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Management Strategy of Vestibular Schwannoma in Neurofibromatosis Type 2

*Haitham Ahmad Odat, †Enrico Piccirillo, †Giuliano Sequino, †Abdelkader Taibah, and †Mario Sanna

**Department of Otolaryngology–Head and Neck Surgery, Jordan University of Science and Technology, Irbid-Jordan; †Department of Otology and Skull Base Surgery, Gruppo Otologico, Paicenza, Italy*

Objectives: To discuss our management strategy of vestibular schwannoma in patients with neurofibromatosis type 2 (NF2).
Study Design: Retrospective study.
Setting: Quaternary referral skull base center.
Methods: Thirty-eight NF2 patients who had undergone 48 operations at the Gruppo Otologico between January 1988 and December 2008. The mean age at time of surgery was 36.3 years (range, 17–65 yr), and the average tumor size was 3.1 cm (range, 0.6–6 cm). There were 27 female and 21 male ears, and 25cases were right side ears, whereas 23 were left sided. The average follow-up time was 3.7 years. Surgical approaches, hearing, and facial nerve functions, as well as hearing rehabilitation and facial nerve reconstruction outcomes, are discussed.
Results: Total tumor resection was achieved in 44 cases (92%). Facial nerve function was postoperatively House-Brackmann grades I–III in 36 cases (77%); it was grade I in 17 cases (35%) and grade II in 8 cases (17%). In 7 cases, hearing preservation was attempted, and a measurable hearing has been recorded in 5 cases (71%). Auditory brainstem implant was inserted in 25 cases, and concomitant cochlear implants were inserted in 5 cases.
Conclusion: Early diagnosis and treatment of bilateral vestibular schwannoma in patients with NF2 will achieve the best outcomes regarding facial nerve, hearing preservation, and postoperative complications. The watchful waiting policy will decrease the chance of reaching these goals. Cochlear implants and auditory brainstem implant have made hearing rehabilitation possible in NF2 patients who had bilateral sensorineural hearing loss. **Key Words:** Auditory brain stem implantation—Cochlear implantation—Hearing preservation surgery—Neurofibromatosis type 2—Vestibular schwannoma.
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Neurofibromatosis 2 (NF2) is a rare multisystem genetic disorder associated with bilateral vestibular schwannomas (VSs), spinal cord schwannomas, meningiomas, gliomas, and juvenile cataracts with a paucity of cutaneous features. It is an autosomal dominant disease, and its gene was mapped to chromosome 22 q 12-2, which codes for a tumor-suppressor protein termed *Merlin* or *Schwannomin*. This protein negatively regulates Schwann cell production. The loss of this protein allows overproduction of Schwann cells (1–3). There are 2 forms of NF2: the severe one (Wishart) with early presentation of the disease and (Gardner), which is the milder form (4).

Address correspondence and reprint requests to Haitham Ahmad Odat, M.D., Department of Otolaryngology–Head and Neck Surgery, Jordan University of Science and Technology, Irbid 22110, Jordan. P.O. Box 3238; E-mail: dr.haithamodat@gmail.com

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Bilateral VS is the most common and well-recognized feature of NF2 leading to significant morbidity (Fig. 1). The average age of diagnosis of NF2 is 25 years; however, many patients present with symptoms before the diagnosis. Symptoms, such as tinnitus, gradual hearing loss, and even vestibular dysfunction, are frequently the initial signs of NF2. Untreated VSs can extend locally and may result in brainstem compression, hydrocephalus, and occasionally, facial nerve palsy. NF2 is a familial syndrome in 50% of cases; for that reason, early screening by magnetic resonance imaging (MRI) and genetic blood testing are advised to diagnose tumors before the symptoms appear (5).

The treatment of patients with NF2 has always presented challenges for neurosurgeons and neurotologists. The timing of the intervention and the selection of surgical approach can be difficult (6).

