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Hearing Preservation Surgery for Vestibular Schwannomas

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Abstract Hearing preservation microsurgery for vestibular schwannomas (VSs) offers favorable hearing preservation rates for small tumors in patients with serviceable hearing. The natural history of VSs is slow growth of 1–2 mm per year; however, some tumors do not grow. Hearing loss can occur gradually or suddenly independent of tumor growth. Management options include observation, microsurgical removal, and radiosurgery. Hearing preservation rates are approximately 72 % with middle cranial fossa microsurgery for small tumors (<10 mm) in patients with good hearing (>70 % word discrimination). Hearing preservation is approximately 50 % with larger tumors not yet touching the brainstem. Longitudinal studies show a 5-year hearing preservation rate for observation of around 50 %. Radiosurgery should not be used as a hearing preservation modality as the 10-year serviceable hearing rates are only around 25 %. Also, observation and radiosurgery do not eradicate the tumor. Normal or near-normal facial nerve function rates are in the 90 % range for all three options. Thus, we favor microsurgical removal for small tumors in young healthy patients.

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Introduction

Vestibular schwannomas (VSs; also known as acoustic neuromas) are benign neoplasms of the eighth cranial nerve (vestibulocochlear nerve). The vast majority of VSs originates laterally in the internal auditory canal (IAC) near Scarpa's ganglia [1]. With continued growth, the tumor extends medially into the cerebellopontine angle (CPA) of the posterior fossa, which can ultimately lead to compression of the brainstem and cerebellum. Given the close proximity to the auditory portion of the eighth nerve, tinnitus and hearing loss are the most common presenting symptoms. Historically, VSs were rarely diagnosed until they were fairly large, and the patient had lost most of their hearing. With the advent of high resolution contrast-enhanced magnetic resonance imaging (MRI), small tumors are often discovered when patients still have serviceable hearing. These patients are potential candidates for treatment of their tumor with the potential for preservation of their hearing. Middle fossa craniotomy and retrosigmoid/suboccipital craniotomy are the two surgical approaches for removal of VSs that allow for possible hearing preservation. This chapter reviews the preoperative evaluation, indications, and surgical approaches for hearing preservation surgery for VSs.

Preoperative Evaluation

MRI

Early identification of small VSs is more common with the refinement, increased resolution, and widespread utilization

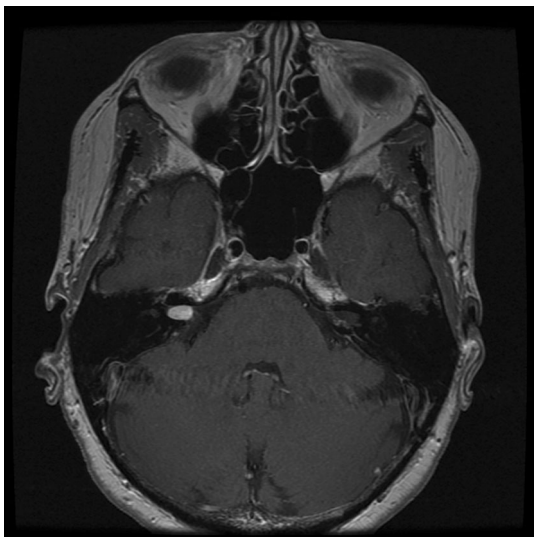

Fig. 1 Axial MRI with contrast showing right internal auditory canal mass (10 mm). There is no extension to the cerebellopontine angle

of MRI. The typical appearance of small VSs is a mass in the IAC that demonstrates contrast enhancement on T1 imaging and no signal on T2 images (Fig. 1). MRI imaging is the gold standard study for detection of a retrocochlear lesion as it is highly sensitive. Auditory brainstem response (ABR) testing often shows abnormalities (absolute wave V latency of >4.5 ms or interaural interpeak latencies for wave I to V of greater than 2 ms) but is not as sensitive as MRI (71 vs. 100 %) [2]. Screening for retrocochlear lesions is controversial, but it is generally agreed upon that those patients under the age of 65 with sudden asymmetrical hearing loss, unilateral tinnitus, unilateral sensorineural hearing loss of >15 dB in 2 consecutive frequencies, or acute vestibulopathy should undergo MRI imaging [3, 4]. Incidental discovery of small VSs in patients without audiologic symptoms also occurs with the increased use of brain MRI for unrelated reasons suggesting that the prevalence of VS maybe has high as 1 in 5,000 persons [5].

