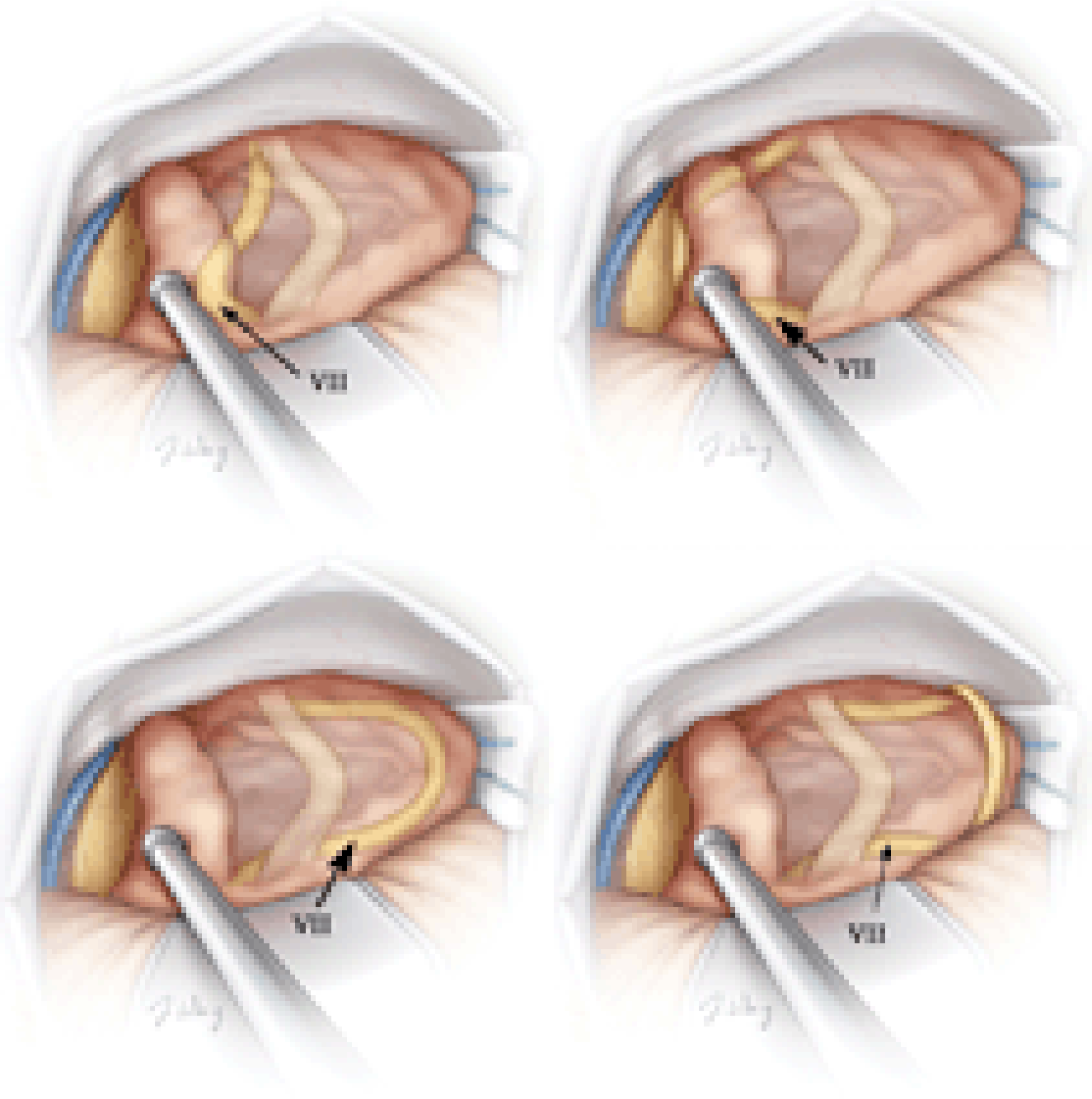
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Introduction

Acoustic neuromas

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Acoustic neuromas (vestibular schwannomas) remain among the most technically challenging of tumors to remove at the cranial base. Over the last 20 years, however, significant advances in skull base microsurgical techniques and intraoperative neuromonitoring have made the possibility of facial nerve and hearing preservation a reality. Better understanding of the natural history, as well as the advent of stereotactic radiosurgery, has increased the number of options and strategic paradigms in the management of these tumors. In the last decade, technological advancements and an emphasis on functional outcomes have raised the bar for acoustic neuroma surgeons with the goal of achieving the best cranial nerve outcomes, oncological control, and posttreatment quality of life for our patients.

In this issue of *Neurosurgical Focus*, we have compiled a broad spectrum of articles dedicated to the subject of acoustic neuromas describing the natural history, conservative management, radiosurgical treatment, resection and postoperative outcomes, and postoperative neuroimaging of these lesions. Contributions from Kondziolka et al., Schmidt et al., Hoa et al., and Thakur et al. review the management of newly diagnosed, small incidental acoustic neuromas and discuss the natural history and treatment dilemmas that are encountered by the acoustic neuroma surgeon.

The next articles focus on functional outcomes after acoustic neuroma treatment. Yashar et al. and Thakur et al. discuss postoperative outcomes after resection of cystic acoustic neuromas. Sonig et al. examine the socioeconomic impact of acoustic neuroma surgery in the United States with respect to discharge disposition and hospital costs. Sun et al. and Gurgel et al. discuss facial nerve outcomes and the neuroanatomical correlation of the House-Brackmann grading system. Ansari et al. provide a systematic review of the postoperative complications associated with each surgical approach.

To achieve the best cranial nerve outcomes after surgery, the acoustic neuroma surgeon should be equipped with the fundamental knowledge and techniques of intraoperative neuromonitoring. Kircher and Kartush, and Oh et al. present comprehensive overviews of intraoperative neuromonitoring techniques and discuss the pitfalls that can be encountered during surgery.

The last section of this issue is devoted to aspects of surgical technique and operative nuances. Chamoun et al., Kulwin and Cohen-Gadol, and Nickele et al. present online video manuscripts on the surgical approaches for acoustic neuromas (retrosigmoid, middle fossa, and translabyrinthine) that serve as useful and practical surgical video atlases. The authors share their operative pearls for successful surgery and include details of patient positioning, surgical incision and opening, tumor dissection and removal, and closure techniques. DeMonte and Gidley review their experience with the middle fossa approach for intracanalicular tumors with excellent hearing preservation rates. In addition, Liu et al. introduce a novel fascial sling technique for dural reconstruction after translabyrinthine approaches to minimize postoperative cerebrospinal fluid leak rates. Lastly, Ginat and Martuza provide an excellent comprehensive review of the interpretation of postoperative neuroimaging after resection of acoustic neuromas.

This edition of *Neurosurgical Focus* covers a wide range of topics that will be of very practical importance to neurosurgeons and neurootologists who treat acoustic neuromas. It is our hope that this collection of papers will provide our readership a better understanding of acoustic neuromas and stimulate new avenues of research. We thank all of the authors who have contributed to this issue and the editorial staff at *Neurosurgical Focus* for their efforts in putting together this exciting issue.
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An update on unilateral sporadic small vestibular schwannoma

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Advances in neuroimaging have increased the detection rate of small vestibular schwannomas (VSs, maximum diameter < 25 mm). Current management modalities include observation with serial imaging, stereotactic radiosurgery, and microsurgical resection. Selecting one approach over another invites speculation, and no standard management consensus has been established. Moreover, there is a distinct clinical heterogeneity among patients harboring small VSs, making standardization of management difficult. The aim of this article is to guide treating physicians toward the most plausible therapeutic option based on etiopathogenesis and the highest level of existing evidence specific to the different cohorts of hypothetical case scenarios.

Hypothetical cases were created to represent 5 commonly encountered scenarios involving patients with sporadic unilateral small VSs, and the literature was reviewed with a focus on small VS. The authors extrapolated from the data to the hypothetical case scenarios, and based on the level of evidence, they discuss the most suitable patient-specific treatment strategies. They conclude that observation and imaging, stereotactic radiosurgery, and microsurgery are all important components of the management strategy. Each has unique advantages and disadvantages best suited to certain clinical scenarios. The treatment of small VS should always be tailored to the clinical, personal, and social requirements of an individual patient, and a rigid treatment protocol is not practical.
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Key Words • **vestibular schwannoma** • **acoustic neuroma** • **observation and imaging** • **microsurgery** • **stereotactic radiosurgery**

Technological advances in neuroimaging have been responsible for changing the face of neurosurgery. This has certainly been true for VS, first described by Eduard Sandifort in the Netherlands in 1777.[3] From what was considered a life-saving surgery (the finger extraction technique) done by Englishman Sir Charles Balance in 1894,[8] VS treatment has evolved toward a more conservative management paradigm.

The common denominator for the evolution of VS treatment has been the focus on patients' quality of life.[111] In the 1990s, achieving tumor control while preserving facial and cochlear nerve function were the ideal goals of surgery. The management of VS has developed further over the past decade, with importance being given to postoperative quality of life.[18,21,61,84,111] This is even more pertinent to patients with small tumors (< 25 mm) or with minimal hearing deficits.[21,61,111] Currently, the treatment options for managing small VS include conservative observation and serial imaging ("wait and scan"), stereotactic radiosurgery and microsurgery.

Formulating the best treatment strategy in patients with small tumors rests on the ability of the physician to understand specific patient and tumor characteristics and to be able to tailor the treatment options to the individual. The variability in the literature with respect to optimal treatment strategies for small VSs makes it even more challenging for the physician to formulate the most suitable plan. The main goal of this review is to outline the most suitable therapeutic option for different cohorts using hypothetical case scenarios and the highest level of evidence available. We list 5 hypothetical clinical situations and extrapolate from the reviewed literature pertinent to each clinical setting. Additionally, we discuss the proposed possible mechanisms of hearing loss in patients with small VSs.

Abbreviations used in this paper: AAO-HNS = American Academy of Otolaryngology–Head and Neck Surgery; CPA = cerebellopontine angle; DPOAE = distortion products of otoacoustic emissions; GKS = Gamma Knife surgery; GRC = Gardner-Robertson Class; PTA = pure-tone average; SDS = speech discrimination score; SRS = stereotactic radiosurgery; VS = vestibular schwannoma.

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What Actually Creates the Treatment Dilemma?

Seth I. Rosenberg started his article on the natural history of acoustic neuroma[86] by quoting from the 1904 article by Fraenkel et al.:[31] "An estimation of the size and character of the growth is nearly always a mere matter of speculation, based on tumor statistics, the rapidity of the clinical course, or associated constitutional conditions." Apart from improvements in estimating tumor location and size, this statement holds true even after 100 years of scientific progress. One of the challenges a physician still has to overcome is that of making a therapeutic decision when a patient with a small VS comes to the clinic with a normal or serviceable hearing level. The parameters that are considered essential for planning the treatment strategy are listed in Table 1.

The factors that make the final decision a daunting task include variability in the data on the natural history of VS,[68,86,118] mechanism of hearing loss,[27,37,57,81,99] and treatment-specific hearing and facial function preservation rates.[96,98,101] These inconsistencies are a major hindrance to the practice of evidence-based medicine. Meta-analyses of pooled data are fraught with considerable assumptions to simplify the nonuniform existent data and achieve statistical power and significance, which can greatly impact their conclusions.[100,101,111,115] These studies should thus be interpreted with caution.

The aim of the current review is not to address statistical "treatment-specific analysis," but rather highlight "case-specific" requirements and extrapolate from the literature in a simplified way for the treating physician.

Mechanism of Hearing Loss in VS

Considering the variability in the data regarding the natural history of VS and the difficulty in predicting hearing outcomes in individual cases, we sought to briefly review the proposed mechanism for hearing loss in VS (Fig. 1).

TABLE 1: Essential parameters to be kept in mind while considering treatment options for small VS*

Parameter
pt age, occupation, SES
tumor location
status of hearing; PTA & SDS
status of CN V and VII function
tumor consistency
solid
cystic
mixed
tumor characteristics
calcification
microhemorrhages
fibrosis
necrosis
any comorbidities
expected impact of the treatment on QALYs
pt's personal preference

* CN = cranial nerve; pt = patient; QALY = quality-adjusted life year; SES = socioeconomic status.

In general, there are 4 patterns of hearing loss:[21,27,37,38,51,57,71,72,81,99] 1) hearing deterioration in "growing" tumors, 2) hearing deterioration in "nongrowing" tumors, 3) early hearing loss, and 4) sudden hearing loss regardless of tumor growth.

Vestibular schwannomas are histologically benign, slow-growing tumors that originate from the Schwann cells of the inferior vestibular nerve in most cases.[44] The location of the tumor's origin is described as the neurilemmal-neuroglial junction (Obersteiner-Redlich zone) within the internal auditory canal.[48,67] Considering the origin of the tumor, the most obvious hypothesis for hearing loss is the mechanical and/or neurotoxic effect that a growing tumor might have on its neighboring neurovascular structures.[57,81] The data, however, do not always show a linear correlation between the rate of tumor growth and deterioration of hearing.[38,39,82,96,104]

Hearing Loss in Patients with Growing Tumors

The "mechanical effect" is thought to involve compression or stretching of the cochlear nerve (conduction block) or compromise of the vascular supply to the cochlea (through occlusion or spasm of the labyrinthine artery).[81] This compromise of the vascular supply can be a result of an increase in intracanalicular pressure or due to direct compression caused by the laterally invading tumor.[7,51] Compression of labyrinthine vessels has been shown to cause degeneration in the organ of Corti and the spiral ganglion (cochlear dysfunction).[22,37,43,71] Of note, the progression of hearing loss in VS is most often gradual,[39,96] although sudden deterioration is also possible.[32,65]

Hearing Loss in Patients with Nongrowing Tumors

Another subset of VS patients develop hearing loss despite the lack of tumor growth on imaging.[38,75] Obviously, the mechanical effect theory fails to explain this category of hearing loss. In these patients, neurotoxic injury, caused either by alterations in the biochemical properties of the inner ear fluid or through accumulation of tumor toxic metabolites, may be responsible for hearing loss. The perilymphatic space and endolymphatic space in the inner ear of patients with VS often stain positive for eosinophilic proteinaceous deposits.[26] The perilymphatic protein levels are reported to be 5–15 times higher than the levels found in the ears of healthy individuals.[93] This rich perilymphatic protein is believed to represent either a by-product of upregulated genetic activity in the VS[109] or simply accumulation of serum proteins by the process of transudation.[93] The correlation between increased proteinaceous material and cochlear dysfunction is still not clear.

Early Hearing Loss

Efferent olivocochlear bundle dysfunction and alteration in biochemical properties of the inner ear fluid seem to be the most plausible causes of early hearing loss in VS patients. Gouveris et al.[37] compared DPOAE am-

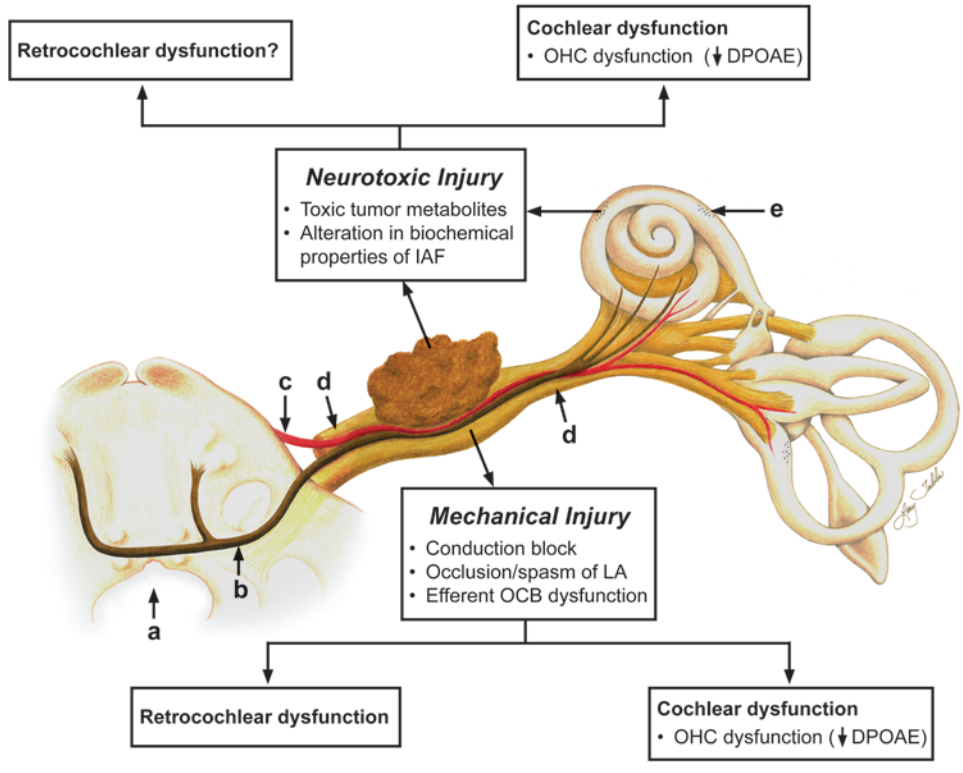

Fig. 1. A schematic showing the possible mechanism of hearing loss in a small VS. a = fourth ventricle; b = efferent olivocochlear tract; c = labyrinthine artery (LA); d = vestibulocochlear nerve; e = proteinaceous deposits in the inner ear fluid due to tumor metabolism; IAF = inner ear fluid; OCB = olivocochlear bundle; OHC = outer hair cells.

plitudes between VS patients with normal, symmetrical hearing and VS patients with mild hearing loss. They reported that the early hearing loss in patients with VS was cochlear in origin, suggested by a consistent finding of decreased DPOAE amplitudes, indicating dysfunction of the outer hair cells. Although the authors did not provide the mean tumor size in their population, the concept of early hearing loss as cochlear in origin was striking. The possibility that a small intracanalicular tumor will impede cochlear blood flow to the level of causing outer hair cell dysfunction seems less likely. On the contrary, the proximity of the passing unmyelinated efferent olivocochlear tracts to the vestibular nerve suggests the etiology to be efferent auditory system dysfunction.[81] The efferent olivocochlear bundle comprises medial fibers that innervate the outer hair cells and lateral fibers that synapse with the dendrites of the inner hair cells. The functions assigned to the efferent system of the ear include noise protection, improvement of signal-to-noise ratio, selective attention, adaptation, and frequency selectivity (through regulation of the micromechanical properties of outer hair cells).[20] Additionally, functionality in the 3D auditory world—localization of sound and speech restoration—is affected in efferent olivocochlear bundle dysfunction.[20]

Animal studies suggest a hearing loss of up to 60 dB related to efferent auditory dysfunction.[73] Pennings et al.[75] followed 47 patients with purely intracanalicular tumors prospectively, without any intervention. At baseline, 12 patients already had PTA greater than 50 dB. This suggests that efferent auditory dysfunction alone may not be responsible for early hearing loss and that alteration in the biochemical properties of the inner ear fluid might also play a role in the initial stages. Recently, van de Langenberg et al.[104] reported that in patients undergoing serial imaging, there was significantly greater deterioration of PTA over time in the ears in which labyrinthine signal hypointensity was seen on T2-weighted MR images than in those that showed a more isointense intense labyrinthine signal[104] ($p = 0.01$). Although nothing in the data suggested an explanation for this finding, the authors hypothesized that biochemical changes, including highly proteinaceous deposits and vascular compromise, might be the reason for the hypointensity.