To observe patients and postpone surgery to keep as much function for as long as possible or to surgically resect 1 tumor, usually the larger one, and to observe the other is not well agreed among authors (7). In this study,

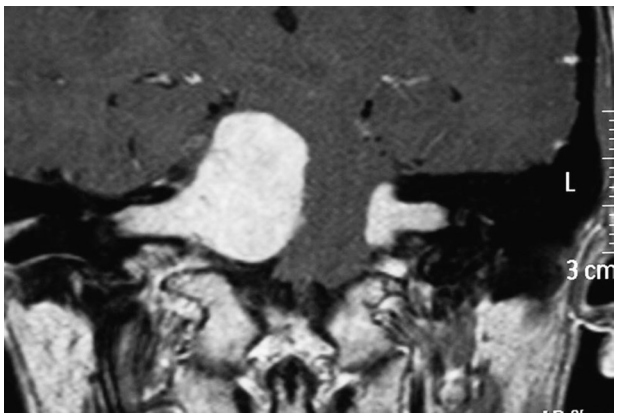

FIG. 1. Coronal T1-weighted MRI with contrast of NF2 patient with bilateral VS.

we discuss our experience in managing NF2 patients. Our results of hearing preservation, hearing rehabilitation, postoperative facial nerve function and rehabilitation, and complications will be discussed.

METHODS

Patient Population

Of the 2,000 VS surgery, which had been operated by the senior author (M. S.), 48 operations (2%) for 38 NF2 patients between January 1988 and December 2008 were included in this article. In 10 patients, bilateral tumor resection was done. There were 27 female and 21 male ears; 25 (52%) were right side ears, whereas 23 (48%) were left sided.

The average age at the time of surgery was 36.3 years (range, 17–65 yr), with standard deviation of 13.4 years. The average hospital stay was 5.4 days (from4 to 9 d), and the mean follow-up time was 3.7 years (ranged from 1 mo to 20 yr).

Tumor Size and Hearing Assessment

The tumors were measured according to the largest cerebellopontine angle component. Preoperative and postoperative hearing was classified according to the Tokyo consensus meeting on systems for reporting results in VS (8).

Measures of speech and sound perception in cochlear implant (CI)– and auditory brainstem implant (ABI)–implanted patients are as follows: Italian version of the Northwestern University Phonetically Balanced Word List (NU 6) and Central Institute for the Deaf Everyday Sentence List (CID sentence) were used to measure speech perception benefits. The speech material presented in hearing-only conditions using a monitored live voice through the sound field at 70 dB and signal to noise ratio of 15 dB (9).

Facial Nerve

Facial nerve function was analyzed using the House-Brackmann (H-B) grading system.

Surgery

Hearing preservation and nonhearing preservation operations were selected according to the tumor size and preoperative hearing status (pure tone audiometry [PTA] and speech discrimination score [SDS]; Fig. 2).

Facial nerve monitoring was applied in all cases, and intraoperative fast auditory brainstem response (ABR) combined with cochlear nerve action potential (CNAP) was used in cases of hearing preservation surgery (HPS). Hearing rehabilitation was performed in 28 cases using ABI or simultaneous CIs.

RESULTS

Forty-eight operations for 38 patients with NF2 were done; average age at time of surgery was 36.3 years (range, 17–65 yr; Tables 1 and 2).

The mean tumor size was 3.1 cm, ranging from 0.6 to 6 cm (standard deviation, ±1.2). Preoperatively, the facial nerve function was H-B grade I in 46 cases (96%), and it was H-B grades I-III postoperatively in 36 cases (77%; Fig. 3).

VS was found to be grade I in 6%, grade II in 27%, grade III in 33%, grade IV in 23%, and grade V in 10%. In 66%, the tumors were equal or less than 3.0 cm, whereas 33% were more than 3.0 cm (Table 3). Total tumor excision was achieved in 44 cases (92%). Subtotal tumor resection was done in 4 cases (8%) because there was a poor cleavage plane between the tumor and the facial nerve, so it was wise to leave a capsular remnant over the nerve to preserve its function. These patients were followed up by annual fat suppression MRI. By an average of 4 years of follow-up time, 1 patient showed tumor regrowth, which was resected by enlarged translabyrinthine-transapical approach (ETLA-TA) approach.

In regard to the tumor size, the facial nerve function was excellent (H-B grades I–II) in 9 cases (56%), and it was H-B grades I to III in 15 cases (94%) when the tumors were less than 2 cm, whereas it was H-B grades I to III only in 68% of tumors larger than 2.0 cm (Table 4). In 7 cases with H-B grade VI after surgery, facial nerve reconstruction has been done. End-to-end anastomosis was done in 2 cases, facial nerve grafting using a sural

