Vestibular schwannoma: Experience of Hospital de Egas Moniz between 2007 and 2021

Original Article

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Abstract

Objective: To report the experience of a tertiary center in the diagnostic approach, therapeutic decision and follow-up of patients with sporadic vestibular schwannoma.
Material and Methods: Retrospective study of patients with sporadic vestibular schwannoma followed at Hospital Egas Moniz between 2007 and 2021. Demographic and clinical data, tumor classification, treatment decision and outcomes of 79 patients were collected from clinical records and the following outcomes analyzed: tumor resection; hearing loss; and degree of facial paralysis.
Results: 28% of the operated patients had a total or near-total resection of the tumor and 72% a subtotal resection. In 7 of each 8 retrosigmoid surgeries resulted in cophosis. Same result occurs in 2 of the 3 middle fossa approaches. No difference was found in the prevalence of facial paralysis between the various surgical approaches.
Conclusion: This is one of the largest series reported nationally. The outcomes evaluated are similar to published international series.

Keywords: Acoustic neuroma; Facial paralysis; Hearing preservation; Radiosurgery; Retrospective studies; Surgery; Treatment outcome; Vestibular schwannoma; Watchful waiting.

Introduction

Vestibular schwannoma (VS) has an annual incidence of 1:100,000 and accounts for 6–7% of all intracranial tumors and 90% of all lesions located in the cerebellopontine angle (CPA).[1] These are neuroectodermal tumors arising from the Schwann cells of one of the vestibular branches of the vestibulocochlear nerve; hence, the term vestibular schwannoma[2] has been used. VSs usually exhibit slow growth at the level of the internal auditory canal (IAC), CPA, cochlea, and/or labyrinth. Larger tumors can compress the brainstem.[1] The most common presenting symptoms are gradual hearing loss (90%) and tinnitus (>60%).

Imbalance, dizziness, and vertigo may also occur. Up to 12% of patients exhibit changes in facial sensitivity due to the compressed trigeminal nerve, and up to 6% present with paresis of the facial nerve, usually patients with a larger vs. Headaches occur as a consequence of hydrocephalus or with larger tumors that cause greater compression of the involved structures.[2,3] The scales most commonly used to classify the tumor according to its size are the House[4], Koos[2], and Samii scales[5]. In addition to the tumor size, the therapeutic approach must consider factors such as the rate of tumor growth, degree of useful hearing, and the patient's age and comorbidities. The probability of total tumor resection with preservation of the auditory function and facial nerve should also be considered. The patient's preference and choice, when correctly informed, is also a relevant factor in the final therapeutic approach or in deciding between various treatments or the different surgical approaches for this disease.[1]

The three main treatment options for patients with sporadic VS are watching or watchful waiting, also called "wait and scan," surgery, and stereotactic radiosurgery (SRS).[6] Most patients with small- and medium-sized tumors have high rates of tumor control and excellent facial nerve outcomes, regardless of the treatment modality.[7]

This study aimed to report the experience of Egas Moniz Hospital in the diagnostic approach, therapeutic options, and follow-up of patients with sporadic VS treated between 2007 and 2021.

Materials and Methods

This was a retrospective study of patients with acoustic neuroma (vestibular schwannoma) treated at Egas Moniz Hospital between 2007 and 2021.

Initially, a search for patients was performed in the electronic database of the Sonho® institution in the period between 2007 and 2021 (15 years) using the various possible diagnostic codes to identify cases of sporadic VS in the International Classification of Diseases (9th and 10th revisions) (Table 1).

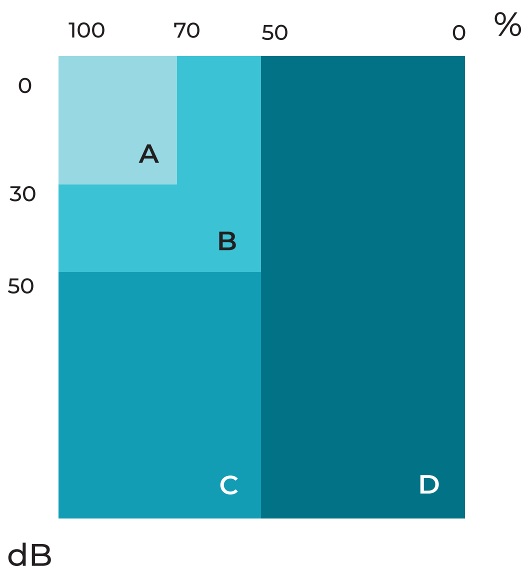
The initial search identified 111 patients. Of these, 32 patients were excluded for the following reasons: 23 patients had different diagnoses (six cases of facial nerve schwannomas, five of neoformations of the geniculate ganglion, three cases of type 2 neurofibromatosis, three cases of iatrogenic facial paralysis, two