Sudden Hearing Loss

Sudden hearing loss is also noted in some VS patients with documented rates between 3% and 23%.[88] The internal auditory artery or the labyrinthine artery (end artery) travels within the internal auditory canal, and it may be possible that vascular insult to it can result in sudden hearing loss.[11,60] However, if only vascular insult was the cause, one would also expect to see vestibular symptoms and cochlear hearing loss, which is not always the case. Further, due to the anatomical distribution of the blood supply to the cochlea, vascular compromise should result in low-frequency hearing loss in these patients, whereas mostly higher-frequency loss is reported in such cases.[51,52] Tumor characteristics, including accelerated tumor growth and an increased number of intratumoral hemorrhages, are thought to cause sudden hearing loss through

nerve compression and rapid accumulation of biochemical toxins.[99] A higher incidence of high- to mid-frequency hearing loss correlates well with the nerve compression theory,[32] but otoacoustic emissions in such patients do not reflect purely retrocochlear hearing loss[71] and suggest that both cochlear and retrocochlear components contribute to sudden hearing loss in such patients.

To summarize, it would be reasonable to state that the exact mechanism of hearing loss in VS patients is likely to be multifactorial. Better understanding of the possible etiologies of hearing loss will help in guiding VS treatment choices, not only on the basis of tumor size and location, but also based on the intricate patterns of inner ear functioning in VS patients at the time of presentation.

In the section below, we present 5 distinct case scenarios and review the literature that is most applicable to formulating a management plan for each hypothetical patient.

Hypothetical Case Scenarios and Review of the Literature

Hypothetical Case 1

> This patient is a 40-year-old man with a small VS (maximum diameter < 25 mm) that was discovered incidentally. The results of his audiometrical examination are within normal limits. Hearing preservation is his primary concern, as he works for a local music band and needs to have normal hearing to be successful. He inquires about the best option(s) for preserving his intact hearing status.

Advancements in neuroimaging have increased the rate of incidental detection of VS. In a recent Rotterdam-based population study, the most common brain lesions discovered incidentally by MRI were asymptomatic brain infarcts, followed by cerebral aneurysms and primary benign tumors.[105] The authors of the study reported the incidence of asymptomatic VS to be 1 in 500 individuals. A recent study of the Surveillance Epidemiology and End Results (SEER) US database identified the incidence rate of VS to be 1.1 per 100,000, with the mean age of detection being 53.1 years and the majority of lesions (84%) occurring in the Caucasian population.[34]

Tumors That Will Grow if Left to Observation. To address the patient's concern about the chances of hearing preservation, knowledge about the chances of the tumor showing growth and the probable growth rate are important. Combining the results of 6 previous meta-analyses shows that tumor growth occurred in 29% to 54% of patients (mean 44.6%).[4,9,89,95,101,113,118] A major drawback in the application of these results is that the mean follow-up time in these studies was only 3 years. A literature review of mostly Level 3 evidence yields impressive variability in growth rates (range 18%–73%).[68] There are very few prospective studies that report tumor growth in the patients followed with observation. Régis et al.[83] reported tumor growth in 77% of their patients (mean follow-up 5.3 years); Hajioff et al.,[39] in their 10-year follow-up prospective study (median duration of follow-up 121 months), reported tumor growth in 78% of patients (< 1 mm/year in 38% and > 1 mm/year in 40%); and Stangerup et al.[96] found that only 17% of intrameatal tumors became extrameatal and that only 28.9% of extrameatal tumors showed growth (mean follow-up 3.6 years). It is difficult to develop a scientific rationale that would explain this variation. Régis et al.[83] pointed out that the definition of tumor growth was relatively personal. For example, Stangerup et al. considered a change in any tumor from intrameatal to extrameatal to be tumor growth, whereas Hajioff et al. defined significant growth as growth of more than 1 mm/year.

Tumor Growth Rates in Major Series. The inability to predict consistently whether a tumor will grow makes predicting the growth rate even more problematic. Tumor growth rate is a parameter about which there is general agreement with respect to treatment strategy decisions. A patient who has a tumor growth rate of more than 2–2.5 mm/year is at a significantly higher risk of hearing loss than a patient with less than 2.5 mm growth per year.[38,39,75,101] This obviously does not account for patients who develop hearing loss without any significant tumor growth (as described earlier in this paper).

Recently, Sughrue et al.,[101] in their systematic review of 34 published studies that included 982 patients (Level 3 evidence), found that in 75.32% of the patients tumor growth was greater than 2.5 mm/year, and the mean tumor growth was 2.9 mm/year. In contrast, a prospective study by the same group showed a growth rate of less than 2.5 mm/year in 83% of patients.[98] The authors of another recent prospective study documented the mean tumor growth in their population to be 2.1 mm/year.[83] A 2005 systematic review by Yoshimoto[118] included 4 prospective studies in which only 29% of tumors showed growth. There is a striking difference between the growth rates documented by older meta-analyses and the 2.9 mm/year reported by Sughrue et al. in their 2010 review.[101] Mean tumor growth rates of 1.8, 1.9, 1.9, 1.2, and 2 (mm/year) were reported in the meta-analyses published in 1998,[89] 2003,[113] 2005,[95] 2005,[118] and 2006,[9] respectively.

Although there is variation in the literature, the results of prospective studies could be interpreted as suggesting that tumor growth is likely to be less than 2–2.5 mm/year in the majority of patients. Table 2 lists various factors that may have prognostic implications for VS growth patterns.[2,5,13,28,39,41,59,62,68,101,104] But there is clearly a need for long-term prospective data documenting annual trends in tumor growth rates.

Hearing Preservation in Wait-and-Scan Group. The most recent meta-analysis, that of Sughrue et al. (2010),[101] reviewed the literature on the natural history of untreated sporadic VS (tumor size < 25 mm). In 982 patients, the overall hearing preservation rate was 54%, with follow-

TABLE 2: Factors that may predict VS growth patterns

Predictors
significant growth (>2 mm/yr)
extension into the CPA
diameter >20 mm
sudden onset or short duration of sensory hearing loss
tinnitus, unsteadiness, & vertigo at presentation

up ranging from 26 to 52 months. Meta-analyses in the past have documented hearing preservation in 49%–63% of patients with a mean follow-up of approximately 3 years.[9,89,94,101,113,118] Sughrue et al. found that the hearing preservation rate was significantly higher for the group of patients demonstrating a tumor growth rate of 2.5 mm/year or less. Interestingly, the overall mean growth rate among 982 patients included in their meta-analysis was 2.9 ± 1.2 mm/year. It is worth mentioning a common finding among the majority of the wait-and-scan group studies, which is deterioration of PTA and SDS over years in almost all the patients presenting initially with some degree of hearing loss.[39,75] Régis et al.,[83] in their cohort of 40 patients followed up for a mean of 43.8 months, reported useful hearing preservation rates of 75%, 52%, and 41% at 3, 4, and 5 years, respectively.

A prospective study from the University of California, San Francisco group (Sughrue et al. 2011[98]) calculated the estimated time to hearing loss among 3 groups of patients—those with intracanalicular tumors, those with 0.1–1 cm extension into the CPA, and those with more than 1 cm extension into the CPA cistern. The estimated median time to hearing loss in these 3 groups was 11.6, 10.3, and 9.3 years, respectively. Further, the authors noted that the median time of hearing loss in patients with a tumor growth rate of more than 2.5 mm/year was 7 years as compared with 14.8 years in those with growth of less than 2.5 mm/year. Hajioff et al.[39] followed a cohort of 72 patients prospectively for a median of 121 months (range 80–271 months); 49% of the patients were advanced in age and 43% made a personal choice for observation. The authors noted that the patients' hearing deteriorated substantially even in cases of nongrowing tumors. The mean deterioration in PTA at 0.5, 1, 2, and 3 kHz was 36 dB, and the SDS deteriorated by a mean of 40%. The overall failure rate in their cohort was 40%, with three-fourths of the failures occurring during the first half of the study.

Another recent noncomparative prospective study followed 186 consecutive patients (median follow-up 43 months), all of which were initially allocated to conservative treatment.[13] At the last follow-up, 40% of the patients required either radiosurgery or microsurgery and the median time to treatment was given as 26 months (SD 17.4 months). Serviceable hearing at 1–3 years and/or beyond 3 years was significantly decreased as compared with baseline hearing status.

Recently, Stangerup et al.[97] reported the results of their 932 VS cases managed with observation; 102 patients (11%) were observed for over 10 years and hearing preservation was seen in 46%. In this study, the author found that patients with 100% SDS at diagnosis have a 75% chance of maintaining good hearing even after 10 years of observation, which was in agreement with various other studies.

Some reproducible conclusions (Table 3) made from a review of literature regarding observation[75,96,101,104] include the following: 1) Patients with an SDS of 100% at diagnosis have higher chances of long-term hearing preservation. 2) Hearing preservation rates are markedly higher for the patients with tumor growth of less than 2–2.5 mm/year. 3) Most tumors demonstrate growth within the first 5 years of diagnosis. 4) Regardless of tumor growth, hearing deterioration is bound to happen without intervention.

TABLE 3: Factors predicting hearing outcomes in the observation and imaging group*

Category & Factor
factors predicting hearing loss
signal hypointensity in the affected labyrinth on T2-weighted MR images
established hearing loss at initial presentation
tumor growth >2.5 mm/yr
factors predicting better hearing outcomes
WRS Class 0 or 1 at initial presentation
initial hearing level up to 10 dB HL at 4000 Hz
tumor growth <2.5 mm/yr

* HL = hearing loss; WRS = word recognition score.

Based on the current literature, a strategy of observation and serial imaging may be continued in a young or middle-aged patient who has normal findings on audiometric examination and a VS that has not demonstrated significant growth. However, if the tumor starts growing, the chance of losing serviceable hearing will increase. Is there a role for early intervention (SRS/MS) in these patients? The chances of hearing preservation in patients treated by means of SRS or microsurgery will be discussed subsequently under *Hypothetical Case 2*; this discussion will also clarify the role of early intervention in Hypothetical Case 1.

It is also worthwhile to mention that even in some academic centers of excellence, where the patients are sent constant reminders for follow-up, the percentage of patients lost to follow-up can be substantial. If we consider health care centers where following up is at the discretion of the patient only and regular follow-up is hindered by numerous factors including socioeconomic status and health insurance issues, it is quite worrisome to imagine the dropout rates from the observation subset of patients.

Hypothetical Case 2

> This patient is a 40-year-old woman with a small VS (< 25 mm) and mild hearing loss (serviceable hearing). Understandably, the patient wants to prevent any further hearing deterioration. She is open to any treatment modality that may stabilize her mild hearing loss or even improve her hearing.

The presence of any symptoms at presentation reflects some significant ongoing biological activity in the tumor causing anatomical and physiological compromise. The possible mechanisms[57,81] through which functional compromise can take place include 1) mechanical compression, 2) neurotoxic effects mediated through either toxic by-products of tumor metabolism or alteration in the inner ear fluid biochemistry, and 3) efferent pathway dysfunction. As previously discussed, there is significant heterogeneity in the data with respect to tumor growth in the absence of treatment; given this heterogeneity, observation and serial imaging is not a reasonable strategy for

patients who present with some hearing loss but want to preserve a serviceable hearing level.

In contrast to the data on growth of untreated tumors, the data available in the literature addressing tumor growth control following SRS demonstrate impressive consistency. The tumor control rate after SRS among the meta-analytical studies[9,47,91,108,113–116] in the last decade (regardless of tumor size) range from 91% to 94.7%, with a mean of 93.4%. Even though microsurgical removal of small VSs is the most effective means of long-term tumor volume reduction, the associated morbidities and lesser rates of hearing preservation, make it a less popular treatment strategy among physicians and patients (details to be discussed later). Of note, stabilizing the tumor through SRS does not preclude the chances of hearing deterioration and there is a subset of patients who experience sudden hearing loss after SRS.[30,40,49,56,69,78,84,85,103,106,112] Nevertheless, the chance of this phenomenon is less in patients in whom the median tumor dose does not exceed 13 Gy.[19,45,52,58,69,108]

Radiobiology of VS

Ionizing radiation induces cell cycle arrest, necrotic cell death, or both in VS.[117] The higher the number of tumor cells in the proliferative phase, the more radiosensitive is the tumor.[53,54] Ionizing radiation arrests the tumor cells in the proliferative phase such that the cell cycle checkpoint signaling (activated by DNA damaging agents) prevents the replication of damaged DNA as well as segregation of aberrant chromosomes, thus achieving tumor control.[117] The radiation also induces double-strand DNA breaks, which leads to apoptosis.[52] In addition to a low proliferative index, another factor causing radioresistance is tumor hypoxia. Rapidly growing tumors will be more hypoxic due to inadequate angiogenesis and therefore may be more radio-resistant.[6]

Lee et al.[53] found that recurrent VSs removed through microsurgery, which were previously treated by Gamma Knife surgery, had lower a proliferation rate than those treated primarily by microsurgery ($p = 0.03$). This suggests that radiosurgery might be able to induce apoptosis in the cells with a high proliferative index and leave behind those with a low proliferative index. Surgical removal alone, especially subtotal resection, might leave behind a mixed population of tumor cells, and subsequently the cells with a high proliferative index might continue to grow.

On occasions, VSs may show a transient increase in size following SRS.[64,77] This phenomenon is reported to occur in 17%–74% of patients undergoing SRS. It may be attributed secondary to radiation-induced tumor necrosis, chronic intratumoral bleeding, or biological response to ionizing radiation. A higher radiation dose has also been described as a possible risk factor for expansion.[43,66,80] Additionally, the growth may also be associated with transient facial spasm and trigeminal nerve dysfunction.[64] This transient expansion is most evident within the 1st year of treatment, and in most cases the tumor volume eventually becomes significantly less than before treatment. Rarely, continued growth after transient expansion is noted. If the growth is persistent and becomes symptomatic, further SRS or microsurgery is usually recommended.

Chances of Hearing Preservation After SRS. Yang et al.,[114] in their 2010 review, quantified data from 45 published studies to investigate hearing preservation rates after SRS. The overall hearing preservation rate (AAO-HNS Class 1 or 2, GRC 1 or 2) was reported to be 51% (mean follow-up 44.4 ± 32 months [SD]). This outcome appeared to be dose dependent. The studies that used a radiation dose of 13 Gy or less had a preservation rate of 60.5%, which was significantly higher than the studies in which the dose was larger than 13 Gy (50.4%). Some recent contemporary retrospective studies[30,52,56,103] using mean tumor margin doses of less than 13 Gy have reported long-term hearing preservation rate ranging from 68% to 93.3%. Details of hearing outcome in the prospective studies are shown in Table 4. The hearing preservation rates in these studies range from 63% to 93% for patients with VSs smaller than 3 cm in maximum diameter. Additionally Niranjan et al.[69] followed 96 patients with intracanalicular tumors prospectively and found that serviceable hearing in 77.5% of the patients with AAO-HNS Grade 1 hearing preoperatively and in 64.5% in those with Grade 2 preoperatively (mean follow-up 42 months). Facial and trigeminal nerve function was preserved in 100% of the patients.

In 2010, Régis et al.[83] published the results of a prospective comparative study on intracanalicular VS, in which they compared patients treated with GKS with the wait-and-scan group. In the 34 patients treated by GKS, the useful hearing preservation rates at 3, 4, and 5 years were 77%, 70%, and 64% in comparison with 75%, 52% and 41% in the wait-and-scan group at the same follow-up points. Also, in that study the chance of maintaining functional hearing and avoiding further intervention was 79% at 2 years' follow-up and 60% at 5 years' follow-up in the GKS group compared with 43% and 14% in the wait-and-scan group.

Hearing Loss After SRS. Gamma Knife surgery for VS has occasionally been shown to cause hearing loss, especially within the first 12 months. Wackym et al.[106] prospectively evaluated 59 patients treated with SRS (mean follow-up 63.76 months). They found that limiting the cochlear dose of radiation to less than 4 Gy can significantly decrease the incidence of hearing loss. They noted a distinct pattern of hearing loss in their cohort of patients, similar to what was reported earlier by Lasak et al.[52] It was consistent with a type of hearing loss resulting from radiation-induced injury to stria vascularis, characterized by a strial presbyacusis–like pattern with hearing loss across all frequencies and relative preservation of speech discrimination ability. Other proposed mechanisms of hearing loss after SRS are damage to spiral ganglion neurons inside the modiolus and ischemic insult to the primary afferent and efferent nerve fibers within the internal auditory canal.[52] Therefore, modulation of the radiation dose to the cochlea below 4 Gy and appropriate shielding techniques employed for protection of the cochlea are reasonable options that may help in decreasing the incidence of hearing loss after SRS.

