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Ipsilateral cochlear implantation in patients with sporadic vestibular schwannoma in the only or best hearing ear and in patients with NF2

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Abstract The aim of this study was to evaluate the cochlear implant (CI) performances in neurofibromatosis type 2 (NF2) patients with bilateral vestibular schwannoma (VS) and in patients with sporadic VS in the only or better hearing ear. All patients with bilateral VS or sporadic VS in the only or better hearing ear who underwent cochlear implantation, either simultaneous to VS surgery or staged after treatment for VS, in the tumor side were chosen for the study. Postimplantation audiometric scores (sound detection, closed-set and open-set discrimination scores) and device use patterns were the main outcome measures. 15 patients were implanted. Eight patients (53 %) were NF2 and seven patients had VS in the only or better hearing ear. One patient was explanted for cerebrospinal fluid leak. In the CI-only condition, the other 14 patients obtained sound detection, 64 % of them achieving open-set discrimination (mean 70 ± 38 %) and 85 % achieving closed-set discrimination (mean 41 ± 33 %). At the last follow-up 10 patients (67 %) were using the CI. Cochlear implantation provides hearing in particular cases of patients with bilateral VS or VS in the only or better hearing ear. As long as anatomic preservation of the cochlear nerve is achieved, cochlear implantation may offer improvement in communication skills for most patients.

Keywords Neurofibromatosis type 2 · Vestibular schwannoma · Cochlear implantation · Hearing outcome

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Introduction

Patients with neurofibromatosis type 2 (NF2) and patients with sporadic vestibular schwannoma (VS) in the better or only hearing ear represent challenging situations for the neurotologic teams, as there is a risk of progression towards profound deafness. Some of these patients are considered candidates for auditory brainstem implantation (ABI) as the method for hearing rehabilitation. Unfortunately, audiological results of ABI in NF2 patients are generally limited to sound awareness and enhanced lip reading, with open-set discrimination achieved in less than 20 % of the cases [1, 2].

Cochlear implantation has recently emerged as a reasonable therapeutic option for patients with bilateral VS or sporadic VS in the only or better hearing ear, when anatomical integrity of the cochlear nerve during tumor excision is maintained. Since Hoffmann reported the first case of ipsilateral cochlear implantation in a patient with NF2 [3], approximately 85 cases have been reported to date [4, 5]. In addition, many case series have been published

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regarding the role of the cochlear implants (CI) in patients with a VS in the only hearing ear [6]. Results of ipsilateral cochlear implantation in NF2 patients and VS in an only hearing ear are promising. Most patients benefit at least from sound awareness and aid lip reading, with many of them achieving open-set discrimination ability, and in certain cases outcomes are comparable to those of post-lingually implanted non-tumor patients. Moreover, cochlear implantation may also play a role when other therapeutic options as wait and scan or radiotherapy (RT) are adopted [7].

Still some controversial issues are need to be clarified. The precise effect of radiotherapy and other demographic, clinical, radiological, and surgical prognostic factors on the long-term audiological outcome is unclear. Similarly, the best time for intervention, selection of the optimal side in NF2 patients and the effect of RT are not well defined. In this article we analyze the hearing outcomes of ipsilateral cochlear implantation in patients with sporadic vestibular schwannoma in the only or best hearing ear and in patients with NF2.

Materials and methods

A multi-institutional, multi-national retrospective study was conducted in 3 tertiary referral care centers. A systematic chart review was carried out on all patients affected by NF2 with bilateral VS and patients affected by sporadic VS in the only or better hearing ear that underwent cochlear implantation as part of their management protocol. Only patients implanted in the same side of the tumor were selected. The study was approved by the local Institutional Review Boards.