Table 1
Diagnostic codes ICD9 and ICD10 used in the study

225	Benign neoplasm of brain and other parts of nervous system
2251	Benign neoplasm of the cranial nerves
3529	Cranial nerve disorder, unspecified
3885	Disorders of the acoustic nerve
9515	Injury to the acoustic nerve (eighth pair) (auditory nerve)
23773	Schwannomatosis
38916	Sensorineural hearing loss, asymmetrical
D333	Benign neoplasm of the cranial nerves
H90A21	Sensorineural hearing loss, unilateral, right ear, with restricted hearing on the contralateral side
H90A22	Sensorineural hearing loss, unilateral, left ear, with restricted hearing on the contralateral side
H933X1	Disorders of the right acoustic nerve
H933X2	Disorders of the left acoustic nerve
H933X9	Disorders of unspecified acoustic nerve
R42	Dizziness and giddiness
S0461XA	Injury to the acoustic nerve, right side, initial encounter
S0462XA	Injury to the acoustic nerve, left side, initial encounter

Figure 1
System of classification of Hearing by the American Academy of Otolaryngology-Head and Neck Surgery (AAO – HNS)

System of classification of hearing		
Class	**Mean hearing threshold**	**Discrimination maximum**
A	≤ 30 dB	≥ 70%
B	> 30 dB ≤ 50 dB	≥ 50%
C	> 50 dB	≥ 50%
D	Any threshold	< 50%



cases of Bell paralysis, two meningiomas, one cholesteatoma of the petroux apex, and one trigeminal nerve schwannoma); nine patients were excluded because they had insufficient clinical information or were lost to follow-up. The final study sample comprised 79 patients. The clinical records of the 79 patients were consulted, and after anonymization, the following demographic and clinical data were retrieved: age, sex, side of the lesion, main presenting symptom, other symptoms, degree of hearing loss, and degree of facial paralysis. To document the manifestations, validated international scales were used whenever available, such as the following: the scale proposed by the American Academy of Otolaryngology-Head and Neck Surgery to classify the hearing loss in the affected ear, based on tone audiometry (mean auditory threshold) and speech audiometry (maximum speech discrimination) (Figure 1); the scale used to classify tinnitus according to the Consensus proposed at the 7th International Conference on Acoustic Neuroma regarding sporadic VS (Figure 2); the scale used to classify imbalance according to the same source (Figure 3); the House-Brackmann scale, used universally to classify the severity of facial paralysis and reconfirmed in the Consensus proposed at the 7th International Conference on Acoustic Neuroma (Figure 4). The Samii scale was used to classify the tumor according to its size, as defined by magnetic resonance imaging (MRI). Figure 5 summarizes the various classifications and their significance.

The main outcomes of the study were as follows: degree of tumor resection (total, near

Figure 2
System of classification of tinnitus according to the 7th International Conference on Acoustic Neuroma

System of classification of tinnitus for Vestibular Schwannoma	
Grade	**Description**
I	Without tinnitus
II	Mild or intermittent, only audible in a silent environment
III	Moderate or persistent, may be audible throughout the entire day
IV	Severe and persistent, interferes with work and sleep

Figure 3
System of classification of imbalance according to the 7th International Conference on Acoustic Neuroma

System of classification of imbalance for Vestibular Schwannoma	
Grade	**Description**
I	Without vertigo or imbalance
II	Mild or occasional
III	Moderate or persistent
IV	Severe and persistent, interferes with life

total, or subtotal); loss of hearing preoperatively and in the immediate postoperative period; degree of facial paralysis preoperatively, in the immediate postoperative period, and at one year of follow-up. All these data were analyzed statistically using version 26 of the IBM SPSS