Audiometry

All patients receive complete audiometric evaluation including air and bone pure-tone thresholds and speech discrimination scores (SDS). Although there are no strict audiometric criteria to define serviceable hearing, the American Academy of Otolaryngology Head and Neck Surgery Committee (AAO–HNS) class A and class B hearing are typically used for determining candidacy for hearing preservation. AAO–HNS hearing class A is defined as the average of the 4 frequency (0.5, 1, 2, and 3 kHz) pure-tone average (PTA) of better than 30 dB and a SDS of better than 70 %, while AAO–HNS Class B is defined as a PTA of better than 50 dB and a SDS of better than 50 % [6]. However, the preservation of word recognition (SDS >50 %) and hence serviceable hearing, regardless of PTA, is a more relevant measure of hearing preservation [7].

Auditory Brainstem Response (ABR)

All patients should have preoperative ABR to confirm that the ABR is of sufficient quality for intraoperative monitoring.

Medical Evaluation

A preoperative medical evaluation by the surgical team should also be performed prior to surgery. Involvement of medical specialists may also be necessary to optimize surgical risk.

Patient Selection

Discovery of a small benign VS in a healthy patient with serviceable hearing leads to a management dilemma. Therapeutic options for patients with small VSs include observation with serial MRI, surgical excision, and stereotactic radiation therapy. Management decisions should be individualized based on patient age and medical condition; the tumor size, location, and growth; the hearing status in both ears; patient preference; and surgeon comfort. Figure 2 presents our typical management algorithm for VSs. For young patients with small tumors and serviceable hearing, we generally favor microsurgery with an attempt to preserve hearing (Fig. 2). Since the goal of radiation therapy is to prevent further tumor growth, we typically reserve stereotactic radiation therapy for older patients with documented tumor growth on serial MRIs.

There are no strict criteria to select patients for hearing preservation surgery. In general, patients considered for hearing preservation surgery should have a PTA of $\leq$50 dB and a SDS $\geq$50 %. Tumor location can impact the chances for hearing conservation. Hearing preservation is more likely in tumors that only involve the medial aspect of the IAC as compared with those that extend into the lateral IAC at the fundus [8].

Tumor size is likely to be the biggest predictor of hearing preservation. The odds of saving hearing are low (0–25 %) once the tumor exceeds 2 cm or touches the brainstem. Attempts to preserve hearing in patients with large tumors often require leaving residual tumor or compromised facial nerve outcomes [9, 10]. Thus, preservation of facial nerve function, rather than hearing, should dictate the surgical approach in patients with these tumors. The translabyrinthine approach allows for lateral identification of the facial nerve at the labyrinthine segment in addition to maximal tumor exposure for removal.