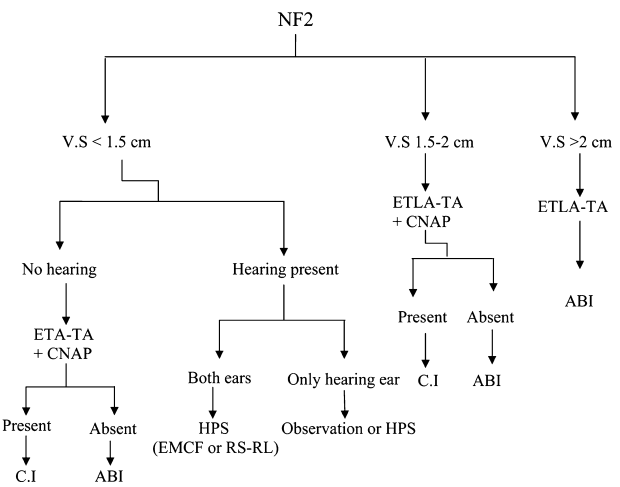

FIG. 2. Management algorithm used at Gruppo Otologico. This scheme represents a general guideline only. Individualized treatment selection is the rule. EMCF, enlarged middle cranial fossa; RS-RL, retrosigmoid-retrolabrynthine.

TABLE 1. *NF2 patients case by case*

Case no.	Age/sex	Ear	Size (cm)	Preoperative PTA	Preoperative SDS	Facial nerve preoperative	Approach	Postoperative PTA	Postoperative SDS	Facial nerve postoperative	HR
1	48/M	R	1.5	65	90	I	RS-RL	90	10	I	
2	49/M	L	1.2	55	100	I	ETLA-TA	120	0	III	ABI
3	65/M	R	1.3	110	0	I	ETLA-TA	120	0	I	
4	20/M	L	2.5	120	0	VI	MTC	120	0	VI	ABI
5	39/M	R	3.8	25	100	I	ETLA-TA	120	0	I	
6	41/F	R	3	120	0	I	ETLA-TA	120	0	I	ABI
7	22/M	L	2.5	120	0	I	ETLA-TA	120	0	I	
8	32/M	R	3.4	70	30	I	ETLA-TA	120	0	VI	ABI
9	33/F	R	3.5	95	10	I	ETLA-TA	120	0	I	ABI
10	34/F	L	3	100	0	I	ETLA-TA	120	0	I	ABI
11	23/F	R	3	35	90	I	ETLA-TA	120	0	III	
12	17/F	L	3.5	120	0	III	ETLA-TA	120	0	VI	ABI
13	17/F	R	2.8	30	90	I	ETLA-TA	120	0	VI	
14	23/F	L	4.5	120	0	I	ETLA-TA	120	0	VI	ABI
15	24/F	R	3.5	120	0	I	ETLA-TA	120	0	III	
16	43/F	L	2.5	120	0	I	ETLA-TA	120	0	II	ABI
17	50/F	L	3.5	120	0	I	ETLA-TA	120	0	VI	
18	31/F	L	3	85	30	I	ETLA-TA	120	0	I	ABI
19	38/M	L	2	50	0	I	ETLA-TA	120	0	III	CI
20	22/F	R	1.3	20	100	I	RS-RL	40	100	I	
21	61/M	R	4.5	95	0	I	ETLA-TA	120	0	VI	
22	28/F	R	2.8	35	100	I	ETLA-TA	120	0	V	
23	44/M	L	1.2	75	100	I	RS-RL	120	0	I	
24	25/F	L	2	50	100	I	TO	120	0	I	ABI
25	26/F	L	2	20	100	I	ETLA-TA	120	0	III	ABI
26	45/M	R	4	70	40	I	ETLA-TA	120	0	I	ABI
27	48/F	L	1	120	0	I	ETLA-TA	120	0	III	CI
28	46/M	R	1	30	100	I	RS-RL	35	90	I	
29	64/F	L	1.5	50	90	I	RS-RL	50	70	II	
30	28/F	R	1.7	20	100	I	ETLA-TA	120	0	III	
31	43/F	L	1.5	90	0	I	ETLA-TA	120	0	III	CI
32	39/M	R	6	100	0	I	TO	120	0	VI	ABI
33	39/M	L	2	85	80	I	TO	120	0	VI	
34	32/M	L	0.6	20	100	I	EMCF	120	0	II	CI
35	41/M	R	4	90	0	I	ETLA-TA	120	0	IV	ABI
36	33/F	R	4	55	75	I	ETLA-TA	120	0	VI	ABI
37	24/M	L	4.5	20	100	I	ETLA-TA	120	0	III	
38	26/M	R	2	20	100	I	RS-RL	100	30	I	CI
39	69/F	R	2.5	65	80	I	ETLA-TA	120	0	I	ABI
40	24/F	R	5	90	0	I	ETLA-TA	120	0	II	
41	25/F	L	2	85	0	I	ETLA-TA	120	0	II	ABI
42	51/M	L	3.5	35	100	I	ETLA-TA	120	0	I	ABI
43	54/M	R	2.5	65	30	I	ETLA-TA	120	0	III	ABI
44	26/F	R	4	100	10	I	ETLA-TA	120	0	II	ABI
45	27/F	L	2.5	120	0	I	TO	120	0	II	ABI
46	19/F	L	3	60	50	I	ETLA-TA	120	0	I	ABI
47	51/M	R	2.5	100	20	I	ETLA-TA	120	0	II	ABI
48	31/F	L	3	45	90	I	ETLA-TA	120	0	III	ABI