Figure 4
House-Brackmann scale for facial paresis

House-Brackmann Scale				
Grade	**Description**	**General**	**Symmetry AT REST**	**Mimicry UNDER EXERTION**
I	Normal	Preserved facial mimicry	Symmetric	With no changes
II	Mild paresis	Mild loss of tone	Symmetrical unchanged tone	**Forehead:** good to moderate **Eye:** complete closure with minimum effort **Mouth:** mild asymmetry
III	Moderate paresis	Evident asymmetry of facial mimicry	Symmetrical unchanged tone, not disfiguring: No paresthesias, contractures or hemi-facial spasms	**Forehead:** good to moderate **Eye:** full closure with effort **Mouth:** asymmetry under maximum exertion
IV	Paresis moderately severe	Evident asymmetry of facial mimicry	Asymmetry Disfiguring loss of tone	**Forehead:** total loss of tone **Eye:** incomplete closure **Mouth:** asymmetry under maximum exertion
V	Severe paresis	Minimum facial mimicry	Asymmetrical	**Forehead:** total loss of tone **Eye:** incomplete closure **Boca:** slight movement
VI	Paralysis	Absence of facial mimicry	Without movement	Without movement

Figure 5
Several classifications of the size of Vestibular Schwannomas according to the 7th International Conference on Acoustic Neuroma

Tumor size (Max diameter in the CPA)	STERKERS	HOUSE	KOOS	SAMII	Tumor description
0 (Intracanal)	Tube type	Intracanalar	Grade I	T1	Limited to the IAC
≤ 10 mm	small	Grade 1 (small)	Grade II	T2	Beyond the IAC
≤ 15 mm		Grade 2 (Medium)		T3a	Tumor occupies the CPA
≤ 20 mm	medium				
≤ 30 mm		Grade 3 (Moderately large)	Grade III	T3b	Tumor occupies the CPA and contacts the stem without compressing it
≤ 40 mm	large	Grade 4 (large)	Grade IV	T4a	Tumor compresses the stem
> 40 mm	very large	Grade 5 (Giant)		T4b	Deviation and marked deformation of the stem and of the IV ventricle under tumor compression

statistics software. The chi-square test (X^2) and respective adjustments were performed in the statistical evaluation of non-quantitative variables; Fisher's exact test and Monte Carlo correction were used for the analysis of tables 2x2 or >2x2, respectively, when the requirements for the expected frequency were not met. Quantitative variables, after normal distribution was excluded, were assessed using the Kruskal-Wallis test, Mann-Whitney test, and Spearman's correlation. Statistical significance was set at $p < 0.05$.

All ethical procedures recommended by the Declaration of Helsinki of the World Medical Association were followed, and all data were anonymized and treated anonymously.

Results

Baseline characteristics

Thirty patients (38%) were men, and 49 (62%) were women. Thirty-five patients (44%) had a tumor in the right ear, and 44 patients (56%) had it in the left ear. The mean and median ages of the patients were 59 years and 61 years, respectively.

The most common presenting symptom was progressive unilateral hearing loss (61 patients [77%]). Seven patients (9%) presented with sudden deafness.

Regarding tinnitus, 38 (48%) patients did not exhibit this symptom, and there was no data on this for nine patients (11%). Tinnitus was mild in 13 patients (17%), moderate in 18 patients (23%), and severe in one patient (1%) (Figure 7).

With regard to imbalance or vertigo, 38 patients (48%) did not exhibit this symptom, and there was no data on this for seven patients (9%). Imbalance was mild or occasional in 22 patients (28%), moderate or persistent in nine patients (11%), and severe and persistent in three patients (4%) (Figure 8).

Regarding auditory deficit, 18 patients (23%) had grade A hearing, 23 patients (30%) had grade B hearing, eight patients (10%) had grade C hearing, and 29 patients (37%) had grade D hearing (Figure 9).

The distribution of patients according to facial mimicry i n the House-Brackmann scale was as follows: 56 (71%) had grade I, seven had grade II, four had grade III, eight had grade IV, three had grade V, and one had grade VI (Figure 10). The distribution of patients according to the Samii classification was as follows: 19 (24%) were in stage T1, 10 (13%) were in stage T2, 15 (19%) were in stage T3a, 16 (21%) were in stage T3b, seven (9%) were in stage T4a, and 11 (14%) were in stage T4b (Figure 11).

