the ear with the tumor seems likely. In this category of patients, various therapeutic options are available: 1) radiologic follow-up; 2) radiotherapy; 3) hearing preservation surgery; 4) tumor removal with preservation of the cochlea and cochlear nerve and simultaneous or subsequent insertion of a cochlear implant (CI); and 5) tumor removal without preservation of the cochlea and/or cochlear nerve and simultaneous or subsequent insertion of an auditory brain stem implant (ABI). Besides these options, cochlear implantation may be performed in the contralateral (deaf) ear, before or after curative surgery on the tumor side.[1,2]

Implantable devices such as the CI and ABI introduced new potential strategies for hearing rehabilitation in patients with neurofibromatosis type 2 (NF2) and bilateral VS. Still, decision-making in the presence of VS (or other, less common lesions of the internal auditory canal [IAC] and/or cerebellopontine angle [CPA]) in an only hearing ear is not easy, and the international literature lacks clear indications and large case series. The aim of this study was to delineate the role of cochlear implantation in the management of patients affected by sporadic VS in their only hearing ear by reporting the outcomes and results from a clinical setting accompanied by an accurate literature review.

MATERIALS AND METHODS

The Gruppo Otologico is a quaternary referral center for neurotology and skull base surgery with a surgical experience of about 2,400 VS and 250 CPA lesions of different histologic types (eg, meningiomas, epidermoids) operated on from 1986 to 2010. Besides this surgical database, more than 350 cases of VS are currently being managed with a wait-and-scan policy. Both databases were retrospectively analyzed to select patients treated for sporadic tumors (ie, not related to NF2) involving

From Gruppo Otologico Piacenza-Roma, Piacenza, Italy. This study was supported by a grant from AINOT (Associazione Italiana Neuro Otologica).

Correspondence: Filippo Di Lella, MD, PhD, Gruppo Otologico Piacenza-Roma, Via Emmanueli 42, 29121 Piacenza, Italy.

the IAC and/or CPA in their only hearing ear. Particular attention was paid to patients managed with a CI before or after tumor surgery and patients currently under a wait-and-scan policy who received a contralateral CI.

The study was approved by the local Institutional Review Board. The clinical investigations were conducted according to the principles expressed in the Declaration of Helsinki. The inclusion criteria were a primary diagnosis of sporadic VS or another IAC or CPA tumor on one side with ipsilateral serviceable hearing (ie, speech discrimination score of greater than 60%) and an absence of a tonal threshold in the contralateral ear. Patients with NF2 were not included in the study population.

The patients' charts and imaging data were collected and carefully reviewed for cause of deafness, length of hearing deprivation, use of hearing aids, and promontory test results. The size of the IAC or CPA mass and the subsequent clinical or surgical management, as well as the histopathologic diagnosis of the tumor, if available, were reassessed.

The Nucleus Promontory Stimulator (model Z10012; Cochlear, Sydney, Australia) was used for all behavioral electrical promontory stimulations. Electrical promontory stimulation was performed to obtain information about the functional integrity of the cochlear nerve both in cases of contralateral deafness (with respect to the tumor side) and in cases in which neural integrity was evaluated on the side of the tumor after curative surgery. Electrical promontory stimulation was completed at 5 frequencies (50, 100, 200, 400, and 800 Hz), with a 50-Hz square-wave stimulus initiated at 0 μA, to ascertain the threshold levels and the maximal acceptable loudness. The protocol used for electrical promontory stimulation is detailed elsewhere.[3]

Postoperative auditory performances were assessed in the auditory-only condition in both closed-set (vowel identification) and open-set formats (disyllabic word recognition, common phrase comprehension) with monitored live voice through the sound field at a level of 70 dB sound pressure level. Hearing results are reported as measured at the last available follow-up visit. In patients who received a CI before curative surgery, masking (white noise) was achieved in the only hearing ear with a loudspeaker placed at a horizontal azimuth of 90°. In these cases, the noise signal intensity was modulated according to the patients' hearing thresholds.

A thorough search of the English-language literature was performed on search engines such as PubMed and Embase with the following 3 searches: "acoustic neuroma" OR "vestibular schwannoma" AND "only hearing ear" OR "single sided deafness" OR "contralateral deafness"; "cochlear implant" AND "acoustic neuroma" OR "vestibular schwannoma"; and "only hearing ear" OR "single sided deafness" OR "contralateral deafness" AND "CPA tumor" OR "skull base tumor."

