The Journal of Laryngology & Otology (2016), ***130***, *606–610.*

doi:10.1017/S0022215116007969

REVIEW ARTICLE

Optimal outcomes for hearing preservation in the management of small vestibular schwannomas

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Abstract

Objective: To undertake a systematic review of the role of microsurgery, in relation to observation and stereotactic radiation, in the management of small vestibular schwannomas with serviceable hearing.

Methods: The Medline database was searched for publications that included the terms 'vestibular schwannoma' and/or 'acoustic neuroma', occurring in conjunction with 'hearing'. Articles were manually screened to identify those concerning vestibular schwannomas under 1.5 cm in greatest dimension. Thereafter, only publications discussing both pre-operative and post-operative hearing were considered.

Results: Twenty-six papers were identified. Observation is an acceptable strategy for small tumours with slow growth where hearing preservation is not a consideration. In contrast, microsurgery, including the middle fossa approach, may provide excellent hearing outcomes, particularly when a small tumour has begun to cause hearing loss. Immediate post-operative hearing usually predicts long-term hearing. Recent data on stereotactic radiation suggest long-term deterioration of hearing following definitive therapy.

Conclusion: In patients under the age of 65 years with small vestibular schwannomas, microsurgery via the middle fossa approach offers durable preservation of hearing.

Key words: Hearing; Vestibular Schwannoma; Acoustic Neuroma; Microsurgery; Middle Cranial Fossa; Radiosurgery

Introduction

Management options for small vestibular schwannomas, defined as those under 1.5 cm in greatest dimension, include observation, microsurgery and stereotactic radiation. The decision-making process for treatment remains controversial, particularly when considering long-term hearing outcomes and patient satisfaction. Indeed, there is no consensus among experts: as a testament to this, national and international meetings discussing vestibular schwannoma often hold panels specifically to discuss the optimal management of small tumours.

Of particular interest in the management of such tumours in patients with serviceable hearing is the impact of treatment on hearing outcome. Various schemes have been proposed to classify hearing in the affected ear. One widely-used classification, set forth by the American Academy of Otolaryngology – Head and Neck Surgery (AAO-HNS), defines class A hearing as a pure tone average (PTA) under 30 dB with a word recognition score exceeding 70 per cent; classes B, C and D represent numerically higher PTA values with lower word recognition scores (Table I).[1] A separate classification, proposed by Meyer *et al.*, focuses on the importance of word recognition score in the serviceability of hearing, and ranges from class I, with a word recognition score of 70–100 per cent, to class IV, with a word recognition score of 0 per cent (Table II).[2] Finally, the Gardner–Robertson hearing scale, ranging from grade I to V, considers both PTA and word recognition score, with grade I corresponding to good hearing and grade V corresponding to no hearing (Table III).[3]

Here, we seek to review the literature on the management of small vestibular schwannomas, and discuss the benefits and alternatives for management, with particular emphasis placed on hearing preservation following therapeutic intervention.

Materials and methods

The Medline database was searched for publications that included the terms 'vestibular schwannoma' and/or 'acoustic neuroma', occurring in conjunction with 'hearing'. Manual screening was first performed to identify publications that included data on small tumours, defined as those under 1.5 cm in maximum diameter. Subsequently, publications reporting on the natural history of small, untreated vestibular schwannomas were selected. Thereafter, publications that considered hearing performance both pre-treatment and

Accepted for publication 21 February 2016 First published online 20 May 2016

TABLE I
AAO-HNS HEARING CLASSIFICATION[1]

Class	PTA hearing level (dB)	Word recognition score (%)
A	≤30	≥70
B	31–50	≥50
C	>50	≥50
D	Any level	<50

AAO-HNS = American Academy of Otolaryngology – Head and Neck Surgery; PTA = pure tone average

post-treatment, following microsurgery or stereotactic radiation, were identified.

The literature search identified 26 papers. Three papers discussed the natural history of hearing during observation of small vestibular schwannomas. Eleven papers discussed hearing after microsurgery for small vestibular schwannomas. Eleven papers discussed hearing after stereotactic radiation for small vestibular schwannomas. One paper discussed hearing after either microsurgery or stereotactic radiation for small vestibular schwannomas.

Across all papers, hearing outcomes were predominantly described using the AAO-HNS classification, the word recognition score classification or the Gardner–Robertson classification. Given the heterogeneity of subject populations and hearing outcomes reported, in addition to the lack of individual patient data in the papers retrieved, pooled analysis was not performed; rather, salient findings were summarised.