Figure 6
Graph representing the degree of tinnitus presented by patients

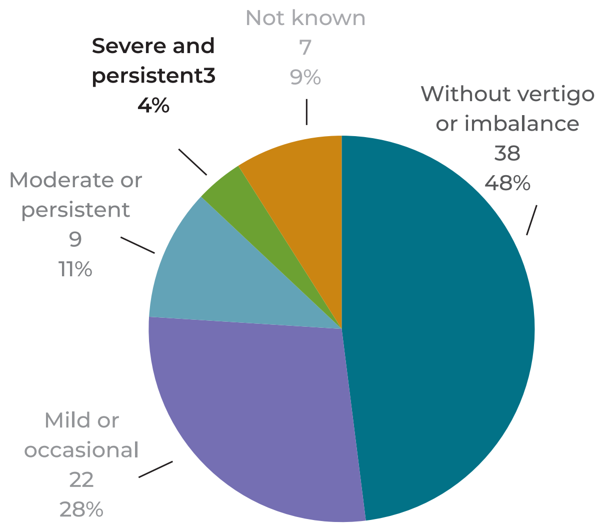


Figure 7
Graph of the tinnitus grades exhibited by the patients

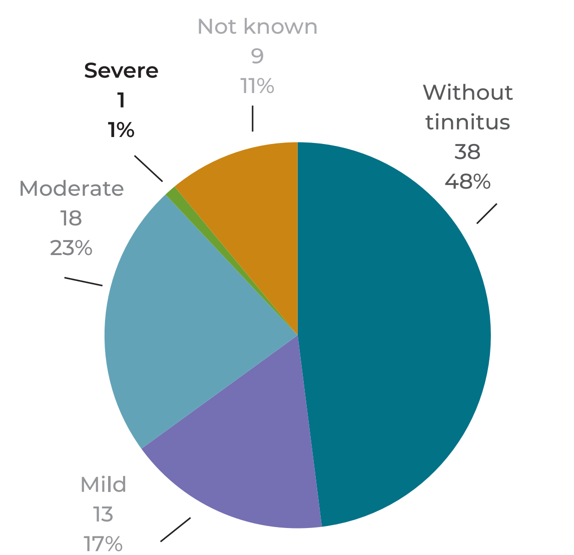


Figure 8
Graph of the vertigo/imbalance grades exhibited by the patients

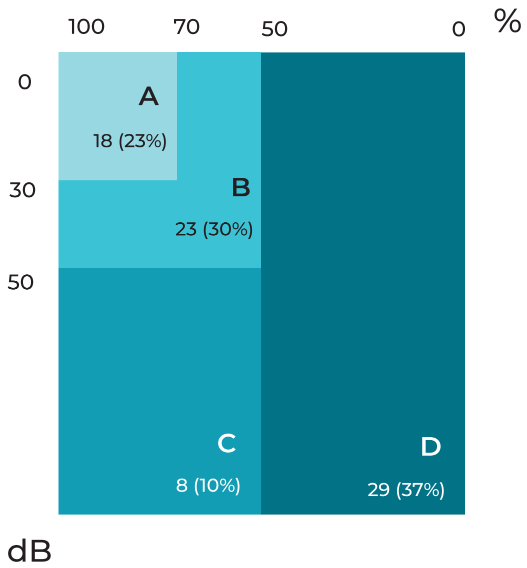
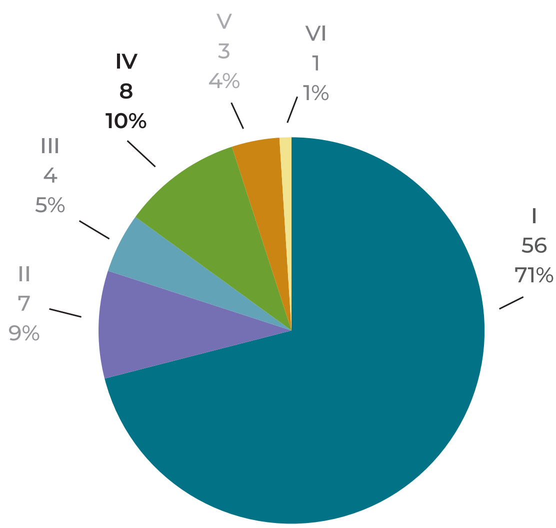


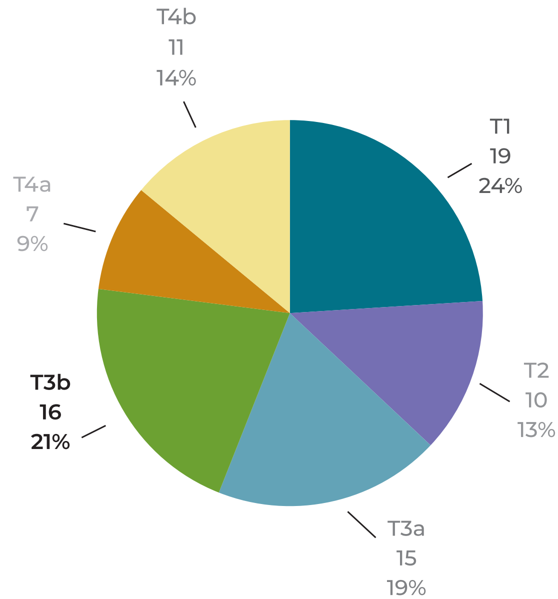
Figure 9
Graph of the auditory deficit exhibited by the patients



