

## Enlarged Translabyrinthine Approach With Transapical Extension in the Management of Giant Vestibular Schwannomas: Personal Experience and Review of Literature

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**Objective:** To describe and analyze the main outcomes achieved in a series of patients with sporadic vestibular schwannoma (VS) larger than 40 mm in extrameatal diameter, defined as giant VS, submitted to microsurgery by the enlarged translabyrinthine approach with transapical extension.

**Study Design:** Retrospective chart review.

**Setting:** Tertiary referral center.

**Patients:** A retrospective chart review was conducted on 2,133 patients who underwent surgery for VS from April 1987 to July 2009. One hundred ten cases of giant VS were elected for analysis.

**Main Outcome Measures:** Extent of removal, residual or recurrent disease, facial nerve integrity during surgery, long-term facial nerve function, and postoperative complications.

**Results:** Total removal was accomplished in 91.8% of cases. In 5 patients (4.5%), total removal was accomplished in 2 stages. Near-total removal was performed in 7 patients (6.3%). The fa-

cial nerve was anatomically preserved in 76.4% of cases. Intraoperative facial nerve reconstruction was performed in 8 cases. Facial nerve function after 1 year of follow-up was House-Brackmann grades I to III in 75% of cases. There were no deaths in this series. Neurovascular life-threatening complications occurred in 2 patients. Cerebrospinal fluid leak was present in 1.8% of cases.

**Conclusion:** Results indicate the enlarged translabyrinthine approach with transapical extension as an elective approach for removal of giant VS. The method permits achievement of a high rate of total removal with low incidence of complications.

**Key Words:** Acoustic neuroma—Enlarged translabyrinthine approach—Giant vestibular schwannoma—Surgery—Transapical extension.

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The management of giant vestibular schwannomas (VSs) has been a challenging task for cranium base surgeons. These huge tumors are frequently related to compressive symptoms over cranial nerves and brainstem, as well as obstruction to the cerebrospinal fluid (CSF) pathway. Microsurgical resection is the treatment of choice for these patients.

House (1) detailed described the translabyrinthine approach (TLA) in 1964. In the following years, some authors pointed out that this method was not suitable for accessing oversized VS because of its constricted exposure (2). Some modifications were further added to the

original approach described by House to obtain a wider surgical view and a better control over the tumor and surrounding structures. The enlarged TLA (ETLA) and the transapical (TA) extension are technical refinements introduced for the treatment of large and giant VS (3–5).

Only few studies have analyzed exclusively outcomes of VS equal or larger than 40 mm, defined as giant tumors. Giant, large, and even medium VS are usually described together, and results of the first group cannot be properly analyzed. When accessing a VS, surgeons expect the approach to be safe, effective, and provide a wide surgical view with a reduced incidence of complications. When managing a giant tumor, these requirements acquire special meaning.

The aim of this study is to analyze a consecutive series of 110 giant VS with special regard to the modifications of the original TLA, extent of tumor removal, residual or recurrent disease, postoperative facial nerve (FN) function, and complications.

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## MATERIALS AND METHODS

We retrospectively reviewed all cases of VS submitted to microsurgical resection at the Gruppo Otorologico in Piacenza and Rome, Italy, between April 1987 and July 2009. Patients were included if they had a VS larger than 40 mm, according to largest extrameatal diameter seen in latest preoperative magnetic resonance imaging (MRI) scan. Patients with neurofibromatosis Type 2 were excluded. Results are described according to the Consensus Meeting on Systems for Reporting Results in Vestibular Schwannoma (6). All patients in this series underwent surgery by the senior author (M. S.) or under his direct supervision. Tumors removed incompletely were grouped as having near-total removal (NTR), when 5% or less of the tumor was unresected, or subtotal removal, when more than 5% was left. Residual or recurrent disease was monitored by gadolinium-enhanced MRI scanning at 2, 12, 36, and 60 months.