Data included patient demographics, tumor size, treatment modality, preimplantation audiometric performance scores, and hearing outcomes after implantation. The hearing status in the non-implanted ear at the time of implantation was also assessed. Tumor size was measured according to the AAOHNS 1995 guidelines [8]. Minimum follow-up was 12 months. Postoperative complications were recorded. Preoperative audiologic measurements included pure tone audiometry (PTA) and maximum speech discrimination score (SDS). For both the implanted and the contralateral side, hearing classes according to the 1995 American Academy of Otolaryngology guidelines were used [8]. Postoperative auditory performances were assessed in the CI-only condition in both closed-set and open-set (dissyllabic word recognition) formats with monitored live voice through the sound field at a level of 70 dB sound pressure level. Hearing results were reported as measured at the last available visit. In patients who received a CI still having residual contralateral hearing, masking (white noise) was achieved in the only hearing ear with a headphone. In these cases the noise signal intensity was modulated according to the patients' hearing thresholds. To show real life hearing, open-set discrimination in the bilateral condition was also measured.

Categorical variables were described by frequency counts and percentages. Continuous variables were summarized with means, standard deviations (SD), and ranges. Groups of patients were compared using the Mann–Whitney test for continuous asymmetrically distributed variables, and Chi-square and Fisher exact tests for categorical values. All tests were two sided, and $p < 0.05$ was considered statistically significant.

Results

Demographic-clinical data: preoperative hearing

This group of patients consisted of seven men and eight women with a mean age of 46 years (range 24–72 years) implanted between 2001 and 2012 (Tables 1, 2). CI was placed in the right side in 4 cases (27 %). Preoperative PTA in the implanted side ranged from 20 to 110 dB with a mean of 61.7 dB. According to the hearing classes it was normal (class A, PTA 0–30 dB, SDS 70–100 %) in 5 cases (30 %) and serviceable (class B, PTA 31–50 dB, SDS 50–100 %) in one case (7 %). The rest of the cases had only measurable hearing or no hearing at all (classes C, PTA 51–100 dB, SDS 50–100 %; or D, SDS 0–49 %). Considering both ears, seven patients had class A at least one side preoperatively (patients 1, 3, 4, 5, 6, 9, 14); that is, they could communicate without any aid before surgery. In four patients hearing class was D bilaterally, not obtaining significant benefit from hearing aids preoperatively (patients 2, 7, 8, 12).

Treatment

Simultaneous tumor resection and CI placement was performed in 11 patients (73 %), all of them undergoing an extended translabyrinthine approach. One of these patients had received RT prior to surgery. In 3 cases, a staged implantation was performed following prior tumor resection by a retrosigmoid approach (2 cases) and a middle fossa approach (1 case). In every patient undergoing surgery, total resection was attempted. One patient underwent cochlear implantation following RT with no surgical resection of her VS. Implant type included devices from Cochlear (7 Nucleus 24), Medel (1 Combi 40+, 3 Pulsar Ti100, 1 Sonata Ti100) and Digisonic (3 Digi Sp). The decision of implantation was individualized and based on the anatomical preservation of the cochlear nerve. Cochlear

Table 1 Patient characteristics

No	Sex	Age (years)	Tumor size (cm)	CI side	Selected CI side	Contralateral condition	Selected hearing side	Preop. Ipsilateral hearing		
								PTA (dB)	SDS (%)	Hearing class
1 NF2	F	50	CI	R	Larger tumor	IC tumor	Worse	45	60	B
2 NF2	F	57	4.0	L	Larger tumor	Previous TL	Better	60	30	D
3 NF2	F	28	1.7	R	Larger tumor	IC tumor	Worse	110	0	D
4 NF2	F	72	1.5	L	Larger tumor	IC tumor	Worse	25	70	A
5 NF2	M	24	1.5	R	Smaller tumor	Large tumor 3.5 cm	Equal	20	100	A
6 NF2	M	33	CI	L	Same size	IC tumor	Equal	20	100	A
7 NF2	F	27	2.5	L	Larger tumor	Previous RS	Better	90	0	D
8 NF2	F	24	2.5	L	Larger tumor	1 cm Tumor	Equal	110	0	D
9 Sporadic	F	52	1.5	L	Tumor side	Meniere HA user	Better	30	80	A
10 Sporadic	M	50	1.0	L	Tumor side	Progressive HL HA user	Better	70	60	C
11 Sporadic	M	53	CI	L	Tumor side	Middle ear surgeries	Better	55	100	C
12 Sporadic	M	42	1.2	L	Tumor side	Traumatic HL Anacusis >10years	Better	70	35	D
13 Sporadic	M	52	CI	L	Tumor side	Progressive HL, HA user	Better	110	0	D
14 Sporadic	M	50	0.5	L	Non-tumor side	Sudden HL	Better	20	100	A
15 Sporadic	F	70	1.5	R	Tumor side	Progressive HL HA user	Better	90	20	D