Treatment

The initial approach was wait-and-scan in 61 patients (51%), surgical treatment in 16 patients (20%), and radiosurgery in one patient (1%) immediately at the time of diagnosis (patient's choice). Of the 61 patients in whom the wait-and-scan approach was used initially, 21 underwent surgery, and two underwent radiosurgery because tumor growth was detected during follow-up.

Figure 10
Graph of the facial paresis grades exhibited by the patients



Three approaches were used in the 37 surgical procedures performed in the following proportion: 22 procedures via the retrosigmoid approach (60%), 12 via the translabyrinthine approach (32%), and three via the middle cranial fossa (8%) (Tables 2, 3, and 4)

Assessment

Regarding tumor resection, of the 25 patients with adequate clinical data, total tumor excision was performed in two (18%) of the 11 procedures performed through the translabyrinthine approach. Moreover, near-total excision was performed in five patients, three (27%) of the 11 translabyrinthine approaches, one (9%) of the 11 retrosigmoid approaches, and one (33%) of the three approaches via the middle cranial fossa. The remaining 18 patients had residual disease.

There was no statistically significant difference ($p>0.05$) in the prevalence of facial paresis among the various approaches up to approximately one year after the surgery. The prevalence of paresis with a grade higher than 3 was 33.3%, 27.3%, and 66.7% for the translabyrinthine, retrosigmoid, and middle cranial fossa approaches, respectively.

There was a statistically significant moderately

Table 2
Patients in whom the surgical approach was via the middle cranial fossa

Staging (Samii)	Age	Sex	Laterality	Hearing (AAO-HNS)	House-Brackmann (immediate postoperative)	House-Brackmann (6-12 months)	Hearing after surgery (AAO-HNS)	Complications	MRI
T1	39	F	Right	A	I	I	D	Vestibular deficit	R
T1	65	M	Right	B	IV	III	B	Exposure keratitis	R
T1	63	F	Left	A	IV	IV	D	-	NTR

F – Female; M – Male; NTR – Near-total resection; R – Residual; T – Total; MRI – Magnetic resonance imaging;
AAO-HNS – American Academy of Otolaryngology-Head and Neck Surgery.

Table 3
Patients in whom the surgical approach was translabyrinthine

Staging (Samii)	Age	Sex	Laterality	Hearing (AAO-HNS)	House-Brackmann (immediate postoperative)	House-Brackmann (6-12 months)	Complications	MRI
T2	64	F	Left	B	V	IV	Abdominal Hematoma; Exposure keratitis	Total
T2	49	M	Right	B	II	I	-	R
T2	54	M	Left	B	II	I	-	R
T3a	43	F	Left	A	I	I	-	Total
T3a	62	F	Right	A	I	I	CSF Fistula	NTR
T3a	62	M	Left	B	I	I	-	NTR
T3a	54	M	Left	B	I	I	-	NTR
T3a	46	M	Left	D	I	I	-	R
T3a	67	M	Left	D	IV	IV	-	-
T3b	61	F	Left	C	II	I	-	R
T4a	55	F	Left	D	V	II	-	R

F – Female; M – Male; NTR – Near-total resection; R – Residual; T – Total; MRI – Magnetic resonance imaging;
AAO-HNS – American Academy of Otolaryngology-Head and Neck Surgery; CSF – cerebrospinal fluid.

positive correlation ($rs = 0.446$, $p<0.05$) between the grade of hearing loss before surgery and tumor size. All 12 patients who underwent the translabyrinthine approach became totally deaf. Two of the three patients selected for the middle cranial fossa approach who had useful hearing also became totally deaf. Regarding the retrosigmoid approach, only nine of these 22 patients with this approach had useful hearing, and only two of these nine patients had their hearing preserved after the surgery.

