Introduction

The management of vestibular schwannomas (VS) has changed radically in recent decades. It has evolved from a monospecialism (mainly surgical) approach to interdisciplinary multimodal management. There is a worldwide trend towards the multidisciplinary management of complex pathologies and diseases (e.g., congenital malformations, oncologic diseases, etc.). The same applies to skull base tumors where collaboration can be expected between disciplines like ENT surgery, neurosurgery, neurology, neuroradiology, radiotherapy, ophthalmology, plastic surgery, physical medicine...

Nowadays, several treatment modalities are available, leading to complex decision-making based on diagnostic findings (mainly imaging) and on the preference of the informed patient. The three main options for the management of vestibular schwannomas are: observation (or wait and scan), surgical removal and stereotactic radiation. In rare cases, one may also consider salvage surgery after failed radiotherapy or radiotherapy after subtotal removal. A shift in the management approach has been observed over ten years, as shown in a study performed in the J. Hopkins Hospital comparing therapeutic management in 1997 and 2007. Nowadays, the wait-and-scan strategy is adopted more often (10.5% in 1997; 28% in 2007), surgery is on the decline (89.5% versus 68%) and radiotherapy is used more often (0% versus 4%). Techniques for hearing preservation surgery, facial reanimation and auditory rehabilitation have also changed dramatically over the past decades.

The multidisciplinary approach has several advantages: pooling of difficult cases, cross-specialty exchanges of ideas and mutual sharing of expertise. As ENT surgeons, we usually establish the diagnosis on the basis of asymmetric hearing loss, vertigo and tinnitus. We are familiar with the auditory and vestibular physiology and temporal bone micro-anatomy. We are also accustomed to dealing with the facial nerve and using endoscopic techniques. Our neurosurgical colleagues are more familiar with intracranial diseases and tumors, the cerebral vasculature, neuronavigation, and stereotactic radiotherapy. Nowadays, modern neuroradiological innovations provide us with detailed information about the exact size and volume of the tumors and about the surgical anatomy, and these factors are important when

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**Multidisciplinary management of vestibular schwannomas: state of the art**

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**Key-words.** Vestibular schwannoma (VS); multidisciplinary management; treatment algorithm; wait and scan for VS; surgery for VS; radiosurgery for VS

**Abstract.** Multidisciplinary management of vestibular schwannomas: state of the art. The preferred setting for the treatment for vestibular schwannoma these days is multidisciplinary and multimodal. A balance has to be struck between a wait-and-scan attitude and a more active approach (surgery or radiotherapy). An initial wait-and-scan attitude is a reasonable management option because many tumors do not grow during a prolonged period of observation. The different surgical approaches may or may not involve attempts to preserve hearing. Stereotactic radiotherapy as treatment of choice is often considered in the elderly patient presenting with a vestibular schwannoma smaller than 2.5 cm with documented growth. This paper will review current treatment modalities and the respective pros and cons. A decisional algorithm as currently adopted by our skull base team is presented at the end of the paper.

choosing the most appropriate approach. The advantage of the multidisciplinary cumulated expertise is a better clinical outcome in patients presenting with a vestibular schwannoma.

**Observation, or wait-and-scan, approach**

Previously, when a diagnosis was established with BERA followed by CT scanning, it was agreed that most tumours grow and that they do this at a rate of between 1-2 mm a year.

Since MRI scanning has become available as a screening method, more and more vestibular schwannomas have been diagnosed. According to a Danish survey the incidence of vestibular schwannomas has increased from 3.1 diagnosed new cases per million per year in 1976 to a peak of 22.8 per million per year in 2004. This was followed by a fall to 19.4 vestibular schwannomas per million per year in 2008. The same study showed that, in particular, the incidence of small vestibular schwannomas diagnosed by MRI had increased. Mean tumour size at diagnosis fell from 30 mm in 1979 to 10 mm in 2008. We observed the same phenomenon later (after 1990), with increasing numbers of diagnoses of intralabyrinthine schwannomas due to improvements in MRI resolution.

In parallel with the identification of more small tumours, hearing acuity at diagnosis has also improved over the years according to Stangerup et al.