No	Preop. Contralateral hearing at implantation			Contralateral hearing at last follow-up			Hearing outcome in the CI only condition			Open SDS in bilateral condition (%)/Hearing mainly based on	Follow-up (mo)	2nd CI
	PTA (dB)	SDS (%)	Hearing class	PTA (dB)	SDS (%)	Hearing class	User	Closed set (%)	Open set (%)			
1 NF2	30	100	A	35	100	B	Yes	100	70	100/Contralateral hearing	21	No
2 NF2	110	0	D	110	0	D	Yes	100	65	65/Ipsilateral CI	124	No
3 NF2	25	100	A	35	80	B	Yes	100	60	100/Contralateral hearing	48	No
4 NF2	20	100	A	35	80	B	Yes	90	45	100/Contralateral hearing	18	No
5 NF2	20	100	A	20	100	A	No	0	0	100/Contralateral hearing	12	No
6 NF2	20	100	A	30	80	B	No	40	0	100/Contralateral hearing	60	No
7 NF2	110	0	D	110	0	D	No[a]	50	0	0/Lipreading	78	No
8 NF2	110	0	D	110	0	D	Yes	50	0	0/Ipsilateral CI	60	No
9 Sporadic	50	70	B	50	70	B	Yes	100	80	100/CI + HA	30	No
10 Sporadic	65	70	C	65	70	C	Yes	100	80	80/CI + HA	14	No
11 Sporadic	110	0	D	110	0	D	Yes	100	65	100/Bilateral CI	46	Yes
12 Sporadic	110	0	D	110	0	D	Yes	100	60	65/Ipsilateral CI	114	No
13 Sporadic	60	80	C	60	80	C	Yes	45	50	80/CI + HA	12	No
14 Sporadic	110	0	D	110	0	D	No	0	0	100/Contralateral CI	12	Yes
15 Sporadic	65	60	C	65	60	C	No, explanted			60/Contralateral HA	14	No

Outcome of cochlear implantation in NF2 patients (1–8), and patients with VS in the only or best hearing ear (Sporadic 9–15)

Hearing classification according to the AAO-HNS 1995 guidelines and their equivalence to traditional hearing criteria. Class A PTA 0–30 dB, SDS 70–100 %, Class B PTA 31–50 dB, SDS 50–100 %, Class C PTA 51–100 dB, SDS 50–100 %, Class D any SDS 0-49 %

CI cochear implant, *HA* hearing aid, *HL* hearing loss, *SDS* speech discrimination score, *IC* intracanalicular. *L* left, *R* right, *TL* translabyrinthine, *RS* retrosigmoid

[a] This patient was user for three years with progressive decline in performance that led to explantation

Table 2 Main clinical, radiological and audiological features of NF2 patients and sporadic VS patients