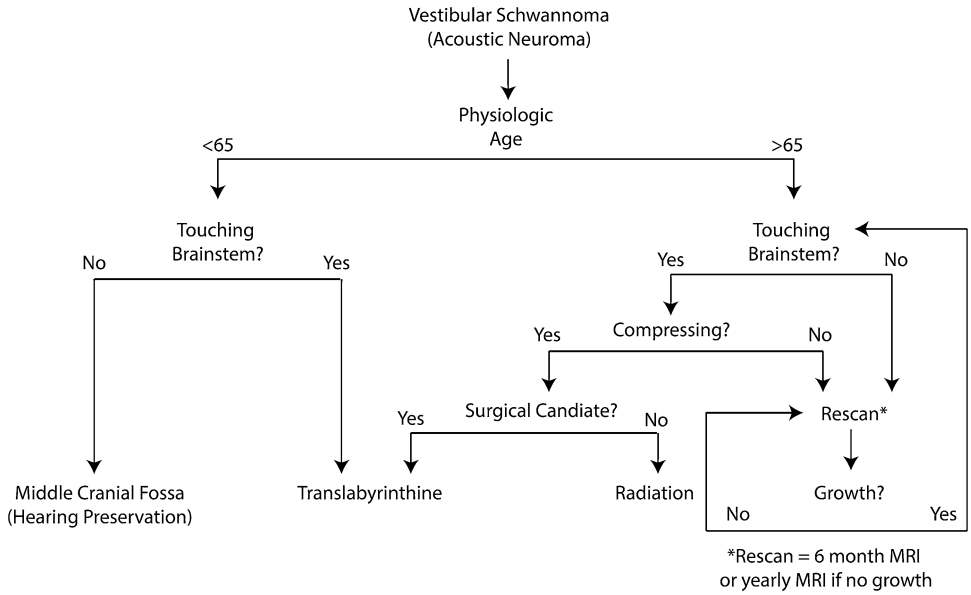

Fig. 2 Management algorithm for vestibular schwannomas (VSs). Patients younger than 65 years old with small VSs are offered middle cranial fossa (MCF) surgical resection. Once VSs touch the brainstem, the chance of hearing preservation is rare, and thus, patients should be offered translabyrinthine approach. Patients older than 65 years of age are typically offered observation with serial imaging. When there is documented tumor growth, or there is compression of the brainstem, treatment is indicated

Knowledge of VS natural history with respect to tumor growth and hearing outcomes is important to understand.

Vestibular Schwannoma Natural History

The natural history of VSs is a slow rate of growth of 1–2 mm per year on average [11•]. Some tumors are quiescent for years, while other tumors can grow quickly. It is still not understood what factors lead to tumor growth. However, it is known that hearing loss can occur without tumor growth [11•].

Given the benign nature of VSs, it is imperative to understand the tumor growth rate and hearing outcomes with long-term observation. Stangerup and Caye-Thomasen [11•] have reported the longest published observational study of VSs. Tumor growth was variable and differed based upon initial tumor size. Approximately, 19 % of all small intracanalicular (restricted to the IAC) tumors had growth during the first 5 years of observation. Conversely, 34 % of larger tumors (those extending from the IAC into the CPA) demonstrated growth. Thus, not all tumors demonstrate growth during observation.

However, it is important to understand that hearing can deteriorate even without tumor growth. In patients whom presented with AAO–HNS class A hearing (word discrimination scores >70 %), only 59 % of patients maintained class A hearing at 5 years and only 46 % of patients maintained class A hearing at 10 years [11•, 12].

Observation of VSs has also been performed in patients over the age of 65 years. Only 28 % of patients required treatment (surgery or radiosurgery) during a 3-years follow-up [13]. Similar to previous observational studies, serviceable hearing (word understanding >50 %) deteriorated in 41 % of patients in the observation group.

Stereotactic Radiation Therapy

The use of stereotactic radiation therapy for VSs has increased over the last 2 decades. The details of this treatment are beyond the scope of this chapter. Briefly, the goal of radiation therapy is to limit tumor growth; it does not eliminate VSs. Reported tumor control rates are quite high, but pre-treatment tumor growth pattern is often not defined. Tumor control rates for tumors that had documented growth prior to radiation are lower, around 70–80 % [14]. While overall tumor control rates out to 10 years are quite high, hearing preservation following stereotactic radiation therapy for small VSs is poor. Long-term analysis shows only 23 % hearing preservation rate at 10 years with stereotactic radiation therapy [15•]. Given the short history of radiation for VSs and the evolving dosing schemes, the long-term (multiple decades) control rate in young patients is still unknown. In addition, there is a small possibility (approximately 0.04 %) of malignant transformation after radiation, and if this occurs, patient survival is often less than 12 months [16]. Also, if a schwannoma fails to respond to radiation, and surgical treatment is necessary, there is a higher risk of facial weakness postoperatively [17, 18]. Thus, we generally reserve stereotactic radiotherapy for those patients unable or unwilling to undergo surgical removal and who have documented tumor growth on serial imaging studies.