During the 1980s, it was noticed for the first time that, in elderly vestibular schwannoma patients (who were not fit to undergo surgery), leaving these tumours alone would not necessarily lead to further growth. Although the percentages for non-growing tumours vary in the literature (from 40% to 76%), most authors agree that a substantial percentage do not grow. In many studies, it is likely that the non-growth rate has been over-estimated because of the relatively short duration of follow-up. Involution has also been observed in some tumours. This may be explained by tumour necrosis caused by intratumoral thrombosis and it may be part of the normal involution of tumours that have reached their maximum growth potential. So the growth pattern may vary from spontaneous involution to rapid growth (up to 2 cm in a year). Unfortunately, few clinical or radiological factors predicting growth have been found so far. According to some authors, initial size is a predictor of later growth. According to Agrawal et al., tinnitus is associated with a threefold increase in the risk of growth.

Failure of the observation policy is defined as the switch from wait and scan to active treatment and this is usually based on significant tumour growth observed with MRI. Other reasons to discontinue the wait-and-scan strategy are incapacitating vertigo or hearing deterioration in cases where hearing preservation surgery has been requested. Various studies report a percentage of failure varying between 0% and 50%. In a large study, El Bakkouri et al. reported a change in therapy from wait and scan to active treatment in 41%: there was a tumour growth of more than 3 mm in 29% and of less than 3 mm in 12%.

A particular concern is the question of whether the conservative approach has a detrimental effect on hearing. Stangerup et al. showed that leaving the tumour alone results in 61% hearing conservation when observed over a period of 3.8 years. The patient’s initial speech discrimination seems to be the best predictive factor for the preservation of good stable hearing at final evaluation. Quarranta found hearing preservation in 60% of cases with a 3.3 year follow-up. However, found a deterioration of PTA from 38 dB to 51 dB and a speech discrimination score (SDS) declining from 66% to 55% during 3.6 years of follow-up. Hypointensity of the intralabyrinthine fluids as observed on T2-W MR images results in significantly faster deterioration of hearing. Similarly, the same hypo-intense signal was found by us to be a negative predictive factor for hearing preservation after surgery. By contrast, some teams promoting the hearing preservation approach recommend early intervention when hearing is still good. The rates of hearing preservation vary according to the size and approach but the benefit is that, if successful hearing preservation is achieved, early intervention leads to enduring and stable hearing. So in young patients with grade 2 tumours (cerebellopontine angle (CPA) portion smaller than 2 cm) the authors would recommend tumour removal and an attempt to preserve hearing before hearing deteriorates because hearing can be preserved in 50-60% of the cases and the facial palsy risk is minimal (0-5%). Some authors also argue that, if growth is faster, facial function is more at risk (some tumours may grow up to 2 cm in one year). Nowadays, this information needs to be transmitted to
our patients, who participate and get involved in the final decision-making.

When a wait-and-scan policy is followed, a second MRI is performed six months after the initial diagnosis and, if no substantial growth (2 mm or less) is observed, this is followed by annual MRI scans. The next question is: how long should we follow up these tumours? Several patterns of growth have been demonstrated, and a tumour that grows may stop doing so, and vice-versa. So some tumours that have been stable for many years can start to grow. Hillman et al.19 pointed to the fact that a substantial number of patients are not followed up for several reasons: in 45% it was a deliberate decision by the patient, in 30% the patient had no clear understanding of the consequences despite adequate explanation and in 25% medical problems took precedence. After an initial period of five years and demonstrated quiescence we repeat MRI at intervals of 2-3 years unless symptoms or audiometry seem to indicate a change in biological behaviour. Kalthoft et al.19 found that when all patients allocated primarily to conservative management were pooled, good facial function was found in 97%, which was significantly better than the result for primary operation (87%).

In conclusion, an initial wait-and-scan attitude may be regarded as a reasonable management option because most of these tumours do not grow during an initial period of observation. Some authors favour it when hearing is already poor and, by contrast, others favour it when hearing is very good. The advantage of this policy is that, in the absence of treatment, serious complications will not occur, or only extremely rarely, and the facial nerve will remain intact in almost all cases. As shown in the decisional algorithm, we adopt this approach for most small tumours (smaller than 1.5 cm in the CPA).