	NF2 patients ($n = 8$)	Sporadic VS patients ($n = 7$)	Total
Age (years)	39.4 (24–72)	52.7 (42–70)	
Sex			
Female	6 (75 %)	2 (28.6 %)	8
Male	2 (25 %)	5 (71.4 %)	7
Follow-up (months)	52.6 (12–124)	34.3 (12–114)	
Tumor size (CPA cm)	1.71 (0–4.0)	0.81 (0–1.5)	
Preoperative hearing			
Ipsilateral			
Mean PTA (dB)	60 (20–110)	63.57 (20–110)	
Mean SDS (%)	45 (0–100)	56.43 (0–100)	
Class A	3	2	5
Class B	1	0	1
Class C	0	2	2
Class D	4	3	7
Contralateral			
Mean PTA (dB)	55.6 (20–110)	81.4 (50–110)	
Mean SDS (%)	62.5 (0–100)	40 (0–80)	
Class A	5	0	5
Class B	0	1	1
Class C	0	3	3
Class D	3	3	6
Approach			
Translabyrinthine	4 (50 %)	7 (100 %)	11
Transmastoid	4 (50 %)	0	4
Surgery			
Simultaneous	4 (50 %)	7 (100 %)	11
Staged	3 (37.5 %)	0	3
No (Radiation)	1 (12.5 %)	0	1
Previous Radiation			
No	6 (75 %)	7 (100 %)	13
Yes	2 (25 %)	0	2
Postoperative hearing			
Closed-set discrimination (%)	66.2 (0–100)	74.2 (0–100)	
Open-set discrimination (%)	30 (0–100)	55.8 (0–80)	
User			
Yes	5 (62.5 %)	5 (71.4 %)	10
No	3 (37.5 %)	2 (28.6 %)	5

Hearing classification according to the AAO-HNS 1995 guidelines and their equivalence to traditional hearing criteria. Class A PTA 0–30 dB, SDS 70–100 %, Class B PTA 31–50 dB, SDS 50–100 %, Class C PTA 51–100 dB, SDS 50–100 %, Class D any SDS 0–49 %

implantation was performed through the round window or cochleostomy after translabyrinthine approach and through standard posterior tympanotomy in the remaining cases. In one case (patient 6) a partial drill out was performed for cochlear ossification after middle fossa removal of VS.

Audiological outcomes

Considering the whole group, all but one patient (patient 15, explanted for cerebrospinal fluid—CSF-leak) had sound detection. In the CI-only condition, 64 % of them achieved open-set discrimination (mean 70 ± 38 %), whereas 85 % achieved closed-set discrimination (mean 41 ± 33 %) (Fig. 1).

NF2 cases

This study includes 8 patients with NF2 (Tables 1, 2). In these patients the larger or equal tumor side was selected for implantation in 7 cases. Only in one case, the smaller tumor was chosen. This patient (patient 5) achieved only sound detection on the implanted side. Of the 8 NF2

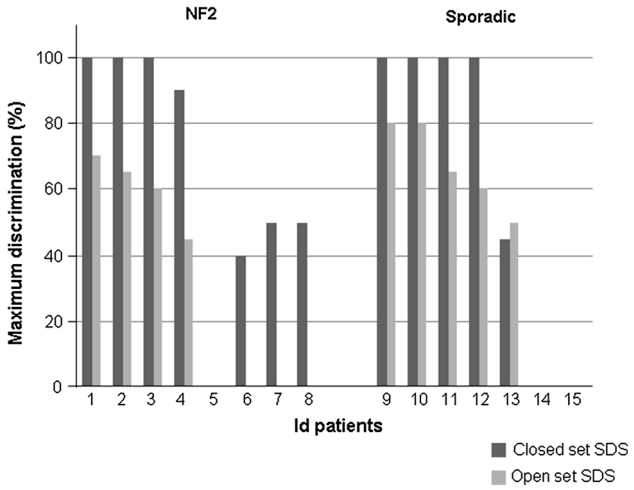

Fig. 1 Hearing outcome with ipsilateral cochlear implantation, both in NF2 patients and in sporadic cases

patients, the side with worse or equal hearing was chosen for implantation in 6 cases. The other 2 cases had better hearing on the implanted side, with one of the patients (patient 7) achieving initially closed-set discrimination, and the other one (patient 2) achieving open-set discrimination. None of the patients received a second implant in this group.