Discussion

The most common clinical presentation of VS in our study was progressive unilateral hypoacusis, which occurred in 61 patients (77%). The mean age at diagnosis was approximately 50 years. A greater access to audiological evaluation and imaging exams, combined with suspicion in patients with any criterion (sudden deafness; unilateral or asymmetrical sensorineural hearing loss), has resulted in an increase in the number of diagnosed tumors, especially smaller-size tumors, and diagnosis at more advanced ages, which traditionally did not occur.[8]

Table 4
Patients in whom the surgical approach was retrosigmoid

Staging (Samii)	Age	Sex	Laterality	Hearing (AAO-HNS)	House-Brackmann (immediate postoperative)	House-Brackmann (6-12 months)	Preserved hearing (AAO-HNS)	Complications	MRI
T3a	50	F	Left	B	II	I	D	Lateral sinus thrombosis	R
T3a	65	F	Right	D	IV	V	With no useful hearing to preserve	-	-
T3b	71	F	Right	A	I	I	B	-	R
T3b	34	M	Right	B	IV	II	D	-	-
T3b	44	F	Right	-	III	III	-	-	-
T3b	81	M	Right	D	I	I	With no useful hearing to preserve	-	-
T3b	25	F	Right	B	I	I	D	-	-
T4a	41	F	Right	B	I	I	C	Thrombosis of the transverse and sigmoid sinuses	-
T4a	52	F	Right	D	I	I	With no useful hearing to preserve	-	R
T4a	71	F	Left	D	VI	VI	With no useful hearing to preserve	-	R
T4a	52	M	Right	A	I	I	D	-	R
T4a	45	M	Left	D	I	I	With no useful hearing to preserve	-	R
T4b	75	M	Right	D	IV	IV	D	Hydrocephalus	R
T4b	18	F	Left	D	III	I	With no useful hearing to preserve	-	R
T4b	70	M	Right	D	IV	III	D	-	NRT
T4b	41	F	Right	D	II	I	D	Thrombosis of the lateral and sigmoid sinuses; Ischemia of the PCerebeloso ; Hemiparesis; Dysphagia	R
T4b	49	F	Left	B	V	V	D	-	R
T4b	59	F	Left	B	III	III	D	-	-
T4b	72	F	Left	C	I	I	D	-	-
T4b	83	F	Left	D	III	III	With no useful hearing to preserve	-	-
T4b	59	F	Right	D	IV	-	With no useful hearing to preserve	-	-
T4b	52	F	Left	D	II	-	With no useful hearing to preserve		-

F – Female; M – Male; NTR – Near-total resection; R – Residual; MRI – Magnetic resonance imaging;
AAO-HNS – American Academy of Otolaryngology-Head and Neck Surgery

The grades of hearing were almost evenly distributed in the sample. Grade D (loss of speech discrimination) included a higher number of patients, which is explained by the tendency of worsening of hearing with the natural course of the disease.

Tinnitus was found in 34 patients (41%) with different degrees of intensity. Unilateral tinnitus is the second most common hearing symptom, is present in 12–60% of patients, and is often accompanied by non-specific symptoms compatible with ear fullness.[9,10,11,12]

Although this tumor originates in one of the vestibular nerves, vestibular complaints are not common because the tumor's slow growth allows adaptation to the chronic progressive vestibular deficit through central mechanisms. In this study, approximately half of the patients (48%) did not have symptoms of vertigo or dizziness.

Although sudden hearing loss is not a common form of presentation, it was observed in seven patients (9%). This less frequent form of presentation occurs in 2–7% of patients, especially in smaller tumors limited to the IAC.[9,10,13,14]

The initial approach in 61 patients (51%) was conservative (wait-and-scan). However, during follow-up, approximately half of these patients (23) ended up undergoing surgery or radiosurgery because they became symptomatic, or the tumors' size and growth became too aggressive.

According to the literature, the proportion of tumors that grow during follow-up varies considerably between 30% and 70% over different periods. This variation may be due to the different methods and criteria used during follow-up.[15,16] According to a systematic review that included approximately 6,000 patients from 53 studies from 1984 to 2014, the mean growth of small- and medium-sized tumors was 33%, with a follow-up of 3.3 years.[7] Other studies showed a tumor growth of 50% over five years.[17,18,19]

After the diagnosis, it is not possible to predict how much and for how long the tumor will grow, and there is no apparent relationship with clinical and demographic factors such as age, sex, tumor size, or symptoms at diagnosis. Tumor growth may be continuous, or it may only occur after a period of inactivity. Tumors with cystic characteristics usually grow more and faster.[20]

Close monitoring of VS with serial MRI and audiological evaluation (watchful waiting or wait and scan) is considered an appropriate strategy for sporadic and asymptomatic VS (degree of recommendation III, level of evidence C).[21] The mean growth of sporadic VSs is 1.1 mm/year in diameter.[22] Studies have shown that with a conservative approach, a growth greater than 2.5 mm/year is significantly associated with a higher rate of hearing loss, compared to lower growth rates (75% and 32%, respectively) with a follow-up period of 26 to 52 months.[23] In this study, age, clinical presentation, and growth of 2.5 mm/year or greater were therapeutic decision factors, especially in borderline tumors.