Surgical intervention

From an historical point of view surgery has been the treatment of choice for vestibular schwannomas. Many technical improvements and refinements have reduced the complication rates. The main approaches to the internal auditory meatus and CPA are:

1) The translabyrinthine route
2) The retro-labyrinthine approach
3) The retrosigmoid approach
4) The middle fossa intervention

1) The translabyrinthine approach

The translabyrinthine approach provides the most direct access to the CPA by traversing the temporal bone and the labyrinthine structures. It is therefore preferred in patients without useful hearing and impaired or absent vestibular function before surgery or when hearing preservation is not an objective, as in large or very large tumours. It can be used for tumours of all sizes and affords the best exposure of the facial nerve from the vertical portion in the mastoid to the root entry zone at the brainstem. An advantage compared to other approaches is the avoidance of the early exposure of brain tissues and avoidance of cerebellar or temporal lobe retraction. Total drilling of all mastoid cells and filling the cavity with abdominal fat after closure of the Eustachian tube and middle ear keeps the rate of cerebrospinal fluid leaks low. It has also been found that there is less post-operative headache by comparison with the suboccipital/retrosigmoid route.18

2) The retro-labyrinthine approach

This route offers a pre-sigmoid retro-labyrinthine route to the internal auditory canal (IAC) and CPA. Although this is considered a narrow approach, Darrouzet et al. claim that the wide removal of bone over the occipital bone, sigmoid sinus, and posterior fossa dura allows good exposure to medially located tumours.19 Pre-operative evaluation of the posterior semicircular canal (PSCC) in relation to the IAC and the level of the jugular bulb is necessary. This is done nowadays with a cone beam CT scan, which results in higher accuracy than conventional high-resolution CT. Peroperative identification of the posterior semicircular canal and clipping of the endolymphatic duct prevents the violation of intralabyrinthine structures. Hearing preservation rates are comparable with the retrosigmoid approach.

3) The retrosigmoid approach

The first procedure described in the literature for a CPA tumour was performed in 1890 by Von Bergmann. Cushing widened the approach into a bilateral occipito-suboccipital approach, allowing more manipulation of the cerebellum. His pupil, Walter Dandy, modified this technique into a unilateral smaller suboccipital craniectomy with partial resection of the cerebellum. Olivecrona transformed Dandy’s approach into a more lateral so called retrosigmoid approach. The real breakthrough was the use of the microscope, which was actually pio-
neered by William House and introduced in neurosurgery by Gazi Yasargil. It was at this point that the term ‘retrosigmoid approach’, as opposed to the sub-occipital approach, became applicable.

The actual retrosigmoid approach consists of a targeted small craniectomy or craniotomy that exposes a part of the transverse and sigmoid sinuses, enabling anterior retraction of the dura and sigmoid sinus. This extra exposure of a few millimetres can make the difference in exposing the CPA without any retraction of the cerebellum. The approach requires the opening of the cisterna magna and CPA cisterns directly after the opening of the dura to prevent contusion and swelling of the cerebellum. This approach provides safe and quick access to the CPA. We use it as our principal approach for vestibular schwannomas when hearing preservation is an objective. It can be used for both large and small tumours. In addition, in large tumours when hearing is good, we try to save any residual hearing or at least the cochlear nerve for possible cochlear implantation later if needed. The drawback can be the removal of the lateral intrameatal part of the tumour. The retrosigmoid tranmeatal approach has been popularised by Samii, and by Magnan in the neuro-otologic community. The tranmeatal approach should be planned on preoperative CT by evaluating the position of the semicircular canals and the extent of bone that can be removed safely in order to expose the internal meatus without compromising hearing. Samii’s group claims a 98% total resection of VS, even extending to the fundus, with functional hearing in 51% of cases and 94% facial nerve preservation (House Brackmann I-II).

Sterkers et al. performed an anatomical study stressing the drawback of the retrosigmoid approach in terms of exposition of the fundus of the internal auditory meatus. This can jeopardise complete tumour removal and hearing preservation. To avoid blind dissection we increasingly use the 4 mm 30° endoscope to control the fundus.

Woodson et al. summarised and demonstrated a very important consensus in the literature, clearly showing that, if hearing is preserved after surgery, hearing preservation is long-term.

We use the retrosigmoid approach as the first-choice approach to hearing preservation surgery. When hearing preservation is an objective, we perform intra-operative monitoring using auditory brainstem response (ABR) and cochlear nerve action potentials (CNAP). There were no significant differences in facial nerve outcome in our series between the translabyrinthine and retrosigmoid approaches. Moreover, electrophysiological tests did not predict facial nerve outcome after vestibular schwannoma surgery. Tumour size therefore remains the best pre-operative prognostic factor of facial nerve function outcome, i.e. outcome is better in smaller lesions.