Results

Observation

Observation is considered a viable management option for patients with small vestibular schwannomas only when there is no evidence of consistent growth or hearing impairment. This strategy has often been advocated for older patients, in whom the risks of microsurgery or stereotactic radiation of vestibular schwannomas may outweigh the associated benefits, particularly in the face of medical co-morbidities.[4] However, it is worth noting that the average growth rate of untreated vestibular schwannomas has been estimated in various studies to lie between 1 and 4 mm per year.[4–6] In one longitudinal study, Charabi *et al.* followed 123 patients over an average follow-up period of 3.4 years; 75 per cent of patients demonstrated deterioration of hearing past serviceable levels and/or substantial tumour growth, necessitating either microsurgical management or stereotactic radiation.[6]

TABLE II
HEARING CLASSIFICATION BASED ON WORD RECOGNITION SCORE*

Class	Word recognition score (%)
I	70–100
II	50–69
III	1–50
IV	0

*As proposed by Meyer *et al.*[2]

TABLE III
GARDNER–ROBERTSON HEARING SCALE[3]

Grade	PTA hearing level (dB)	Word recognition score (%)
I: Good	0–30	70–100
II: Serviceable	31–50	50–69
III: Non-serviceable	51–90	5–49
IV: Poor	90–100	1–4
V: Deaf	0	0

PTA = pure tone average

Furthermore, while observation may seem an attractive option for select patients, the natural history of vestibular schwannomas suggests that hearing preservation with observation only is far from guaranteed, even with small tumours. In another longitudinal study, Stangerup *et al.* observed that 88 per cent of patients with a word recognition score of 100 per cent retained a class I score for hearing (according to criteria set forth by Meyer *et al.*[2]) after a median follow-up time of roughly four years.[7] In patients with even slightly impaired hearing at presentation, with a word recognition score ranging between 90 and 99 per cent, a significantly lower fraction – only 55 per cent – retained a speech discrimination score of 70 per cent or better. Based on these data, the authors suggested that observation may be an acceptable alternative leading to hearing preservation only if hearing is completely intact at the time of presentation.

An additional relative contraindication to observation, when hearing preservation is desired, is documented tumour growth. Sughrue *et al.* noted that the best predictor of hearing preservation was not tumour size at presentation, but rather the rate of tumour growth: tumours enlarging greater than 2.5 mm/year were associated with a significantly lower rate of hearing preservation.[8] It is worth noting that deterioration of auditory function may occur even in the absence of imaging evidence of tumour growth, as noted by Walsh *et al.*, although the mechanism for this remains unclear.[9] In elderly patients, a small amount of growth may be tolerated if the size of the tumour is quite small and it is felt that the patient may not need intervention during their life span.

Together, these points suggest that observation should be recommended only in select patients with small tumours who experience no decline in hearing and demonstrate very slow tumour growth. When hearing begins to deteriorate, or consistent tumour growth is documented, microsurgery or stereotactic radiation is usually indicated.

Stereotactic radiation

Over the past two decades, stereotactic radiation has emerged as an alternative management option for

https://doi.org/10.1017/S0022215116007969 Published online by Cambridge University Press

vestibular schwannomas. Specific radiosurgical modalities include Gamma Knife®, CyberKnife® and, more recently, fractionated linear accelerator ('LINAC') treatment. While historical published studies hold that Gamma Knife stereotactic radiation allows for excellent hearing preservation, newer studies demonstrate that hearing preservation rates are at best initially comparable to surgery, and hearing outcomes have now been shown to further deteriorate on a long-term basis following stereotactic radiation.

In one study, Prasad *et al*. noted that among all patients treated with Gamma Knife stereotactic radiation, hearing deteriorated in 60 per cent of patients after a mean follow-up period of six years.[10] In a related meta-analysis, Yang *et al*. observed an overall hearing preservation rate of 57 per cent after stereotactic radiation treatment of any kind.[11] Meijer *et al*. studied the long-term effects of linear accelerator based stereotactic radiation, administered in either a single fraction or fractionated regimens, and noted hearing preservation rates of 61 to 75 per cent at five years.[12] This finding closely paralleled a study by Spiegelmann *et al*., who reported an actuarial rate of hearing preservation of 71 per cent.[13] A more guarded outlook was offered by Paek *et al*., who followed 25 patients with vestibular schwannomas with initial serviceable hearing.[14] After an average dose of 12.0 Gy, 52 per cent of patients had preserved serviceable hearing (Gardner–Robertson grade I or II), and only 36 per cent of patients retained their pre-radiation Gardner–Robertson hearing grade.[14]

