

Vestibular schwannoma between 1 and 3 cm: Importance of the tumor size in surgical and functional outcome



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ABSTRACT

Objective: The aim of the study was to compare the surgical and functional outcomes of the microsurgical osteoplastic retro-sigmoid approach in a semi-sitting position in two groups of patients with vestibular schwannomas (VSs) ranging from 1 to 3 cm in size.

Method: A 5-year retrospective evaluation was made of these two groups of patients with VS: Patients with VS sizes 1–<2 cm in maximal intra/extrameatal diameter ($n = 292$) were assigned to group “A” and a matched group of patients with VS between 2 and 3 cm in size ($n = 154$) were assigned to group “B”.

Results: Significant differences in postoperative outcomes ($p < 0.05$) were found for facial nerve function of House–Brackmann grade I (94% group A vs. 78% group B) and preservation of preoperative hearing (51% group B vs. 34% group A).

Patients with tumors sizes ranging between 1 and <2 cm exhibited total tumor removal with significantly higher facial nerve preservation and hearing function preservation rates compared with patients with tumors 2–3 cm in size.

Conclusion: Even a small increase in tumor size correlated with a significant reduction in good hearing and facial preservation postoperatively, which implies that tumor removal should be performed at the earliest stage possible. Furthermore, these results contradict recommending the wait-and-see approach for intra/extrameatal tumors.

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1. Introduction

The goal of every surgery should be to avoid life-threatening events and to find ways to preserve nerve function, and thus improve patient's quality of life [3]. Surgery for vestibular schwannoma (VS) remains one of the most demanding surgical procedures at the same time that microsurgery is trending as the primary tool for managing patients with VS. Consequently, there is a pressing need to better identify and measure factors influencing functional outcomes in microsurgery patients.

VS, often called acoustic neuroma [1–3], is currently the preferred name for a tumor that arises not from the acoustic nerve but from the schwann-cell sheath of the pars superior of the vestibular nerve (not the cochlear portion) [4]. These tumors are histologically benign [3] and diagnosis can often be made by

clinical history alone. The prevalence in adult males and females is about equal [5].

Symptoms are closely associated with tumor size and often start with a triad of unilateral sensorineural hearing loss, tinnitus, and balance difficulties (gait disturbances) [4,5]. Yet, large tumors can still be present but show surprisingly few symptoms and almost no signs except for hearing loss. Even 4- and 5-cm tumors may have virtually no physical findings and few associated symptoms. In addition, a subset of patients has central neurofibromatosis, bilateral VS, and multiple intracranial meningiomas as well as other neoplasms.

Time of diagnosis, clinical experience, and tumor size were identified as factors significantly influencing the postoperative outcomes regarding hearing and facial nerve function. Several authors report hearing preservation of about 50% after removal of small tumors and even better results have been reported for completely intracanalicular lesions [3,6].

Therefore, the philosophy of the senior author (A.S.) is that in all cases of VS, regardless of tumor size, complete surgical removal and preservation of facial nerve function should be the goal and the greatest effort to preserve functional hearing should always be made.

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Table 1

The distribution of patients in two different groups in our study depending on the tumor size. We have compared the surgical and complication results in these two groups of acoustic neuromas.

Group	A	B
Tumor size	1–< 2 cm	2–3 cm
Population	292	154
Mean size	1.7 cm	2.6 cm

There are many treatment options in management of acoustic neuromas including wait-and-see [10–18], microsurgery, fractionated stereotactic radiotherapy [7], gamma knife surgery [8], and radiosurgery [9]. The question remains as to which of these methods is the best option for management of acoustic neuromas. In this study, the focus is on microsurgery as a treatment method and the evaluation of tumor size as a postoperative functional determinant (groups A and B) (Table 1).

Because the results are presented in standardized form (House–Brackmann scale for facial function and Gardner and Robertson classification for hearing function), they can be used for comparisons between the various methods used to treat VS.

2. Methods

2.1. Study design

Between 1999 and 2010, the senior author (AS) excised more than 1000 VS via the suboccipital retrosigmoid approach and semi-sitting position for patients in two different clinics (Table 2). In addition, between 2002 and 2010, 627 cases of VS were surgically treated at the Clemens Hospital in Münster.