Hearing Outcomes After Microsurgery. Recently, Sughrue et al.[100] performed a systematic review to highlight the hearing preservation rates after microsurgical re-

TABLE 4: Summary of results of prospective studies comparing microsurgery and radiosurgery in small VSs (< 3 cm)*

Study & Details
Régis et al., 2002
no. of pts: SRS 97, MS 110
tumor size: Koos Stage II & III
FU: 4 yrs
functional hearing preservation (GRC 1 or 2) in pts w/ GRC 1 hearing preop: SRS 70%, MS 37.5%
(overall functional hearing preservation rate [GRC 1 or 2]: SRS 40%, MS 5%, p < 0.0001)
intact facial motor function postop: SRS 100%, MS 53%, p = 0.00005
ocular problems postop: 27% in SRS group vs 83% in MS group (p < 0.0001)†
hypesthesia: SRS 4%, MS 29%, p = 0.0009
no return to work: SRS 1%, MS 34%, p = 0.00016
change in daily life postop: SRS 9%, MS 39%, p = 0.00017
% of pts suffering from postop tinnitus, vertigo, & imbalance: no statistically significant difference btwn SRS & MS groups
Pollock et al., 2006
no. of pts: SRS 46, MS 36
tumor size: <3 cm
FU: mean 42 mos (range 12–62 mos)
AAO-HNS Class A or B hearing preservation at last FU: SRS 63%, MS 5%, p < 0.001
AAO-HNS Class A preservation at last FU: SRS 50%, MS 0%, p < 0.01
HB Grade 1 facial movement preservation at last FU: SRS 96%, MS 75%, p < 0.01
Dizziness Handicap Inventory score: SRS 8.4%, MS 16.5%, p = 0.02
% of pts w/ postop tinnitus & headache & associated changes in QOL: no statistically significant difference btwn SRS & MS groups
QOL: significantly better overall QOL in SRS group than MS group at 3 mos, 1 yr, & 3 yrs
Myrseth et al., 2009
no. of pts: SRS 60, MS 28
tumor size: <25 mm
FU: 2 yrs
GRC 1 hearing preservation: SRS 68%, MS 0%, p = 0.0009
HB Grade 1 facial nerve function preservation: SRS 98.3%, MS 53.6%, p = 0.0009
QOL: measured by GBI at 2 yrs, significantly better in SRS group than MS group
% of pts w/ postop tinnitus, vertigo, & unsteadiness: no statistically significant difference btwn SRS & MS groups
Sughrue et al., 2010‡
no. of pts followed prospectively in the MS group: 204 (tumor diameter <3 cm in 129)
FU: median 10.2 yrs
AAO-HNS Class A or B hearing preservation at 10 yrs: SRS 93%, MS 68%
HB Grade 1 or 2 facial nerve function preservation at 10 yrs: SRS 100%, MS 76%
freedom from any tumor growth in first 10 yrs: SRS (<13 Gy) 82%, MS 89%
freedom from need for add'l therapy: SRS 96%, MS 91%
freedom from any tumor growth in first 20 yrs: MS 86% (20-yr data not available for SRS)

* add'l = additional; FU = follow-up; GBI = Glasgow Benefit Inventory; HB = House-Brackmann; MS = microsurgery; QOL = quality of life.

† Also reported by Tamura et al.

‡ Sughrue et al. compared their data collected prospectively for patients undergoing microsurgery with their previously published data on radiosurgery in young patients.

section of VS. In the selected 49 published articles, which included 998 patients, an overall hearing preservation rate of 52% was reported. Interestingly, the breakdown of the results according to tumor diameter revealed hearing preservation in the different subgroups as follows: 64% in patients with tumors less than 1 cm in diameter; 61% in those with tumors 1–1.5 cm in diameter; 32% in those with tumors 1.5–2 cm in diameter; and 37% in those with tumors 2–2.5 cm in diameter ($p < 0.0001$). A very significant drop in hearing preservation was observed when the tumor diameter increased beyond 1.5 cm. Comparison of the largest systematic reviews of pooled data to date shows that hearing preservation rates achieved by microsurgery in patients with tumors less than 1.5 cm in size are similar to those observed in the literature for SRS. Nonetheless, the results of these systematic reviews are based largely on Level 3 or 4 evidence and should be interpreted cautiously. The prospective comparative studies that compare the results of small VSs treated with SRS or microsurgery at a single institution provide probably the best available evidence from

which conclusions can be drawn (Table 4), suggesting that SRS is the best management strategy, considering CN VII and VIII preservation rates and postprocedural quality of life. Moreover, similar results have been demonstrated by other retrospective studies.[46,63,78,79,84,113] That being said, the results of a prospective nonrandomized comparative study (Level 2 evidence) are contrary to the aforementioned one.[24] At almost 3 years' follow-up, there was no statistically significant difference across the various health dimensions pertaining to quality of life (SF-36 questionnaire) for the observation, SRS, and microsurgery groups. Similar results were reported in a multicenter cross-sectional study with a maximum follow-up of no more than 5 years.[14] One must remember that long-term follow-up data (> 10 years) in an SRS group are still not widely available.

Koos et al.[50] reported remarkable results in their group of patients with small VSs treated by microsurgery. Good hearing (using the 50/50 rule) was maintained in 100% of patients with Koos Stage 1 tumors and in 87% of patients with Koos Stage 2 tumors. Of patients presenting initially with House-Brackmann Grade 1 or 2 facial nerve function, 88% had House-Brackmann Grade 1 function 18 months postoperatively. These results have not been widely reproducible.

To summarize, if a young or middle-aged patient presents with serviceable hearing loss, following a wait-and-scan strategy is unlikely to do any good with respect to the ongoing active tumorigenesis and the patient's hearing status. Considering the chances of stabilizing or improving hearing status, freedom from future treatment at 10 years' follow-up, and the quality of treatment, the literature suggests that such patients may benefit the most from SRS. Since data for 20-year follow-up are not widely available for SRS (as opposed to microsurgery), one would have to clearly inform patients of the potential long-term failures that have not yet been well described.

Hypothetical Case 3

> This patient is 40-year-old woman with a small VS (< 25 mm) and a serviceable hearing level. The patient works as a newscaster or a professional model, and therefore, apart from hearing preservation, her primary concern is to maximize the chances of preservation of normal facial movement.

Facial Nerve Outcomes After Microsurgery. A recent systematic review addressing facial nerve outcomes after microsurgery examined data from 79 articles pertaining to a total of 11,873 patients.[102] Although the inclusion criteria were not specific for tumor size, the authors were able to compare the facial nerve preservation rates in 2 subgroups: studies with a mean tumor size < 20 mm (90%) and studies with a mean tumor size > 20 mm (67%). A previous meta-analysis done in 2003 yielded similar results.[113] In the studies representing Class II evidence for treatment of small VSs, the postoperative rate of facial nerve preservation ranged from 53% to 76% (Table 4), remarkably worse than the results reported in systematic reviews that largely represent data from retrospective case series.[102] It is evident that smaller VSs treated with microsurgery have better hearing and facial nerve preservation rate than larger VSs (Fig. 2). Class II or higher evidence for intracanalicular VS managed through microsurgery is lacking. Recently, Noudel et al.[70] conducted a systematic review that included 35 studies of microsurgical resection of intracanalicular VS. They reported that the average hearing preservation rate (AAO-HNS Classes A and B or GRC I and II) for intracanalicular VS was 58% with a retrosigmoid approach and 62% with a middle fossa approach. Interestingly, facial nerve preservation (House-Brackmann Grade 1 or 2) was as high as 91% in the retrosigmoid approach group as compared with 77% in the middle fossa approach group, although the difference was not statistically significant ($p > 0.05$).

Facial Nerve Outcomes After SRS. Recently, Yang et al.[116] quantified the existing data to evaluate facial nerve preservation after SRS, and the results correlated with what one would intuitively expect. The authors reported an overall facial nerve preservation rate of 96.2%; for the population with tumors of less than 1.5 cm^3 in volume, the rate was 99.5%. They also found that reduction of the radiation dose to less than 13 Gy was associated with significant improvement in facial nerve preservation rates. In all but one of the comparative prospective studies, facial nerve preservation rates were much higher in the SRS treatment groups than in the microsurgery groups. The one exception was the study by Di Maio and Akagami,[24] who reported no statistically significant difference in the incidence of facial weakness between the microsurgery and SRS groups.

To summarize, in a young or middle-aged patient who presents with serviceable hearing and in whom preservation of facial function is crucial (for example, in someone who works as a newscaster or professional model), the literature suggests the superiority of SRS over observation or microsurgery.

Hypothetical Case 4

> This patient is a 40-year-old man with a small VS (< 25 mm) and with a nonserviceable hearing level. He wants the best chance to stabilize or restore hearing function and emphasizes his desire to choose an intervention that will provide him with best quality of life.

Currently the data in the literature are equivocal with respect to the choice between microsurgery and SRS for patients with small VSs who present with a nonserviceable hearing level. In patients who have already lost serviceable hearing it is worthwhile to consider tumor control and treatment-related morbidity factors that influence quality of life. In patients presenting with profound hearing loss, comparative prospective studies have not found any statistically significant difference among those undergoing SRS and those undergoing microsurgery in terms of quality of life.[36] Whitmore et al.[111] recently reported the results of a comprehensive quantitative, statistically driven study designed to determine which treatment modality yields the best quality of life at 5- and 10-year follow-up in patients with VSs smaller than 25 mm in diameter who present with some residual hearing. Even though the study was limited by the assumptions the authors had to make to come up with the best possible comparative model, it provided useful insights into treatment-related morbidity

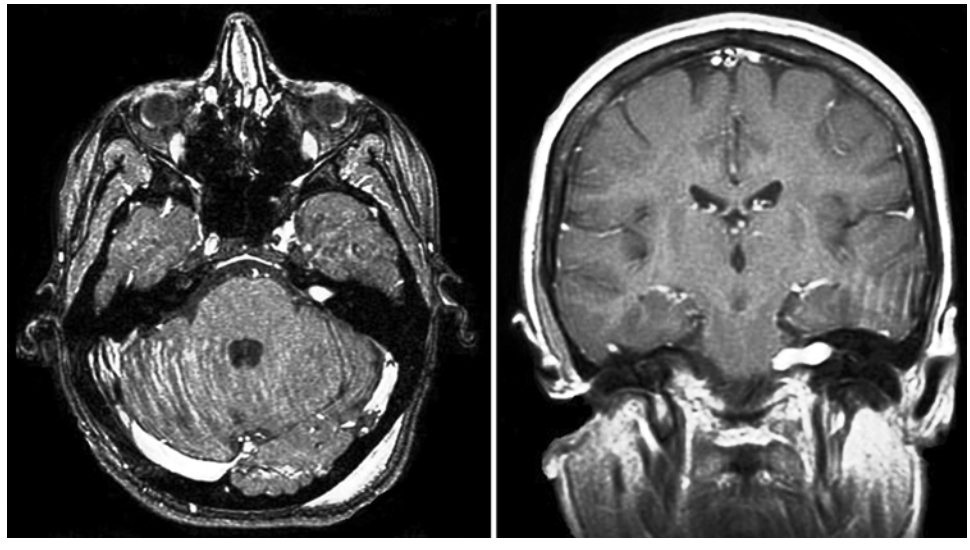

Fig. 2. Contrast-enhanced T1-weighted MR images demonstrating small VSs with diameters of 1 cm **(left)** and 2 cm **(right)**. Systematic review of mostly Class 3 evidence suggests that the chances of facial nerve preservation after microsurgery is significantly higher in tumors 1.5 cm or less in diameter than in those with a diameter of more than 1.5 cm.

parameters and their influence on postoperative quality of life. The authors concluded that at 5 years posttreatment, patients who underwent radiosurgery had a better quality of life than those treated with either microsurgery or managed with observation. At 10 years, quality of life in both the radiosurgery and microsurgery groups was significantly better than in the observation group, but there was no statistically significant difference in the quality of life between the SRS and microsurgery groups. The authors also found that hearing loss had a greater overall negative impact on quality of life than other morbidities, including facial weakness and numbness. This finding was also documented by a previous prospective study[24] (Level 2 evidence). Some studies have also shown that disabling imbalance has the maximum negative impact among complications involving CN VII and VIII (that is, audiofacial complications, including facial nerve palsy and hearing loss) and nonaudiofacial complications. In these studies, microsurgical resection was associated with the alleviation of the disability and improvement in quality of life for the majority of patients.[55]

Based on the current review of the literature, in patients presenting with a nonserviceable hearing level, SRS does not seem to provide any additional advantage over microsurgery. There is no significant difference in the quality of life; incidence of nonaudiofacial complications, including tinnitus, vertigo, and unsteadiness; and tumor control among the 2 treatment modalities, although there is a trend toward better results in patients complaining of imbalance in the microsurgery group. Some authors have reported comparable results of facial nerve preservation with microsurgical excision (especially for tumors less than 1.5 cm in diameter).[24,87,102] Certainly, surgical experience is an important factor that needs to be taken into account when analyzing and comparing outcomes of surgical treatment of small VS. At the hands of some experienced surgeons, facial nerve outcomes are excellent in patients treated for small VS, and it is worthwhile to consider microsurgery as a safe alternative in patients with nonserviceable hearing.[24,87,102]

Also, in a patient whose hearing status is out of the equation, the small but real risk of radiation-induced malignancy may be considered (especially if the patient is young).[23,90] Additionally, Di Maio et al.[24] showed in their prospective study that patients in the observation and radiation groups may have a greater psychological burden associated with harboring a tumor than those treated with surgery. Therefore, when improvement in hearing status is unlikely, microsurgery is certainly a valid treatment option.

Hypothetical Case 5

> This patient is a 65-year-old man (or other adult with significant comorbidities), with a small VS (< 25 mm) and serviceable hearing loss. He seeks advice regarding the best possible way to stabilize the hearing loss and maintain an optimal quality of life for the coming years.

There seems to be a general consensus on the treatment strategy for these patients in the literature. If the patients' symptoms are minimal, the support for observation with serial imaging is immense. There is a great chance in this age group that active treatment may not be necessary.[39,110] Furthermore, in the studies that address long-term follow-up for conservatively managed cases, patients 65 years or older represent a substantial percentage in the cohort.[12,39] For patients in this age group who are symptomatic or in whom rapid tumor growth (> 2.5 mm/year) is documented, SRS seems to provide the best audiofacial outcomes and postoperative quality of life (Table 4). The nonaudiofacial postoperative morbidities, such as dysequilibrium, vertigo, and tinnitus, can usually be managed by appropriate rehabilitative services.

Special Considerations

Sudden Hearing Loss in Small VS

The percentage of cases in which patients present

with sudden hearing loss is relatively low (0.8%–3.7%).[15] The chances of restoration of sudden hearing loss in VS patient through radiosurgery are not well documented. Most of the authors studying sudden hearing loss describe their treatment protocol as a combination of microsurgery and medical management with corticosteroid agents and vitamin B_{12}.[32,42,88] Friedman et al.[32] reported measurable hearing preservation after microsurgery in 73% of their patients with VSs less than 2 cm in diameter who presented with sudden hearing loss. Even though there remains a small chance of measurable-to-complete recovery with just observation or with medical therapy alone, microsurgery may provide the best chance of stabilizing or improving hearing in patients with small VSs presenting with sudden hearing loss.[11,32,42,65,88]

Cystic Small VS

Cystic lesions account for approximately 4%–15% of all VS cases.[22,76] They are characterized by rapid growth and therefore may result in a shorter duration of symptoms related to the vestibulocochlear and facial nerves and may also be seen in certain cases of small VS. Management of small VS with a cystic component is contentious.[10,16,17,22,33,74,92,94,107] There is a severe lack of comparative data (even Level 3 or 4 evidence) to support the superiority of surgery or SRS or radiotherapy as a treatment strategy. Treatment of these lesions by means of either microsurgery or SRS is challenging and fraught with risks. Enlargement of the cyst following radiation, even after several years, has been noted to occur, and in some cases emergent surgical decompression may be required to avoid permanent cranial neuropathy.[22,35,74,92] Results of surgery, however, are also known to be unfavorable for cystic VS, and surgical treatment is associated with higher rates of morbidity.[10,17,33,94,107] At this point, the data in support of SRS or radiation therapy for treatment of cystic VS are too scant to even consider these treatments as reasonable alternatives to surgery.[92] That being said, SRS and radiotherapy as well as surgery would have to be evaluated with better study designs and longer follow-up.

Syndrome-Associated Small VSs

Apart from being sporadic, VSs are known to develop in patients with neurofibromatosis Type 2. They may be unilateral or bilateral. Various studies have highlighted their unpredictable and unique natural history, growth patterns, and treatment response.[1,25,29] Discussion of neurofibromatosis Type 2 is beyond the scope of this article.

Limitations of this Review

Comprehensive systematic analyses currently existing in the literature have a restricted clinical applicability due to the numerous inconsistencies, selection biases and lower level of evidence. For this reason, this review also has its limitations in suggesting a standard algorithm based on the limited higher form of evidence, even though every effort was made to customize answers to patient-specific requirements based on the level of evidence existing in the literature.

Conclusions

Even after more than 100 years since the first VS surgery, the controversy in treatment strategy remains. More large-scale prospective randomized trials may help us to better understand the "best" therapeutic option for certain patient scenarios. To achieve statistical significance for comparing outcomes for observation, radiosurgery and microsurgery for small vestibular schwannomas, a cohort of over 900–1000 patients followed for over 10 years is required. The logistics of obtaining the required sample size is challenging. Due to the paucity of high-level evidence to compare the treatment groups, proponents of each option will presumably stick with their current preference. We have attempted to extrapolate from the literature to specific patient scenarios. The existing evidence suggests that the treatment of small VS should always be tailored to the individual patient rather than being a generalized treatment protocol. We hope that our case-specific review may serve as a guide for neurosurgeons, neuro-otologists, and radiation oncologists treating this ever-increasing patient population and may encourage focused prospective randomized trials.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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The approach to the patient with incidentally diagnosed vestibular schwannoma

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With the increasing prevalence and decreasing cost of MRI scans, incidental discovery of vestibular schwannoma (VS) has become more common. Scarce literature exists regarding management of the tumors in those patients with incidentally discovered VSs, and clear guidelines for management do not exist. In this review, the authors examine the available literature for insights into management of incidentally diagnosed VS and provide an algorithm for their management.
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Key Words • **incidental vestibular schwannoma** • **natural history** • **management**

The majority of patients in whom VS is diagnosed present with symptoms including hearing loss, tinnitus, disequilibrium, or vertigo; rarely they may also present with symptoms of fifth or seventh cranial nerve dysfunction, or with symptoms referable to hydrocephalus such as headache. With the increasing availability and decreasing cost of MRI scans, the "incidental" discovery of VS may become more common.[18] We use the term "incidental diagnosis" of VS here to refer to the diagnosis of VS in asymptomatic patients who undergo radiological imaging for reasons other than a suspected VS. By definition this excludes patients undergoing MRI for asymmetrical hearing loss or dizziness, but does not exclude patients with subjectively "normal" hearing who nevertheless have audiometric evidence of mild hearing loss.