RESULTS

Patients. A total of 8 patients were found who fit the inclusion criteria; 2 more patients (patients 6 and 8) who did not strictly fit the inclusion criteria were added to the series because of their pertinent history. Thus, there were 6 women and 4 men; their mean age was 49 years (range, 25 to 76 years). In all patients, the tumor diagnosis was made during clinical and radiologic evaluation of progressive hearing loss in the only hearing ear. Complete demographic and clinical management data are presented in Table 1. The audiological results from the most recent follow-up visit are presented in Table 2. In all cases, a CI was placed contralateral to the tumor side; the mean follow-up after cochlear implantation was 40 months (range, 11 to 67 months).

This series included patients with different causes for the hearing loss in their nontumor ear. Three of these patients (patients 1, 2, and 4) had had anacusis since early childhood that was presumably of congenital origin; patient 3 had profound sensorineural hearing loss as a consequence of having had viral parotitis at 7 years of age. Patients 5, 6, 7, and 8 had sudden deafness or progressive sensorineural hearing loss before the diagnosis of tumor in their only hearing ear was made. Patients 9 and 10 lost the hearing on the nontumor side after surgical procedures in the middle ear. Patients 6 and 8 underwent primary treatment elsewhere and did not strictly fit the inclusion criteria. A VS in the only hearing ear of patient 6 had been removed 6 years before our consultation, and patient 8 received a diagnosis of VS during preoperative evaluation for a CI and was referred after implantation. As already mentioned, both patients were included for their pertinent history.

Surgical Management. Contralateral CI placement was performed as primary treatment before tumor removal (early implantation) in 7 patients (patients 1, 3, 4, 5, 7, 8, and 10); patient 8 has been included even though the implantation had been performed elsewhere 3 years earlier. All patients who underwent tumor removal (patients 1, 4, 7, 8, and 10) routinely use their CI with subjective benefit and objectively measurable increases in auditory performance (median disyllabic word and sentence

TABLE 1. DEMOGRAPHIC, CLINICAL, AND AUDIOLOGICAL FINDINGS OF PRESENTED PATIENT SERIES

Pt	*Sex*	*Age*	*Right Ear*	*Left Ear*	*Side/Duration of Deafness/ Cause*	*Tumor Side/ Histology/ Dimensions/ Behavior*	*EPS Side*	*CI Side and Date of Surgery/Use*	*CPA or IAC Tumor Management*
1	F	41 y	Anacusis	45 dB PTA, 100% SDS	R/anacusis since early childhood/ unknown	L/VS/1.5 cm/ growing	+R	R Apr 2006/ sleeper→user	L RS-RL, no hearing preservation Jan 2007, L CI Mar 2007
2	F	61 y	Anacusis	35 dB PTA, 80% SDS	R/anacusis since early childhood/ unknown	L/VS/3 cm (CPA)	+R	R Jun 2004/ user	R TLA Jul 2002
3	F	46 y	25 dB PTA, 100% SDS	Anacusis	L/anacusis since late childhood/ parotitis	R/meningioma (NC)/2 cm/stable	+L	L Jan 2010/ sleeper	R wait-and-scan, stable hearing with no tumor growth after 14 mo
4	M	46 y	Anacusis	60 dB PTA, 70% SDS	R/anacusis since early childhood/ unknown	L/VS/3 cm	+L	R Dec 2010/ sleeper→user	L TLA Sep 2011
5	F	48 y	Anacusis	35 dB PTA, 85% SDS	R/12 mo/sudden deafness	L/VS (NC)/ intracanalicular/ stable	+R	R Jun 2005/ sleeper→user	L wait-and-scan, slight hearing worsening with no tumor growth after 60 mo
6	M	41 y	35 dB PTA*, 85% SDS	Anacusis, ABI†	L/21 y/sudden deafness	R/VS/2 cm	+L	R Jan 2010/ user	L RS + ABI‡ 2003
7	M	50 y	Anacusis	20 dB PTA, 100% SDS	R/22 y/sudden deafness	L/VS/1 cm/ growing	+R	R Dec 2009/ sleeper→user	L TLA + CI Jan 2010
8	F	76 y	Severe SNHL; benefit with HA*	Anacusis, CI†	L/progressive SNHL/unknown	R/VS/2 cm/ growing	NA	L Jun 2007‡/ user	R TLA subtotal removal Jul 2009
9	F	57 y	40 dB PTA, 90% SDS	Anacusis	L/40 y/radical mastoidectomy	R/VS/3 cm (previous RT)/ growing	+L	L Nov 2004/ user	R TLA Jan 2004
10	M	25 y	Anacusis	50 dB PTA, 80% SDS	R/10 y/radical mastoidectomy	L/VS/3 cm/ cystic	+R	R Sep 2006/ sleeper→user	L TLA + ABI Nov 2006