For the candidate patients, re- and postoperative neuro-radiological data, operative reports, and follow-up data for up to 8 years were reviewed. Demographics recorded for each patient included age, gender, initial symptoms or signs, neurological status before and after surgery, tumor size, surgical complications, and neurological status at discharge and at follow-up.

The completeness of tumor removal was assessed intraoperatively by visual observation at the end of surgery and early after surgery with imaging studies (MRI with contrast) 1 day after surgery (Fig. 2). Facial nerve function was assessed according to the House–Brackmann scale and hearing level was classified according to the Gardner Robertson classification. Matching criteria (Table 3) were then used to stratify patient characteristics. All surgeries were performed by the same surgeon at the same institution using identical standards to further reduce statistical bias. After applying matching criteria, the pool of candidates ($n = 424$) was then apportioned into two groups. Patients with tumor size 1–<2 cm were

Table 2

The distribution of surgically treated patients with vestibular schwannomas in two different centers.

Group 1	1997–2002	420
Group 2	2002–2010	572 + 54 ^a
		1046

^a54 patients were excluded from the study (see Table 3).

Table 3

Our exclusion criteria and the distribution of our excluded patients.

The patients with NF I	1
The patients with NF II	6
Recurrent tumor after Gamma knife surgery	7
Recurrent tumor after radiotherapy	12
Recurrent tumor after microsurgery	27

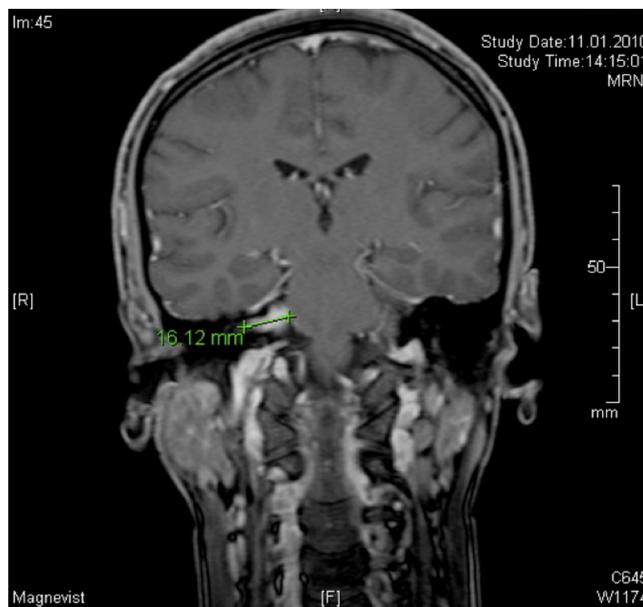


Fig. 1. The definition tumor size in our study is based on maximal intra/extrameatal diameter.

assigned to group “A” ($n = 292$) and patients with tumor size of 2–3 cm were assigned to group “B” ($n = 154$) (Table 1) (Figs. 1 and 2).

In the literature, the use of grade III Gardner–Robertson as a measure of functional hearing is controversial. Various reports as well as experts consider using grade I–III as a measure of good hearing function. Based on expert opinion and such reports, we also considered Grade I–III as a measure of good hearing function [10].

To confirm the grading, we asked all of our grade III patients about their ability to hear stereo sound and answer the telephone with their problem ear. As a result, all could answer the telephone and hear stereo sound. We did not use the Speech Discrimination Test because our patient data was not complete enough for meaningful statistical analysis.

2.2. Patient demographics

The average age for patients in group A was 50.2 years (range 28–65 years); there were more men (54%) than women (46%). The left side was involved in 40% of the cases and the right side in 60% of the cases. The mean follow-up duration was 63 months (range 9–88 months). The mean age of patients in group B was 53.7 years, 57% were male and 43% female. The left side was involved in 44% of the cases and the right side in 56% of the cases. The mean follow-up duration was 67 months (range 12–70 months) (Table 4). The chi-square and Fisher exact tests were applied to determine if the proportion of surgical and functional outcomes between the two groups were equal. A p -value < 0.05 indicated a statistically significant difference.

Table 4

The comparison of group A and B in our study.