Although a relatively large body of literature exists concerning the natural history of patients with VS with regard to hearing and tumor progression (including those with good hearing), few studies look specifically at tumors that are diagnosed incidentally.[10,12,18] These patients therefore present a challenge to the surgeon, in that recommendations made to the patient are at best extrapolations from data describing mixed populations of both asymptomatic and symptomatic patients with "good hearing." At the present time, however, there is insufficient evidence to suggest that incidentally discovered tumors behave differently from those in patients with minimal symptoms and/or minimal hearing loss.

Abbreviations used in this paper: ABR = auditory brainstem response; IAC = internal auditory canal; RVR = reduced vestibular response; SDS = speech discrimination score; SRS = speech recognition score; VS = vestibular schwannoma.

The goal of the present paper is to assist the surgeon in counseling the asymptomatic patient with VS. For purposes of patient counseling we summarize the evidence available about the natural history of VS (with regard to growth and hearing loss) and the implications of tumor size, hearing status, and other prognostic factors for predicting tumor behavior and hearing preservation of the incidentally diagnosed VS. We conclude with an algorithm for the management of incidental VS based on these considerations.

Methods

Literature Review

A systematic search was performed using the PubMed and MEDLINE databases to identify papers dealing with an incidentally found acoustic neuroma or VS. We first searched the databases by going to the main site and performing a search using the key words "Acoustic Neuroma Incidental," which yielded 23 results. We then limited these results to English-language articles, and this narrowed it down to 21 papers. To ascertain that all papers on the subject were found, we alternatively searched using the key words "Vestibular Schwannoma Incidental," and this resulted in 26 articles. After limiting this search to English-language articles, it yielded 23 papers. To make our search as comprehensive as possible and to ensure that we would not to miss any relevant articles, we therefore alternately used the key words "Vestibular Schwannoma Asymptomatic" and "Acoustic Neuroma Asymptomatic," which returned 57 and 54 articles, respectively. After transferring all of these articles to an EndNote library, we came to a total of 78 articles.

To further ensure that we found all articles on the topic, a search of MEDLINE for articles published between 1948 and April 2012 was performed. The key words "Acoustic Neuromas" and "Incidental" (7 results), "Vestibular Schwannoma" and "Incidental" (0 results), "Acoustic Neuroma" and "Asymptomatic" (16 results), and "Vestibular Schwannoma" and "Asymptomatic" (5 results) were used for the query. Four additional articles were identified in these results but yielded papers not written in English, so they were excluded from our analysis. Next, we systematically reviewed the remaining 78 articles to determine which papers met our criteria for discussing incidental VSs. We excluded case reports and papers that analyzed patients who presented with symptomatic VSs. We eventually narrowed these 78 articles to the 9 papers ultimately used in our analysis.

The quality of evidence in the selected articles was categorized according to the US Preventive Services Task Force criteria for ranking evidence (Table 1).[9] Articles were reviewed for data on methodology (retrospective vs prospective), number of patients, tumor size on discovery, and presenting symptoms, if applicable.

Results

As stated above, we use the term "incidental" VS to refer to lesions that are identified in asymptomatic patients who undergo radiological imaging for reasons other than suspected VS. Some have suggested that this group of patients is more appropriately referred to as "asymptomatic at presentation," because these patients may acknowledge a symptom that they have ignored but that is attributable to the VS.[18]

A total of 9 studies were identified that specifically included incidental tumors. Case reports were excluded; thus all the studies were retrospective case series. Table 2 shows the data for the retrospective case series on incidental VS.[2,10,12,13,16,19,23,26,27] The studies did not consistently describe how the workup was conducted, but of the ones that did, there seemed to be a trend toward a higher likelihood of MRI being the definitive modality. Most of the patients were older than 50 years of age. The average tumor size was 13 mm at the time of detection.

TABLE 1: Hierarchy of research design*

Level of Evidence	Description
I	evidence obtained from at least 1 properly randomized controlled trial
II-1	evidence obtained from well-designed controlled trials w/o randomization
II-2	evidence obtained from well-designed cohort or case control analytic studies, preferably from >1 center or research group
II-3	evidence obtained from multiple time series w/ or w/o the intervention—dramatic results in uncontrolled experiments (such as the results of the introduction of penicillin treatment in the 1940s) could also be regarded as this type of evidence
III	opinions of respected authorities, based on clinical experience, descriptive studies & case reports, or reports of expert committees

* System proposed by Harris et al.

Investigators have attempted to quantify the frequency of incidental VS. Studies based on autopsy reports have noted an incidence as high as 1%.[18] Recently, Lin and colleagues[12] attempted to estimate the prevalence of incidental VS from an intracranial MRI database of 46,414 patients and noted a 0.02% estimated prevalence of incidental VS. The authors suggest that, whereas incidental VS may be less prevalent than autopsy studies suggest, incidental VS may actually be more prevalent than suggested by epidemiological studies. Similarly, recent studies in Denmark estimated the incidence of VS to be 19.4 VSs per million per year as of 2008,[23] whereas another recent study estimated the prevalence of VS in asymptomatic patients at 0.2%.[28]

Stangerup and Caye-Thomasen[21] noted that the annual number of diagnosed cases of VS in Denmark rose from 7.8 cases per 1 million per year in 1976 to 23 cases per 1 million per year in 2004 in a population of 5.4 million. Over this entire period, tumor size at diagnosis declined from a median of 35 mm to 10 mm in 2009, and the median age of patients at diagnosis slowly increased from 50 to 60 years. The authors suggest that if decreasing tumor size and increasing incidence of VS was the result of easier access to MRI units, then the median age at time of diagnosis would be expected to decrease simultaneously. The authors suggested that easier access to MRI units resulted in more elderly patients being offered an MRI study. This study confirms that incidental VS is becoming an increasingly more prevalent diagnosis and highlights the need to have a more rational approach to management.

Review of the Literature

Evidence-Supported Guide for Discussion With Patients With Asymptomatic VS

The statements below are supported by our review of the literature and are intended as evidence-based "talking points" for discussion with patients receiving a diagnosis of asymptomatic VS. These statements are summarized in Table 3.

Approximately Two-Thirds of Tumors Do Not Grow During an Observation Period of Approximately 5 Years (Class III). An understanding of the natural history of VS aids management decision making for incidental VS. Nikolopoulos and colleagues[15] have recently reviewed the literature for VS growth and found that as much as 75% of tumors (range 6%–75%) exhibited no growth during the follow-up period (the mean growth rate in millimeters/year ranged from 0 to 10.3 mm/year, and mean follow-up ranged from 19 months to 5.5 years). Fucci and colleagues[6] at the House Clinic reported on 119 VSs that were observed over a mean duration of 2.5 years (range 5 months–8 years) with serial MRI studies, noting that 36 tumors (30%) grew > 2 mm during the observation period. Recently, Agrawal and colleagues[1] reported on 180 patients with VSs, noting that 65 patients (36%) exhibited growth of their tumor, defined as ≥

TABLE 2: Review of the literature for incidentally diagnosed VS*

Authors & Year	Study Design	No. of Cases	No. of VSs	Mean Age	Avg Tumor Size at Detection	Presenting Sx/Reason for Imaging Study†
Lustig et al., 1998	retro	546	4	NA	NA	1) eval for breast cancer mets; 2) MS; 3) slurred speech; 4) weakness
Tos et al., 1999	retro	NA	970	NA	NA	NA
Anderson et al., 2000	retro	24,246	17	56 yrs	8 pts <1 cm; 6 pts 1–2 cm; 3 pts >2 cm	NA
Nutik & Babb, 2001	retro	433	NA	54.0 yrs (M), 54.9 yrs (F)	13.4 mm (M), 16.2 mm (F)	NA
Lin et al., 2005	retro	46,414	9	58 yrs	13.8 mm	1) seizures; 2) contralat HFS; 3) juvenile angiofibroma; 4) breast cancer mets; 5) Horner syndrome; 6) SDH; 7) dizziness; 8) TIA
Smouha et al., 2005	meta-analysis	1,345	7	62 yrs	11.8 mm (900 of 1,345 pts)	
Stangerup et al., 2010	retro	2,283	NA	55.4 yrs	16.4 mm	NA
Varughese et al., 2012	retro	4.8 mil	178	NA	0.71 cm^3	1) balance problems (58 of 178); 2) tinnitus (129 of 178); 3) vertigo (77 of 178); 4) hearing: Gr A (52), Gr B (50), Gr C (20), Gr D (54), 2 missing data
Jeyakumar et al., 2007	retro	121	15	55.7 yrs (symptomatic grp), 52.8 yrs (incidental grp)	1.5 cm (symptomatic grp), 1.1 cm (asymptomatic grp)	1) neck & ear pain; 2) blurry vision; 3) exotropia; 4) family Hx of brain aneurysms; 5) head trauma; 6) contralat ear sudden hearing loss; 7) migraine HAs; 8) visual changes, ataxia, contralat ear hearing loss, ipsilat deafness since birth; 9) contralat hearing loss, HAs, bilat ear pressure; 10) parkinsonism; 11) periorbital numbness, HAs; 12) lt facial twitching & numbness, fatigue, HAs, lt hand tremor, ataxia, visual disturbances, tinnitus, MRI done because demyelinating disease suspected; 13) migraines; 14) sinusitis, long-term ipsilat hearing loss thought to be due to noise damage (gunfire); 15) tremor, unsteady gait

* The evidence level in all studies was Grade III according to the US Preventive Services Task Force system (see Table 1). Abbreviations: avg = average; eval = evaluation; Gr = Grade; grp = group; HA = headache; HFS = hemifacial spasms; Hx = history; mets = metastasis; mil = million; MS = multiple sclerosis; NA = not available; pts = patients; retro = retrospective; SDH = subdural hematoma; Sx = symptoms; TIA = transient ischemic attack.

† Numbers followed by parenthesis represent the cases of VS.

TABLE 3: Summary of evidence statements*

Evidence Statement	References
Approximately 2/3 of tumors do not grow during an observation period of approximately 5 yrs	1, 6, 15, 27
Approximately 50% of patients may maintain their hearing during an observation period of approximately 5 yrs	22
Tumors exceeding 1.5–2 cm in maximal dimension have a higher probability for growth	1, 6, 10, 15, 20, 23
Initial hearing loss, even small degrees of loss, may predict a greater chance of loss of good hearing over time	8, 17, 21
The presence or absence of a normal RVR & ABR, presence of fundal fluid, & tumor size may help one to counsel patients on prognosis	4, 7
Approximately 2/3 of patients will retain serviceable hearing following hearing preservation surgery	5, 11, 28
Approximately 50% of patients will retain serviceable hearing following radiosurgery	29

* The evidence level in all studies was Grade III according to the US Preventive Services Task Force system (see Table 1).

1 mm per year in maximal tumor dimension, over a mean follow-up of 32 months. Not unexpectedly, Agrawal and colleagues also noted that larger tumor size at presentation was associated with a greater likelihood of tumor growth in the future, with a 1-mm increase in tumor size increasing the risk of tumor growth by 20%. The available studies regarding the natural history of VS growth have been extensively reviewed by Nikolopoulos and colleagues.[15] Importantly, occurrence of growth within the 1st year may be an early indicator of the need for active intervention.[29]

Approximately 50% of Patients May Maintain Their Hearing During an Observation Period of Approximately 5 Years (Class III). Stangerup and colleagues[23] examined the long-term hearing preservation of 932 patients in Denmark with VS who underwent a "wait and scan" approach. Of these patients, 178 possessed American Association of Otolaryngology–Head and Neck Surgery Class A hearing at diagnosis, with 91 patients (51%) maintaining Class A hearing during the observation period (mean observation period 4.7 years, range 0.5–21 years). Similarly, approximately 54% of patients with Class A or B hearing at diagnosis maintained Class A or B hearing during the observation period. Furthermore, of the 491 patients with good hearing (defined as an SDS of ≥ 70%) in the tumor-affected ear at diagnosis, 290 patients (59%) maintained good hearing at the last evaluation, with a mean of 4.7 years of observation.

Tumors That Exceed 1.5–2 cm in Maximal Dimension Have a Higher Probability for Growth (Class III). As Nikolopoulos and colleagues[15] have discussed, the pattern of VS growth can be highly variable, with periods of stability alternating with periods of growth, instead of steady growth with time. In addition, no reliable predictors of tumor growth have been borne out by all studies, with the exception that some studies have found large initial tumor size to be a predictor of future growth.[1,6,20,25] Although incidentally discovered VSs tend to be smaller than symptomatic VSs, the presence or lack of symptoms may not necessarily predict initial tumor size.[1,10]

Initial Hearing Loss, Even Small Degrees of Loss, May Predict a Greater Chance of Loss of Good Hearing Over Time (Class III). Stangerup and colleagues[22] noted that, whereas 81% of patients with 100% SDS at diagnosis (108 patients) maintained good hearing, 55% of patients with even small (1%–10%) initial speech discrimination loss at diagnosis (78 patients) maintained good hearing over a mean observation time of 3.9 years. This effect was even further magnified in patients with an SDS of 70%–79% at diagnosis (64 patients), with only 38% maintaining good hearing. This study is supported by Remenyi and colleagues,[17] who noted that patients with a good initial SRS fared better with respect to hearing preservation, in contrast to patients with a poor initial SRS, with good and poor being defined as an SRS consistent and inconsistent with degree of sensorineural hearing loss by using Arthur Boothroyd word lists. The likelihood of loss of good hearing over time with even a small (1%–10%) decline in SDS is not insignificant, with > 50% of patients losing good hearing over the observation period. This becomes more significant as the initial SDS score declines. Finally, hearing loss may continue to progress despite lack of tumor growth.[8] In addition to being important in counseling patients, this information helps the surgeon begin to build a framework for management of the incidentally discovered VS.

Abnormal ABR Latency, Inferior Vestibular Nerve Origin, and Opacification of the Fundus of the IAC by Tumor are Poor Prognostic Indicators for Hearing Preservation (Class III). The use of ABR and electronystagmogram in addition to MRI studies may allow for prognostication of hearing preservation in select patients who choose to proceed with a hearing preservation microsurgical approach. Specifically, Brackmann and colleagues[4] reported that in addition to better preoperative hearing, a short intraaural wave V or absolute wave V latency on ABR and tumor origin from the superior vestibular nerve on MRI studies were associated with higher rates of hearing preservation in patients undergoing middle fossa craniotomy resection for VS. It has been proposed that a hypoactive RVR may predict superior vestibular nerve origin. However, Brackmann and colleagues did not find the presence of a hypoactive RVR to be significantly correlated with a greater likelihood of hearing preservation. However, a normal RVR in a patient whose imaging does not clearly delineate the origin of the tumor may portend a lower likelihood of hearing

preservation, or at least heighten suspicion for an unusual anatomical configuration. Goddard and colleagues[7] noted that the presence of fundal fluid and tumor origin from the superior vestibular nerve were predictive of better hearing outcomes, with both factors having a higher correlation with preservation of the patient's American Association of Otolaryngology–Head and Neck Surgery hearing class and with preservation of serviceable hearing. These prognostic factors aid in counseling patients about surgical treatment and may aid in convincing an appropriately selected patient to proceed with microsurgical resection.

Approximately Two-Thirds of Patients Will Retain Serviceable Hearing Following Hearing Preservation Surgery (Class III). Friedman and colleagues[5] reported long-term (5 years) preservation of serviceable hearing in 16 (70%) of 23 patients who underwent middle fossa resections for intracanalicular tumors (1.1 ± 0.4 cm; mean ± SD) who exhibited serviceable hearing in the immediate postoperative period. Woodson and colleagues[30] reported on 23 (88%) of 26 patients with VS who underwent middle fossa resection and who maintained a word recognition score of > 70% at > 5 years of follow-up. Woodson and colleagues concluded that initial postoperative findings are predictive of long-term hearing results. Kutz and colleagues[11] recently reported serviceable hearing preservation in 24 (63.2%) of 38 patients with preoperative serviceable hearing. These authors noted that patients with VSs ≤ 10 mm compared with patients with VSs > 10 mm were more likely to exhibit serviceable hearing preservation (73.3% vs 25%). Although these surgical results represent the outcomes from some experienced groups and cannot necessarily be generalized to all surgeons, these results do give a sense of what is possible and achievable in appropriately selected patients.

Approximately 50% of Patients Will Retain Serviceable Hearing Following Radiosurgery (Class III). In a recent meta-analysis, preservation of serviceable hearing, defined as a speech reception threshold < 50 dB and an SDS > 50% occurs in approximately 51% of patients following stereotactic radiosurgery, regardless of radiation dose, tumor size, or patient age.[31] This meta-analysis of 4234 patients identified 1322 who had serviceable hearing prior to stereotactic radiosurgery for VS. This rate is similar to the natural history of hearing loss in sporadic VS. Stereotactic radiosurgery in this meta-analysis consisted of single-shot treatment with a mean margin dose of 14.2 ± 2.4 Gy (range 11.5–21.5 Gy). The mean tumor size, for those studies in which it was available, was 3.9 cm. The mean follow-up was 44.4 ± 35 months (the mean margin dose and follow-up are expressed ± SD). In this meta-analysis, 542 patients received an average radiation dose of ≤ 13 Gy, and 671 patients received an average radiation dose of > 13 Gy. The lower-dose (≤ 13 Gy) and higher-dose (> 13 Gy) groups had hearing preservation rates of 60.5% and 50.4%, respectively (p = 0.0005).

