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Strategy for the surgical treatment of vestibular schwannomas in patients with neurofibromatosis type 2

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ABSTRACT

Objective: Guidelines for appropriate management of vestibular schwannomas in NF2 patients are controversial. In this paper we reviewed our experience with patients with NF2 for the results of surgical treatment with particular reference to hearing and facial nerve preservation.

Methods: We included in the study 30 patients (16 women and 14 men) with the diagnosis of NF2 treated in our department between 1998 and 2014 who underwent surgery for vestibular schwannoma removal with a follow-up for at least 1 year. In 3 cases, the vestibular schwannomas were unilateral. Six patients with bilateral vestibular schwannomas underwent unilateral procedure. Therefore, 51 acoustic tumors were studied in 30 patients.

Results: No operative death we noted. Significant deterioration to the non-functional level occurred in 19 out of 22 cases with well-preserved preoperative hearing. Only three ears maintained their preoperative good hearing. Hearing was preserved in cases of small schwannoma not exceeding 2 cm. Among 21 patients who underwent bilateral operations hearing was preserved in 3 out of 7 cases when smaller tumor or better hearing level side was attempted at first surgery. In contrary none of the 14 patients retained hearing when the first operation concerned the worse-hearing ear. Among 14 tumors up to 2 cm there was only one case of moderately severe facial nerve dysfunction (House–Brackmann Grade IV) in the long follow-up.

Conclusion: Early surgical intervention for vestibular schwannoma in NF2 patient is a viable management strategy to maintain hearing function and preserve facial nerve function.

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1. Introduction

Neurofibromatosis type 2 is an autosomal dominant syndrome caused by mutation of the NF2 tumor suppressor gene, located at 22q12.2 [1,2]. The incidence is approximately one case in 25,000 live births, and the prevalence is 1 case in 100,000–200,000 people [3]. The syndrome has the highest spontaneous mutation rate of any human genetic disorder and approximately half of the cases represent new spontaneous mutations [4]. The hallmark of this disease is the development of bilateral vestibular schwannomas, which occurs in 90–95% of patients [5,6]. Other central and peripheral NF2-associated tumors include schwannomas of non-vestibular cranial, spinal, and peripheral nerves, meningiomas, spinal ependymomas and optic gliomas. Moreover, patients demonstrate non-neoplastic ocular manifestations.

This disorder is one of the most devastating and formidable conditions with increased morbidity rates and early death. This is due to these tumors' significant intracranial tumor burden and bilateral location, progressive growth despite its histologically benign nature and the capacity to produce further handicap by spinal cord compression.

This condition most commonly manifests itself with hearing loss. Guidelines for appropriate management of vestibular schwannomas in NF2 patients are controversial. Management strategy aims to preserve hearing and facial nerve function. Treatment options include observation, radiosurgery, and surgery. The clinical decision regarding the timing of the intervention is very difficult. For many neurosurgeons the risk of bilateral hearing loss secondary to attempted surgery is the reason for the suspension of the surgical removal of these lesions until they reach sufficient size to induce hearing loss, other cranial neuropathy, or brainstem compression [7]. Surgical removal of the tumor, however, only when it reaches a significant size (watchful waiting policy) reduces the chances of useful hearing preservation. In this paper we reviewed our experience with patients with NF2 for the results of surgical treatment with particular reference to hearing and facial nerve preservation.

2. Materials and methods

2.1. *Patient population*

34 patients with neurofibromatosis type 2, as defined on the basis of the modified National Institute of Health (NIH) Consensus Panel Criteria [8], were surgically treated at our institution between 1998 and 2014. We have retrospectively reviewed the clinical records, neuroimaging studies, and follow-up data of the treated patients. We included in the study patients with the diagnosis of NF2 who underwent surgery for vestibular schwannoma removal and were observed for at least 1 year. Two patients were excluded because they were lost to follow-up. The other two patients were operated on due to intracranial meningioma and their vestibular schwannomas had been treated previously at another institution. These patients were also excluded from the study. A total of 30 patients met the inclusion criteria and were suitable for analysis. In 3 cases, the vestibular schwannomas were unilateral. Six patients with bilateral vestibular schwannomas underwent unilateral procedure: two patients with the only serviceable ear, no tumor progression and hearing stable on that side were not qualified for surgery and the other four patients did not agree to have bilateral surgery. Therefore, 51 acoustic tumors were studied in 30 patients. In five patients with bilateral tumors first vestibular schwannoma had been operated on at the referring institution. In 4 out of five such cases first operation had been non-radical. In all cases in this subgroup, facial nerve function deteriorated to House–Brackmann grade VI and hearing function was lost. One of these patients was admitted to the department with additional cranial nerve IX and X deficits present after the first surgery. We did repeated procedure on these patients and complete surgery was achieved in each case.