ABI indicates auditory brainstem implant; CI, cochlear implant; EMCF, enlarged middle cranial fossa; ETLA-TA, enlarged tanslabrynthine-transapical; F, female; HR, hearing rehabilitation; L, left; M, male; MTC, modified transcochlear; R, right; RS-RL, retrosigmoid-retrolabrynthine; PTA, pure tone audiometry; SDS, speech discrimination score; TO, transotic.

nerve was done in 4 cases, and hypoglossal-facial anastomosis was the selected approach in 1 case. H-B grade III was the end result in 3 (43%) of the reconstructed cases on the last follow-up visit.

A useful preoperative hearing (classes A, B, and C) was present in 19 cases (40%) with average tumor size of 2.5 cm. In 29 cases (60%), no useful hearing among the cases was found, with average tumor size of 2.7 cm. We could not find a relation between the tumor size and preoperative hearing loss (Table 5).

HPS was performed in 7 cases, and a useful postoperative hearing (classes A, B, and C) has been maintained using the retrosigmoid-retrolabrynthine and enlarged middle fossa approaches in 4 cases (57%), and in 5 cases (71%), a measurable hearing has been recorded. Although the tumor size and preoperative hearing are cardinal issues in selecting hearing or non-HPS, it is evident that the tumor size is not a predictor for perioperative hearing level and/or functional outcome in HPS (Table 6).

TABLE 2. *General characteristics of 48 operated ears for 38 patients with neurofibromatosis type 2*

	Average	Range	SD
Age	36.3	17–65	13.4
Size (cm)	3.1	0.6–6	1.2
Preoperative PTA	73.1	20–120	35.1
Preoperative SDS	45.9	0–100	44.5
Postoperative PTA	114.1	35–120	19.4
Postoperative SDS	6.3	0–100	21.5

PTA indicates pure tone audiometry; SD, standard deviation; SDS, speech discrimination score.

TABLE 3. *Tumor sizes according to the Tokyo consensus meeting*[a]

	Grade I	Grade II	Grade III	Grade IV	Grade V
Intrameatal	(0.1–1 cm)	(1.1–2 cm)	(2.1–3 cm)	(3.1–4 cm)	(>4 cm)
No. of cases	3	13	16	11	5

[a]New and modified reporting systems from the consensus meeting on systems for reporting results in vestibular schwannoma (7).

Non-HPS was considered in 41 cases (85%), and it has been chosen either because of large tumor size or significant preoperative hearing loss. The ETLA-TA was used in 36 cases (75%), transotic in 4cases (8%), and modified transcochlear in 1 case (2%). Hearing rehabilitation was performed by implantation of 25 ABI for 24 patients (Fig. 4) and 5 simultaneous CI.

The patients received ABI (24 Nucleus 24M ABI, 1 Digisonic SP ABI) at the removal of the first VS (14 times), at the removal of the residual first tumor (2 times), or at the removal of the second tumor (9 times). Two of these patients received ABI at first and second sides. First-side operation was done in another center, and therefore, it will not be calculated. Both patients had dislocation of their first ABI. We implanted a second ABI at the contralateral ear with better results.

We have seen variable results including some very good users (4 with more than 50% speech discrimination) and 3 with even telephone use and 75% to 100% speech discrimination. The overall results are unpredictable; 19 of the 23 patients we could follow up on are users, 8 have sound recognition, 11 have some kind of word recognition, and only 8 have speech recognition (Table 7) (9).