Sporadic cases

This study includes 7 patients with VS in the only or better ear (Tables 1, 2). Hearing class in the contralateral ear was class B in one case, class C in 3 cases, and class D in 3 cases. The tumor side was initially chosen for implantation in 6 out of the 7 cases. Two patients received a second implant, which was placed on the tumor and non-tumor side, respectively (patients 14 and 11). These two cases will be described in detail. Patient 14 had a 20-year anacusis due to a sudden hearing loss on the right side and a growing VS with 5 mm in the CPA on the left side. He received a right CI with acceptable outcome. One year later he underwent a translabyrinthine approach on the left side with tumor resection and simultaneous cochlear implantation. He achieved sound detection with no open-set abilities with his left CI. However, there was a marked increase in the right CI performances after contralateral tumor removal, which led to telephone use. The other patient (patient 11) had a long-term right deafness due to several tympanoplasties performed more than 25 years before. In 2007 he developed a sudden left hearing loss with partial recovery. Mean PTA threshold was 35 dB. Magnetic resonance imaging (MRI) showed a 10 mm intracanalicular left VS. One year later his hearing deteriorated with a mean PTA threshold of 50 dB. In 2009, he underwent a translabyrinthine approach on the left side with tumor resection and simultaneous cochlear implantation. SDS was 65 %. Two years later, right cochlear implantation was performed. He achieved excellent results with SDS 79 % and sentences 99 %. The results with both implants are SDS 88 % and sentences 100 %. He is using both implants daily.

Impact of variables on the audiological outcome

No differences between NF2 patients and patients with VS in the only or better ear (sporadic group) were found in terms of gender, age at implantation, or preoperative hearing. As expected, preoperative contralateral hearing was poorer in the sporadic group, with 86 % of the patients having contralateral classes C or D while only 37 % of the NF2 patients had contralateral classes C-D (Fisher test, $p = 0.026$).

The percentage of patients achieving open-set discrimination with the ipsilateral CI was 50 and 71 % for the NF2 and the sporadic group, respectively. The difference was no statistically significant (Fisher test, $p = 0.3$). There was an association between age and achieving open-set ability. Mean age for patients who had postoperative open-set discrimination was 50.7 years (SD 11.7) whereas mean age for those who had no open-set capability was 31.6 years (SD 10.9) (Mann–Whitney $p = 0.013$). Other factors as gender, preoperative hearing, or simultaneous vs staged implantation, had no impact on SDS. Interestingly, no correlation was found between tumor size and hearing results. Two patients underwent RT, one as the only treatment and the other one previous to surgery. None of the patients receiving RT achieved open-set discrimination.

Follow-up and use of the device

At the last follow-up (mean 44 months, range 12–124), 10 patients (67 %) were using the CI, 5 NF2 patients and 5 of the sporadic VS patients. Among the 5 non-users, one patient (patient 7) achieved sound detection, and the implant helped her lip reading. She was very happy with her device although she did not achieve word recognition. 6 months following surgery she needed higher stimulation levels, which increased during the next months. She was a daily user of the CI during almost three years, until she achieved no benefit with it. She was explanted in May 2009. Another patient (patient 15) was explanted because of persistent CSF leak before obtaining any benefit from the device. Patients 14 and 6 do not use the ipsilateral CI because of not achieving open-set abilities, patient 14 also having facial stimulation at higher intensity. His communication relies on contralateral CI with which he is able to use the telephone. Patient 5 has only sound detection with no closed or open discrimination with his CI. As he still has

good hearing in the contralateral side, he is not taking advantage of the implant.

Communication abilities

Considering global communication abilities, hearing was mainly based on the ipsilateral implantation in 2 out of the 5 NF2 patients who were daily users (Table 1). The other 3 patients used the implant daily, although contralateral hearing was class A or B. In this group, in bilateral condition, 5 patients had 100 % SDS, one had 65 %, and 2 patients had no word discrimination at all. In the sporadic group, hearing was based exclusively on the ipsilateral CI in one patient, on bimodal stimulation in 3 patients, on bilateral CI in one patient, and on contralateral CI or hearing aid in 2 patients (Table 1). In this group all patients achieved at least 60 % of open-set discrimination in the bilateral condition.