PTA — pure tone average; SDS — speech discrimination score; EPS — electrical promontory stimulation; CI — cochlear implant; CPA — cerebellopontine angle; IAC — internal auditory canal; VS — vestibular schwannoma; RS-RL — retrosigmoid-retrolabyrinthine approach; TLA — translabyrinthine approach; NC — not confirmed at histopathologic examination; ABI — auditory brain stem implant; RS — retrosigmoid approach; SNHL — sensorineural hearing loss; HA — hearing aid; NA — not available; RT — radiotherapy.

*Data from another department.

†Environmental sound detection.

‡Performed in another department.

recognition scores of 68% and 54%, respectively). In this group, patients 1, 4, and 7 reported a remarkable increase in their CI performance after tumor removal with loss of hearing in what had been their only hearing ear.

Patient 1 received a second CI after tumor removal (retrosigmoid approach without hearing preservation), but although the cochlear nerve was spared, no results were obtained at activation. The same patient did well with her first implant for more than 60 months (word and sentence recognition scores reaching 90%), but more recently presented with a sudden significant drop in performance accompanied by dizziness and spells of vertigo. She currently uses her CI, but her auditory performance is significantly lower than it was a few months earlier. Patient 4's CI was activated in February 2011, and he initially could identify 35% of vowels in closed set but had no open-set abilities. His CI performance increased both subjectively and objectively after tumor removal, reaching a 25% score on the disyllabic word test and a 20% score on the sentence recognition test (September 2011). Patient 7 underwent tumor removal through a translabyrinthine approach with cochlear nerve preservation and simultaneous cochlear implantation. Patient 8 was included in this group of patients even though she underwent implantation elsewhere 3 years earlier. The VS in her only hearing ear was initially managed conservatively; her residual hearing on the tumor side allowed for bimodal stimulation for about 16 months,

TABLE 2. AUDITORY PERFORMANCES AS EVALUATED AT LAST AVAILABLE FOLLOW-UP

Pt	Sex	Follow-Up	Age	Implant Type/Side	User or Sleeper Device	Device Use	Disyllabic Word Recognition	Sentence Recognition	Telephone Use	Subjective Comments
1	F	67 mo	41 y	Digisonic SP/R	User	Daily	65% (90% at 48 mo)	15% (90% at 48 mo)	No (yes at 48 mo)	Excellent benefit; performance improved after tumor removal; decrease in performance at 60 mo
				Digisonic SP/L	Nonuser*					
2	F	60 mo	61 y	Nucleus 24/R	User	Daily	90%	65%	Yes (close relatives)	Sufficient benefit
3	F	22 mo	46 y	Sonata ti 100/L	Sleeper		90%†	40%†		No benefit from implant; maintains normal hearing in tumor ear
4	M	11 mo	46 y	Digisonic SP/R	User	Daily	25%	20%		Marked subjective benefit; performance improved after tumor removal
5	F	60 mo	48 y	Nucleus 24/R	User		90%†	90%†		Excellent benefit; performance improved after progression of hearing loss on tumor side
6	M	18 mo	41 y	Digisonic SP/R	User	Daily	90%	90%	Yes (close relatives)	Excellent benefit
7	M	14 mo	50 y	Digisonic SP/R	User	Daily	90%	85%	Yes (close relatives)	Excellent benefit; performance improved after tumor removal
				Digisonic SP/L	Nonuser‡					
8	F	48 mo	57 y	Nucleus 24/L	User	Daily	90%	90%	Yes (close relatives)	Excellent benefit
9	F	60 mo	76 y	Nucleus 24/L§	User	Daily	90%	85%	Yes (close relatives)	Excellent benefit
10	M	48 mo	25 y	Digisonic SP/R	User	Daily	70%	75%	Yes	Excellent benefit
Mean		40 mo	49 y				79%	66%		
Median							90%	75%		

*No auditory sensation from implant in tumor ear.

†Contralateral masking applied.

‡Environmental sound detection only from implant in tumor ear.

§Implanted in another department.

at which point tumor growth and hearing deterioration imposed a change in management strategy.