Hearing Preservation Microsurgery Approaches

There are two microsurgical approaches to VSs for hearing preservation. The middle cranial fossa (MCF) approaches the tumor from superior, while the retrosigmoid (RS)

approaches the tumor from posterior. The hearing preservation and facial nerve outcomes of each approach will be discussed. The hearing preservation rate appears to be slightly higher with MCF approach compared with RS [19].

Middle Fossa Craniotomy

Middle fossa (MF) craniotomy approach for VSs surgery was popularized by Dr. William House in 1961. This approach is best suited for small tumors that extend less than 1 cm into the CPA and is our preferred surgical approach for hearing preservation.

The great advantage of the MF approach is that it provides exposure of the entire length of the IAC, including the fundus, obviating the need for blind tumor dissection in the lateral IAC. Middle fossa craniotomy offers the highest rate of hearing preservation and allows for identification of the facial nerve as it exits the IAC. This permits the surgeon to develop a plane between the facial nerve and the tumor in a location where the nerve is not displaced by tumor. The temporal lobe retraction and the drilling and exposure of the IAC are performed extradurally, and there is a very low incidence of persistent, postoperative headaches. Cerebral spinal fluid (CSF) leaks are also rare with this approach.

The main disadvantage of the middle fossa approach is that the surgeon has to work past the facial nerve to remove the tumor. Fortunately, long-term facial nerve outcomes remain excellent and comparable to those obtained with the translabyrinthine approach [20, 21••]. The second disadvantage of the middle fossa approach is that it provides limited exposure of the posterior fossa unless the superior petrosal sinus is sectioned, restricting the use of this approach to cases with less than 2 cm tumor extension into the CPA. However, hearing preservation in tumors with significant extension into the posterior fossa is unlikely and the choice of which approach to use should not jeopardize facial nerve function or risk tumor recurrence for a low likelihood of hearing preservation. Therefore, we often recommend translabyrithine removal of these tumors even if the patient has good hearing.

Hearing preservation rates with middle fossa (MF) tumor removal are excellent in patients with small tumors. At our institution, 72 % (65 of 90 patients) with a VS of $\leq$10 mm retained serviceable hearing [21••]. Approximately 9 % of patients (8 of 90) had improvement in hearing after MF tumor removal. Hearing preservation rates decrease as tumor size increased. When the VS is larger (11–15 mm), hearing preservation was achieved in about 50 % of patient [21••]. Postoperative facial nerve function was excellent with 94 % of patients having normal (House Brackmann HB I) or near-normal (HB II) facial function [21••]. Others have demonstrated hearing preservation rates of 65–73 % and excellent facial nerve outcomes [22–25]. If hearing is saved, the durability of long-term hearing preservation is excellent [7, 22, 26].

MF tumor removal is relatively safe. Infection and seizure rates are around 1 %. Postoperative CSF leak rate is approximately 5 % and is most often successfully treated with lumbar drain and 5 days of bed rest. No patients had intracranial hemorrhage, stroke, or long-term seizure or word-finding disorders [21••].

The technical details of the middle fossa approach to removal of VSs have been described in detail [27]. Briefly, the patient is positioned in the supine position with the head turned to the side with the table turned 180° from the anesthesiologist. Facial nerve and ABR monitors are placed. The patient receives antibiotics, dexamethasone (10 mg), and mannitol (0.5 g/kg), and the patient is hyperventilated prior to craniotomy to reduce intradural pressure as the temporal lobe is elevated.

A temporal skin incision is incised down to the temporalis fascia, which is harvested for lateral skull base reconstruction. An anterior-inferiorly based temporalis muscle flap is then raised to identify the root of the zygomatic arch bone. Leaving a cuff of muscle and fascia along the muscle's insertion on the squamous portion of the temporal bone facilitates closure.