Postoperative imaging

Postoperative imaging policy following implantation was variable among the patients. Six out of the 7 non-NF2 patients underwent postoperative CT scans. Of the 8 NF2 patients, 6 underwent at least one 1.5 tesla MRI, all of them with the internal magnet in place. Different grades of artifacts were found depending on the CI manufacturer and the sequences used.

Discussion

Outcomes of cochlear implantation in patients with sporadic VS in the only or best hearing ear and in patients with NF2

Our study confirms literature data reporting that most patients undergoing cochlear implantation in the same side of a VS, either after surgical removal or after RT treatment, benefit from their device and are users. Closed-set discrimination was possible in 86 % of the patients with a mean discrimination score of 70 ± 38 %, whereas open-set discrimination was achieved by 64 % of patients with a mean postoperative score of 41 ± 33 %. At the last follow-up, 10 out of 15 patients were CI users. However, it should be noted that the preoperative hearing and tumoral status of the patients varied widely in this series. 6 out of the 15 patients had no hearing on the contralateral ear and for 3 out of them, the VS was small with still useful preoperative hearing (patients 11, 12, 14). Two of these patients achieved open-set discrimination. On the other hand, four patients had small tumors and good contralateral hearing (patients 1, 4, 5, 9), three of them achieving open-set discrimination. The reasons for not using the device at the last follow-up were also variable. Of the five non-users, one was explanted due to CSF leak (patient 15), three due to not achieving open-set discrimination (patients 5, 6, 14), two of them with contralateral good hearing, and the last patient (patient 7) had deterioration of hearing following initial acceptable results.

Since 1992, when Cueva et al. [9] described the electrical promontory results in six patients following VS resection, and 10 years later Hoffman described the first case of cochlear implantation in a patient with NF2 [3], about 85 cases of ipsilateral cochlear implantation in patients with VS have been reported to date, both in NF2 and non-NF2 patients [4–6, 10, 11]. When including both populations, the present study represents the largest multi-institutional series, with 15 cases. In general, the hearing results of cochlear implantation in patients with VS may seem disappointing when compared to traditional postlingual CI users, whereas they compare favorably with those undergoing ABI [12]. However, recent studies show that approximately 60 % to 70 % of CI users in VS ears achieve open-set abilities [4, 13]. It has been suggested that while cochlear implantation outcomes may decrease with time due to long-term loss of axons, the audiological performance of ABI user will improve in the years following implantation. In our study, one patient (patient 7) showed a decrease in performance, which led to explantation 2 years after surgery. This patient had undergone RT 5 years before implantation (See "Results"). High charge levels may indicate loss of axons, which can occur even several years following RT. Carlson et al. [4] described a very similar case. The authors wonder if delayed radiation injury, unidentified device malfunction or increasing tumor burden could explain the loss in performance. No other patients showed any decrease in their CI performance in our series. Moreover, an increase in performance occurred in patient 14 after contralateral tumor resection. Interestingly, two patients in this study maintain their open-set capability after more than 10 years of follow-up. This finding suggests that hearing outcomes may remain stable with time, at least in non-irradiated patients undergoing total tumor resection.

Factors affecting outcome

As it has also been demonstrated in previous studies, hearing results after implantation in an ear with VS vary dramatically [14]. Several prognostic factors have been proposed, including duration of deafness, prolonged time between VS resection and implantation, cochlear ossification, and hearing in the contralateral ear [4, 14, 15]. Most series analyzing prognostic factors have a small number of cases and include only NF2 patients. In our study, the age

of patients who achieved postoperative open-set discrimination was significantly higher than those who did not achieve this capability. This finding may be related to the fact that age at implantation was higher in the sporadic group compared to the NF2 group, although the difference was not significant probably due to the small cohort size. Interestingly, no difference was found between the NF2 and the sporadic VS groups in terms of audiological outcomes. Other factors as preoperative hearing, tumor size, radiation or treatment approach (simultaneous vs staged) had no impact on the audiological outcome. The presence of contralateral residual hearing has been stated as a negative prognostic factor for cochlear implantation in patients with VS. Some authors [14, 15] have suggested that integration of the signal from the CI with the contralateral natural hearing may be a difficult task for some patients. In our study as well as in the recent paper by Lloyd et al. [16] contralateral hearing had no impact on the CI performance.