There were 16 women and 14 men in the cohort. The mean age at the time of surgery was 25.7 years (range 13–50 years). Tumor sizes ranged from 8 to 50 mm, with a mean of 28 mm. The neurotological manifestations of vestibular schwannomas were most commonly progressive hearing loss, tinnitus and dizziness. The average duration of symptoms before admission was 3.9 years. Hearing levels were measured by pure-tone average (PTA) and speech discrimination score (SDS) and were classified using Gardner–Robertson hearing scale [9]. Most ears had non-serviceable hearing (Gardner–Robertson Grade III-V) before surgery (Table 1). Pre- and postoperative facial nerve function was assessed using the House–Brackmann scale [10]. Two patients had Grade II and one had Grade IV House–Brackmann function before surgery.

Our treatment philosophy assumed that the optimal strategy for each NF2 patient is to maintain as much useful hearing function as possible for as long lifetime as possible. Different from non-NF2 patients the resections of vestibular schwannomas in NF2 patients and their completeness were not a fundamental goal of treatment in favor of extending good quality of life period, specifically preserving unilateral hearing at least. NF2 is not a one uniform illness but comprises several subtypes of tumor of the brain, spinal cord and peripheral nerves and as the spontaneous clinical course of the disease varies from patient to patient it may need different treatment strategies. We should always remember that treatment of patients with NF2 necessitates surgical treatment of other than vestibular schwannoma tumors and the need for observation of asymptomatic tumors. Planning treatment strategies we are guided by the principle that time of deafness should be as short as possible. However, hearing restoration with the auditory brainstem implants (ABIs) precludes the observation of other tumors using MRI. This is particularly important in cases of intramedullary tumors, which should be monitored closely.

Depend on vestibular schwannoma extension, related necessity of brain stem decompression and preoperative auditory function as well as the presence of symptomatic intracranial meningioma and/or spinal tumors, the surgical strategy i.e., the indication, the order of tumors removal and the timing of tumor resection should be planned individually in each patient. In some cases treatment was started with the removal of other symptomatic tumors (likewise non-vestibular schwannoma, intracranial meningioma or spinal tumor) or these tumors were attempted in second stage after surgery of

Table 1 – Postoperative hearing results as a function of the preoperative hearing status.

Preoperative hearing status[a]	No. of cases Postoperative hearing status[a]					
	I	II	III	IV	V	Total
I	2	1	3	3	3	12
II			1	2	7	10
III					10	10
IV					7	7
V					12	12
Total	2	1	4	5	39	51

[a] According to Gardner–Robertson scale.

the first vestibular schwannoma. Then the second vestibular tumor surgery had to be postponed. In case of asymptomatic but large spinal tumor of the cervical region our management strategy was to start with spinal cord decompression. In cases where spinal canal tumor was found that required observation, brainstem or cochlear implant assumption was postponed until control MRI evaluation of any tumor growth was performed.

The surgical management of intracranial meningiomas and spinal tumors in NF2 patients will be presented in a separate paper.

One of the management options was the surgical resection of the larger tumor with dead ear or non-serviceable hearing with subsequent auditory brainstem implant implementation to assist learning of a new type of auditory function by still hearing ear on the side of a smaller tumor. Preoperatively some patients had useful hearing preserved on both sides. In some of them the first operation was performed on the smaller tumor and better-hearing ear. In the remaining patients, treatment was initiated at larger tumor and worse-hearing ear. When hearing has been preserved at the first operation the next stages of treatment become easier. If surgery on the first side with preoperative serviceable hearing was performed with subsequent loss of auditory function then auditory brainstem implant was used. In most cases ABI procedure was performed during second side vestibular schwannoma resection when it came to total deafness. For patients with bilateral hearing loss who had intact cochlear nerve, cochlear implant for auditory rehabilitation was offered. Our experience in applying the ABI procedure is presented in the subsequent work.

2.2. *Surgical approach*

In 46 cases retrosigmoid approach was used and 2 ears of patients were treated via a translabyrinthine approach. Three cases of small tumor did undergo middle fossa excision as the first step of treatment. Intraoperative facial nerve monitoring was used in all cases. Hearing function was monitored intraoperatively using auditory brainstem response (ABR) assessments or transtympanic electrocochleography (TT-ECochG) in selected cases of preoperatively serviceable hearing.