The FN function was assessed according to the House-Brackmann (HB) Grading Scale (7). Postoperative outcomes were scheduled after at least 1-year follow-up. All cases were directly evaluated by the senior author or his associates. Complications, such as neurologic dysfunctions, cerebellar symptoms, and cranial nerve deficits, are described only if present exclusively after surgery.

Statistical significance was calculated with Fisher's exact test when required.

## SURGICAL TECHNIQUE

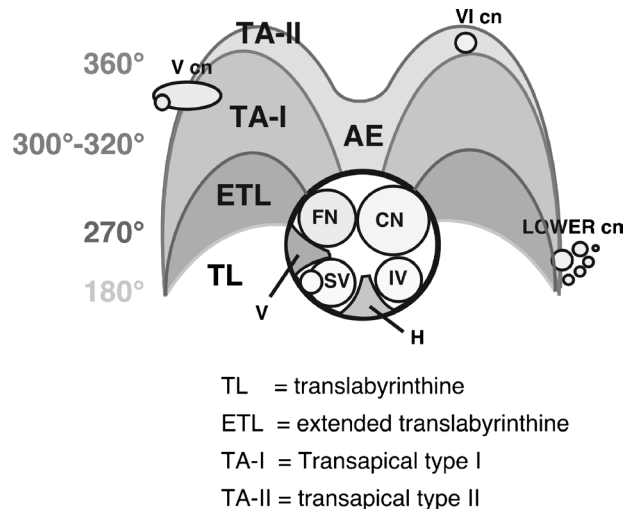
The ETLA includes a broad mastoidectomy in which bone anterior and posterior to the sigmoid sinus and around the middle fossa is completely removed (8). When necessary, a high placed jugular bulb and surrounding periosteum are pressed inferiorly with oxidized cellulose (Surgicel, Ethicon) and bone wax (9). The internal auditory canal (IAC) is exposed approximately 270 degrees of its circumference.

The principle of the TA extension is an increase of bone removal around the IAC from 300 to 360 degrees (Fig. 1). In large and giant VS with anterior extension, the circumference of bone removal around the IAC can reach up to 320 degrees (TA extension Type I). Cases in which the IAC is drilled up to 360 degrees (TA extension Type II) are reserved for patients with meningiomas of the posterior surface of the petrous bone (8).

The superior ampullary nerve has been our main anatomic guide to identify the FN. It is identified at the level of superior semicircular canal's ampulla and detached from its bony canal. The FN is separated from the superior ampullary nerve by the widest part of the vertical crest, lowering the possibility of trauma (10).

For continuous intraoperative monitoring of the FN, we use the Nerve Integrity Monitor (NIM Response 2.0; Medtronic Xomed). Two pairs of subdermal needle electrodes are placed 5 mm apart in the orbicularis oculi and orbicularis oris muscles. In our protocol, we investigate the pattern of burst and train responses during the last step of tumor removal combined with intraoperative stimulation of the nerve at minimal threshold stimulation. We are not used to perform intraoperative mapping of the FN.

Tumor debulking is gradually accomplished. When a large piece of tumor is coagulated, its capsule is detached



**FIG. 1.** Diagram illustrating the extension of traditional TLA to ETLA with TA extension by increasing the drilling around the IAC. AE indicates anterior extension; H, horizontal crest; IV, inferior vestibular nerve; SV, superior vestibular nerve; V, vertical crest.

from surrounding structures and then excised. Finally, only a small layer of tumor and capsule is attached to neurovascular structures that lie anteriorly. The cleavage plane is found, and removal is accomplished (8). The incus is disarticulated and removed from the tympanic cavity using a right angle pick. Subsequently, attic, middle ear, and Eustachian tube are plugged with dry periosteum. A careful inspection of the mastoid with a small hook will ensure that all open cells can be closed with bone wax.

Obliteration of the operative site is accomplished by packing it completely with long strips of abdominal fat in such a manner that part of the strips protrudes into the cerebellopontine angle (CPA). No attempt is made to suture the dural edges.