Group	A	B
Tumor size	1–< 2 cm	2–3 cm
Number	292	154
Mean age	50.2 year	53.7 year
Mean F/A	63 month	67 month
Male	54%	57%
Female	46%	43%

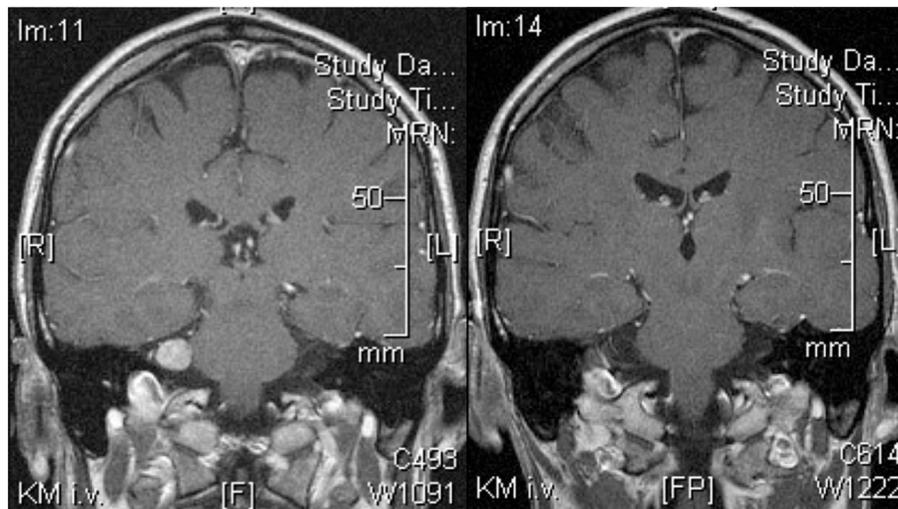


Fig. 2. Pre and postoperative imaging study in a patient selected from our series of Vestibular Schwannomas with intra/extrameatal size of 1.6 cm.

2.3. Surgical strategy

The standard retrosigmoid approach is often preferred by neurosurgeons and has many advantages. One major advantage is the flexibility of the approach with regard to tumor size and extracanalicular extension. The semi-sitting, supine, supine oblique park bench and lateral oblique positions have been used for suboccipital removal of acoustic neuromas [25]. All patients in our series were operated on in the semi-sitting position (Fig. 3).

Surgery was performed with the patient under general anesthesia and in skeletal fixation in a true semisitting position. An area approximately two fingers-breadth posterior to the mastoid was identified for the incision. An excision was made extending from just superior to the external auditory canal down to approximately 2 cm below the occiput. Clean dissection is important because a layered closure can help prevent postoperative incisional CSF leaks [26].

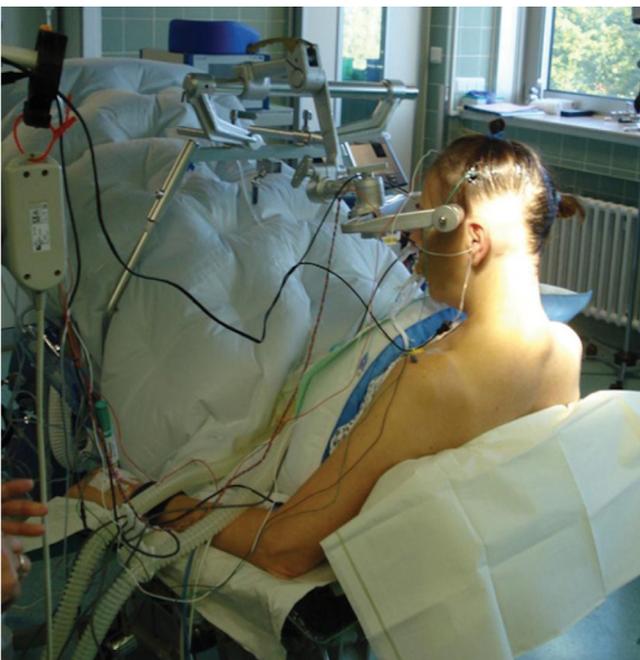


Fig. 3. The semisitting position in operation of a left-sided vestibular schwannoma via an osteoplastic suboccipital approach.

We placed the bur hole in the most superior part of the incision above the asterion, and then performed the occipital craniotomy with high speed drilling over the sigmoid sinus, using the craniotome on the opposite side to remove the bone. We paid close attention to any air cells that became visible during the opening procedure and applied bone wax when identified.