2.3. *Patient's follow-up*

The scope of resection was evaluated based on the intraoperative observation and the result of postoperative brain magnetic resonance imaging (MRI). Postoperative hearing data and facial nerve function were obtained on discharge from hospital and at follow-up hospital stay one year after operation. The postoperative follow-up period ranged from 18 months to 15 years (mean 5.3 years).

3. Results

3.1. *Operative results*

Total or near-total tumor removal was obtained in all cases, as confirmed by postoperative MRI. Schwannoma diagnosis was established based on tumor histopathology. In four cases tumor histology revealed mixed type of tumor (schwannoma plus meningioma).

No operative death we noted. In six patients with extremely large tumors new postoperative paresis in previously normal cranial nerves IX and X occurred and two of these patients required temporary tracheotomy and percutaneous endoscopic gastrostomy (PEG). Cranial nerves IX and X palsy, however, subsided in three patients and significantly diminished in the other three during follow-up. A new abducens nerve deficit occurred in 2 patients and proved permanent. Five patients suffered from postoperative cerebellar ataxia but showed gradual improvement in the follow-up. One patient required ventriculo-peritoneal shunt placement because of the hydrocephalus development in the postoperative course. Two patients experienced a cerebrospinal fluid leak which was resolved with lumbar drainage.

3.2. *Hearing preservation*

Significant deterioration to the non-functional level occurred in 19 out of 22 cases with well-preserved preoperative hearing. Only three ears maintained their preoperative good hearing. Hearing was preserved in cases of small schwannoma not exceeding 2 cm. Tables 1 and 2 present postoperative hearing results for the preoperative hearing level and different tumor size groups.

Among 21 patients who underwent bilateral operations the first operation on vestibular schwannoma was performed on the side of larger tumor in 14 patients and on the smaller tumor in 7 cases. The auditory function was then preserved in 3 patients and in 3 other patients the cochlear nerve was preserved anatomically. In 4 patients ABI operation was

Table 2 – Postoperative hearing results as a function of the preoperative tumor size.

Preoperative tumor size (mm)	No. of cases Postoperative hearing status[a]					
	I	II	III	IV	V	Total
≤10		1				1
>10 ≤ 20	2		4	1	5	12
>20 ≤ 30				3	13	16
>30				1	20	21
Total	2	1	4	5	39	51

[a] According to Gardner–Robertson scale.

Table 3 – Hearing preservation in 21 patients operated on for bilateral vestibular schwannoma depending on the first attempted tumor side.

First attempted tumor side	No. of patients		
	Serviceable hearing	Non-serviceable hearing	Total
Smaller tumor and/or better hearing status	3	4	7
Larger tumor and/or worse hearing status	0	14	14

Table 4 – Postoperative hearing results as a function of surgery timing after vestibular schwannoma diagnosis.

Surgery timing after vestibular schwannoma diagnosis	No. of cases Postoperative hearing status[a]					
	I	II	III	IV	V	Total
≤6 months	2	1	3	1	18	25
>6 months ≤ 1 year			1	2	8	11
>1 year ≤ 2 years				2	4	6
>2 years					9	9
Total	2	1	4	5	39	51

[a] According to Gardner–Robertson scale.

performed at the end of the procedure. The second side tumor surgery was performed with subsequent loss of hearing in each case and cochlear nerve preservation was achieved only in one case. In 11 patients ABI was inserted in the second stage of the procedure. One patient had bilateral ABI insertion.

In summary, in the group of 21 patients operated on both sides 3 patients preserved hearing on one side, 4 patients had cochlear nerve preserved unilaterally and 14 patients underwent ABI insertion. In 4 patients who had some measurable hearing postoperatively and confirmed eighth cranial nerve preservation by an intraoperative monitoring, cochlear implants were inserted in the next step. Hearing preservation was noted only when smaller tumor and better hearing level side was attempted at first surgery (Table 3). There was significant relationship between surgery timing and the ability to preserve hearing: better results were obtained when patients were treated within 6 months of the diagnosis of vestibular schwannoma (Table 4).

In all three patients with unilateral vestibular schwannoma hearing function of the affected ear was lost after surgery. Of the 6 patients with bilateral tumors who underwent only unilateral operations, in 4 cases ear was dead preoperatively and in two other patients hearing was lost after surgery; in 2 patients ABI surgery was performed. Four patients from this subgroup exhibited progressive hearing loss on the second side but refused surgical procedure and the other two still retain useful hearing and are being monitored.