The transotic approach is an anterior prolongation of the ETLA, at the expenses of the cochlea. The middle ear is removed, and the external auditory canal is closed in 2-layered blind sac. The FN is skeletonized and maintained in place (8,11,12). We use this approach exclusively in cases of high jugular bulb and coincident dominant sigmoid sinus. The transcochlear approach combines the removal of external auditory canal and middle ear with posterior rerouting of the FN and removal of the cochlea (12). This approach is indicated in cases of residual tumor with preoperative FN deficit.

## RESULTS

### Patients

Between April 1987 and July 2009, 2,133 patients underwent surgery for VS at the Gruppo Otorologico (Table 1). The study group included 110 patients (59 women and 51 men) who presented an extrameatal tumor sized at least 41 mm. These patients represent 5.1% of all cases. The mean age was 42.5 years (range, 18–80 yr). The right side



remaining patients, 101 presented Grade I, and 5 presented Grade II before surgery.

The FN was anatomically preserved in 81 (76.4%) of the 106 cases. Considering both groups (FN preserved or interrupted), 88 patients had completed the minimum required 1-year follow-up at the time this study was being performed and were available for analysis of long-term outcomes. Results were compared with those obtained in patients with VS equal or smaller than 40 mm (Table 2). In patients with VS equal or smaller than 10 mm, the incidence of Grade I was superior to 70%, and Grade VI was observed in less than 0.5% of cases. Considering the group with giant VS, 8% of the patients reached Grade I after 1 year, whereas 25% of patients presented Grade VI. From a statistical perspective, patients with giant VS presented worst outcomes in comparison to patients with smaller tumors ( $\chi^2$ ,  $p < 0.001$ ).

The FN was interrupted during surgery in 25 patients (23.6%). In 8 cases, an intraoperative reconstruction was performed (end-to-end anastomosis in 1 case; sural nerve graft in 7 cases). Twelve patients were submitted to delayed hypoglossofacial anastomosis. The remaining patients refused any additional treatment.

### Postoperative Complications

There were no deaths related to the surgery.

One patient presented a CPA hematoma in the early postoperative period that required immediate evacuation. In another patient, an asymptomatic subdural collection was seen in a postoperative MRI scan. Hemorrhagic infiltration of the brainstem was diagnosed in a third patient with a transitory dysfunction of the VIth, IXth, and Xth cranial nerves. In the last 2 cases, blood collections were reabsorbed in 3 to 6 weeks.

One patient presented a persistent rhinoliquorrea that required closure of the middle ear and the Eustachian tube with perichondrium. A second patient presented CSF leak through the retroauricular incision, requiring reopening of the wound and reinforcement of the muscular suture.

Six patients presented postoperative VIth cochlear nerve deficit that was recovered completely in 5 of them. Lower cranial nerve dysfunctions were seen in 3 cases, all of them transitory.

One patient developed postoperative cerebral edema, with signs of increased intracranial pressure that required external ventricular shunt.

**TABLE 2.** Long-term (1 year) facial nerve function in 1,808 cases (%)

Size	Intrameatal <11 mm	11–20 mm	21–30 mm	31–40 mm	>40 mm	
Cases	274	406	506	389	145	88
HB grade						
I	76.5	73.1	48.2	22.6	16.5	7.9
II	8.9	9.6	13.6	9.3	8.2	7.9
III	13.8	15.8	29.4	47.3	35.9	46.6
IV	0.4	1.0	3.8	8.7	11.0	9.1
V	—	—	1.1	2.3	6.9	3.4
VI	0.4	0.5	3.9	9.8	21.5	25.0

**TABLE 3.** Postoperative complications (110 cases)

Complication	Cases	%	Comments
Mortality	0	0	
Cerebrospinal fluid leak	2	1.8	Revision surgery in both cases
Subcutaneous cerebrospinal fluid collection	3	2.7	Conservative management
Meningitis	0	0	
Vascular			
Cerebellopontine angle hematoma	1	0.9	Revision surgery
Subdural hematoma	1	0.9	Spontaneous involution
Brainstem hematoma	1	0.9	Spontaneous involution
Cranial nerve dysfunction			
VIth	1	0.9	Permanent
VIth	9	8.2	Permanent in 1 patient
IXth or Xth	5	4.5	Spontaneous involution in all cases
Cerebellar symptoms	5	4.5	Spontaneous involution in all cases
Cerebral edema	1	0.9	Required ventriculoperitoneal shunt
Abdominal hematoma	2	1.8	Required revision surgery

A general overview of postoperative complications is seen in Table 3.