Simultaneous CI (Nucleus 24 Contour) was inserted in 5 cases (10%; Table 8). Four patients achieved good levels of auditory performance in open-set tests, scoring from 55% to 100% in comprehension tests at the 1-year follow-up. Two of these patients reached hearing outcomes similar to the best of standard postlingual adult implantees; also, they were able to easily understand phrases during telephone conversations (all the CI patients have been using their devices daily, and 4 have been telephone users). In 1 patient, the open-set score was 0%, and the bisyllabic word recognition was 10%. He finds the implant useful only for lip-reading and detecting environmental sounds.

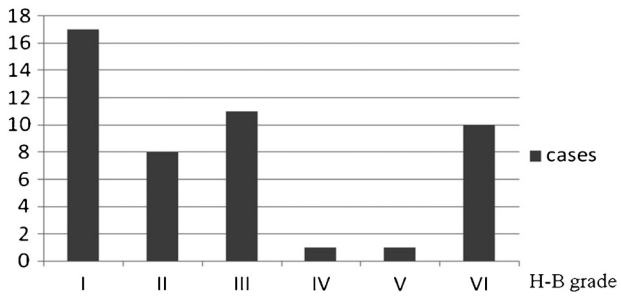

FIG. 3. Postoperative facial nerve function according to the H-B grading system.

Postoperative complications occurred in 6 cases (13%) (Table 9). In the 3 cases of postoperative lower cranial nerve palsy, the tumor sizes were larger than 4 cm, and they were compensated without surgical intervention (10).

Three of our patients were sent for radiotherapy, and during the follow-up time, 2 of them showed regrowth of their tumors (one after 3.5 yr and the other after 10 yr) and underwent complete surgical resection with facial nerve grade VI after surgery. The third patient was a 20-year-old male patient who had malignant transformation of his tumor 4 years after gamma knife radiotherapy. Surgical resection was done, and he died 3 months later (11).

DISCUSSION

NF2 is a devastating disease with multiple intracranial and extracranial tumors. Bilateral VS is the commonest feature of the disease. Well-accepted guidelines for the management of NF2 disease are still controversial (6,7), and the treatment changes widely as a result of the wide variety of tumor sizes and clinical presentations. Associated symptoms (brainstem compression or hydrocephalus), hearing status, and presence of other intracranial tumors must be considered in the management of these tumors.

Observation

The policy has been to wait for hearing deterioration before proceeding to the tumor removal (watchful waiting policy) (12). We did not find significant relationships between tumor size and the ability to preserve hearing. Neither preoperative PTA values nor SDSs were significantly related to the amount of hearing change. There were no other identifiable preoperative predictive factors, including preoperative hearing. Although only 7 patients underwent HPS, we could be able to preserve a significant hearing in 3 of them. In our experience in solitary VS surgery, we found that it is possible to preserve the cochlear nerve anatomically, but it is difficult to preserve the hearing in most of cases when the tumor is larger than 1.5 cm. However, individualized treatment should always be selected.

We agree with other authors (6,13) that watchful and scanning should no longer be considered the standard of care because this policy decreases the chance of hearing

TABLE 4. *Comparison of tumor sizes and facial nerve function*

	Tumor grade											
	Grade I		Grade II		Grade III		Grade IV		Grade V			
	0.1–1 cm		1.1–2 cm		2.1–3 cm		3.1–4 cm		>4 cm		Total	
House-Brackmann grade	Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post
I	3	1	13	6	15	6	10	4	5		46	17
II		1		1		4		1		1	0	8
III		1		5		3	1	1		1	1	11
IV								1				1
V						1						1
VI				1	1	2		4		3	1	10
Total	3	3	13	13	16	16	11	11	5	5	48	48

Pre indicates preoperative; Post, postoperative.

preservation and gives time for tumors to grow, which makes the surgery more difficult later on. Patients who undergo surgery when their tumors have reached a substantial size makes it difficult to preserve hearing and, in some cases, to preserve the facial nerve. Generally, VSs among patients with NF2 are more aggressive and tend to invade the cochlear nerve more than those of unilateral VSs, theoretically making hearing preservation more difficult (6).

In few situations in which patients have medical contraindications for surgery and/or they are affected by tumors in the only hearing ear, they are assessed routinely to ensure that brainstem compression or hydrocephalus does not result. MRI is performed 6 months after the diagnosis, and then, annual MRI scans are performed to document tumor size. Surgical intervention is considered when life-threatening complications occur, the tumors become excessively large (increasing the perioperative morbidity), or the hearing becomes unserviceable (5,6,13–15).

