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Original research article

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

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ARTICLE INFO

*Article history:*
Received 10 May 2015
Accepted 20 June 2015
Available online 3 July 2015

*Keywords:*
Vestibular schwannoma
Hearing preservation
Neurofibromatosis type 2
Surgical management

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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http://dx.doi.org/10.1016/j.pjnns.2015.06.008

## 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