Of the 2 patients who did not undergo tumor removal, 1 (patient 3) preserved a stable normal hearing threshold with no evidence of tumor growth, and the CI was therefore maintained inactive or as a "sleeper." The other (patient 5) had no increase in tumor dimensions, but showed progression of the hearing loss. Her auditory performance with the CI improved after her hearing decreased, and she is currently a daily user of the device.

Contralateral cochlear implantation was performed after tumor removal in 3 cases (patients 2, 6, and 9). Patient 6 had been previously managed elsewhere with surgical removal of the tumor and simultaneous ABI placement through a suboccipital approach. His ABI performance never reached open-set abilities and progressively declined over time, whereas his CI performance has been quite impressive since activation, and the patient is now a daily CI user without any need for the ABI. All patients in this group routinely use their CI, and their median disyllabic word and sentence recognition scores were 90% and 80%, respectively. There were no major or minor complications after the surgical procedures.

Literature Review. A thorough search of the English-language literature retrieved a few case reports in which VS in the only hearing ear was managed conservatively or with radiotherapy.[4-7] Our group, in 1994, reported on 5 patients who were treated conservatively and pointed out the possibility of co-

TABLE 3. REVIEW OF LITERATURE ON PATIENTS AFFECTED BY VS IN ONLY HEARING EAR AND MANAGED WITH CI OR ABI INSERTION

Authors	*Year*	*No. of Pts*	*Case No.*	*Age*	*Hearing (Cause)*	*Promontory Stimulation*	*First Treatment*	*Second Treatment*	*Results With CI (Follow-Up)*
Thedinger et al[8]	1993	3	1	49 y in 1989	L profound SNHL on PTA (labyrinthectomy for Meniere' disease 14 y earlier); R 65 dB SRT and 16% SDS (2.7-cm VS)	+L	L CI	R TLA tumor removal 3 mo later	Good; CID sentences 171/200 (open set); discrimination 38% (6 mo)
Talbot et al[9]	1994	1	1	69 y in 1994	L total HL on PTA (childhood mumps); R residual hearing on PTA (2-cm VS)	NA	R CI	L TLA tumor removal	NA
Shin et al[10]	1998	3	1	66 y in 1994	L total HL on PTA (Meniere's disease); R progressive moderate SNHL on PTA (grade II VS)	+L	Radiologic follow-up, no growth after 6 mo	R CI	Good; 98% open-set sentence discrimination (12 mo)
			2	62 y in 1994	L total HL on PTA (childhood); R progressive profound SNHL on PTA (4-cm VS)	+L	MF tumor removal	L CI 3 mo later	Poor; 13% open-set sentence discrimination (14 mo)
			3	44 y in 1992	L total HL on PTA (childhood); R progressive profound SNHL on PTA (1-cm meningioma)	+R	MF decompression and subsequent TLA tumor removal	R CI 7 mo later	Good; 70% open-set sentence discrimination (48 mo)
Arístegui and Denia[11]	2005	1	2	45 y in 2002	L total HL on PTA (craniocencephalic trauma in childhood); R progressive SNHL on PTA (1.5-cm VS)	NA	L TLA tumor removal and simultaneous CI		Good; 90% to 100% speech discrimination (6 mo)
Ramsden et al[12]	2005	2	1	45 y in 1996	L total HL on PTA (congenital); R 30-40 dB SNHL on PTA and 100% SDS at 70 dB (2-cm VS)	+L –R	R SO tumor removal, HL	L CI 4 mo later	Poor; CI useful for environmental sound detection and lipreading (12 mo)
			2	49 y in 1999	L total HL on PTA (congenital); R moderately severe high-frequency SNHL on PTA and 0% SDS (2-cm VS)	+L	L CI	L TLA tumor removal 22 mo later	Poor; CI useful for environmental sound detection and lipreading (12 mo)
Grayeli et al[2]	2008	3	1	71 y	Solitary VS on one side and contralateral HL on PTA with serviceable hearing, HA	NA	Simultaneous TLA tumor removal and ABI placement		20% Sentence recognition; no ABI use (26 mo)
			2	51 y	Solitary VS on one side and contralateral SNHL on PTA (sudden deafness) with serviceable hearing, CI	NA	CI on sudden deafness side	TLA tumor removal and and ABI placement	No ABI use; CI use with 70% sentence recognition (21 mo)
			3	69 y	Solitary VS on one side and contralateral total HL on PTA (petrous bone cholesteatoma)	NA	Simultaneous TLA and ABI placement		70% sentence recognition with ABI + lipreading (35 mo)