Once the bone was removed, the dura was opened in a cruciate fashion superiorly and laterally (parallel with the sigmoid sinus) 1 cm away from the junction of the sigmoid and transverse sinuses. This exposure identified the inferior margin of the transverse sinus and the medial margin of the sigmoid sinus. We routinely opened the cisterna magna to allow CSF to drain, and from this point on, retractors were used more to protect the cerebellum than to retract brain tissue.

Once the posterior aspect of the tumor was exposed, the next step in the procedure was to stimulate the seventh nerve to be certain it is not spread over the posterior aspect of the tumor. Once the surgeon visually inspected the tumor and made certain that the seventh nerve was not in an aberrant position, the next step was to identify the plane between capsule and arachnoid. An extracapsular dissection was then carried out, gradually exposing the posterior, superior, inferior, and then medial margins of the tumor.

For our tumor series, because all tumors were smaller than 3 cm in diameter, the internal auditory canal (IAC) was always drilled away first. For drilling the IAC, we preferred to raise small dural flaps, which were subsequently replaced, and allowed the canal to be drilled completely away. The posterior wall of the canal was removed, and, depending on the anatomy, the dissection was carried superiorly and inferiorly to expose more of the tumor [27,28].

After drilling the IAC, we removed the tumor from the lateral side up to medial side using ultrasound-guided aspiration, microsissors, and microcup forceps. Once internal debulking of the tumor was deemed adequate, the tumor capsule was gently moved into the field while maintaining the arachnoid plane.

Adhesions between tumor and dura at the margins of the porus make it extremely difficult to dissect the seventh nerve free at this level. Therefore, a facial nerve stimulator was used to verify that there was no involvement of the capsule with the facial nerve [28–31].

Frequently there are air cells in the drilled margins of the petrous bone. This area is filled with muscle tissue and fibrin once tumor removal is complete in order to prevent postoperative internal CSF leakage. Meticulous hemostasis was maintained. The dura was

Table 5

The comparison of postoperative facial nerve preservation rate at discharge time in both groups.

Group	A (%)	B (%)
Grade I (Facial excellent preservation)	94	78
Grade II	6	20
Grade III	0	2
Facial anatomical preservation	100	100

Table 6

The pre and postoperative Hearing results in group A based on Gardner–Robertson Scale.

	I	II	III	IV
Preop	33%	26%	35%	6%
Postop	18%	16%	17%	49%

Table 7

The pre and postoperative hearing results in group B based on Gardner–Robertson Scale.

A	I	II	III	IV
Preop	27%	31%	29%	13%
Postop	12%	15%	7%	66%

Table 8

The comparison of functional hearing preservation in both groups based on good hearing function (first line) or serviceable hearing preservation (second line).

Group	A	B
Hearing functional preservation (I, II, III G and R) ^a	51%	34%
Preservation of Serviceable Hearing (I, II G and R)	34%	27%

^aIn the literature it is controversy to use grade III Gardner–Robertson as a functional hearing. Various literatures as well as expert people consider using grade I–III as a good functional hearing. Based on expert opinion and such literatures, we also consider Grade I–III as a good functional hearing [10] (See Section 2).

closed in a watertight fashion. At this point, we replaced the bone and we believe this method (osteoblastic instead of osteoclastic) reduces the postoperative headache and neck pain, and CSF leakage (incision leakage) that are common after VS surgery in a semisitting position (Table 12) [32].

3. Results

The mean tumor size in Group A was 1.7 cm and in Group B was 2.6 cm. Total removal was achieved in all patients in both groups. The anatomical integrity of the facial nerve was preserved in 100% in both groups.

At time of discharge, 94% of patients in group A had excellent (H&B Grade I) and 6% had good (H–B Grade II) facial nerve function. In 51% of patients in group A, the preoperative hearing level was preserved. There was no occurrence of newly developed lower cranial nerve dysfunction in any of the patients.

In comparing the two groups, a significant difference was found for the following outcomes: excellent facial nerve function (H&B grade I) (94% in group A vs. 78% in group B) and preservation of preoperative hearing (51% in group A vs. 34% in group B) (Tables 5–8).

Postoperative facial paresis was directly evaluated before discharge on the 7th day of hospitalization and after 6 months. Differences between postoperative facial paresis grade at discharge and 6 months after surgery are summarized in Tables 9 and 10.