Discussion

Management Considerations

We propose an algorithm for management of the incidentally diagnosed VS (Fig. 1).

Given the natural history of growth and the greater likelihood of smaller tumors in patients with incidentally discovered VS, an initial period of observation to assess for tumor growth is a reasonable initial treatment option. Consideration should be given to the patient's preferences, including but not limited to hearing preservation. If observation is the elected initial approach after a full discussion with the patient, assessment with serial MRI studies is recommended. Despite the fact that some studies indicate that growth usually occurs within the first 5 years of observation, monitoring MRIs beyond this time period is necessary because these tumors can continue to grow slowly and/or unpredictably over time.[10]

Subsequent decision making depends on a combination of factors, including but not limited to patient symptomatology, significant tumor growth (> 2 mm/year), and patient preferences. Initial workup in addition to imaging should include an audiogram and may include an ABR and videonystagmography study for prognostication and counseling purposes. Whereas the mean tumor growth rate varies 1 and 2 mm/year for all tumors and 2–4 mm/year for those tumors that grow, some tumors may exhibit exceptional growth that exceeds 18 mm/year.[15] Furthermore, Mick and colleagues[14] have noted that the majority of tumors identified as growing continue to grow on subsequent observation.Demonstrated significant growth (> 2 mm/year) is the rationale for intervention, either by a microsurgical or radiosurgical approach, according to many studies.[1,3,25]

After follow-up imaging, tumors are divided into subgroups of stable and growing tumors. Tumors that presented incidentally may also of course become symptomatic over time. We believe that treatment is indicated for incidentally discovered tumors that become symptomatic or exhibit significant growth. However, the timing of intervention depends not only on initial tumor size and demonstrated growth or symptoms, but is also influenced by the patient's age and general medical condition as well as the patient's and surgeons' preferences.

Parameters for Intervention

Stable Tumors. Incidental tumors with relative stability (≤ 2 mm/year) on serial imaging that remain asymptomatic may safely continue to be observed; microsurgery may be offered in patients who begin to experience progressive onset of symptoms, including progressive hearing loss or imbalance for example.

Growing Tumors. Intervention is indicated in those tumors that exhibit significant growth on follow-up imaging (> 2 mm/year). These patients may be treated with either microsurgery or radiosurgery, depending on such factors as tumor size, patient age, and hearing status. Patients without hearing who have a growing VS and who elect microsurgical resection should be offered a resection via the translabyrinthine approach. Patients with serviceable hearing can be offered a hearing preservation approach. For small tumors ≤ 1.5 cm, in general we favor the middle fossa approach for microsurgical resection. With respect to the retrosigmoid approach, we reserve this approach for smaller tumors with minimal extension into the IAC (less

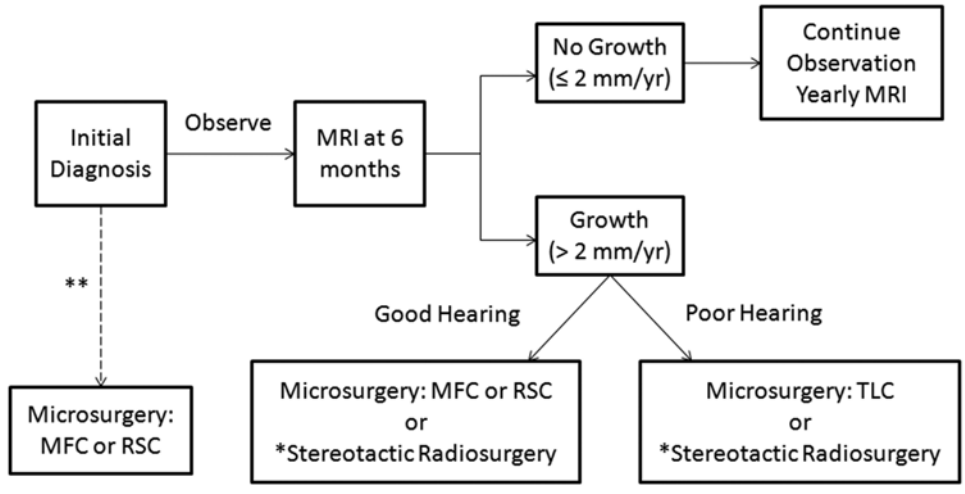

Fig. 1. Algorithm for management of incidental VS. Use of stereotactic radiosurgery *(single asterisks)* can be considered in elderly patients, in patients in poor general medical condition, and in patients expressing a preference for radiosurgery. We do not consider stereotactic radiosurgery a hearing preservation approach. Thus, although we may offer the approach as an option to our patients, the individual is counseled that the likelihood of hearing loss is similar to continued observation. In carefully selected patients, microsurgical resection *(double asterisks)* can be considered without prior observation, with the purpose of preserving good hearing. Patients wishing to preclude future hearing loss are particularly appropriate candidates for this approach. MFC = middle fossa craniotomy; RSC = retrosigmoid craniotomy; TLC = translabyrinthine craniotomy.

than one-half of the proximal IAC) with a predominantly cerebellopontine angle component, in which the patient desires an attempt at hearing preservation. A discussion of the relative advantages and disadvantages of the middle fossa approach versus the retrosigmoid approach for hearing preservation is beyond the scope of this paper.

Microsurgery Versus Radiosurgery. A discussion of the extensive literature on microsurgery and radiosurgery for VS is beyond the scope of this review. Although we favor observation for most incidentally discovered tumors, treatment at the time of initial diagnosis may be reasonable for tumors that at the time of diagnosis are sufficiently large to have mass effect on the cerebellar peduncle and/or brainstem. Most patients with incidental VS will, however, have smaller tumors and desire hearing preservation. Therefore, if treatment is contemplated, consideration for the relative odds of hearing preservation with treatment (that is, microsurgery or radiosurgery) versus observation is advised.

Summary Remarks

Our review of the literature provides evidence on which to inform our recommendations for management of incidentally diagnosed VS. Admittedly, there is a dearth of high-quality evidence (that is, Class I or II) to guide clinical decision making in the treatment of incidental VS; however, we do believe that careful evaluation of the available Class III data can aid in patient counseling. Clearly, there will never be Class I evidence (that is, randomized, blinded data) on which to base these clinical decisions. This review is not intended to be an exhaustive or comprehensive review of the literature—no attempt at meta-analysis has been made, and the data presented should not be interpreted as such. Rather, this is a highly selected review of the literature consisting of high-quality retrospective studies and expert opinion.

Approximately two-thirds of VS tumors do not grow, and approximately 50% of patients maintain their hearing during an observation period of approximately 5 years. Incidentally discovered VSs tend to be smaller, so observation is a reasonable option if a patient follows up with an initial Gd-enhanced MRI study at 6 months and then, as long as no growth occurs initially, MRI studies yearly. With significant growth with or without symptoms, microsurgical resection may be offered as a treatment option. This position is strengthened by a recent report from Sughrue and colleagues[24] in which it was noted that patients who had tumors with growth rates > 2.5 mm per year tended to have worse hearing results versus those who had tumors with growth rates ≤ 2.5 mm per year.

Initial hearing loss predicts a greater chance of loss of good hearing over time and provides support for microsurgical resection via a hearing preservation approach without initial observation, because hearing loss may continue to progress despite lack of tumor growth. Large initial tumor size may be a predictor of future growth, so observation for tumors exceeding 1.5–2 cm is not recommended given the higher probability for growth.

We believe that only significantly growing tumors (> 2 mm/year) should be treated with radiosurgery. Stereotactic radiosurgical treatment carries a small risk of malignant transformation, which must be discussed with patients. Microsurgical resection is generally more difficult after radiation, due to the formation of significant adhesions between the facial nerve and tumor, and it presents increased risk of facial nerve injury.

Conclusions

An understanding of the natural history of VS growth and changes in hearing associated with these tumors over time is essential to the management of the incidentally diagnosed VS. Furthermore, an understanding of the con-

sequences and associated caveats of the various treatment options is an essential part of managing this disease and counseling patients with VSs. Finally, a proper understanding of all the aspects discussed in this paper will enable appropriate management of expectations in patients with incidentally diagnosed VS.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Lekovic, Hoa, Schwartz. Acquisition of data: Lekovic, Hoa, Drazin, Hanna. Analysis and interpretation of data: all authors. Drafting the article: Lekovic, Hoa, Drazin, Hanna. Critically revising the article: Lekovic, Hoa, Drazin, Schwartz. Reviewed submitted version of manuscript: Lekovic, Hoa, Drazin. Approved the final version of the manuscript on behalf of all authors: Lekovic. Study supervision: Lekovic.

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The impact of comorbidities, regional trends, and hospital factors on discharge dispositions and hospital costs after acoustic neuroma microsurgery: a United States nationwide inpatient data sample study (2005–2009)

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Object. Hospitalization cost and patient outcome after acoustic neuroma surgery depend on several factors. There is a paucity of data regarding the relationship between demographic features such as age, sex, race, insurance status, and patient outcome. Apart from demographic factors, there are several hospital-related factors and regional issues that can affect outcomes and hospital costs. To the authors' knowledge, no study has investigated the issue of regional disparity across the country in terms of cost of hospitalization and discharge disposition.

Methods. The authors analyzed the Nationwide Inpatient Sample (NIS) database over the years 2005–2009. Several variables were analyzed from the database, including patient demographics, comorbidities, and surgical complications. Hospital variables, such as bedsize, rural/urban location, teaching status, federal or private ownership, and the region, were also examined. Patient outcome and increased hospitalization costs were the dependent variables studied.

Results. A total of 2589 admissions from 242 hospitals were analyzed from the NIS data over the years 2005–2009. The mean age was 48.99 ± 13.861 years (± SD), and 304 (11.7%) of the patients were older than 65 years. The cumulative cost incurred by the hospitals from 2005 to 2009 was $948.77 million. The mean expenditure per admission was $76,365.09 ± $58,039.93. The mean total charges per admission rose from $59,633.00 in 2005 to $97,370.00 in 2009.The factors that predicted most significantly with other than routine (OTR) disposition outcome were age older than 65 years (OR 2.22, 95% CI 1.411–3.518; $p < 0.001$), aspiration pneumonia (OR 16.085, 95% CI 4.974–52.016; $p < 0.001$), and meningitis (OR 11.299, 95% CI 3.126–40.840; $p < 0.001$). When compared with patients with Medicare and Medicaid, patients with private insurance had a protective effect against OTR disposition outcome. Higher comorbidities predicted independently for OTR disposition outcome (OR 1.409, 95% CI 1.072–1.852; $p = 0.014$). The West region predicted negatively for OTR disposition outcome. Large hospitals were independently associated with higher hospital charges (OR 4.269, 95% CI 3.106–5.867; $p < 0.001$). The West region had significantly higher ($p < 0.001$) mean hospital charges than the other regions. Patient factors such as meningitis and aspiration pneumonia were strong independent predictors of increased hospital charges ($p < 0.001$). Higher comorbidities (OR 1.297, 95% CI 1.036–1.624; $p = 0.023$) and presence of neurofibromatosis Type 2 (OR 2.341, 95% CI 1.479–3.707; $p < 0.001$) were associated with higher hospital charges.

Conclusions. The authors' study shows that several factors can affect patient outcome and hospital charges for patients who have undergone acoustic neuroma surgery. Factors such as younger age, higher ZIP code income, less comorbidity, private insurance, elective surgery, and the West region predicted for better disposition outcome. However, the West region, higher comorbidities, and weekend admissions were associated with higher hospitalization costs.
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Key Words • **comorbidity** • **regional hospital factor** • **microsurgery** • **discharge disposition** • **hospital cost** • **Nationwide Inpatient Sample** • **acoustic neuroma**

Acoustic neuromas (vestibular schwannomas) account for 6% of all intracranial tumors, with an incidence rate of approximately 1 in 100,000.[30] The management of these benign tumors is challenging and includes different treatment modalities.[3,4,9,12,27,28] The correct management decision depends on several factors, such as patient age,[25,29,39] tumor size,[23,33] solid or cystic tumor,[17,21,22,26,34] surgical approach,[10,31] growth rate of the tumor, and the magnitude of hearing loss.[11,13,33] Resection, especially in experienced hands, has consistently shown good results with very low morbidity.[32] However, the relatively recent introduction of radiosurgery as a viable alternative treatment modality has prompted a review of the outcomes and associated cost for patients undergoing resection.[1]

Abbreviations used in this paper: CAD = coronary artery disease; CCS = Clinical Classifications Software; GKS = Gamma Knife surgery; LOS = length of stay; NF2 = neurofibromatosis Type 2; NIS = Nationwide Inpatient Sample; OTR = other than routine.

Various factors determine outcomes in surgically treated patients, including facial nerve function,[6,15] hearing preservation rate, and the patient's Karnofsky Performance Scale score, and several studies have rightly focused on these aspects. Few studies have assessed in depth the relationship between demographic features such as race,[18] sex,[24] insurance status,[2] and patient outcome. Some studies had focused on the relationship between volume and provider caseloads and patient outcome.[18,35] Apart from demographic factors, there are several hospital and regional considerations that can impress upon the outcomes. For the most part, however, these variable considerations have largely remained unaddressed. Additionally, any discussion on the efficacy of excision of acoustic neuromas is incomplete without an analysis of the associated costs. There are various studies assessing the costs associated with open surgery, but they have done so with the intention of comparing the costs with those of radiosurgery and a conservative "wait-and-watch" policy.[37,38] These studies, however, do not delve into the various patient- and hospital-related factors that may be associated with an increase in cost of surgical treatment.

To our knowledge, no study has examined in depth the affiliation of hospital, region, and comorbidity factors and hospital charges. To address this gap in the literature, we analyzed the US national database to understand the socioeconomics of acoustic neuroma surgery and to elucidate various factors (demographic, hospital, and regional) responsible for escalating the total hospitalization charges pertaining to inpatient admissions for the excision of acoustic neuromas.

Methods

The NIS is the largest all-payer inpatient care database that is publicly available in the US and contains data on 5–8 million hospital stays from about 1000 hospitals sampled to approximate a 20% stratified sample of US community hospitals. The NIS data from 2005 to 2009 were analyzed using CCS and ICD-9-CM codes to extract the data. We used the single-level ICD-9-CM Code 0401, which is coded as "craniotomy for acoustic neuroma" to extract data from 25,669 hospitals.

As several factors can affect the outcomes and hospital costs, the following categorical variables were generated from the database to further aid in our analysis. 1) Patient age older than 65 years. 2) Patients with CSF diversion procedures (ventricular shunts). This variable was constructed using ICD-9-CM Codes 0231, 0232, 0233, 0234 0235, 0239, 0242, and 0243. 3) Patients with meningitis. This variable was constructed using CCS Code 76 (except meningitis caused by tuberculosis or sexually transmitted disease). 4) Patients with CSF otorrhea. This variable was constructed using ICD-9-CM Code 388.61 for CSF otorrhea. 5) Patients and CSF rhinorrhea. This variable was constructed using ICD-9-CM Code 349.81 for CSF rhinorrhea. 6) Patient undergoing concomitant GKS. This variable was constructed using ICD-9-CM Codes 9230 and 9239 for stereotactic radiosurgery. 7) Patients with aspiration pneumonia. This variable was constructed using CCS Code 129 for aspiration pneumonitis. 8) Patients with other respiratory diseases. This variable was constructed using CCS Codes 125 (acute bronchitis), 126 (other upper respiratory infections), 127 (chronic obstructive pulmonary disease and bronchiectasis), 128 (asthma), 130 (pleurisy, pneumothorax, and pulmonary collapse), 131 (respiratory failure, insufficiency, and arrest [adult]), 132 (lung disease due to external agents), and 133 (other lower respiratory disease). 9) Patients with CAD. Postoperative morbidity due to CAD was assessed using CCS Codes 100 (acute myocardial infarction), 101 (coronary atherosclerosis and other heart disease), 107 (cardiac arrest and ventricular fibrillation), and 108 (congestive heart failure, nonhypertensive). 10) Patients with NF2. This variable was constructed using ICD-9-CM Code 23772. 11) Comorbidity index. The following comorbidities were included in the study as provided by the NIS database: AIDS, alcohol abuse, deficiency anemias, rheumatoid arthritis/collagen vascular diseases, chronic blood loss anemia, congestive heart failure, chronic pulmonary disease, coagulopathy, uncomplicated diabetes, diabetes with chronic complications, drug abuse, hypertension, hypothyroidism, liver disease, lymphoma, fluid and electrolyte disorders, metastatic cancer, other neurological disorders, obesity, paralysis, peripheral vascular disorders, pulmonary circulation disorders, renal failure, solid tumor without metastasis, peptic ulcer disease excluding bleeding, valvular disease, and weight loss. The comorbidity accumulation indices of individual patients ranged from 0 to a maximum of 12. Percentiles were calculated. Patients above the 75th percentile (comorbidity index of 2 or more) formed the high comorbidity index cohort. 12) Hospital charge. The coded variable "total charges" in the NIS data was used to assess the cost incurred by the hospital. Cost percentiles were created, and the 75th percentile position was at $89,600.00. Hospital charges higher than this value formed a cohort of patient that had highest financial implications on the hospital. This cohort was compared with others (cohort with hospitalization charges below the 75th percentile). 13) Final outcome. The coding of the discharge disposition was uniform from 1998 to 2006. New codes have been added since then. For a uniform analysis of data from 1998 to 2009 across all states, the NIS has collapsed transfers, including skilled nursing facility, intermediate care, and other type of facility into 1 category ("DISPUniform"). In our study, 10 patients or fewer had a discharge disposition to short-term care; thus, a separate analysis of this cohort was not possible, and we have analyzed the dichotomized disposition outcome in our study. Discharge disposition to home was "routine" disposition outcome (as defined in the NIS database), and all others, such as transfer to short-term hospital, skilled nursing facility, intermediate care, home health care, against medical advice, and mortality, were considered OTR discharge disposition.[19,23]

Hospital Factors

To understand the impact of hospital factors on patient outcome and complications, the following coded variables in the NIS database were studied. 1) Region of the hospital. In the NIS database, hospitals are divided into the following regions: Northeast, Midwest, South, and West. Figure 1 shows the details of the states included in various regions. 2) Hospital ownership. The data were analyzed based on the following parameters: government, private and non-

profit, private, and investor owned. We collapsed the categories into government and private. 3) Hospital location. The location of a hospital was divided into rural and urban areas. 4) Hospital bedsize. In the NIS database, hospitals are classified on the basis of bedsize as small, medium, and large. The details are provided in Table 1. Different regions have different definitions of hospital bedsize. 5) Teaching status of the hospital. The hospitals are divided into nonteaching and teaching in the NIS database. 6) Weekend admission. This variable was studied to analyze the relationship between outcome and weekend admission.[8] 7) Median household income quartile for a patient's ZIP code. In the NIS, the patient's ZIP code median income is provided. The median household income quartiles for a patient's ZIP code are $1–$38,999, $39,000–$47,999, $48,000–$62,999, and $63,000 or more.