3.3. *Facial nerve function*

Immediately after surgery facial nerve function was normal in 2 cases. In 19 cases postoperative facial nerve function was moderately impaired (Grade II and III on the House–Brackmann scale) and early rehabilitation of the nerve was introduced. In another 19 cases facial nerve function was Grade IV and the eye was protected with a humid chamber. Anatomical continuity of the facial nerve was not compromised in any of these cases as confirmed by intraoperative facial nerve monitoring and patients showed gradual improvement of the nerve function during postoperative rehabilitation. In 8 out of 11 cases with severe postoperative facial nerve palsy (Grade V and VI) a golden plate had been implanted into the upper lid in the early postoperative period in order to facilitate eye closure and then hemihypoglossal-facial nerve anastomosis was performed usually 1 month after tumor removal. Facial nerve function after the anastomosis procedure was satisfactory in all but one case. In another case a facial plastic surgery was performed because of long-lasting

Table 5 – One year postoperative facial nerve function as a function of the preoperative tumor size.

Preoperative tumor size (mm)	No. of cases Postoperative facial grade[a]						
	I	II	III	IV	V	VI	Total
≤10	1	1					2
>10 ≤ 20	5	3	3	1			12
>20 ≤ 30		10	4			2	16
>30	1	4	11	3		2	21
Total	7	18	18	4	0	4	51

[a] According to House–Brackmann scale.

facial neuropathy. In the two other patients whose first operation had been performed at another institution and the facial nerve was then permanently damaged underwent there tarsorrhaphy and were not qualified for restoration of facial nerve continuity.

Long-term facial nerve function results are shown in Table 5.

Among 14 tumors up to 2 cm there was only one case of moderately severe facial nerve dysfunction (House–Brackmann Grade IV) in the long follow-up. On the other hand we achieved unsatisfactory results of facial nerve preservation (House–Brackmann Grade IV-VI) in 8 out of 37 larger tumors exceeding 2 cm.

3.4. *Tumor recurrence*

There was vestibular schwannoma recurrence in 5 cases. Median time to recurrence was 8.8 years (range 5–14 years). Tumor recurrence was noted in 3 patients. In two patients there was bilateral tumor regrowth: one patient is qualified for second surgery of the two tumors and the other patient underwent complete re-surgery on one side and remains asymptomatic while observing a second side small tumor regrowth. The third patient who had undergone unilateral procedure refused surgery both for the second side progressive tumor growth as well as for tumor recurrence after first surgery.

4. Discussion

4.1. *Treatment options and hearing preservation*

Neurofibromatosis type 2 (NF2) is a heritable tumor predisposition syndrome that leads to the development of multiple intracranial tumors. The pathognomonic lesion of NF2 is vestibular schwannoma appearing bilaterally. In this retrospective study we examined the cases of 30 adolescents and adults with NF2 who had undergone vestibular schwannoma surgery. The main goal of treatment in patients with this condition in view of the danger of bilateral deafness with its serious consequences is to preserve as much hearing as possible which is a tremendous advantage in the quality of the patients' life. This is especially important because of the young age of the NF2 patients and knowing that these tumors will continue to grow over time [11,12]. Early detection of NF2 allows those individuals with small tumors and useful hearing the advantage of greater ability to preserve hearing. Better preoperative hearing is related to better postoperative hearing [13–15]. Unfortunately, in our series nearly 60% of ears had non-serviceable hearing and only 4% of tumors did not exceed 1 cm preoperatively. Thus, hearing was preserved in only 3 cases. Slattery et al. [13] reported that difference in average tumor size between the groups in which hearing was preserved was 0.6 cm. In their study hearing was preserved in 55% of cases, however, most patients (81%) had very good hearing before surgery. On the basis of their results Brackmann et al. [11] proposed that early surgical intervention for patients with vestibular schwannomas and NF2 may prevent the occurrence or progression of hearing loss. Similarly, Sami et al. [16] advocate for active management strategy of early surgery in patients who have NF2 if the chances of functional hearing preservation are realistic. In patients with similar tumor sizes their recommendation for surgery depends on the superior quality of preoperative ABR on one side, as the chances of successful hearing preservation are better on that side. It should be noted that besides tumor size and audiometric data, patient's preoperative ABR is very important predictive factor for the auditory outcome. Tumor parameters influencing the chances of hearing preservation such as tumor consistency and adhesion at the tumor plane and the fundus of the internal auditory canal might be obtained by the patient's ABR [17].

The policy of proactive surgical management of these tumors assumes that given the benefits of preserving useful auditory function on at least one side, the decision about surgery on the side of better hearing is worth taking, despite the risk of reduced postoperative quality of life in case of hearing lost. Described above philosophy of treating vestibular schwannomas in NF2 patients is contrary to the traditional approach of withholding intervention from this patient group either the tumor enlarges significantly or the patient becomes symptomatic [7]. So-called watchful waiting policy points that the majority of NF2 intracranial tumors including vestibular schwannomas grow in a ''saltatory'' pattern, with periods of growth alternating with quiescent periods [18]. In this approach these tumors should not be surgically treated simply because they exist, as they may not grow for considerable periods of time [19,20]. Dirks et al. [18] analyzed the rate of growth and the growth pattern of various intracranial tumors in NF2 patients and recommend to operate only on tumors that are progressively symptomatic. In their opinion tumor progression alone, without the development of progressive symptoms, should not be used as an indication for surgery.