CSF leak developed in 4.5% of patients in group A and in 4% in group B, but the difference was not statistically significant. Approximately 2% of all the patients underwent lumbar drainage because of postoperative CSF leakage and wound revision was necessary

Table 9

The improvement of facial paralysis up to discharge time (7th postoperative day) after HES (hydroxyethyl starch) infusion.

	Grade II to I	Grade III–II
Group A	1%	0%
Group B	3%	0.5%

Table 10

The improvement of facial nerve paralysis from discharge day up to first follow up after 6 months.

	Grade II to I	Grade III–II
Group A	1%	0%
Group B	7%	1%

Table 11

The distribution of different types of CSF Leak in both groups of patients.

CSF leak	A	B
Rhinoliquorrhea	2%	2%
Incisional CSF leak	2.5%	2%
Ear CSF leak	0%	0%

Table 12

The comparison of different complications in both groups.

Group	A	B
Wound revision	1%	1%
CSF leak	4.5%	4%
Lumbar drainage	3%	3%
Headache	8%	9%
Neck pain	7%	7%
Hydrocephalus	0%	0%

in only 1% of all patients (Tables 11 and 12). The perioperative mortality rate in both groups was 0%.

In our series, we did not observe hydrocephalus or vascular complications. During surgery in the semi-sitting position, we usually use intraoperative Transesophageal Echocardiography (TEE) but in the series described herein we used intraoperative Doppler Sonography. For surgical removal of VS less than 3 cm, we observed three cases (<1%) with postoperative radiographically-relevant air embolism in chest X-Ray (bilateral air space opacification or ‘white out’ because of increased vascular permeability similar to ARDS) but none of these patients experienced neurological deficit and none was hemodynamically unstable during surgery. By CT scan (in the first 24 h) we observed pneumocephalus in 37% of patients, and in 12% of the cases, pneumocephalus led to late extubation (>3 h).

4. Discussion

Determining the overall prevalence of VS in the general population can be challenging because of the deceptive nature of the symptoms; however, understanding the prevalence of acoustic neuromas and their growth rate are important in order to predict outcomes and determine treatment options. Increased tumor size complicates tumor resection and decreases the chance of preserving good hearing and facial nerve function. Consequently, expected growth rate plays a key role in determining whether continued observation or surgical intervention is indicated. Moreover, the proximity of acoustic neuromas to the brain stem raises concerns that continued growth could lead to brain stem compression.

The growth pattern of acoustic neuromas is variable and incompletely understood. In a study published by Nikolopoulos et al. as many as 75% of tumors showed no growth, which has supported a ‘wait-and-rescan’ policy for many patients even though there are

no reliable predictors of tumor behavior, and some tumors grow rapidly. Primary longitudinal studies are needed to better define the natural history of these tumors to limit unnecessary interventions [43].

A review of published studies with at least 50 patients indicated that the proportion of tumors exhibiting growth during observation ranges from 30% to 85% and that the growth rate also varies from 0.4 to 2.4 mm/yr [10–19]. Stangerup et al. published a series of 552 patients observed for an average of 3.6 years and found that tumors showing growth did so in the first 5 years after diagnosis. Interestingly, they also found that intrameatal and extrameatal tumors had a statistically significant different rate of growth with 17% of intrameatal and 29% of extrameatal tumors showing growth within 4 years of diagnosis [3]. This interesting finding emphasizes that the growth of intrameatal and intra/extrameatal tumors are different. Furthermore, the findings show that a measurable proportion of small VS, particularly pure intrameatal tumors, will not grow during follow up. Thus, we excluded patients with pure intrameatal VS because these tumors have a completely different natural history.

In view of current evidence, time of diagnosis and tumor location with respect to the IAC play an important role in the evolution of VS [11–19]. Not so well understood is the evolution of VS in patients with neurofibromatosis type 2 (NF2). VS tumors in these patients frequently exhibit unpredictable growth rates with more rapid growth in younger patients [20–23]. Slattery et al. found that the average growth rate (measurement of the tumor's greatest diameter) was 1.3 mm/yr in patients with NF2 [24]. Therefore, we also excluded patients with NF because of the great difference in the natural history of these tumors (Table 4).

Nevertheless, despite the fact that slow-growing tumors with no neurological effect can often be monitored for signs of progression clinically and with serial radiographic studies, our results indicate (and previous literature suggests) that the sooner the tumor is excised after diagnosis, the more positive the outcome prognosis for facial nerve and hearing function. However, despite our findings, we believe continued observation is an acceptable option for many patients with small-sized tumors, particularly intrameatal tumors and VS in elderly patients [41–43].