### DISCUSSION

Most cranium base surgeons deal with giant VS in their practice. In referral centers, this is particularly true. Retrosigmoid approach (RSA) and TLA are established methods for removal of these tumors. Authors who adopt the TLA emphasize the advantages of minimal cerebellar retraction and identification of the FN before its involvement with the tumor. Some authors draw attention to the necessity of extending bone removal until the posterior fossa dura, which allows retraction of the sigmoid sinus (13,14). This refinement can be considered a decisive step forward regarding the effectiveness of the ETLA.

Anderson et al. (15) proposed a combined method for removal of VS larger than 40 mm, in which RSA and TLA are used in 1 single stage. The authors indicate the limitations of the singular approaches as the difficult management of a high jugular bulb. In our experience, the ETLA overcomes these limitations without additional morbidity. Patni and Kartush (16) proposed a staged strategy for large VS, in which the tumor is partially removed by RSA, and a second surgery is performed 4 to 6 months later by the TLA. Our opinion is that combined strategies expose patients to risks inherent to each isolated approach.

Sluyter et al. (17) proposed the association of the TLA with transtentorial extension. Of the 120 patients, 12 presented postoperative aphasia, and 4 required permanent medication for seizures. We do not consider the transtentorial extension a suitable option for removal of large or giant VS because of risks of permanent neurologic deficits secondary to retraction of the temporal lobe.



**TABLE 4.** Review of literature: overall results

Main author	Year	Cases	Size (mm)	Giant VS	Total removal	FN integrity	Approach
Tos	1989	400	All sizes	149	98.7%	89.2% <sup>a</sup>	TLA
Briggs	1994	167	≥40	167	95.2%	91.6%	TLA
Lanman	1999	190	≥30	68	96.3%	93.7%	TLA
Sluyter	2001	120	≥20	48	91.7%	80.8%	TLA-TT
Pareschi	2002	71	>25	19	100%	78.9% <sup>a</sup>	ETLA
Mamikoglu	2002	81	≥30	29	95.1%	85.2%	TLA
Sanna	2004	175	≥30	45	85.1%	84.8%	ETLA + TA/TC/TO
Roland Jr.	2004	56	≥30 <sup>b</sup>	<sup>d</sup>	73.2%	92.8%	TLA (84% of cases)
Patni	2005	34	>30	<sup>d</sup>	98% <sup>c</sup>	100%	RSA + TLA (staged)
Anderson	2005	71	≥30	29	95.7%	<sup>d</sup>	RSA + TLA (combined)
Mehrotra	2008	62	>40	62	90.3%	87.1%	RSA
Godefroy	2009	50	>25	<sup>d</sup>	26%	96%	TLA
Samii	2009	50	>40	50	100%	92%	RSA
This study	2010	110	>40	110	91.8%	76.4%	ETLA + TA/TC/TO

<sup>a</sup>Result achieved in VS equal or larger than 40 mm.

<sup>b</sup>The authors consider the intracanalicular portion of tumor.

<sup>c</sup>The authors consider total and near total removals.

<sup>d</sup>Data not available or unclear.

ETLA indicates enlarged translabyrinthine approach; FN, facial nerve; RSA, retrosigmoid approach; TLA, translabyrinthine approach; TC transcochlear approach; TO, transotic approach; TT, transtentorial; VS, vestibular schwannoma.

Of the 13 series analyzed (13–25), in 3 articles, the authors described exclusively the outcomes of VS equal or larger than 40 mm (Table 4). Samii et al. (23) and Mehrotra et al. (24) used the RSA; Mehrotra et al. (24) presented the higher incidence of complications in all series analyzed (Table 5), opposed to good results described by Samii et al. (23). This may indicate the importance of personal experience in achievement of satisfactory outcomes. Briggs et al. (13) used the TLA as the preferred method; outcomes described reveal a high incidence of total removal, FN integrity, and a low incidence of complications.