4) The middle fossa approach

The middle fossa approach is used by a few centres for small tumours restricted to the IAC and when hearing preservation is a possibility, in other words when the pure-tone average is 30dB or better and the speech discrimination score exceeds 70%. This strategy affords a good approach to the fundus of the IAC and is selected when the tumour is situated laterally. However, since the facial nerve is the first structure encountered after the opening of the IAC, there is a higher risk of facial nerve injury, at least of a temporary nature, than in the other approaches. This is why we do not use this approach for vestibular schwannomas because facial nerve preservation is considered to be a higher priority.

Radiosurgery and stereotactic radiotherapy for VS

Radiation kills tumour cells by inducing extensive DNA damage. In classical radiotherapy, therapeutic efficacy is based on enhanced DNA repair capacity after radiation in normal cells compared to tumour cells. In radiosurgery, it is possible to selectively irradiate a sharply defined target, inducing tumour cell death and largely sparing the surrounding normal tissue. Lars Leksell (°1907-†1986) developed the application of single dose stereotactic radiotherapy to intracranial lesions. In a Gamma knife system, 201 Cobalt-60 sources are positioned in a sphere with the beams directed to a single focus point. The target point is defined by a stereotactic frame fixed on the patient’s head. The accumulation of these multiple beams creates a radiation volume.

An alternative is the Linac system, in which a linear accelerator is used to generate X-rays. These high-dose X-rays are sent through a heavy lead cone to shape a beam that can be rotated around the head of the patient so that the intersection of all the beams is again the intracranial lesion of the patient. The target is again localised using a stereotactic frame on the patient’s head but this tar-
get is fixed and the radiation source is moved in circles around the target. The beam can also be shaped by using lead leaflets known as ‘multileaf collimators’ (MLC). The final biological effect of both systems is very comparable. The tumour margin dose varies between 12 to 14 Gray.

The main problem in the overwhelming amount of literature is that many variables have been confounded in the cohorts of patients treated with radiosurgery. Some patients were treated at the time of diagnosis; others had undergone surgery (with partial resection and/or recurrence); follow-up was very short in some patients; hearing was not evaluated in all studies, etc.

We can distillate some clear facts. Follow-up periods vary from 2 years to 14 years. Most studies show tumour control of around 95% during follow-up. Facial nerve function is preserved in most studies in 95-97%. Hearing preservation is related to the direct dose to the cochlea. So VSs growing deep into the fundus involve a higher risk for hearing after radiosurgery. There are still some doubts about the preservation of serviceable hearing in the long term. Finally, tumour size exceeding 2.5 cm is not a good indication for radiosurgery.

More recently, there has been a rise in the use of fractionated stereotactic radiotherapy for VS. In this treatment, the total radiation dose of approximately 58 Gray is achieved with a fractionation of $5 \times 1.8$ Gray/week. The aim is to treat VSs larger than 2.5 cm in diameter.

In our department, we use radiosurgery as the first-choice treatment for elderly patients presenting with VSs smaller than 2.5 cm in which there is documented growth. Radiosurgery can be offered to younger patients with tumours smaller than 2.5 cm in cases of medical co-morbidity or after counselling of the patient.

**VS treatment strategy**

The main concern should be to provide the best treatment for each individual patient in order to maintain maximum physical and mental function in the long term. The challenge is to define and incorporate the different variables into this treatment algorithm. Variables include patient variables such as symptoms, age at presentation, social and professional context, medical condition and psychological condition. Tumour variables are tumour volume, extension, consistency, growth rate and location. Each patient variable can combine with a different tumour variable and vice-versa and therefore result in completely different treatment options.

In general, treatment can be “passive” or “active”. The questions to be addressed in passive treatment are 1) the investigational interval, 2) how and for how long to evaluate and 3) when should passive treatment switch to an active strategy? Which factors – growth but also hearing status – will be taken into account? Once the active treatment option has been adopted, it is important to determine whether ‘active’ stands for surgery or stereotactic radiotherapy or a combination of the two. If surgery is proposed, the type of surgery (total, subtotal) and the approach (translab, retrosigmoid) should be considered.

Even after “active” treatment patients return to “passive” treatment status, a decision will be required again about the follow-up interval, and tumour regrowth or recurrence should result in the repeat consideration of “active” treatment options.

In our department we have developed a treatment algorithm as a guideline for VS treatment (Figure 1). It will be clear that, in some cases, the algorithm is aborted for different reasons such as patient preference, general medical condition etc. Even so, we find it useful, for ourselves and our patients, to use this structured treatment strategy. This strategy

![Figure 1](image)

**Figure 1**

also results in a solid cohort of patients, who are treated using a more or less identical strategy, something that can be of scientific value over a period of time.

References


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