SRT — speech reception threshold; CID — Central Institute for the Deaf; HL — hearing loss; MF — middle fossa; SO — suboccipital approach.

chlear implantation in the contralateral deaf ear.[1] Only a small number of articles are available on long-term experience with a CI or ABI, and each presents 3 or fewer cases (Table 3).[2,8-12] Thedinger et al[8] were the first to describe a case in which insertion of a contralateral CI was performed before the removal of VS in the only hearing ear, in 1989; other case reports followed.[9,10] In 2005, Ramsden et al[12] reported on 2 patients with presumed congenital hearing loss on one side and VS in their only hearing ear; a CI was inserted in the congenitally deaf ear in one case before, and in the other case after, removal of the tumor. In both cases, the auditory results can be considered poor in comparison to the standard performance of those with postlingual cochlear implantation. Ramsden et al[12] speculated on a possible decrease in neural tissue in the spiral ganglion and cochlear nuclei of congenitally deaf ears that might

render CI and ABI worthless in the auditory rehabilitation of such patients. In light of their results, they propose ABI placement during tumor removal and reserving cochlear implantation in the nontumor ear as a secondary option to be used if the ABI results should be disappointing.[12] Grayeli et al,[2] in 2008, reported on ABI positioning in 3 patients affected by VS in their only hearing ear. Only 1 of the 3 patients was a daily ABI user, whereas the ABI results were poor in the other 2. Remarkably, 1 of these 2 ABI nonusers had previously received a contralateral CI, obtaining good auditory performance.[2] We also found reports on intracapsular tumor removal[13] and middle fossa decompression surgery[14] as means of delaying hearing loss in the only hearing ear, but nowadays, these strategies play a very limited role.

DISCUSSION

The contemporary management of tumors affecting the central auditory pathway should ideally be complete surgical removal with facial nerve and hearing function preservation. Vestibular schwannoma in the only hearing ear is an uncommon scenario, with a prevalence of about 0.3% in our experience.[15] Each patient needs to be treated by taking into account many parameters such as the tumor dimensions, the age and preferences of the patient, surgical risks, alternative treatments, and quality-of-life outcomes. A conservative attitude (hearing preservation surgery, radiotherapy, wait-and-scan policy) should be adopted in cases of small lesions (less than 1.5-cm cisternal component), in elderly patients (more than 65 years of age) or patients with a poor performance status, and in the presence of good or serviceable hearing. Progression of the hearing loss should be expected in about 30% of cases as a consequence of the natural tumor history and radiotherapy, and in more than 60% of cases after hearing preservation surgery.[16-18]

Younger patients are exposed to the risks of both tumor growth and hearing deterioration and should be treated aggressively. It is mandatory for the neurotologic team to adopt a strategy that reduces the chances of total bilateral deafness. When VS is diagnosed on one side with serviceable or good hearing and there is no useful hearing in the opposite ear, an electronic device such as a CI or ABI should be considered as part of the management plan.

The patients described in this report represent, to the best of our knowledge, the largest series in the existing English-language literature from a single center. Their clinical management can be summarized as follows.

Contralateral CI placement as primary treatment before tumor removal (early implantation) was counseled and carried out in 7 patients (patients 1, 3, 4, 5, 7, 8, and 10) because of young age and the possibility of hearing loss progression in the tumor (and only hearing) ear (patients 3 and 5); young age and rapidly declining hearing (patients 1 and 7); or large tumor dimensions (patients 4 and 10). Patient 8 received a CI at another institution, 3 years before tumor removal. Six patients (patients 1, 4, 5, 7, 8, and 10) are daily users of the CI, and 5 of them have shown good auditory performance (patients 4, 5, 7, 8, and 10).

Contralateral CI placement following tumor removal was performed in 3 patients. It was the patient's choice in 1 case (patient 2). Another case had large tumor dimensions with signs of intracranial hypertension requiring immediate tumor removal (patient 9). One patient (patient 6) had previously undergone surgery elsewhere for simultaneous removal of the tumor and ABI placement through a suboccipital approach and received a contralateral CI 6 years later at our institution.