Statistical Analysis

Discharge disposition and the hospital charge were the dependent variables that were studied. Demographic, patient, hospital factors, and the dependent variable were first analyzed by univariate analysis, followed by a multivariate binary logistic regression model, which generated the probability value, odds ratio, and confidence interval. To study the intergroup differences between the mean of total hospitalization charges, we plotted a box plot to determine the distribution of the data (Fig. 2). As the data (cost incurred by a hospital) had significant outliers, we used the nonparametric Kruskal-Wallis 1-way ANOVA to determine the statistical significance ($p < 0.05$). Statistical analysis was done using IBM SPSS Statistics 20 and JMP statistical software, version 9. Mean values are presented as ± SD.

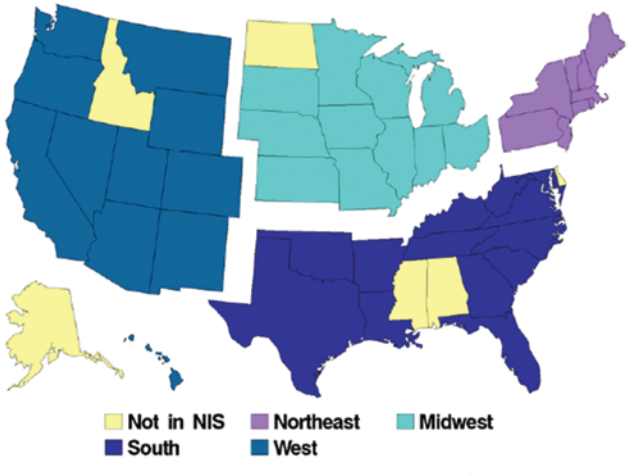


Fig. 1. All states, by region, as provided by the NIS data. *Northeast:* Connecticut, Maine, Massachusetts, New Hampshire, New Jersey, New York, Pennsylvania, Rhode Island, and Vermont. *Midwest:* Illinois, Indiana, Iowa, Kansas, Michigan, Minnesota, Missouri, Nebraska, North Dakota, Ohio, South Dakota, and Wisconsin. *South:* Alabama, Arkansas, Delaware, District of Columbia, Florida, Georgia, Kentucky, Louisiana, Maryland, Mississippi, North Carolina, Oklahoma, South Carolina, Tennessee, Texas, Virginia, and West Virginia. *West:* Alaska, Arizona, California, Colorado, Hawaii, Idaho, Montana, Nevada, New Mexico, Oregon, Utah, Washington, and Wyoming. *The following states are not included in the NIS data:* Alabama, Alaska, Delaware, Idaho, Mississippi, and North Dakota.

TABLE 1: Hospital bedsize by region as provided in the NIS database

Location & Teaching Status	Hospital Bedsize*		
	Small	Medium	Large
Northeast			
rural	1–49	50–99	100+
urban, nonteaching	1–124	125–199	200+
urban, teaching	1–249	250–424	425+
Midwest			
rural	1–29	30–49	50+
urban, nonteaching	1–74	75–174	175+
urban, teaching	1–249	250–374	375+
South			
rural	1–39	40–74	75+
urban, nonteaching	1–99	100–199	200+
urban, teaching	1–249	250–449	450+
West			
rural	1–24	25–44	45+
urban, nonteaching	1–99	100–174	175+
urban, teaching	1–199	200–324	325+

* Values given are the number of beds.

Results

A total of 2589 admissions from 242 hospitals were analyzed from the NIS data from 2005 to 2009. The mean patient age was 48.99 ± 13.861 years (range 8–91 years), 304 patients (11.7%) were elderly (> 65 years), and there were more female patients (1309 patients [52.3%]) than male patients (47.6%). The predominant race was white (1432 patients [55.31%]). However, in the database, the coded variable "race" has missing values for 28.7% of admissions; thus, race was not included in analysis. The demographic details are given in Table 2. Univariate analysis of these demographic variables was done (Table 3). Age older than 65 years and female sex were associated significantly with OTR discharge disposition ($p < 0.001$ and $p = 0.003$, respectively). Most patients were covered by private insurance (1942 patients [75%]). Other than routine discharge disposition was seen in 127 Medicare patients (38.1%) and 45 Medicaid patients (36.3%). The difference was statistically significant ($p < 0.001$; Table 3).

Patient Factors and Outcome

As described in *Methods*, new categorical variables were created for analysis from the database. Ventricular shunt placement was performed in 36 patients (1.39%), and it was significantly associated with OTR discharge disposition. Of the patients who underwent ventricular shunt placement, 27.7% were nonelective admissions. This difference was significant ($p = 0.001$), and in all of these patients ventricular shunt placement was done prior to surgery during the same hospital stay. Meningitis occurred in 16 patients (0.6%) and was significantly associated with OTR discharge disposition. Rhinorrhea was seen in 43 patients (1.6%). Ten patients or fewer were noted to have CSF

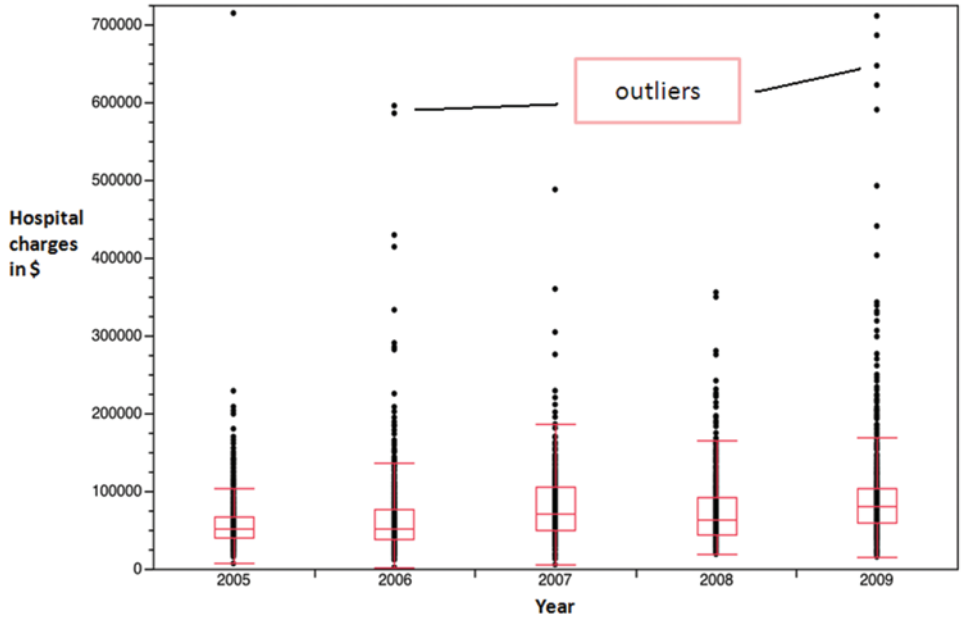


Fig. 2. Box plot showing the distribution of hospital cost across 2005–2009. The data had several outliers; therefore, a nonparametric test using the Kruskal-Wallis test was used to compare the means.

otorrhea as a complication. Therefore, this complication is not displayed in tabulated format, in accordance with the NIS reporting guidelines. Patients with a high comorbidity index had significantly more OTR discharge dispositions (574 patients [22.1%]). Aspiration pneumonia as a complication was seen in 0.88% of admissions (23 patients), and its presence was significantly associated with OTR discharge dispositions. Respiratory diseases and CAD were seen in 328 (12.6%) and 103 (3.9%) patients, respectively, and their associations with OTR discharge disposition were significant ($p < 0.001$). Other factors such as the presence of seventh cranial nerve paresis, tinnitus, decreased hearing, and keratoconjunctivitis were studied, but their association with OTR discharge disposition was not significant ($p > 0.1$) and, therefore, was not included in the regression analysis. There were 81 patients (3.1%) with NF2, and NF2 status was not associated with OTR disposition outcome (Table 3).

Demographic and patient factors with $p < 0.1$ were included in multivariate regression analysis. The highest odds of OTR disposition outcome were associated with the presence of aspiration pneumonia. An OTR disposition outcome was 16.08 times for likely for patients with aspiration pneumonia than for those without it (OR 16.085, 95% CI 4.974–52.016; $p < 0.001$). Similarly, meningitis was associated with high odds for OTR disposition outcome (OR 11.299, 95% CI 3.126–40.840; $p < 0.001$). The model was adjusted for age, sex, comorbidities, and ventricular shunt procedures. Each of these variables predicted independently for OTR disposition outcome. Female sex was associated with OTR disposition outcome (OR 1.340, 95% CI 1.055–1.702; $p = 0.016$). Patients with private insurance when compared with those receiving Medicare and Medicaid had a protective effect against OTR disposition outcome (OR < 1; Table 4). Patients who lived in areas with a ZIP code income of $38,999.00 or less had a significantly OTR disposition outcome when compared with other categories (when compared with ≥ $63,000, OR 1.39; $p = 0.032$). We analyzed 28 comorbidity variables (details given in *Methods*) to form the comorbidity index. A higher comorbidity index independently predicted for OTR disposition outcomes (OR 1.409, 95% CI 1.072–1.852; $p = 0.014$).

Hospital Factors and Patient Outcome

Hospital factors and their impact on discharge disposition were analyzed as well. There were 33 admissions (1.2%) on a weekend (Saturday or Sunday), which was significantly associated with OTR discharge disposition ($p = 0.010$). Nonelective admission had significant association with OTR disposition outcome ($p = 0.013$). Hospital ownership had no bearing on the patients' outcome ($p = 0.2$). Rural hospitals shared 0.15% (39 admissions) of the caseload and were associated with significantly more OTR discharge dispositions ($p = 0.005$). Analysis on the basis of region showed that the West region had significantly better outcomes (950 [90.6%], $p < 0.001$; Table 5). Admission on a weekend was an independent predictor of OTR discharge disposition (OR 2.204, 95% CI 1.016–4.782; $p = 0.045$). The OR was 2.2 times more when compared with admission on weekdays. Nonelective admissions also had a significant impact on discharge disposition (OR 1.491, 95% CI 1.071–2.077; $p = 0.018$) (Table 6). Univariate analysis earlier showed that the West region was significantly associated with routine discharge disposition ($p < 0.001$). We found a significant association between the regions and the discharge dispositions. The association was significant when the West region was compared with the Northeast, South, and Midwest regions. The regression model predicted negatively for OTR disposition outcome. The odds of an OTR discharge disposition were 0.199, 0.624, and 0.542, respectively (Table 6).

TABLE 2: Demographics of the patients who underwent excision of acoustic neuromas*

Characteristic	No. of Admissions
sex	
male	1192
female	1309
primary expected payer (uniform)	
Medicare	333
Medicaid	124
private	1942
self-pay	99
no charge	8
other	78
race (uniform)†	
white	1432
black	71
Hispanic	184
Asian or Pacific Islander	82
Native American	13
other	62
missing	745
total	2589
median household income quartile for pt's ZIP code	
$1–38,999	350
$39,000–47,999	552
$48,000–62,999	689
≥$63,000	910
pt age (yrs)	
<65	2285
>65	304

* Data regarding sex and primary payer are missing in 88 and 5 cases, respectively. Abbreviation: pt = patient.
† Race had significant missing variables and hence was not analyzed further.

TABLE 3: Univariate analysis of patient and demographic factors affecting discharge disposition (final outcome)*

Categorical Variables & Subcategories	No. of Admissions		p Value
	OTR	Routine	
primary expected payer (uniform)			<0.001†
Medicare	127	206	
Medicaid	45	79	
private	219	1723	
self-pay	8	91	
no charge	2	6	
other	12	66	
pt age (yrs)			<0.001†
<65	288	1997	
>65	125	179	
CSF diversion			<0.001†
not done	390	2163	
done	23	13	
meningitis			<0.001†
absent	401	2172	
present	—	—	
rhinorrhea			0.229
absent	405	2141	
present	—	—	
comorbidity index			<0.001†
low	258	1757	
high	155	419	
aspiration			<0.001†
absent	394	2172	
present	19	4	
respiratory disease			<0.001†
absent	310	1951	
present	103	225	
CAD			<0.001†
absent	378	2108	
present	35	68	
NF2			0.209
absent	396	2112	
present	17	64	
hospital ownership			0.438
private	12	80	
government	401	2096	

* — = not displayed according to data user agreement with the Healthcare Cost and Utilization Project. (Data are not to be represented in any given cell of tabulated data if ≤ 10.)
† Significant difference.

Cost of Hospitalization

The NIS database coded variable "Total Charges" was analyzed. The cumulative cost incurred by the hospitals when performing 2589 acoustic neuroma excisions between 2005 and 2009 was $948.7 million (after adjusting for discharge weights). The mean expenditure was $76,365.09 ± $58,039.93. The mean total charges per admission rose from $59,633.00 in 2005 to $97,370.00 in 2009. Box plot analysis of yearly hospital charges showed significant outliers (Fig. 2); therefore, a nonparametric test to assess the difference of the means was used. The difference in the groups was significant ($p < 0.001$; Fig. 3).

The NIS database permits analyses based on regions (Fig. 1). We analyzed whether there is any disparity in hospital charges across these regions. The mean hospital charge per admission in the West region was $88,761.00 and that in the Southern region was $65,329 (Fig. 4). This difference was significant ($p < 0.001$). The difference in hospital charges between large hospitals and small and medium hospitals was significant ($p < 0.001$; see Table 8). The mean hospital charge of large hospitals was $80,130.00 and that for small hospitals was $61,960.00. The difference in mean hospital charges between teaching ($73,219) versus nonteaching ($88,564) hospitals was

TABLE 4: Outcome of multivariate binary logistic regression analysis*

Covariate	B Value	p Value	OR (95% CI)
age >65 yrs	0.801	0.001†	2.228 (1.411–3.518)
CSF diversion	1.701	<0.001†	5.482 (2.480–12.118)
meningitis	2.425	<0.001†	11.299 (3.126–40.840)
rhinorrhea	0.333	0.447	1.395 (0.591–3.292)
higher comorbidity index	0.343	0.014†	1.409 (1.072–1.852)
aspiration	2.778	<0.001†	16.085 (4.974–52.016)
other respiratory disease	0.477	0.003†	1.611 (1.175–2.209)
CAD	0.301	0.244	1.352 (0.814–2.244)
NF2	0.438	0.166	1.549 (0.834–2.877)
private insurance status compared w/ others			
Medicaid	−0.744	0.001†	0.475 (0.302–0.748)
Medicare	−1.121	<0.001†	0.326 (0.210–0.505)
female sex	0.293	0.016†	1.340 (1.055–1.702)
ZIP code income <$38,999.00 compared w/ others			
$39,000–47,999	0.248	0.194	1.281 (0.882–1.862)
$48,000–62,999	0.039	0.831	1.039 (0.729–1.482)
>$63,000	0.328	0.032†	1.388 (1.341–1.983)

* The dependent variable was discharge disposition (final outcome); demographic and patient factors were the covariates.
† Significant difference.

TABLE 5: Univariate analysis of hospital factors and discharge disposition (final outcome)*

Variable	No. of Admissions (%)		Total	p Value
	OTR	Routine		
admission day				0.010†
weekday	402 (15.7)	2154 (84.3)	2556 (100.0)	
weekend	11 (33.3)	22 (66.7)	33 (100.0)	
admission status				0.013†
nonelective	60 (21.1)	225 (78.9)	285 (100.0)	
elective	353 (15.3)	1951 (84.7)	2304 (100.0)	
hospital bedsize				0.008†
small	16 (18.2)	72 (81.8)	88 (100.0)	
medium	57 (11.4)	444 (88.6)	501 (100.0)	
large	338 (16.9)	1658 (83.1)	1996 (100.0)	
hospital ownership				0.270
private	12 (13.0)	80 (87.0)	92 (100.0)	
government	401 (16.1)	2096 (83.9)	2497 (100.0)	
teaching status of hospital				0.21†
nonteaching	62 (12.8)	422 (87.2)	484 (100.0)	
teaching	349 (16.6)	1752 (83.4)	2101 (100.0)	
location of hospital				0.005†
rural	13 (33.3)	26 (66.7)	39 (100.0)	
urban	398 (15.6)	2148 (84.4)	2546 (100.0)	
hospital region				<0.001†
Northeast	143 (34.4)	273 (65.6)	416 (100.0)	
Midwest	81 (14.3)	485 (85.7)	566 (100.0)	
South	90 (16.1)	468 (83.9)	558 (100.0)	
West	99 (9.4)	950 (90.6)	1049 (100.0)	

* Data are missing from hospital bedsize, teaching status of hospital, and location of hospital in 4 cases.
† Significant difference.

TABLE 6: Outcome of multivariate binary logistic regression analysis*

Covariate	B Value	p Value	OR (95% CI)
weekend admission	0.790	0.045†	2.204 (1.016–4.782)
nonelective op	0.400	0.018†	1.491 (1.071–2.077)
hospital bedsize			
medium	0.603	0.061	1.827 (0.971–3.436)
large	0.391	0.188	1.479 (0.826–2.646)
hospital location	0.080	0.823	1.083 (0.538–2.181)
teaching status	0.132	0.411	1.141 (0.833–1.565)
hospital region compared w/ West			
Northeast	−1.615	<0.001†	0.199 (0.149–0.266)
South	−0.472	0.003†	0.624 (0.456–0.854)
Midwest	−0.613	<0.001†	0.542 (0.399–0.736)

* The dependent variable was final outcome, and hospital factors were the covariates.
† Significant difference.

significant ($p < 0.001$); however, between rural and urban hospitals it was not significant ($p = 0.24$; Table 7).