While some reported rates of initial hearing preservation after stereotactic radiation approximate results reported with microsurgery, more recent research highlights the potentially deleterious long-term impact of stereotactic radiation on long-term hearing. In a recent study, Carlson *et al*. noted that more than 60 per cent of patients with tumours under 1 cm in greatest diameter and serviceable hearing pre-operatively (AAO-HNS class A or B) developed non-serviceable hearing (AAO-HNS class C or D) at a mean of 4.2 years following treatment.[15] At 10-years' follow up, only 23 per cent of patients retained serviceable hearing.[15] In a similar study of patients with serviceable pre-treatment hearing, defined as Gardner–Robertson grade I or II, Hasegawa *et al*. reported five-year hearing preservation rates of 64 per cent in grade I patients and only 24 per cent in grade II patients.[16] As an additional point of caution, stereotactic radiation may be associated with a risk of malignant transformation, albeit rare.[17,18]

The observed hearing impairment following stereotactic radiation may be attributed to several factors. First, radiation may cause direct cochlear inflammation and injury.[19] Another hypothesis suggests that as the mechanism of tumour control with stereotactic radiation is likely to be radiation-induced vasculitis, this treatment may impair the vascular supply to the tumour; this radiation effect may damage the vascular supply to the cochlear nerve, and lead to neuropathy and hearing loss. This hypothesis is corroborated by the observation that hearing loss is generally not acute, but rather develops over 6 to 24 months, and often progresses thereafter.[20] Alternatively, stereotactic radiation can cause direct neuropathy to the cochlear nerve.[19] Regardless of the specific mechanism, the aggregate literature demonstrates that stereotactic radiation poses a significant risk to hearing following treatment.

Microsurgery

When a patient has serviceable hearing pre-operatively, the decision to pursue hearing-preserving surgical management depends on various factors, including the status of the contralateral ear and tumour size. When hearing preservation is attempted, either middle fossa or retrosigmoid-suboccipital craniotomy approaches may be considered.

The middle fossa approach, developed at the House Clinic by Dr William House in the 1960s,[21] is particularly advantageous, as exposure of the entire internal auditory canal allows for early visualisation of the cochlear nerve in conjunction with the vestibular and facial nerves. In contrast, the retrosigmoid approach often does not allow for optimal exposure of the lateral internal auditory canal, placing the cochlear and facial nerves at risk, and, in certain cases, not allowing total tumour removal from the fundus of the internal auditory canal.[22]

In a retrospective analysis of 151 patients who underwent middle fossa craniotomy, Slattery *et al*. noted that hearing was preserved in 68 per cent of patients, with 52 per cent of patients demonstrating post-operative hearing within 15 dB and speech discrimination within 15 per cent of the pre-operative score.[22] In this report, no correlation was observed between tumour size and hearing preservation.[22] More recent data from the House Clinic suggest that when controlled for tumour size, the middle fossa approach for hearing preservation is superior to the retrosigmoid approach.[23]

In a similar study, Wang *et al*. reported 103 patients who underwent a middle fossa approach for vestibular schwannoma resection.[24] At initial post-operative follow up, AAO-HNS class A hearing was preserved in 67 per cent of patients; among patients presenting initially with class B hearing, 77 per cent were class B or better. At a follow-up time of five years, 65 per cent of patients with initial post-operative class A hearing remained class A, while 66 per cent of patients with initial post-operative class B hearing were graded class B or better.[24] Kutz *et al*. reported 38 patients with class A or class B hearing according to AAO-HNS criteria pre-operatively; following middle cranial fossa resection, 73 per cent of patients retained class A or B hearing.[25]

The decision to pursue microsurgical resection of a vestibular schwannoma might be influenced by any observed deterioration in hearing status that may

https://doi.org/10.1017/S0022215116007969 Published online by Cambridge University Press

foreshadow either long-term hearing impairment or, more importantly, potentially worse hearing outcomes after stereotactic radiation. As noted above, Stangerup *et al*. observed that even a small decline in hearing during observation predicted statistically significantly worse hearing outcomes, with only 55 per cent of such patients retaining serviceable hearing.[7] Furthermore, pre-treatment hearing strongly predicts long-term hearing following stereotactic radiation, as noted by Carlson *et al*.: patients who presented with AAO-HNS class B hearing carried a 2.4-fold increased risk of non-serviceable hearing as compared to AAO-HNS class A patients.[15] Taken together, these data suggest that because hearing impairment portends poorer hearing outcomes following stereotactic radiation, microsurgical resection is the preferred approach when hearing impairment becomes apparent.