They calculated that prophylactic resection of growing but asymptomatic tumors will lead to additional unnecessary surgeries over a definite period of time. Similarly, Peyre et al. [19] reported unpredictable vestibular schwannoma growth rates in NF2 patients, lack of correlation between growth rates and hearing outcome and demonstrate that most patients preserve some degree of hearing and most tumors grow slowly in approximately 5-year follow-up. It seems, however, that in case of vestibular schwannoma this approach reduces the chances of long-term hearing preservation. Many patients can initially avoid surgical intervention but looking at the long term with tumors growing slowly and hearing worsening gradually they end up facing surgical treatment with little chance of good hearing outcome. Nevertheless, observation without surgical procedure might be useful option for patients with non-serviceable hearing or who have a tumor in their only hearing ear [21].

Another option for the treatment of NF2 is stereotactic radiosurgery. This treatment modality is proposed for small and middle-sized tumors. Radiosurgery eliminates many of the risks of resection [22]. Recent radiosurgical series of patients with NF2 reported the hearing preservation rate of 67% and the actuarial tumor control rate of 67% at 5 years [23,24]. NF2-associated intracranial tumors, however, both schwannomas and meningiomas, seem to be less sensitive to stereotactic radiosurgery than their sporadic counterparts [18]. Another drawback of radiosurgery is the inability to provide brainstem decompression in patients with large tumors who have significant symptoms from compression.

4.2. *Auditory rehabilitation*

In our series complete bilateral hearing loss was frequent consequence of vestibular schwannoma management. 20 patients were offered auditory brain implants or cochlear implants which are options for hearing remediation. Auditory rehabilitation becomes a significant aspect in the care of patients with NF2 because most of them will lose hearing bilaterally. Hearing aids are usually less effective in NF2 cases because speech discrimination in these patients has been compromised and they require implanted devices for hearing improvement. Vestibular schwannomas frequently destroy the cochlear nerve and auditory rehabilitation in these individuals relies on auditory brainstem implants (ABIs) which bypass the external, middle, and inner ear and directly stimulate the cochlear nucleus. Placement of the ABIs have the potential to recover some degree of auditory perception, however, it is rarely adequate for speech understanding and mainly serve to enhance lip reading [25,26]. Nevertheless Matthies et al. [27] have reported recently that open-set speech recognition in pure auditory mode is feasible in patients with ABIs. In case of an intact cochlear nerve following vestibular schwannoma removal alternative strategy to restore hearing is cochlear implantation. Cochlear implants directly stimulate the cochlear nerve within the cochlea and typically result in open-set speech recognition and ability to converse by telephone [28,29], thus improving social and professional functioning. This suggests that there will be an increasing role for cochlear implantation in hearing rehabilitation.

4.3. *Facial nerve function*

Preservation of facial nerve function is critical in NF2 patients for social and quality of life reasons. The level of facial function preservation was satisfactory in the study group and was obviously related to the tumor size. Anatomic preservation of the facial nerve is the primary goal of vestibular schwannoma treatment both for NF2 patients as well as for patients with sporadic tumors. Facial nerve continuity is to be compromised in various situations [14]. The facial nerve could be lost in previous surgery that was performed elsewhere. In case of circumscribed schwannoma resection usually led to facial nerve discontinuity. Sometimes there could be no cleavage plane between the vestibular, cochlear, and facial nerves and the facial nerve continuity could be easy lost in these cases. Besides, the facial nerve could also show tumorous changes. In the case of permanent damage to the facial nerve it should be reconstructed intracranialy at the cerebellopontine angle by sural grafting, reanimated by hemihypoglossal-facial anastomosis or patient should undergo facial plastic surgery [30–33].

5. Conclusions

Clinical decision regarding when to operate on NF2 patients with vestibular schwannomas relay on the realistic chances of hearing preservation. It seems that the initiation of treatment from small tumors with preserved hearing offers the best chance of hearing preservation. In this situation, it is safe to continue further treatment of other tumors or observe tumor growth using MRI.

Conflict of interest

None declared.

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None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; EU Directive 2010/63/EU for animal experiments; Uniform Requirements for manuscripts submitted to Biomedical journals.

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