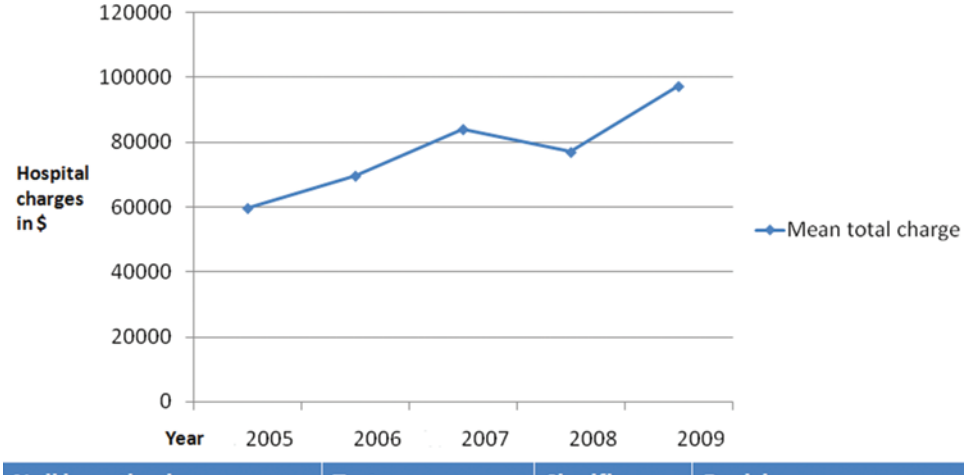
Hospital factors were further analyzed by univariate (Table 8) followed by multivariate regression analysis. Only those factors with $p < 0.1$ were included in the analysis. Large hospital bedsize was independently associated with higher hospital charges (OR 4.269, 95% CI 3.106–5.867; $p < 0.001$). The West region had significantly higher mean hospital charges when compared with other regions. Regression analyses also showed a significantly higher odds ratio with significant values ($p < 0.001$) when compared with other regions (Table 9). Interestingly, government or federal ownership had a protective effect against higher hospitalization charges (OR < 1 with a negative trend [OR 0.306, 95% CI 0.181–0.518; $p < 0.001$]).

Admission on a weekend was significantly ($p = 0.032$) associated with higher cost in univariate analysis (Table 8). The mean LOS for patients who were admitted over the weekend (8.7 ± 13 days) was greater than that for patients admitted on a weekday (5.4 ± 6.2 days). However, logistic regression analysis did not show a significant impact on overall cost incurred ($p = 0.07$; Table 9). On bivariate Spearman nonparametric correlation analysis, LOS had a significantly high correlation with hospital cost incurred ($p < 0.001$). As we intended to study the predictors of higher hospital cost, LOS was not chosen as a covariate in the regression model on account of its significant correlation with hospitalization cost.

Patient factors also had an impact on hospital charges. Postoperative meningitis and aspiration pneumonia were very strong independent predictors of increased hospital charges ($p < 0.001$). The ORs were 18.82 and 12.2, respectively (Table 10). A higher comorbidity index (OR 1.297, 95% CI 1.036–1.624; $p = 0.023$) (Fig. 5) and presence of NF2 (OR 2.341, 95% CI 1.479–3.707; $p < 0.001$) was associated with higher hospital charges (Table 10).

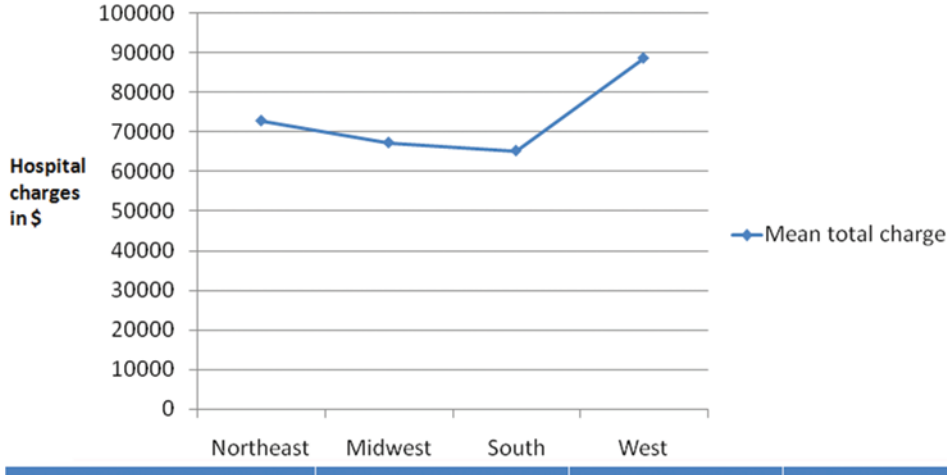
Discussion

Management of acoustic neuromas has evolved during the past 20 years. Treatment modalities include conservative management,[3] aggressive resection,[33] GKS,[4,12] and subtotal resection followed by stereotactic radiosurgery.[16] Several factors have been documented that are associated with outcomes, such as solid tumor versus cystic tumor, size of the lesion, and age, but there are very few studies that have focused on the demographic, patient, and hospital-related



Null hypothesis	Test	Significance	Decision
The distribution of total charges is the same across the years	Independent samples Kruskal-Wallis Test	<0.001	Reject the null hypothesis

Fig. 3. Graph showing that the mean total hospitalization charges steadily increased over the years. Total hospitalization charges increased from $59,633.00 in 2005 to $97,370.00 in 2009. The Kruskal-Wallis test showed that the difference in means was significant.



Null hypothesis	Test	Significance	Decision
The distribution of total charges is the same across the region	Independent samples Kruskal - Wallis Test	<0.001	Reject the null hypothesis

Fig. 4. Graph showing the mean hospital charges across the regions. The South region had the lowest hospital charges/admissions, and the West region had the maximum hospital charge. The Kruskal-Wallis test showed that the difference in means was significant.

factors that affect the discharge disposition and cost of hospitalization. In their analysis of NIS data from 1996 to 2000, Barker et al.[2] found a significant association of factors such as race, insurance status, and income status of the place of residence. Similar associations were found by McClelland et al.[18] in their study on the NIS from 1994 to 2003 (Table 11). We compared the discharge disposition of the elderly population with others. The elderly population had a significant OTR disposition outcome, suggesting that the younger population has better outcome.[14,39] However in their series, Pulec et al.[29] did not find any significant difference in the outcome between the 2 population groups. Patients with private insurance and higher household income had significantly better outcomes after surgery. Barker et al.[2] found similar observations in their series. These factors predicted independently for good outcome in our regression model. This trend has remained the same since the last decade when the NIS data were first analyzed.[2]

TABLE 7: Independent samples Kruskal-Wallis test results, showing significant difference between the mean of various groups

Variable	Mean Total Charges ($)	p Value
hospital bedsize		<0.001*
small	61,960	
medium	61,705	
large	80,130	
hospital location		0.241
rural	76,151	
urban	76,140	
hospital region		<0.001*
Northeast	72,910	
Midwest	67,384	
South	65,329	
West	88,761	
teaching status		<0.001*
nonteaching	88,564	
teaching	73,219	
comorbidity index		<0.001*
low	72,413	
high	89,828	

* Significant difference.

In our analysis, the operative mortality rate was 0.5% (13 patients). In addition to these demographics, we studied other patient-related factors that were not studied earlier. Aspiration pneumonia is one of the most feared complications after surgery, as lower cranial nerve dysfunction makes the patient vulnerable. It was the strongest predictor of in-hospital morbidity in our study ($p < 0.001$). In addition, other respiratory disease also significantly predicted for morbidity ($p = 0.003$; Table 4).

In one of the largest surgical series, Samii and Matthies[33] reported meningitis in 1.2% of cases. In our analysis, it was 0.6% and a predictor of OTR disposition outcome (Table 4). Hydrocephalus is an important cause of increased morbidity. This complication has been reported to range from 2.3% to 3.2% in the literature.[18,33] We studied the variable "ventricular shunts" separately, as this variable could serve as surrogate marker of hydrocephalus requiring treatment. Ventricular shunts were associated with OTR discharge disposition ($p < 0.001$). The occurrence of meningitis was significantly seen in patients who presented for acute care and underwent a diversion procedure before undergoing definite surgery. A CSF fistula was seen in 51 patients (1.9%) but was not associated

TABLE 8: Univariate analysis of hospital factors affecting cost of the treatment*

Variable	No. of Admissions (%)		Total	p Value
	Higher Cost (75th percentile)	Lower Cost		
admission day				0.032†
weekday	669 (26.2)	1887 (73.8)	2556 (100.0)	
weekend	14 (42.4)	19 (57.6)	33 (100.0)	
type of admission				0.072
nonelective	86 (30.2)	199 (69.8)	285 (100.0)	
elective	597 (25.9)	1707 (74.1)	2304 (100.0)	
hospital bedsize				<0.001†
small	52 (59.1)	36 (40.9)	88 (100.0)	
medium	54 (10.8)	447 (89.2)	501 (100.0)	
large	574 (28.8)	1422 (71.2)	1996 (100.0)	
hospital ownership				<0.001†
private	61 (66.3)	31 (33.7)	92 (100.0)	
government	622 (24.9)	1875 (75.1)	2497 (100.0)	
hospital teaching status				<0.001†
nonteaching	201 (41.5)	283 (58.5)	484 (100.0)	
teaching	479 (22.8)	1622 (77.2)	2101 (100.0)	
hospital location				0.265
rural	— (20.5)	— (79.5)	— (100.0)	
urban	672 (26.4)	1874 (73.6)	2546 (100.0)	
hospital region				<0.001†
Northeast	89 (21.4)	327 (78.6)	416 (100.0)	
Midwest	84 (14.8)	482 (85.2)	566 (100.0)	
South	82 (14.7)	476 (85.3)	558 (100.0)	
West	428 (40.8)	621 (59.2)	1049 (100.0)	
South region				<0.001†
no	601 (29.6)	1430 (70.4)	2031 (100.0)	
yes	82 (14.7)	476 (85.3)	558 (100.0)	
hospital bedsize				<0.001†
other	631 (25.2)	1870 (74.8)	2501 (100.0)	
small	52 (59.1)	36 (40.9)	88 (100.0)	

* — = not displayed according to data user agreement with the Healthcare Cost and Utilization Project. (Data are not to be represented in any given cell of tabulated data if ≤ 10.)
† Significant difference.

with OTR disposition outcome. Additionally, ventricular shunts also escalated the hospital cost. We also analyzed the outcome of patients with NF2 and found that the disease was not a predictor of OTR disposition outcome. One reason could be that 97.5% of patients with NF2 were younger than 65 years. Since fewer than 10 patients with NF2 were older than 65 years, a detailed statistical analysis of this cohort could not be done.

Hospital Factors and Outcome

Most of the available literature is about single-surgeon[32,33] or single-institution experiences. Rarely is acoustic neuroma surgery mentioned in the context of a geographical location[35] or country.[2] The single-surgeon/single-institution experience series serve as the gold standard to understand the outcomes in terms of the cranial nerve or other focal neurological deficits. The NIS database gives the opportunity to study several demographic, patient, and hospital-related variables in large numbers for better statistical prowess. Patients admitted on weekends had a 2.2 times higher odds of OTR disposition outcome than patients admitted on weekdays (Table 6), and patients who needed acute care (nonelective) had significant OTR disposition outcomes, even after adjusting for other factors.

We analyzed the discharge dispositions and hospitalization cost to understand whether a regional disparity exists. The NIS data sample has been statistically structured to allow analysis in terms of regions and not on the basis of states. The West region had significantly more routine dispositions than other regions. Regression analysis also showed that the West region had a protective effect (< 1 OR) against OTR disposition outcome when it was compared with other regions (Table 6). This disparity persisted despite adjusting for other patient factors. Non-

TABLE 9: Outcome of multivariate binary logistic regression analysis*

Covariate	B Value	p Value	OR (95% CI)
large hospital bedsize	1.451	<0.001†	4.269 (3.106–5.867)
West region compared w/ others			
Northeast	1.000	<0.001†	2.719 (2.021–3.658)
Midwest	1.448	<0.001†	4.257 (3.214–5.637)
South	1.489	<0.001†	4.432 (3.332–5.894)
nonteaching hospital	0.257	0.058	1.293 (0.992–1.686)
federal/government ownership	−1.185	<0.001†	0.306 (0.181–0.518)
admission on weekend	0.732	0.073	1.078 (0.876–1.036)

* The dependent variable was higher hospitalization charges (hospital charge > 75th percentile), and the hospital factors were the covariates.
† Significant difference.

TABLE 10: Outcome of multivariate binary logistic regression analysis*

Covariate	B Value	p Value	OR (95% CI)
elderly population	−0.030	0.837	0.970 (0.729–1.292)
meningitis	2.935	<0.001†	18.817 (4.205–84.198)
rhinorrhea	1.040	<0.001†	2.829 (1.531–5.229)
comorbidity index	0.260	0.023†	1.297 (1.036–1.624)
aspiration	2.498	<0.001†	12.155 (3.516–42.027)
respiratory disease	0.587	<0.001†	1.799 (1.384–2.339)
CAD	0.217	0.344	1.243 (0.793–1.948)
NF2	0.851	<0.001†	2.341 (1.479–3.707)

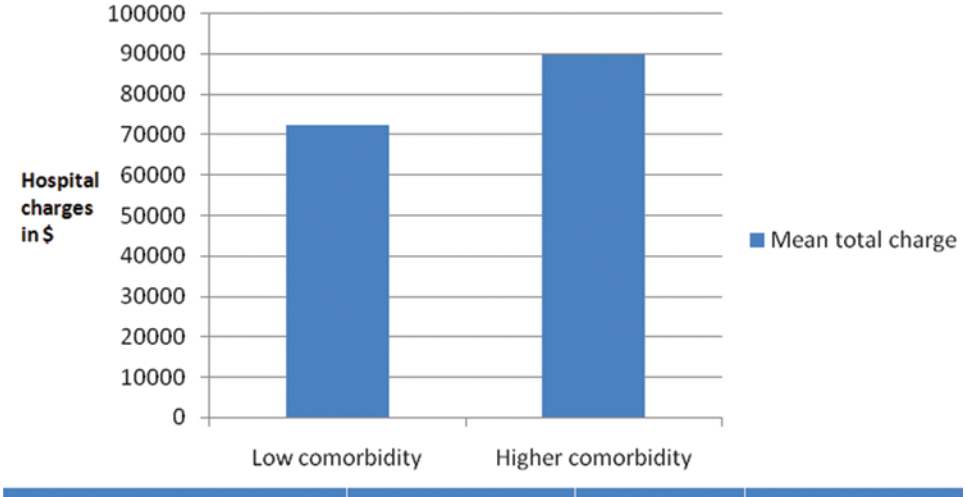
* The dependent variable tested was higher hospitalization cost, and patient factors were the covariates.
† Significant difference.

teaching status and rural location of the hospital were not associated with OTR discharge disposition.

Hospital Factors and Treatment Cost Incurred

We performed a detailed analysis of various factors that could escalate the cost of hospital LOS. Factors such as hospital region, rural or urban setting, small or large hospital size, teaching or nonteaching centers, and comorbidity index were analyzed in a multivariate binary logistic regression model, which was adjusted for age, ventricular shunt procedures, and other complications. The cost of acoustic neuroma surgery has increased from $25,800 in 1996[2] to $76,365.09 in 2009 (current study). The results showed that there is regional disparity in the hospital charges across the US. The South region had significantly fewer mean hospital charges ($p < 0.001$; Fig. 4). One reason for the disparity could be the low cost of infrastructure in the southern region.

Teaching hospitals had a lower hospitalization cost than nonteaching hospitals. The difference was significant ($p < 0.001$; Table 8). However, in the regression model after adjusting for other factors, the association of nonteaching hospitals with higher hospitalization cost was not significant (Table 9). Few earlier studies showed that teaching hospitals incurred a higher hospitalization cost than nonteaching hospitals, but these studies only took orthopedic patients into account.[20,36] The results of our study are interesting because they show a decrease in hospitalization cost for neurosurgical patients treated at teaching hospitals. However, for a larger reach the data need to be studied over



Null hypothesis	Test	Significance	Decision
The distribution of total charges is the same across the comorbidity index	Independent samples Kruskal-Wallis Test	<0.001	Reject the null hypothesis

Fig. 5. Graph showing that patients with a low comorbidity index had lower hospitalization costs than those with a higher comorbidity index. The Kruskal-Wallis test showed that the difference in means was significant.

TABLE 11: A brief summary reviewing results in the literature in regard to acoustic neuroma surgery*

Parameter	McClelland et al., 2011	Barker et al., 2003
NIS database time period	1994–2003	1996–2000
no. of excisions	4886	2643
multivariate analysis of adverse discharge disposition (p value)		
age	0.0019	0.07
primary payer	0.017	<0.001
high surgeon volume	<0.0001	0.004
female sex	0.034†	NS
race	0.26	NS
admission type (routine/others)	0.27	0.02
median income	0.7	0.002
hospitalization cost (p value)		
high-volume hospital	not commented	0.006‡
surgeon caseload	not commented	0.09

* NS = not significant.
† After exclusion of markers of advanced disease.
‡ Charges were significantly lower at higher-volume hospitals.

a longer time and take into account varied neurosurgical procedures to understand the hospitalization cost (teaching vs nonteaching hospitals) for neurosurgical patients.