Variability in auditory performance depends ultimately on the status of the cochlear nerve. The mechanisms underlying hearing loss in patients with VS include vascular compression of the internal auditory artery, intratumoral bleeding, biochemical changes in the inner ear, and direct compression of the cochlear nerve. Surgery may also cause mechanical or thermal injury in the cochlear nerve or the labyrinthine artery. It has been suggested that a vascular compromise causing hearing loss may lead to successful outcomes following CI, whereas significant neuronal injury due to tumor growth, RT or surgical trauma may be associated with poorer outcomes [14]. In recent years different electrophysiological tests have been used to determine intraoperative candidacy for CI. Nevertheless positive responses do not assure good CI performance whereas patients with negative responses may obtain some benefit from the device [11, 17]. Therefore, the decision of implantation is usually taken on an anatomical basis.

The role of radiation

The impact of RT on the audiological outcomes after cochlear implantation is not clear. It has been shown that hearing loss after RT occurs due to stria vascularis injury [18]. These findings suggest that CI performance following RT would compare favorably to outcomes after surgery. In the present study only 2 patients received RT, one of them showing a decrease in performance which led to explantation with the time, and none of them achieving open-set abilities. Lustig et al. [14] described two patients undergoing cochlear implantation after RT to their VS. One patient achieved open-set discrimination while the other one only had sound detection. Trotter et al. [19] reported 3 NF2 patients with VS in the only hearing ear treated with RT. Although the 3 patients were daily users, the authors emphasize the possibility of malignant transformation as well as the importance of tempering the expectations of long-term hearing in these patients. In a recent study, patients who had RT had worse discrimination results when compared to those with no treatment. While all patients who were implanted with no treatment to their tumors received significant benefit, only 1 out of 6 patients undergoing RT achieved open-set discrimination [5]. The impact of previous RT on ABI outcomes has been also evaluated. Siegbahn et al. [20] have recently reported the results of 20 NF2 patients undergoing ABI. Nine of them had undergone gamma knife treatment before the ABI surgery. Seven of these patients reported benefit from their implant and were users, and two patients were categorized as part-time users. In the study by Grayeli et al. [21] about 31 ABI recipients, three patients had a previous irradiation on the side of the ABI. These patients were among excellent ($n = 1$) and good ($n = 2$) performers. RT had no obvious adverse effect on implant function in these two studies.

Surgical considerations

To increase the probability of a successful cochlear implantation, subtotal tumor resection has been proposed, especially in NF2 patients. In a recent review of the literature, Carlson et al. [4] described 3 patients undergoing less-than-total resection followed by cochlear implantation, all of them achieving open-set abilities. In our study, all the patients—in both the NF2 and the sporadic groups—underwent total tumor resection, audiological outcomes being similar to those reported in previous studies as a group [16]. The fact that even partial resection does not assure a superior outcome, the need for life-long tumor surveillance and increased tumor growth rates in NF2 patients, and the difficulty of performing imaging studies following cochlear implantation suggest that complete or at least near-total tumor resection should be generally attempted when possible. In this study six NF2 patients underwent postoperative MRI. MRI with a CI is becoming more frequent in the last years. Much of the concern over this procedure revolves under issues related to the magnet. Main concerns are potential displacement or heating of the device, the artifact generated by the magnet, demagnetization of the internal magnet, and electrical interference with the CI electronics [22]. While removal of the magnet was necessary some years ago, it has been recently demonstrated that MRI scanning without magnet removal is safe and well tolerated. Walton et al. [23] have reported 76 MRI scans performed in 13 patients with CI or ABI without magnet removal. Using appropriate MRI sequences, the scans were well tolerated in the majority of

patients, and it was possible to study not only the contralateral but also the ipsilateral internal auditory canal (IAC) and cerebellopontine angle (CPA). According to their classification, the view of the ipsilateral IAC-CPA was unimpaired (grade 0) in 85 % of head scans, distorted (grade 1) in 13 %, and entirely obscured by artifact (grade 2) in 2 % of the cases.