Ipsilateral cochlear implantation was performed in 2 patients (patients 1 and 7). Patient 1 underwent a retrosigmoid-retrolabyrinthine approach for removal of a left-sided tumor in order to preserve hearing. Anatomic maintenance of the cochlear nerve was achieved, but after the operation the patient had anacusis. However, her auditory scores with use of the right CI that was positioned a few months earlier reached 85% on open-set speech comprehension, and after 12 months from the initial fitting, she was able to use the telephone with her family. At the patient's request, left promontory stimulation was performed 3 months after surgery and showed positive responses; it was then decided to perform a left cochlear implantation. In December 2007, a CI was placed in the left ear, but no auditory sensations were felt by the patient at activation; raising of the signal intensity led to facial nerve stimulation, and because of the reasonable performance obtained with the right implant, it was decided not to use the one on the left side. Patient 7 underwent a translabyrinthine approach for removal of right-sided VS. The decision to operate was based on the radiologic evidence of tumor growth and on the results obtained with the left CI placed 6 months earlier (sentence recognition score, 70%; speech comprehension score, 65%). The cochlear nerve was spared, and a CI was simultaneously placed. The second implant was activated in March 2011, and the patient showed only sound detection abilities while reporting a marked subjective improvement of right CI performance. In both of these cases, ipsilateral implantation was performed as a secondary procedure because of the

good results obtained with contralateral CIs and the fact that the functional status of the cochlear nerve is unpredictable after surgical manipulation even if anatomic integrity is maintained.

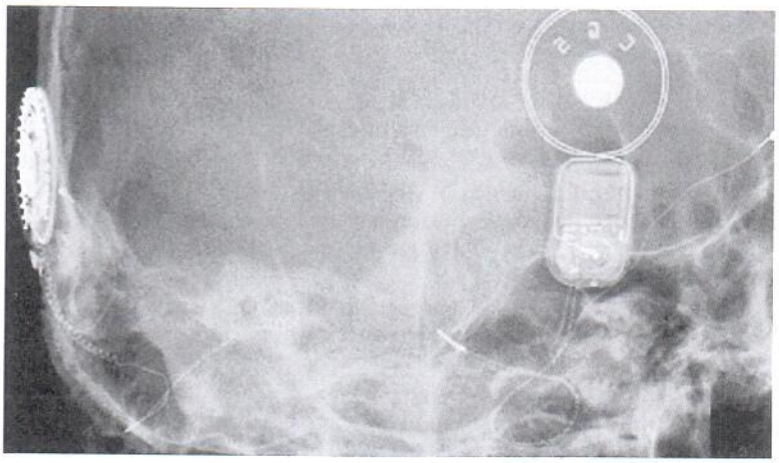
Analysis of the presented case series showed positive effects following placement of a CI in the nontumor ear. Implanting a CI in patients before or after contralateral tumor removal led to good CI results, with overall median word and sentence recognition scores of 90% and 75%, respectively. Except for patient 3, who retains good hearing in the tumor ear and has no benefit from electrical hearing, the remaining patients are daily CI users with subjectively reported advantages to their social life. Patient 1 experienced an unexplained deterioration in CI performance 60 months after CI placement and is currently under evaluation. Accurate masking was performed when we evaluated individuals with good hearing on the tumor side (patients 3 and 5); the masking may nevertheless have been inadequate to completely exclude the influence of their only hearing ear on their auditory performance.

Long-term deafness and even congenital hearing loss cannot be considered as contraindications to insertion of a CI in the nontumor ear. Except for patient 5, the length of deafness in the study group was at least 10 years. Four patients with evidence of complete hearing loss since early (patients 1, 2, and 4) or late childhood (patient 3) have undergone implantation; still, it is difficult to define as congenital a long-term deafness in adult individuals evaluated 30 or 40 years after their first diagnosis. The results for these patients are fairly good — especially patients 1 and 2 — as are the auditory performances of patients who were deaf from other causes and had long-standing hearing deprivation.

A remarkable increase in CI performance was seen in some patients after tumor removal and subsequent loss of hearing in what had been their only hearing ear (patients 1, 4, and 7). Patient 5 experienced a significant improvement in CI performance associated with progression of the hearing loss on the tumor side. From these data, one could theorize that the auditory cortex gives precedence to the best hearing pathway, or that the improvement of the deprived auditory pathway via electrical stimulation is hampered by the normal hearing. By the same token, once the hearing starts to deteriorate, the performance of the CI seems to improve dramatically.

Although CI insertion in the nontumor ear shortly before or after tumor removal may avoid total bilateral deafness and reduce the time needed for hearing rehabilitation, inserting a CI in a patient being followed with a wait-and-scan policy with no evidence of tumor growth (early implantation with devices maintained inactive or as a "sleeper") may be a matter of debate. It could be years before auditory performance in the only hearing ear declines, and — as a major drawback of early implantation — this strategy could lead to the presence of a non–state-of-the-art device.