Radiotherapy

Stereotactic radiotherapy in NF2 patients is still controversial. It should be carefully considered because radiation exposure may induce or accelerate tumors in a patient with an inactivated tumor suppressor gene or rarely induce malignant transformation in addition if surgical resection is needed; it is much more complex with decreased chances of hearing and facial nerve preservation (3,11), although some authors consider it safe and effective in tumor control as well as low morbidity (16,17).

We agree with others that radiotherapy is not superior to microsurgery regarding tumor control and perioperative morbidity (6,13,15). In the present study, 2 of our patients showed regrowth of their tumors after radiotherapy (one after 3.5 yr and the other after 10 yr), and they had facial paralysis (H-B grade VI) after operations. Also, we had 1 case (20 yr old) who had had malignant transformation of his tumor 4 years after gamma knife radiotherapy; then, he underwent surgery and died 3 months later (11).

TABLE 5. *Preoperative hearing in relation to the tumor size*

Hearing class[a]	No. of cases preoperative (%)	Average tumor size
A	6 (13)	2.4 (0.6–4.5)
B	6 (13)	2.6 (0.8–3.5)
C	7 (15)	2.6 (1.2–6)
D	6 (13)	2.4 (0.8–4)
E	12 (25)	2.9 (1–4.5)
F	11 (23)	2.7 (1.2–5)

PTA indicates pure tone average at 0.5, 1.0, 2.0, and 4.0 kHz (15).

[a]Hearing classification according to the Tokyo consensus meeting: class A (PTA, 0–20; SDS, 100%–80%), class B (PTA, 21–40; SDS, 79%–60%), class C (PTA, 41–60; SDS, 59%–40%), class D (PTA, 61–80; SDS, 39%–20%), class E (PTA, 81–100; SDS, 19%–0%), and class F (PTA, >100).

Microsurgery

Decisions regarding the timing of the intervention and the type of surgical approach can be very difficult (6). The surgery should be carefully discussed with the patient and his/her family.

Although partial removal is not recommended by some authors (6), sometimes, it is mandatory to do subtotal resection in cases where intraoperative hearing monitoring indicates cochlear nerve function deterioration and when there is severe adherence of the tumor to the facial, cochlear nerves, intracranial vessels, or brainstem. In such situations, we leave a small piece of tumor in contact with these structures to avoid major complications.

Total Tumor Resection

The goal of surgery in patients with NF2 should be complete tumor removal but not at the expense of functional impairment (18).

We were able to totally remove VS unilaterally or bilaterally in 96% of cases; 92% in 1 stage, whereas in 2 cases (4%), 2-stage tumor resection was done via ETLA-TA and TO approaches as their tumors where large (5 and 6 cm) and adherent to the brainstem without cleavage plane. During dissection of the tumors from the brainstem, the patients had bradycardia, so we decided to stage the surgery. Samii et al. (7,15) have reported total removal in 87.5% and 85%.

According to preoperative hearing status and tumor size, total tumor removal could be done via either hearing

TABLE 6. *Hearing preservation surgery*

Approach	Tumor size (cm)	PTA preoperative	SDS (%) preoperative	PTA postoperative	SDS (%) postoperative
1. RS-RL	1.5	65	90	90	10
2. RS-RL	1.3	20	100	40	100
3. RS-RL	1.2	75	100	120	0
4. RS-RL	1	30	100	35	100
5. RS-RL	1.5	50	90	50	70
6. EMCF	0.6	20	100	120	0
7. RS-RL	2	30	100	100	30

EMCF indicates enlarged middle cranial fossa approach; PTA, pure tone average; RS-RL, retrosigmoid-retrolabrynthine approach; SDS, speech discrimination score.

preservation or nonhearing preservation operations. In Figure 2, we describe a general scheme for surgical management of NF2. However, individualized treatment selection depends on numerous factors including the level of serviceable hearing, the depth of tumor extension into the internal auditory canal, the size of the tumor, and the experience and familiarity of the surgeon (19).

Hearing Preservation Surgery

Most of the recent articles talked about hearing and facial nerve preservation in NF2 patients (6,7,13–15,20,21) because these are the most disabling outcomes either of the disease itself or of the surgery performed for excision of bilateral VS .There is a general agreement among authors that early diagnosis and intervention are mandatory to achieve those goals (6,7,13–15,18,20–22) because the chances of preserving hearing and facial functions decrease as time goes on. Such general agreement is related to the fact that the pattern of growth of tumor in NF2 is variable, which makes it difficult to predict their behaviors, and the delay in diagnosis may cause early loss of hearing and facial nerve function. Because early diagnosis allows detection of the tumors while they are small and hearing function is still good, they could be relatively easily resected with low chances of perioperative complications.