A 4.5 $\times$ 4.5 cm temporal craniotomy centered over the zygomatic root is created with 4 mm burrs. The bone flap is carefully elevated from the dura. Extradural elevation of the temporal lobe is performed to expose the floor of the middle fossa. The greater superficial petrosal nerve (GSPN), arcuate eminence, and petrous ridge are identified. The IAC lies at the bisection of an angle formed by the greater superficial petrosal nerve and the arcuate eminence. The House-Urban middle fossa retractor is positioned along the margins of the craniotomy, and the blade of the retractor is placed along the true petrous ridge directly medial to the IAC.

Using diamond burrs, the bone over the arcuate eminence is removed to "blue-lining" the superior semicircular canal. The IAC is exposed medially to laterally. Medially the IAC is exposed for a full 270°. Laterally, dissection is limited posteriorly by the ampullated end of the superior canal and anteriorly by the cochlea. The facial nerve is followed laterally, and the labyrinthine portion of the facial nerve is identified and fully decompressed to accommodate for any postoperative swelling that may occur. Bill's bar separates the superior vestibular nerve from the facial nerve at the fundus, and it is fully exposed.

ABR monitoring is then performed during tumor removal. The dura of the IAC is opened. The facial nerve is identified and dissected sharply from the tumor. The tumor is then microdissected from the IAC while preserving the

facial nerve, cochlear nerve, and labyrinthine blood supply. Both vestibular nerves are removed with the tumor.

After tumor removal, a small piece of temporalis muscle or fat is placed in the dural defect of the IAC. The temporalis fascia is placed over the middle fossa floor, and the retractor is removed. The bone flap is replaced followed by multilayer closure of the muscle, galea, and skin.

Retrosigmoid Craniotomy

The retrosigmoid approach creates a panoramic view of the posterior fossa from the tentorium cerebelli to the foramen magnum. It is frequently the approach most familiar to neurosurgeons. There is no temporal lobe retraction like the MF approach, but the lateral cerebellum must be retracted. With respect to hearing preservation surgery for small VSs, the main disadvantage is the limited lateral exposure of the IAC. The posterior semicircular canal and crus commons limit exposure of the lateral IAC from a retrosigmoid approach. Since dissection of the tumor in the lateral IAC is done blindly, there is a higher risk of leaving tumor behind.

Hearing preservation rates are lower than with the middle fossa approach [28], and the incidence of CSF leak and persistent, postoperative headache are higher in the retrosigmoid approach. The postoperative headaches may be related to the intradural drilling of the IAC or muscle attachment to the dura.

Because of the limited exposure of the lateral IAC in the retrosigmoid approach, we prefer the middle fossa approach in hearing preservation cases with tumors less than 2 cm. The retrosigmoid approach is best suited for patients with good hearing and whose tumors are larger than 2 cm and have limited lateral extension in the IAC. However, the likelihood of hearing preservation in such tumors is lower that that achieved in patients with small tumors approached through the MF.

The retrosigmoid approach has been described in detail [29]. Briefly, the patient is placed in the "park-bench" or standard supine position, and the bed is rotated 180° from the anesthesiologist. Facial nerve and ABR monitors are placed.

A curvilinear incision is made three to four finger breadths behind the postauricular crease. Skin flaps are raised in the subcutaneous plane for 2 cm and the muscle and periosteum are incised in a stair step fashion 1 cm anterior to the skin incision. Cervical muscles are detached, and an anteriorly based flap is elevated forward 1.5 cm anterior to the sigmoid sinus exposing the mastoid and its tip, and a posteriorly based flap is elevated to the occiput. This exposes the craniotomy site.

A 4 × 4 cm bony window is made in the suboccipital region using either a high-speed drill with 4 mm burrs or a craniotome. The sigmoid sinus forms the anterior limit of the window, and the transverse sinus forms the superior margin. The bone flap is set aside and used for reconstruction at the end of the procedure. This helps restore the skull contour and limit postoperative headaches. Mastoid air cells are usually opened in this approach and require obliteration with bone wax and/or abdominal fat at the end of the procedure to prevent CSF leakage. The dura of the posterior fossa is opened beginning 2–3 mm posterior to the junction of the transverse and sigmoid sinuses, reflected posteriorly, and held in position with stay sutures.