The literature additionally suggests that initial post-operative hearing is relatively durable following microsurgical resection, and is therefore predictive of long-term hearing. In a study of patients who underwent middle cranial fossa resection of a vestibular schwannoma, Woodson *et al*. observed that 38 of 42 patients demonstrated preservation of class I hearing, both immediately post-operatively and at the latest follow up.[26] In 29 patients with more than 5 years of follow up, post-operative word recognition score based class I to II hearing was maintained in 28 patients.[26] In another study, Friedman *et al*. demonstrated that of 23 patients with serviceable hearing immediately following middle fossa surgery, 70 per cent retained serviceable hearing at 5 years or longer.[27] Betchen *et al*. reported that 30 (86 per cent) of a series of 35 patients maintained immediate post-operative hearing over a mean follow up of 7 years.[28] These studies reinforce two concepts: first, that the middle fossa approach potentially offers excellent preservation of hearing, and, second, that initial post-operative hearing is highly predictive of long-term hearing, allowing for more definitive patient counselling following surgery.

Various studies have focused on predictors of hearing results following middle cranial fossa surgery. In one study, Brackmann *et al*. reported that pre-operative hearing status and auditory brainstem response findings, specifically shorter interaural wave V latency and shorter absolute wave V latency, were associated with preservation of hearing following middle fossa resection.[29] In another study, Goddard *et al*. noted that the presence of fundal fluid on magnetic resonance imaging (MRI) pre-operatively was associated with hearing preservation; furthermore, tumours intra-operatively noted to arise from the superior vestibular nerve were also associated with higher rates of hearing preservation.[30] These pre-operative indicators may assist with patient counselling prior to determination of management.

When comparing middle fossa craniotomy to other microsurgical approaches, middle fossa craniotomy has been associated with more favourable hearing outcomes as compared to the retrosigmoid approach for small tumours.[31] A disadvantage of the middle fossa approach is brain retraction, which can potentially cause significant neurosurgical morbidity, particularly in the elderly. In our practice, however, brain retraction time in the middle fossa approach can be reduced to 30 minutes with the retractor in place. Following this, cerebrospinal fluid is drained from the posterior fossa cistern, the retractor is removed, and reinforced silicone elastomer or bone wax is used to protect the temporal lobe during the final drilling of the lateral internal auditory canal. By performing the conclusion of the approach without the retractor in place, temporal lobe manipulation is minimised.

For patients presenting with significant hearing loss, microsurgery via a translabyrinthine approach offers wide exposure with excellent facial nerve preservation, with over 90 per cent of patients presenting with small tumours demonstrating House–Brackmann grade I or II function one year following resection.[32] In other words, while the middle fossa approach offers an excellent chance of retaining residual hearing, the translabyrinthine approach is the preferred 'gold standard' when hearing at presentation is impaired or poor.

Finally, the microsurgical approach offers advantages over stereotactic radiation when considering surveillance for residual or recurrent tumours following definitive therapy. Following microsurgical management, it is common practice among neurotologists to perform MRI surveillance at one-, three- and five-year intervals post-operatively; at five years, if no growth or residual disease is observed, no further imaging is warranted.[33,34] In contrast, follow up after stereotactic radiation is often performed at more frequent intervals – even within the first year after therapy – and may extend well beyond five years following therapy, leading to an increased burden on the patient and greater healthcare costs.[35]

Discussion

The management options for small vestibular schwannomas (defined as those less than 1.5 cm in greatest dimension) include observation, stereotactic radiation and microsurgery. In patients with serviceable hearing at presentation, hearing preservation is of utmost importance for many patients. However, the development of auditory symptoms, including high frequency hearing loss and tinnitus (which may signal an increase in tumour size or the onset of compression), may portend an eventual decline in hearing and necessitate definitive therapy if long-term hearing preservation is a high priority.

Long-term data for stereotactic radiation management have recently become available; most of the reports suggest that Gamma Knife stereotactic radiation is associated with a hearing preservation rate approximating 23 per cent at 10-years' follow up.[15] These data will likely have implications for other stereotactic radiation techniques, including CyberKnife and fractionated linear accelerator treatment. In comparison, various studies have reported durable hearing preservation rates ranging from 68 to 77 per cent following a

https://doi.org/10.1017/S0022215116007969 Published online by Cambridge University Press

middle fossa craniotomy approach for tumour resection,[22,24,25] and others suggest that immediate postoperative hearing is highly predictive of long-term hearing.[25–27] Based on these considerations, we feel that for small vestibular schwannomas, the management option most appropriate for patients desiring durable hearing preservation is microsurgery via a middle fossa approach.

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Dr E P Wilkinson takes responsibility for the integrity of the content of the paper
Competing interests: Dr Wilkinson receives research support from Cochlear Americas and from MED-EL USA, and serves as a consultant for Nurotron, but no source had any involvement in this project.