Implantation prior to contralateral tumor removal leads to very satisfactory results; 5 patients (patients 1, 4, 7, 8, and 10) had undergone implantation in expectation of tumor excision and did well with a CI after they lost the hearing on the tumor side. Our experience with CI placement in patients followed with a wait-and-scan policy is less encouraging. Patients 3 and 5 underwent implantation in consideration of their young age and the possibility of declining hearing on the tumor side. In these patients, the tumor dimensions remained stable (after 16 and 60 months of observation, respectively), and no tumor removal was undertaken. Patient 3 maintained a normal tonal threshold and did not use the CI, whereas patient 5 had a slight worsening of her hearing loss and is a daily CI user, reporting subjective benefit from the device.



(Patient 6) Postimplantation radiograph shows evidence of cochlear implant in right cochlea and auditory brain stem implant on left side.

TABLE 4. SURGICAL MANAGEMENT OPTIONS FOR VS (OR OTHER CPA TUMOR) IN ONLY HEARING EAR

Options *before* removal of vestibular schwannoma
Hearing aid in that ear if surgery can wait
Cochlear implant on contralateral side
Options *during* removal of vestibular schwannoma
Hearing preservation (small vestibular schwannoma)
Anatomic preservation of cochlear nerve and simultaneous placement of cochlear implant in same ear
Ipsilateral placement of auditory brain stem implant when none of above-mentioned strategies are any longer possible
Options *after* removal of vestibular schwannoma
Cochlear implant on contralateral side
Cochlear implant on same side if cochlear nerve is spared
Ipsilateral or contralateral placement of auditory brain stem implant when none of above-mentioned strategies are any longer possible

Patients managed for VS need long-term follow-up with magnetic resonance imaging. Device displacement, heating, and magnet demagnetization have been indicated as problems following cochlear implantation. The magnet in the CI creates an artifact on the imaging, and scanning needs to be done with a head bandage (up to 1.5 T). None of our patients experienced any problem (CI dislocation or malfunction) during or after magnetic resonance imaging up to 1.5 T. Contralateral visualization is not hampered by artifacts or distortion caused by the CI.[19,20]

Some authors have speculated as to the use of an ABI in this clinical setting.[12] An ABI represents the only available means of hearing restoration in NF2 patients when there is no possibility of preserving the auditory nerve or the cochlea, but the auditory results are disappointing in comparison to those obtained with a CI. The majority of ABI users have environmental sound detection abilities, but only a lucky few obtain speech recognition abilities. Still, there are no definite prognostic factors that will predict postoperative ABI performance.[21] There have been only a few reports on brain stem implantation in nontumor patients, and although the auditory results seem better than those in NF2 patients, indications for an ABI apart from NF2 are exceptionally rare.[22] In patient 6 of this series, the right side was completely normal from an anatomic point of view (ie, cochlea and cochlear nerve present and intact), and cochlear implantation should have been performed before tumor excision and ABI insertion on the tumor side (see Figure).

It should be emphasized that in the presence of a patent and undamaged cochlea and preserved cochlear nerves, use of a CI should be attempted before an ABI is even proposed. A promontory stimulation test gives a useful indication as to the integrity of the cochlear nerve, even if the test results do not correlate with the final hearing level.[23] Electrical promontory stimulation is an encouraging prognostic factor for future CI performance and is a valuable means of testing the already-operated ear to ascertain cochlear nerve function, as in patients 9 and 10 in our series. Table 4 gives an outline of available surgical options for patients with VS in the only hearing ear.

In summary, cochlear implantation on the nontumor side is useful in selected patients with VS in the only hearing ear; recent and significant hearing deterioration and tumor growth represent the main indications.[2] In patients who maintain serviceable hearing and have no signs of tumor growth, CI placement should be postponed.

CONCLUSIONS

Data from the presented series suggest that cochlear implantation on the nontumor side is useful in selected patients with VS in the only hearing ear. Once the decision to remove the tumor in the only hearing ear is made, CI insertion should be performed first, allowing rapid auditory rehabilitation. Recent and significant hearing deterioration and tumor growth represent the main indications.

REFERENCES

1. Naguib MB, Saleh E, Aristegui M, Mazzoni A, Sanna M. Decision making in acoustic neuroma management: the only hearing ear. Skull Base Surg 1994;4:32-6.