Cerebellopontine angle exposure is begun by draining CSF from the cisterna magna and CPA cistern. Arachnoid adhesions in the cistern are lysed, which decompress the posterior fossa and allow the cerebellum to fall medially and posteriorly. Premature cerebellar retraction, prior to CSF decompression, risks massive cerebellar swelling. The cerebellum is covered by a protective pad or cottonoids and retracted posteriomedially. This exposes the CPA and the 7th and 8th cranial nerve complex with the tumor emanating from the porus acousticus. The trigeminal nerve is seen superiorly and the lower cranial nerves (IX, X, and XI) inferiorly.

Prior to drilling the IAC, the dura of the posterior face of the petrous ridge is incised, and superior and inferior dural flaps are elevated. Bone removal over the IAC proceeds from a medial to lateral direction using a high-speed drill. Bony troughs 3–4 mm in diameter are then created above and below the IAC. The goal is to provide 180°–270° exposure of the IAC circumference. The posterior semicircular canal limits lateral exposure of the IAC. The dura of the IAC is then opened exposing the intracanicular portion of the tumor and the neural structures of the IAC.

The tumor and both vestibular nerves are then microdissected from the canal while preserving the facial and cochlear nerves and the labyrinthine blood supply. Intracapsular debulking of the tumor may be necessary prior to dissection of the capsule from the facial and cochlear nerves.

The IAC defect is inspected, and any air cells are meticulously sealed with bone wax. A small muscle plug or small piece of abdominal fat is used to plug the IAC defect. The dural edges are then reapproximated in a water-tight fashion with 4–0 braided nylon. The edges of the craniotomy are inspected, and any opened air cells are sealed with bone wax. If a significant amount of mastoid air cells were opened, these are best sealed with a fat graft. The bone flap is repositioned, and the soft tissues are reconstructed in layers. A sterile pressure dressing is applied.

Postoperative Care

Postoperatively, the patient is monitored in an intensive care unit for 24 to 48 h. For patients undergoing middle fossa craniotomy, the pressure dressing is changed on the first postoperative day. The pressure dressing is used until

the fourth postoperative day. Patients often have intense vertigo during the initial postoperative period which is treated with steroids and anti-emetic medications. Most patients are out of bed sitting in a chair on the first postoperative day and begin ambulation the following day. Early ambulation accelerates vestibular compensation.

Cerebrospinal fluid leaks are rare but occur 1–5 % of the time following middle fossa approaches and from 5–15 % following retrosigmoid craniotomy. A reservoir test is performed on postoperative day 2 or 3 to test for a CSF leak. Initial management of a postoperative CSF leak includes a lumbar drain and elevation of the head of the bed. Early recognition and management of CSF leaks are almost always successful. Only rarely re-exploration of the defect with additional fat obliteration is required to treat a postoperative CSF leak.

Patients are discharged from the hospital once they are able to walk safely and take care of themselves without difficulty. This usually occurs on the fourth or fifth day following surgery. Most patients return to routine daily activities within 1–2 weeks following surgery but will notice early fatigue that persists for 1–3 months after surgery. Return to full-time employment therefore usually requires 4–6 weeks of convalescence.

Patients are seen in clinic at 1 month after surgery with audiogram, a baseline, postoperative MRI, and examination of the wound and ear. MRI imaging is repeated at 1 and 5 years following surgery to monitor for tumor recurrence.

Conclusion

Modern diagnostic and imaging techniques often detect VSs while they are still small and causing minimal symptoms. For those patients with serviceable hearing and a small VS, it is practical to treat the tumor while preserving hearing. The middle fossa craniotomy approach is used most often for the surgical removal of tumors with attempted hearing preservation, while the retrosigmoid approach is reserved for larger, medially positioned tumors. Translabyrinthine craniotomy is the preferred approach for patients with poor hearing or large tumors. Nonsurgical options, including observation with serial imaging and stereotactic radiotherapy, are appropriate for selected patients.

Compliance with Ethics Guidelines

Conflict of Interest Rick F. Nelson, Bruce J. Gantz and Marlan R. Hansen declare that they have no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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