### Hearing Preservation

A common criticism of the TLA is the sacrifice of any residual hearing function. According to Samii et al. (23), hearing preservation is possible even in patients with giant VS. In their series of 50 giant VS treated by the RSA, 9 patients had preoperative serviceable hearing. In only 3 cases (6% of the entire series), it was possible to maintain hearing at functional levels, according to the

New Hannover Classification. In contrast, other authors affirm that hearing preservation is extremely unlikely in patients with VS larger than 3 cm (18,19). Facing the published results and our personal experience, we sustain the policy of using the RSA only in patients with VS smaller than 15 mm and hearing levels class A or B, according to Sanna's classification (6).

### Surgical Technique

Extent of exposure has always been a major concern regarding lateral cranium base surgery. With the ETLA, the operating field is enlarged at the expenses of bone removal. Most of the retraction is performed over the sigmoid sinus. A high jugular bulb can be securely decompressed and pushed inferiorly. The TA extension highly raised the effectiveness of the ETLA, providing an excellent control of anterior extension of the tumor. When properly performed, the ETLA with TA extension allows removal of tumors of any size.

Transcochlear and transotic approaches provide better anterior extension compared with the ETLA alone. After

**TABLE 5.** Review of literature: postoperative complications (%)

Main author	Deaths	Vth	Vlth	LCN	Atassia	CPA hematoma	Meningitis	CSF leak <sup>a</sup>
Roland	—	—	7.1	—	—	3.6	—	3.6
Lanman	—	4.7	3.7	—	0.5	1.6	3.7	1.1
Mehrotra	11.3	—	—	19.3	—	1.6	6.5	33.8 <sup>b</sup>
Samii	—	—	—	6.0	—	—	—	2.0
Sanna	0.6	0.6	6.8	0.6	—	1.7	—	2.3
Briggs	—	—	—	1.8	7.8	1.8	7.2	3.6
Pareschi	—	—	—	1.4	—	—	4.2	2.8
Godefroy	—	—	—	—	—	—	5.9	1.9
Mamikoglu	—	—	—	6.2	—	1.2	3.7	4.9
This study	—	0.9	5.4	2.7	4.5	0.9	—	1.8

<sup>a</sup>Requiring revisional surgery.

<sup>b</sup>The authors do not specify how many cases required revisional surgery.

CPA indicates cerebellopontine angle; CSF, cerebrospinal fluid; LCN, lower cranial nerve.

the introduction of the TA extension, these methods have rarely been performed in our center.

### FN Outcomes

The ETLA offers the advantage of allowing direct access to FN before its involvement with the tumor, at a constant anatomic location (19). When necessary, the approach provides an excellent view to perform its immediate reconstruction. In these cases, sural nerve interposition is our procedure of choice. The end-to-end anastomosis is not indicated because traction on nerve fibers impedes adequate functional recovery. In the RSA, these types of reconstruction are not possible. We limited the use of hypoglossofacial anastomosis to cases in which the FN is not identified at brainstem.

We have observed that a silent electromyography pattern during the last step of the tumor removal procedure is more likely to be associated with poor long- and short-term outcome than one in which considerable spontaneous or mechanically elicited activity is found.

### Completeness of Removal

Incomplete removal may result either from unpredictable intraoperative circumstances or from a planned NTR. Our philosophy has always been to perform total removal. Nevertheless, planned partial removals have been considered in elderly patients, supported by the general belief that VS are benign slow-growing tumors (26). In cystic VS, there is a trend to leave the capsule unresected, avoiding surgical trauma to underlying brainstem or vital vessels (27).

Patients submitted to a 2-staged strategy form a group in which the tumor is extremely adherent to the brainstem. Tumor debulking is related to excessive bleeding and vital sign changes, as bradycardia or raising arterial pressure. In these cases, staged strategy could be considered a life-saving intraoperative decision. Our policy is to perform a second stage 4 to 6 months later, by the same approach.