SRS is considered a valid alternative to watchful waiting in patients with small tumors and asymptomatic cases (Koos I and II, Samii T1-T3a) because it halts tumor growth and preserves the function of the nerve in the long term. However, there is a small risk of deterioration that will affect the patient's quality of life (degree of recommendation II, level of evidence B)

SRS is a non-invasive treatment modality that uses the delivery, in a single session or several sessions, of a fraction of high-dose radiation with extreme precision to well-localized targets with well-defined edges, usually intracranially. A fundamental feature of this technique that distinguishes it from conventional external radiotherapy (ERT) is obtaining a high-dose gradient beyond the edges of the lesion—there is maximum sparing of radiation to the normal tissues adjacent to the lesions as a result of using multiple targeted beams.[24]

Most studies with SRS in the last decade have reported tumor control rates from 92% to 98% with a follow-up period between 3 and 10 years.[25-30] There is a tendency for progressive hearing loss after SRS: a study with 44 patients with VS who had useful hearing and were treated between 1997 and 2002, with a follow-up of 9.3 years, had useful hearing rates of 80, 55, 48, 38, and 23% at 1, 3, 5, 7, and 10 years, respectively.[31]

In this study, all patients selected for SRS had useful hearing. In one patient, there was an attempt to control the growth of the tumor (Koos III – Samii T3b) with SRS, but they ended up undergoing surgery. Another patient with a tumor (Koos II – Samii T3a) and vestibular symptoms preferred to be primarily treated with SRS. However, they also had to be operated because of tumor growth. Finally, one patient with a tumor (Koos II – Samii T2) who underwent SRS remains under follow-up and has not needed surgery yet. The few patients selected for SRS were a limitation of this study.

Most patients with medium-size tumors exhibit auditory or vestibular symptoms. Facial paresis is rare, even in these patients, and if present, the differential diagnosis should be facial schwannoma. In this study, facial paresis was only observed in patients with large tumors who had to undergo surgery. Owing to the symptoms and size of the tumor, the approach in patients with medium-size tumors should be therapeutic, either by surgery or by SRS (level of evidence C)[32]

The risks are lower with SRS; however, only surgery has curative intent by totally removing the tumor. Subtotal resection to preserve function may be an option with subsequent SRS (degree of recommendation III)[33]

The choice of surgical approach depends on the patient's level of hearing and preference, characteristics of the tumor, and the surgeon's experience. The experience of the surgical team is an important factor that affects the final outcome. Therefore, VS should only be treated at specialized centers with a high volume of surgical procedures (degree of recommendation IV)[34,35,36]

Surgery may be considered even for small tumors if there is cystic degeneration or if the main objective of the treatment is cure (degree of recommendation III, level of evidence C).[37,38,39] The objective of the surgery should be total or near-total (NTR) resection because the size of the residual disease correlates with the likelihood of recurrence (degree of recommendation III, level of evidence B).[40] For large tumors (Koos IV and Samii T4a and T4b), surgery is the treatment of choice to remove the symptomatic or potentially fatal lesion causing compression of the brainstem.[41] The suboccipital or retrosigmoid (retromastoid) approach is preferred by neurosurgeons and is especially indicated for tumors located in the CPA or those causing a significant degree of compression of the brainstem. This approach allows the removal of tumors of various sizes and, in theory, offers the possibility of preserving hearing. However, the results of this study and other studies show that hearing preservation is not achieved in most cases. This approach offers convenient visualization of the brainstem, cranial nerves, and neighboring vascular structures but requires some retraction of the cerebellum. Furthermore, access to the fundus of the IAC is limited, from where a variable volume of residual tumor is often not removed.[42,43]

The translabyrinthine approach is performed by otorhinolaryngologists with otoneurosurgical experience working with a team of neurosurgeons. It allows the removal of tumors of all sizes. This approach allows access to the IAC and visualization of the entire course of the facial nerve, including the portion located in the Fallopian canal after labyrinthectomy has been performed. This approach has the advantage of offering convenient access to the tumor without the need to retract the occipital or temporal lobes.[44,45] Traditionally, this approach is said to result in a complete loss of the inner ear's function and is thus not appropriate for patients with residual useful hearing who require a route that preserves hearing. However, even the routes that theoretically preserve hearing often end up not doing that. Thus, the translabyrinthine approach is a route that may be offered to patients with useful hearing but with little chance of preserving it by any possible approach (patients with large tumors, for example).