4.1. Facial (seventh) nerve preservation

In the majority of patients, the issue of greatest concern is seventh nerve function [33,34]. The percentages of functional and anatomical facial nerve preservation in our series of acoustic neuromas are shown in Table 8.

These outcomes are in agreement with those of other experienced acoustic neuroma surgeons today [31].

There are different mechanisms of facial nerve injury during acoustic neuroma surgery: direct trauma, stretching, vascular injury, and thermal effect [35]. Recent literature described the role of vasospasm in postoperative facial paresis and found positive results with the use of nimodipine and hydroxyethyl starch (HES) [6–8]. We also have used HES infusion in patients with facial paresis in our series immediately after surgery and continuing for 10 days; this treatment has had very positive effects in these patients. We observed a 4% recovery of facial nerve paresis (H&B Grade change after 10 days) after IV HES infusion. Table 9 shows the effect of HES infusion in our patients with facial paresis after the surgery.

Recently, several series using lower marginal doses of radiation from 12 to 13 Gy have produced excellent results in terms of tumor control, facial nerve preservation, and trigeminal nerve preservation. Regarding facial weakness and numbness, Beegle et al. reported 4.6% new-onset facial weakness and 3.6% new-onset facial numbness in a series of 390 patients. Lowering the marginal dose of radiation reduced the rate of facial nerve complications in this series [36]. These results are comparable with our results

of facial nerve function preservation when using the retrosigmoid approach.

In our series of acoustic neuromas (between 1 and 3 cm) we did not have any incidence of intraoperative anatomic or physiologic transection.

4.2. Cochlear (eighth) nerve preservation

Currently, there are two methods of hearing monitoring in acoustic neuroma surgery: auditory brainstem response (ABR) and direct cochlear nerve monitoring. ABRs provide intraoperative feedback for cochlear nerve function but its limitation is a 2–3-min lag time during recording. On the other hand, direct cochlear nerve monitoring provides more feedback and can improve the hearing preserving results [37]. Danner and coworkers found a hearing preservation rate of 41% with the use of ABRs and 71% with direct cochlear nerve monitoring for tumors less than 1 cm [38].

Strauss et al. showed in a postoperative study that the use of nimodipine and HES for an average of 9 days after surgery increased the rate of hearing preservation in comparison to control group; this finding suggests there is a vasoactive role in hearing loss [39].

Paek et al. reported maintenance of serviceable hearing in 52% of patients treated with a prescription dose of 12 Gy with a median tumor volume of 3 cm [3,8]. Iwai et al. reported maintenance of serviceable hearing in 56% [40]. These results compare favorably with the results seen in our series using the retrosigmoid approach.

We performed only ABR monitoring in our series and we did not use direct cochlear monitoring. Moreover, we did not routinely use nimodipine. The hearing preservation results for both groups are shown in Tables 6–8.

4.3. Surgical position

There has been a general decline in the use of the semisitting position in posterior fossa surgery out of fear of a VAE (Venous Air Embolism). However, there is no statistical relevant evidence justifying the abandonment of this surgical position. Feigl et al. showed in a prospective study that under meticulous anesthetic and neurosurgical cooperation, even patients with a PFO (patent foramen oval) could be operated on in the semisitting position with only a very low risk for VAE [44].

5. Conclusion

The goal of VS treatment should be a one-stage complete tumor removal and the preservation of neurological function, because this is the main determinant of the patient's quality of life. This objective was achieved safely and more successfully when the retrosigmoid approach in the semi-sitting position was used to excise tumors 1–< 2 cm in size. Surgery for tumors in this size range achieved complete tumor removal with significantly greater facial nerve preservation and hearing function conservation compared to surgery for tumors 2–3 cm in size.

The current literature reports a range of tumor growth of 0.4–2.4 mm/yr; thus, any delay in tumor removal may lead to a change in tumor size. Because our results indicated that even a small increase in tumor size significantly correlates with a possible reduction of facial nerve conservation and hearing preservation postoperatively, tumor removal should be performed at an early stage, especially for young patients with VS between 1 and 2 cm in size. Furthermore, our findings contraindicate recommending the wait-and-see approach at least for intra/extrameatal tumors.

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