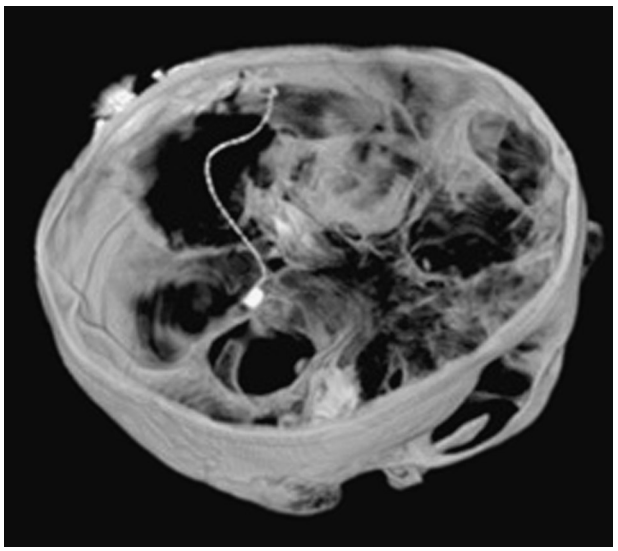

FIG. 4. Three-dimensional computed tomographic scan of the postoperative result after placement of a right-side ABI.

Patients with only hearing ears represent a great challenge for surgeons. Proper counseling and explanation of pros and cons must be discussed carefully with patients. We prefer to observe these patients with annual MRI scan and audiometric examinations. Once their hearing starts to deteriorate or the tumor size increases (>0.2 cm/yr), we advise HPS. Should the hearing could not be preserved but present CNAP, then CI could be simultaneously implanted, whereas if the CNAP could not be preserved, then ABI is recommended (23,24). As VSs among patients with NF2 are more aggressive and tend to invade the cochlear nerve, we attempt hearing preservation when the tumor size is inferior to 1.5 cm and hearing is present. In cases where VS is greater than 2 cm, it is difficult in most cases to preserve the hearing. However, CI in preserved cochlear nerve function and ABI when it is lost are the available options.

In our study, only 7 cases underwent HPS because the majority of cases (67%) had large size tumors (>2 cm), and 60% of the cases had no useful hearing before the operations. By using retrosigmoid-retrolabrynthine and enlarged middle fossa approaches, we were able to preserve some hearing in 5 cases (71%) and useful hearing in 4 cases (57%).

The hearing preservation success rate varies widely in literatures from 24% to 70 % (6,7,13–15,20,21). Samii et al. (7,15) reported a 57% success rate using retrosigmoid approach in 30 NF2 cases where the tumor sizes were less than 3 cm. Slattery reported a 55% hearing preservation rate by using the middle fossa approach in 47 pediatric NF2 cases (13). Brackmann reported 65% hearing preservation rate in 28 operated ears (6).

Nonhearing Preservation Surgery

Most NF2 patients present either when the tumor is too large for hearing preservation or the hearing loss is already at a significant level and hearing cannot be preserved.

In the present study, non-HPS was considered in 41 cases (85%) for the same indications previously mentioned. The ETLA-TA approach was used in 36 cases (75%), transotic in 4 cases (8%), and modified transcochlear in 1 case (2%).

TABLE 7. *Electrodes and auditory outcomes*

No.	Electrode activation	Electrode used	User	Sound	Words	Sentence	Speech
1	21	21	Y	100	90	100	100[a]
2	14	19	Y	100	70	72	80[a]
3	20	18	Y	100	80	70	75[a]
4	13	13	Y	70	0	0	0
5	17	19	Y	60	0	0	0
6	11	10	Y	90	30	15	20
7	11	13	Y	80	10	0	0
8	17	12	Y	100	30	35	30
9			[c]				
10	5	10	Y	90	65	35	40
11	7	6	Y	80	10	0	0
12a	0	0	N				
13	0	0	N	0	0	0	0
14	8	8	Y	0	0	0	0[b]
15	0	0	N	0	0	0	0
16	21	21	Y	100	60	65	70[a]
17	13	13	Y	40	0	0	0
18	0	0	N	0	0	0	0
19	12	12	Y	85	20	10	0
20	5	5	Y	60	0	0	0
21	0	0	N	0	0	0	0
22	17	17	Y	75	0	0	0
23	9	9	Y	70	0	0	0
12b	11	11	Y	85	20	15	20
24	21	20	Y	65	0	0	0

N indicates no, Y, yes.
[a]Patient uses telephone.
[b]Auditory brainstem implant ''sleeper.''
[c]The patient died.