### Postoperative Complications

In the last years, rates of mortality in most studies have been drastically reduced (Table 5). Deaths usually result from neurovascular disorders, such as arterial occlusion, perioperative bleeding, or trauma to brainstem. Patients with larger tumors are more exposed to risk. Giant VS are extremely adherent to underlying structures, and the anatomy is always distorted.

In this series, neurovascular life-threatening complications occurred in 2 cases. One patient developed a cerebral edema followed by intracranial hypertension. A second patient had a CPA hematoma requiring immediate intervention. The ETLA offers a unique advantage in these cases. Once the fat strips are removed, the CPA is promptly accessed, and the hematoma can be evacuated. Another remarkable advantage of the ETLA is the minimal cerebellar retraction, which reflects our low incidence of postoperative ataxia.

Most cases of CSF leak have been successfully managed with supportive measures. Cases requiring reintervention are described as 1% to 5% in most studies (Table 5). In the present series, 2 patients (1.8%) needed surgical management. Our recent studies (28) revealed an overall incidence of 0.55% of CSF leak requiring revisional surgery in more than 1,800 cases of VS operated by the ETLA. Our incidence greatly fell after we adopted the method of closing the dural defect with abdominal fat strips instead of sutures of the dura.

Lower cranial nerve deficits are usually temporary but related to a long hospital stay because of problems concerning swallowing disorders and aspiration pneumonia. Removal of bone between IAC and jugular bulb permits an excellent control over the region of these nerves. Therefore, we do not perform intraoperative monitoring of the lower cranial nerves in cases of VS removed through ETLA.

From a clinical perspective, oversized VS are associated to a higher incidence of cranial nerve deficits and cerebellar ataxia. Life-threatening complications, such as CPA hematoma or trauma to the brainstem, have an

**TABLE 6.** Postoperative complications according to preoperative vestibular schwannoma size in 2,133 patients of 1,941 removed through enlarged translabyrinthine approach (%)

Size	Intrameatal	1–10 mm	11–20 mm	21–30 mm	31–40 mm	>40 mm
Cases	296	484	603	465	175	110
Deaths	—	—	—	0.2	0.6	0
CSF	0.7	1.0	0.2	0.9	1.7	1.8
Meningitis	—	—	0.3	0.2	—	—
CPA hematoma	—	—	—	0.2	1.7	0.9
Subdural hematoma	—	0.4	—	0.4	0.6	0.9
Brainstem hematoma	—	—	—	—	1.1	0.9
Vth CN dysfunction	—	—	—	0.2	0.6	0.9
VIth CN dysfunction	—	—	0.2	1.3	4.6	5.4
LCN dysfunction	—	0.2	0.3	1.0	1.1	2.7
Cerebellar ataxia	—	0.2	0.3	1.0	2.3	4.5
Cerebral edema	—	0.2	0.3	0.4	—	0.9
Postural instability	0.3	2.0	0.7	0.6	1.7	5.4
Abdominal hematoma	0.3	0.2	0.2	—	—	1.8

CN indicates cochlear nerve; CPA, cerebellopontine angle; CSF, cerebrospinal fluid; LCN, lower cranial nerves.

incidence of approximately 1%, even in patients with giant tumors (Table 6).

### CONCLUSION

The advantages of the ETLA with TA extension, in comparison to the RSA, include the following: 1) easy identification of the FN in an invariable position in the fundus of the IAC; 2) FN can be dissected from its emergency in the brainstem up to the fundus, allowing intraoperative reconstruction in cases of anatomic interruption; 3) management of the high jugular bulb with the possibility of lowering it and achieving optimal results over lower cranial nerves; 4) minimal or no cerebellar retraction; and 5) in case of CPA hematoma in the postoperative period, the approach permits rapid control of bleeding and prompt evacuation.

The outcomes achieved in this series indicate that ETLA with TA extension is the elective approach for removal of giant VS. In our experience, the method permits the achievement of a high rate of total removal of tumors with extremely low incidence of major complications.

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