Intervention planning in NF2 patients

Several patients with NF2 will progress to bilateral deafness in the long term. Interestingly, 4 out of the 5 NF2 patients with useful contralateral hearing showed some grade of deterioration from the date of implantation to the last follow-up. Therefore, early surgical intervention for VS in these patients when the cochlear nerve can be spared is an important consideration to allow for possible cochlear implantation. In these NF2 cases, the largest tumor is usually operated first, CI being placed simultaneously—if a translabyrinthine approach is performed—, or sequentially if a hearing preservation approach is performed. Most surgeons initially treat the VS on the side that has the poorest SDS. Successful implantation allows the surgeon to be more aggressive with treatment of the contralateral tumor [14], instead of observing its growth. If contralateral hearing is useful, the possibility of a "sleeper" implant may be considered [4]. In this situation, the CI is placed in anticipation of complete hearing loss in the future in the contralateral ear, and the patients do not usually use the implant until contralateral hearing is lost [16]. In this study, two of the NF2 patients (patients 5 and 6) were non-users at the last follow-up due to the fact that contralateral hearing was 100 and 80 %, respectively. The performance will probably increase as contralateral hearing deteriorates. However, patient 1 was also implanted with a 100 % discrimination score at the contralateral side. Although she was advised that the implant was a sleeper, the patient is using it daily and gaining considerable benefit from the device. This finding reflects that she has no difficulty in merging the different signals from the two ears. Nevertheless, the optimal timing for implantation of a sleeper CI may be controversial as it depends not only on contralateral hearing, but also on ipsilateral hearing, tumor size, and patients' preferences. In this study the largest tumor side was approached first in all but one case, and the side with the worse or equal hearing was chosen for implantation in 6 out of the 8 cases. Although only 2 patients relied mainly on their ipsilateral CI for communication, it is likely that more patients will become active users as contralateral hearing deteriorates with the time.

Intervention planning in patients with sporadic VS in the only or best hearing ear

The presence of a VS in the only or best hearing ear represents a clinical challenge. In this group, only two patients were non-users, including the patient who was explanted due to CSF leak. The main difficulty in these cases is to decide the optimal time for therapeutic intervention. Tumor resection may lead to bilateral total deafness, whereas a wait and scan policy may lead to tumor growth, VIII nerve compression, and deterioration of hearing. Cochlear implantation appears as a reasonable option for these cases. Deciding to implant first the deaf side or the tumor side is a difficult task, which must be discussed with the patient. In the study by Di Lella et al. [6] on ten patients with VS in the only hearing ear, three patients reported a remarkable increase in their CI performance after tumor removal, what implied loss of hearing in which had been their only hearing ear. In the study by Mukherjee et al. [5] one patient was implanted in the non-tumor side, first doing poorly on that side and then requiring implantation on the tumor-affected side, without treating the tumor. In the present study the two possible strategies were considered. As a general policy, in a deaf ear with no contraindications and no tumor it seems more reasonable to implant this one first, before VS removal. However, a long duration of deafness and a deterioration of hearing on the tumor side hearing may be reason to implant the tumor side first. In any case the patient should be informed about both possible approaches and discuss the pros and cons with the surgeon.

Conclusions

In conclusion, cochlear implantation in patients with bilateral VS or VS in the only or best hearing ear may be an effective support for communication, providing open speech perception in selected cases. In light of our results, and data from previous studies, cochlear implantation should be considered the default strategy in VS patients with an intact ipsilateral cochlear nerve. Proper preoperative counseling and realistic expectations are mandatory. Although it is difficult to establish recommendations or protocols, even in patients with NF2 and large tumors, the possibility of sparing the cochlear nerve should be considered.

Conflict of interest The authors declare that there is no conflict of interests regarding the publication of this paper.

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