Facial Nerve Function

Facial nerve preservation is a priority in dealing with NF2 patients because they often are young and bilaterally affected. Brackmann et al. (6) reported facial nerve preservation (grades I–III) in 92.5% immediately after surgery and 99.9% on long-term follow-up. Samii et al. (7,15) reported an 89% success rate. The results by Slattery et al. (13) and Macnally et al. (20) were 81% and 88%, respectively. Facial nerve function of grades I to III was preserved in 94% of our cases, with mean follow-up time of 3.7 years.

Hearing Rehabilitation

With the knowledge that the majority of the patients with NF2 will progress to bilateral deafness sooner or later, a hearing rehabilitation is usually necessary (23,25). NF2 patients are rarely CI candidates, although if the cochlear nerve is preserved, they could greatly benefit from this (10,26–29). We routinely use intraoperative fast ABR and CNAP simultaneously to monitor cochlear nerve function in the retrosigmoid-retrolabrynthine approach. In the setting of preserved cochlear nerve with negative fast ABR, but preserved CNAP, the option of a CI can be considered. However, where the fast ABR and the morphology and amplitude of the CNAP are significantly degraded at the brainstem, the placement of an ABI should be strongly considered at the time of tumor removal (9,24).

Simultaneous CI was performed in 5 patients, 4 achieved open-set speech recognition abilities comparable to those of standard adult postlingual implant patients, whereas the remaining patient reported benefits only in environmental sound detection and lip-reading. We prefer simultaneous CI because rapid and progressive osteoneogenesis can occur into the cochlea after VS resection, mainly in the labyrinthectomized ear (30), although successful implantation has been performed up to 18 months after labyrinthectomy (31). Whether the benefits of cochlear implantation in patients with NF2 remain stable overtime requires multi-institutional, prospective trials. Currently, the report on long-term results demonstrated that hearing performance did not deteriorate over an extended postoperative time course (28).

Usually, individuals with NF2 who have operations to remove their VSs lose their cochlear nerves and are not suitable for CI (25). This was the case in 24 of our patients were 25 ABI were implanted. Speech perception results do not match with the good results seen in modern cochlear implantation, but the auditory sensations provided by ABI can be very helpful in facilitating oral communication of the NF2 patient. The overall outcome is that the patient benefits at least an increased awareness of their surroundings, and when asked in a questionnaire, many say they benefit greatly from their ABI (25,32). We have seen the same variable results including some very good users (4 with more than 50% speech discrimination) and 3 with even telephone use and 75% to 100% speech discrimination.

In our center and in concordance with the majority of other centers (25,32,33), we recommend implantation at the time of the first-side tumor removal. A small percentage will not respond to their ABI, and these patients could have a second chance at the second removal. Patients should be carefully counseled because the results

TABLE 8. *Hearing results in cochlear implant patients*

	Vowel identification (%)	Consonant identification (%)	Bisyllabic word recognition (%)	Sentence recognition (%)	Common phrases comprehension (%)
Case	12 mo	12 mo	12 mo	12 mo	12 mo
1	100	100	80	90	100
2	100	100	72	81	86
3	100	100	50	50	55
4	40	39	10	0	0
5	100	100	78	85	93

Mo indicates months.

TABLE 9. *Postoperative complications*

Complication	No. of cases	Comment
Lower cranial nerve palsy	3	Conservative management
VIth cranial nerve palsy	1	Temporary
Postauricular seroma	1	Aspiration and compressive dressing
Abdominal hematoma	1	Wound was opened and a drain was inserted
Cerebrospinal fluid leak	0	—
Death	0	—

can vary a lot and the device tuning and rehabilitation can take much longer than patients expect.

CONCLUSION

Early diagnosis and intervention of NF2 will achieve the best outcomes regarding facial nerve, hearing preservation, and postoperative complications. The watchful waiting policy will decrease the chance of reaching these goals and should not be considered the standard policy of treatment. Stereotactic radiotherapy is controversial and should be carefully considered in patients with NF2. CI and ABI development have made hearing rehabilitation possible in NF2 patients who had bilateral sensorineural hearing loss because they provide good support in the communications skills of these patients.

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