2. Grayeli AB, Kalamarides M, Bouccara D, Ambert-Dahan E, Sterkers O. Auditory brainstem implant in neurofibromatosis type 2 and non–neurofibromatosis type 2 patients. Otol Neurotol 2008;29:1140-6.

3. Vincenti V, Pasanisi E, Guida M, Di Trapani G, Sanna M. Hearing rehabilitation in neurofibromatosis type 2 patients: cochlear versus auditory brainstem implantation. Audiol Neurootol 2008;13:273-80.

4. Chu SM, Young YH. Ipsilateral and contralateral vestibular schwannomas in unilateral long-term deaf ears. Eur Arch Otorhinolaryngol 2007;264:693-6.

5. Pensak ML, Tew JM, Keith RW, Vanloveren HR. Management of the acoustic neuroma in an only hearing ear. Skull Base Surg 1991;1:93-6.

6. Branch MP, Hirsch BE. Management of unilateral acoustic neuroma in a 72-year-old patient with contralateral congenital deafness. Otolaryngol Head Neck Surg 2002;127:483-6.

7. Tschudi DC, Linder TE, Fisch U. Conservative management of unilateral acoustic neuromas. Am J Otol 2000;21:722-8.

8. Thedinger BA, Cueva RA, Glasscock ME III. Treatment of an acoustic neuroma in an only-hearing ear: case reports and considerations for the future. Laryngoscope 1993;103:976-80.

9. Talbot A, Tonkin J, Fagan P, Platt-Hepworth S. Vestibular schwannoma in an only hearing ear. J Laryngol Otol 1994;108: 983-5.

10. Shin YJ, Fraysse B, Sterkers O, Bouccara D, Rey A, Lazorthes Y. Hearing restoration in posterior fossa tumors. Am J Otol 1998;19:649-53.

11. Arístegui M, Denia A. Simultaneous cochlear implantation and translabyrinthine removal of vestibular schwannoma in an only hearing ear: report of two cases (neurofibromatosis type 2 and unilateral vestibular schwannoma). Otol Neurotol 2005; 26:205-10.

12. Ramsden R, Khwaja S, Green K, O'Driscoll M, Mawman D. Vestibular schwannoma in the only hearing ear: cochlear implant or auditory brainstem implant? Otol Neurotol 2005; 26:261-4.

13. Lownie SP, Drake CG. Radical intracapsular removal of acoustic neurinomas. Long-term follow-up review of 11 patients. J Neurosurg 1991;74:422-5.

14. Gadre AK, Kwartler JA, Brackmann DE, House WF, Hitselberger WE. Middle fossa decompression of the internal auditory canal in acoustic neuroma surgery: a therapeutic alternative. Laryngoscope 1990;100:948-52.

15. Dispenza F, De Stefano A, Flanagan S, Romano G, Sanna M. Decision making for solitary vestibular schwannoma and contralateral Meniere's disease. Audiol Neurootol 2008;13:53-7.

16. Massick DD, Welling DB, Dodson EE, et al. Tumor growth and audiometric change in vestibular schwannomas managed conservatively. Laryngoscope 2000;110:1843-9.

17. Stangerup SE, Caye-Thomasen P, Tos M, Thomsen J. Change in hearing during "wait and scan" management of patients with vestibular schwannoma. J Laryngol Otol 2008;122: 673-81.

18. Lin VY, Stewart C, Grebenyuk J, et al. Unilateral acoustic neuromas: long-term hearing results in patients managed with fractionated stereotactic radiotherapy, hearing preservation surgery, and expectantly. Laryngoscope 2005;115:292-6.

19. Crane BT, Gottschalk B, Kraut M, Aygun N, Niparko JK. Magnetic resonance imaging at 1.5 T after cochlear implantation. Otol Neurotol 2010;31:1215-20.

20. Wackym PA, Michel MA, Prost RW, Banks KL, Runge-Samuelson CL, Firszt JB. Effect of magnetic resonance imaging on internal magnet strength in Med-El Combi 40+ cochlear implants. Laryngoscope 2004;114:1355-61.

21. Sanna M, Di Lella F, Guida M, Merkus P. Auditory brainstem implants in NF2 patients. Results and review of the literature. Otol Neurotol 2012;33:154-64.

22. Sanna M, Khrais T, Guida M, Falcioni M. Auditory brainstem implant in a child with severely ossified cochlea. Laryngoscope 2006;116:1700-3.

23. Neff BA, Wiet RM, Lasak JM, et al. Cochlear implantation in the neurofibromatosis type 2 patient: long-term follow-up. Laryngoscope 2007;117:1069-72.