The middle cranial fossa approach may be considered in patients with small tumors that require an approach that preserves hearing. It offers access to the IAC superiorly and craniotomy above the zygomatic process. Extradural dissection up to the arched eminence and the superior edge of the temporalis rock are necessary. Patient selection is the key for this approach because tumors extending to the fundus of the IAC or below the falciform crest of the IAC are more difficult to address using this approach, especially relative to those that do not reach the fundus of the IAC, which are the best indication for the use of this approach.[44,45,46,47]

The auditory outcomes of the three approaches demonstrate that patients who undergo surgery through the translabyrinthine approach become inevitably deaf, but the cophosis rates of the middle cranial fossa and retrosigmoid approaches are two-thirds and seven-eighths, respectively. These rates vary according to the study but are invariably high, even in international centers of excellence.

The degree of tumor resection, categorized into total, near-total (if less than 5x2x2 mm), and subtotal (5x2x2 mm or more), is related to the probability of relapse.[48,49,50,51] A study with 116 patients showed recurrence rates of 3.8%, 9.4%, and 27.6% in VS treated with total, near-total, and subtotal resection, respectively.[52] In this study, the translabyrinthine approach yielded the best results in tumor excision and was thus deemed the most controlled and precise approach for tumor resection.

As vs typically exhibits slow growth, several patients can compensate the peripheral vestibular deficit through central mechanisms. Therefore, few patients report dizziness or balance problems as the tumor grows. The degree of caloric asymmetry—documented vestibular deficit present preoperatively—is an important parameter because it allows the prediction of the intensity of vertigo immediately after surgical resection of the tumor. The intensity of the vertigo increases with decreasing deficit caused by the tumor before surgery. Regardless of the result of this evaluation, most patients benefit from vestibular rehabilitation.[53] It was demonstrated in a small study that cervical vestibular evoked myogenic potential (cVEMP) tests, together with caloric tests, help predict whether there is tumor involvement of both vestibular nerves or only the inferior nerve.[54] This is controversial because some authors have reported that there is no correlation between the cervical and ocular vestibular evoked myogenic potential (cVEMP and oVEMP) tests, even in combination with caloric tests, regarding the nerve origin of the tumor.[55] Moreover, several studies have shown high degrees of sensitivity and specificity for the nerve origin of the tumor when these tests are associated with the video head impulse test (vHIT).[56, 57] When a patient with VS presents with unilateral vestibular deficit in the caloric tests, the remaining assessment by videonystagmography (VNG) can provide information on the state of compensation. For example, abnormal eye movements in the form of spontaneous and positional nystagmus and/or in the head impulse test indicate that the vestibular deficit is not physiologically compensated.

MRI is the diagnostic method of choice in patients with suspected VS. T1 weighted imaging with intravenous administration of gadolinium is the gold standard.[58,59] The brainstem auditory evoked potentials should also be evaluated when trying to preserve hearing. Greater changes in the evoked potentials indicate a lower probability of preserved hearing, even by using a potentially hearing preserving approach (degree of recommendation III, level of evidence B).[60, 61,62]

This study has some limitations: its retrospective nature; conducted in a single center; a lack of evaluation of the treatment results with respect to the quality of life; a few patients underwent SRS.

Conclusion

This is one of the largest studies on the treatment of VS at the national level.

The results of this study confirm what has been reported by other authors and advocated in international consensus documents—the primary objective of the treatment of VS should not be complete excision of the tumor at all costs but avoidance of complications and preservation of the facial nerve and auditory functions for as long as possible. Therefore, the physicians who help patients in deciding on the best treatment for their disease should use the best evidence and treat the patients and provide counseling in which all options, including watchful wait, surgery, radiosurgery, or combinations of the above (sequentially or planned according to the progression of the primary or previously treated disease) are explained and made available. To this end, a collaboration among otorhinolaryngologists, neurosurgeons, and physicians with experience in radiosurgery is necessary.

Conflicts of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

Data Confidentiality

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

Protection of humans and animals

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the 2013 Helsinki Declaration of the World Medical Association.

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Availability of scientific data

There are no datasets available, publicly related to this work.

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