To further understand the disparities, we analyzed the difference of the mean between groups as outlined earlier in *Results*. As there were significant outliers that could be due to multiple procedures or complications during a single admission for a patient, nonparametric Kruskal-Wallis 1-way ANOVA was done (Fig. 2). It has been shown that comorbidities can influence hospitalization charges.[5,7] A higher comorbidity index > 2 comorbidities) also had a significant impact on the overall hospitalization cost (Fig. 5 and Table 7). We constructed a binary dependent variable, higher hospital charge, which comprised higher hospital charges (> 75th percentile). The other hospital factors, higher comorbidity index, and patient factors were adjusted for in the analysis. The West region predicted for higher cost than the other regions (Table 9). Among the patient factors, aspiration pneumonitis had the highest odds for higher hospital charges ($p < 0.001$; Table 10) and for OTR disposition outcome (OR 16.085, 95% CI 4.974–52.016; $p < 0.001$).

Study Limitations

There are limitations of our study given that it is a retrospective analysis, and there could be coding errors or underreporting of events in the database. Moreover, preoperative presentation could not be separated from a postoperative complication. Factors such as grade of seventh cranial nerve paresis, hearing function, size of the lesion, cystic or solid nature of tumor, extent of resection, use of GKS, and Karnofsky Performance Scale score could not be studied under a separate category. Thus, a discharge disposition as "routine" is not synonymous with "good resection," and "OTR discharge disposition" does not necessarily mean "poor resection." Discharge disposition is dependent on several factors, and a good surgical outcome does not necessarily translate into a good discharge disposition. Moreover, the NIS database cannot be used to comment purely on surgical outcomes; hence, we have analyzed the dichotomized disposition outcome in our study. Similarly, a good surgery with minimal operative complications could not be analyzed separately, as there is no separate code of postoperative complications in the NIS database. There are some restrictions of NIS database usage, such as not reporting events for which there are 10 or fewer patients. We were unable to understand the rare cohort of patients who acutely worsened after GKS because there were 10 or fewer patients; therefore, the results cannot be reported.

Conclusions

There are several factors that can affect a patient's discharge disposition and hospitalization charges. It is not possible for each and every patient to have access to the "best" care provider or the "best" surgeon. The results of our analysis are important as they provide a broader picture of outcome of acoustic neuroma surgery and the cost incurred across the country. The finding of regional disparity in hospital charges is particularly important in the current scenario of passing of the Patient Protection and Affordable Care Act and the impact of medical tourism on the revenues generated by our health system. More studies are needed to understand the regional disparity of various neurosurgical procedures in terms of outcomes and hospital cost.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Sonig. Acquisition of data: Sonig, Khan, Wadhwa, Thakur. Analysis and interpretation of data: Sonig. Drafting the article: Sonig. Critically revising the article: Sonig. Reviewed submitted version of manuscript: Sonig, Khan, Wadhwa, Thakur. Approved the final version of the manuscript on behalf of all authors: Nanda. Statistical analysis: Sonig. Administrative/technical/material support: Nanda. Study supervision: Nanda.

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Incidental vestibular schwannomas: a review of prevalence, growth rate, and management challenges

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With the relatively recent increase in the use of MRI techniques, there has been a concurrent rise in the number of vestibular schwannomas (VSs) detected as incidental findings. These incidental VSs may be prevalent in up to 0.02%–0.07% of individuals undergoing MRI and represent a significant portion of all diagnosed VSs. The management of these lesions poses a significant challenge for practitioners. Most incidental VSs tend to be small and associated with minimal symptoms, permitting them to be managed conservatively at the time of diagnosis. However, relatively few indicators consistently predict tumor growth and patient outcomes. Furthermore, growth rates have been shown to vary significantly over time with a large variety of long-term growth patterns. Thus, early MRI screening for continued tumor growth followed by repeated MRI studies and clinical assessments throughout the patient's life is an essential component in a conservative management strategy. Note that tumor growth is typically associated with a worsening of symptoms in patients who undergo conservative management, and many of these symptoms have been shown to significantly impact the patient's quality of life. Specific indications for the termination of conservative management vary across studies, but secondary intervention has been shown to be a relatively safe option in most patients with progressive disease. Patients with incidental VSs will probably qualify for a course of conservative management at diagnosis, and regular imaging combined with the expectation that the tumor and symptoms may change at any interval is crucial to ensuring positive long-term outcomes in these patients. In this report, the authors discuss the current literature pertaining to the prevalence of incidental VSs and various considerations in the management of these lesions. It is hoped that by incorporating an understanding of tumor growth, patient outcomes, and management strategies, practitioners will be able to effectively address this challenging disease entity.
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Key Words • **incidental lesion** • **vestibular schwannoma** • **vertigo** • **acoustic neuroma** • **tumor growth** • **conservative management** • **hearing preservation** • **quality of life**

Vestibular schwannomas account for up to 10% of all primary brain neoplasms[39] and represent a largely heterogeneous group of tumors with a wide variety of clinical manifestations, growth patterns, and patient outcomes. Typically, patients with these lesions present with unilateral high-frequency sensorineural hearing loss, although they can also present with headache, tinnitus, vertigo, and balance problems.[20] With the increased use of advanced radiographic imaging, particularly contrast-enhanced MRI, a greater number of VSs are diagnosed in patients without any of the symptoms typically associated with these tumors, and they are often detected incidentally after MRI for other suspected intracranial lesions. Recently, numerous studies have been undertaken to try to characterize the prevalence of VSs in patients who are asymptomatic and whose VSs are diagnosed due to an incidental finding.[1,10–14,26,33,37]

Defining the prevalence of incidental VSs as well as managing them has become a challenge for neurosurgical and neurootological practice. An increasing percentage of patients with these lesions are presenting to these practice settings without asymmetrical hearing loss or any other suggestive symptoms. In fact, among patients seen for a VS on MRI, the tumor was an incidental finding in 5%–12% of those patients.[15,26] Furthermore, the wide variety of tumor growth rates and interventional outcomes associated with incidental VSs make the long-term management of these lesions an issue that is much debated. In this paper, only sporadic VSs not associated with neurofibromatosis Type 2 will be discussed.

Abbreviation used in this paper: VS = vestibular schwannoma.

We reviewed the latest research with regard to the prevalence and presentations of incidental VSs. In this paper, we discuss how tumor growth rates and clinical outcomes relate to the management of these lesions, including the benefits and pitfalls of conservative management techniques. Our goal is to elucidate how the latest research can help to guide the clinician to effectively manage the increasing number of patients likely to present with incidental VSs.

Epidemiological Data

Early attempts to define the prevalence of VSs in the general population were made in cadaveric studies in both the pre- and post-MRI era. These studies involved the serial dissection of numerous temporal bones from deceased patients and ultimately suggested a prevalence of around 0.8%–2.4% (Table 1).[13,14,33] Since VSs had not been diagnosed in these patients' lifetimes and the patients may not have undergone a workup for symptoms related to VSs, in theory these early study results could correlate well with the actual prevalence of asymptomatic incidental VSs. However, the numerous limitations in studying only cadavers make it difficult to generalize these results to the entire population. Primarily, the fact that these studies were conducted in patients who were no longer living results in an obvious selection bias for older patients with other significant comorbidities. Furthermore, cultural differences, changes in the practice of medicine, and changes in our understanding of VSs throughout the time periods in which these studies were conducted probably meant that a number of these tumors were undiagnosed in life, adding further bias to the results. As a consequence, these values probably represent a gross overestimation of the actual prevalence of incidental VSs within the current population. Regardless, the relative consistency of these findings suggests that the results are not entirely baseless and may represent a close estimate of the prevalence of VSs in older populations at the time that these studies were conducted.

In contrast to those early cadaveric studies, more recent attempts to elucidate the prevalence of VSs in patients not suspected of having the disease have taken advantage of modern advances in imaging technology.[1,14] With the advent of MRI, researchers can retrospectively analyze large imaging databases, enabling them to include a wider variety of patients and yield sample sizes significantly greater than what was possible with cadaveric studies (Table 2). It has been shown that VSs account for approximately 0.2% of incidental MRI findings.[38] Two studies in particular have provided the best illustration of the prevalence of incidental VSs in patients not suspected of having the disease. In the first of these studies, Anderson et al.[1] found 17 patients with unsuspected VSs after reviewing 24,246 MRI studies obtained for reasons other than ruling out a VS or evaluating patients with tinnitus, sensorineural hearing loss, or vertigo. This yielded a VS prevalence of 0.07% in patients in whom no VS was suspected. More recently, Lin et al.[14] analyzed 46,414 MR images obtained for reasons other than audiovestibular signs, revealing a much lower prevalence of 0.02%, or only 8 patients with a positive reading. These latter authors determined that the difference between the two studies was statistically significant, ultimately attributing the discrepancy to numerous factors. They cited the increase in the number of MRI studies, the shorter time period analyzed by Anderson et al., and differences in the criteria of accepted MRI studies as possible reasons for this discrepancy. Moreover, Lin and colleagues specifically excluded studies on the internal auditory canal, whereas Anderson and associates did not. Imaging studies of the internal auditory canal are usually conducted to rule out a VS, even though it may not have been specified in the imaging database. Thus, the prevalence of 0.02% proposed by Lin et al. probably represents the best estimation of incidental VSs to date.

Interestingly, the study by Lin et al.[14] also revealed a number of other patient variables associated with finding a VS on MRI. The number of patients presenting with VSs, whether symptomatic or not, was not statistically significant for a sex difference; however, when the study was limited to just those patients with an incidental finding, men represented a significantly larger proportion of the study group. The authors postulated that this could be the result of behavioral differences; that is, men may be less likely to seek treatment or consult a physician for audiovestibular concerns. This point was illustrated by a male patient who presented with an incidental finding but later reported profound unilateral deafness on further questioning after receiving the MRI results. Actually, 5 of the 8 patients determined to have incidental findings later reported significant audiovestibular symptoms that were not initially related when their histories were taken. This finding implies that while the Lin et al. study shows a lower prevalence of incidental VSs as compared with that in the Anderson et al.[1] study, the actual number of truly asymptomatic patients with positive findings could be even lower than suggested if one assumes that all patients with symptoms can be ruled out by taking a thorough history. Regardless, both of these studies were within a

TABLE 1: Prevalence of incidental VSs in studies of temporal bones at autopsy

Authors & Year	No. of Bones	No. of VSs	VS Prevalence (%)
Hardy & Crowe, 1936	250	6	2.4
Leonard & Talbot, 1970	490	4	0.8
Stewart et al., 1975	893	5	0.9
Karjalainen et al., 1984	298	0	0
Lin et al., 2005	746	5 (4)*	1.0

* One cadaver had bilateral VSs.

TABLE 2: Prevalence of VSs in studies using MRI for diagnosis

Authors & Year	No. of Cases	No. of VSs	Prevalence (%)
Selesnick et al., 1993	161*	0	0
Anderson et al., 2000	24,246	17	0.07
Lin et al., 2005	46,414	9	0.02

* Study performed prospectively.

similar order of magnitude compared with the cadaveric studies and probably represent a closer assumption of the real prevalence of incidental VSs.

Despite the prevalence of incidental VSs within the general population, in neurosurgical practice these lesions represent a significant fraction of the patients presenting with positive MRI findings. In a study conducted by Jeyakumar et al.,[11] among 121 patients with diagnosed VSs, the lesions were incidentally discovered in 15 (12.3%) of them. In direct contrast to the findings of Lin et al.,[14] Jeyakumar and colleagues showed that incidental VSs were diagnosed in 4 times as many women than men, although the authors did not offer an explanation for this finding. Additionally, they showed that there was no age difference between those presenting with symptomatic VSs and those with incidental VSs. By putting all of these studies in the context of neurosurgical practice, it can be determined that patients with incidental VSs represent a significant portion of the neurosurgery patient base and that a diagnosis of VS will probably result from an incidental finding in a notable percentage of patients with VS. Therefore, neurosurgeons must have an intimate understanding of the tumor characteristics and treatment choices for incidental VSs, as they are likely to encounter this disease entity in their practice.

Growth Patterns and Natural History

Recently, an extensive body of literature has emerged discussing the natural history and growth patterns of VSs in both symptomatic and asymptomatic patients. A thorough understanding of the variety of characteristics associated with tumor progression is crucial in determining the proper care of patients who are asymptomatic with incidental findings. We are not aware of any studies specifically focused on the growth rates of incidental or asymptomatic tumors by themselves; however, numerous studies have been conducted to characterize the natural history and growth patterns of VSs that were not subjected to any initial intervention. An understanding of growth patterns in these tumors is paramount to guiding long-term treatment and follow-up decisions.[30]

In general, VSs tend to be a relatively slow-growing lesion as compared with other more malignant intracranial tumors, with many showing little to no growth after diagnosis. A recent meta-analysis conducted by Nikolopoulos et al.[22] showed that the rate of tumor growth varies significantly among patients who undergo conservative management. In 41 of the studies included in that review, the number of tumors demonstrating growth varied from 6% to 73%, with the rate of individual tumor growth varying significantly as well, from 0.3 to 4.8 mm/year. While the mean growth rate varied between 1 and 2 mm/year for all tumors, the growth rate was between 2 and 4 mm/year when only tumors that grew were considered. Moreover, some studies in the meta-analysis documented exceptional growth rates in excess of 15 mm/year. Nikolopoulos and colleagues also demonstrated that tumors showing only continuous growth represented a minority of the lesions, accounting for only 15%–25% of the tumors across the studies. Another meta-analysis conducted by Yoshimoto[41] also reported a wide range of variability among studies, with 15%–85% of tumors showing growth. This author also demonstrated an overall growth rate of 1.2 mm/year. Taking into consideration all studies in the meta-analysis, he established that an average of 46% of tumors showed growth. However, he also indicated that the percentage of tumors that were determined to grow was greatly affected by the type of study conducted. The average percentage of growing tumors was 39% in the studies in which MRI alone was used and only 29% when considering only prospective studies. This finding suggests that the number of tumors that grow may actually be lower than initially predicted, as higher-quality studies are likely to be more representative of real growth characteristics. Interestingly, both of these reviews also showed that a significant portion of tumors actually regress, which occurred in 8%–22% of tumors.[22,41]

Studies have shown that "tumor growth" is a broad descriptive term used to identify what is actually a largely heterogeneous group of tumor growth patterns. Three studies in particular have attempted to characterize VSs into specific groups based on their growth patterns (Table 3).[22,24,28] What these studies have shown is that there is not a one-size-fits-all description that characterizes tumor growth patterns. Furthermore, how the growth rate at one interval fits into the entire natural history of the tumor is largely unpredictable.[22,41] Thus, it is crucial that practitioners appreciate the variability in individual tumors and do not use a growth rate from one interval to define the rate for future intervals. For example, just because a patient presents with a small incidental tumor that reduced in size on the first follow-up scan does not mean that future scans are unnecessary. At any moment the growth rate of that tumor can change.

There have also been attempts to define independent patient and tumor characteristics that can predict tumor growth. For example, Yoshimoto[41] proposed that larger tumors represent a lower risk for future VS enlargement. Solares et al.[30] also suggested that tumor size plays a role in defining tumor growth, showing that 5-year no-growth rates vary significantly among intracanalicular, Grade I

TABLE 3: Literature survey of studies examining tumor growth patterns on follow-up imaging in patients who underwent conservative management*

Growth Pattern	Shin et al., 2000	Nikolopoulos et al., 2010	Rosenberg, 2000
reduction only	5	7	8
reduction followed by growth	20	NA	NA
stability only	20	50	35
stability followed by growth	NA	16	13
growth only	15	25	21
growth followed by stability	NA	3	13
growth then reduction	40	NA	10

* Values expressed in %. Note that the specific radiological definitions used for growth varied slightly among the studies. Moreover, the exact names for growth patterns were altered for simplification, and not all patterns were accounted for in every study. Abbreviation: NA = not applicable.

tumors, and tumors above Grade I (89.8%, 73.9%, and 45.2% respectively). These latter authors also suggested that when analyzed separately based on tumor size, women had a significant difference in growth rates, showing growth in 90.9% of VSs smaller than 10 mm, as compared with 62.3% of VSs larger than 10 mm. However, as illustrated in the extensive meta-analysis conducted by Nikolopoulos et al.,[22] there is marked variability in the significance and outcomes associated with independent predictive variables across studies. While some indicated that the growth pattern within the 1st year was a significant indicator of tumor behavior, other studies directly contradicted this assertion.[5,24] The same was true when initial tumor size was used to predict growth, that is, with contradictory evidence suggested across studies.[7,18,22,23,25] However, Nikolopoulos and colleagues did assert that cystic tumors tend to have a higher growth rate (approximately 3.7 mm/year) as compared with solid tumors. Regardless, no other consistent indicators of growth were revealed across the multiple studies: not age, initial lesion size, duration of symptoms, tumor laterality, or patient sex. However, the authors did admit that there were several limitations to their review, including variability in follow-up, different use of imaging studies, multiple publications from different institutions, and even the inclusion of patients with neurofibromatosis Type 2 in some studies in which a clear distinction was not specified. Thus, further large-scale prospective studies are needed to effectively elucidate possible predictors of growth as well as potential mechanisms leading to growth or regression.

Management Selection Strategies

Currently, 3 basic options exist for managing newly diagnosed VSs: 1) microsurgical removal; 2) radiation therapy, including stereotactic radiosurgery and stereotactic radiotherapy; and 3) conservative management. Specific treatment decisions at the time of VS diagnosis are poorly standardized at this point, with extensive variation in protocols among practice settings, but in general they have been determined based on tumor size, associated symptoms, and specific patient indicators. Details on the efficacy of and outcomes for primary interventional therapies, namely the use of microsurgery and radiation therapy at the time of diagnosis, have been well characterized in the literature.[19,20,34,36] Recently, however, conservative management via serial radiological studies has become increasingly popular in patients with smaller tumors, especially when they are minimally or completely asymptomatic.[22,34,41] The reasoning behind adopting a "wait-and-scan" approach is based on two fundamental ideas. First, a high proportion of the tumors do not grow following diagnosis. Second, there is sparse evidence to indicate that the treatment of a nongrowing lesion is beneficial. In contrast, surgery or radiotherapy can result in significant complications and is best avoided when unnecessary.[20] Therefore, conservative treatment in patients with smaller tumors ultimately enables them to avoid the possible complications of primary intervention while allowing practitioners to closely monitor their status over time.

As discussed above, existing data on tumor growth rates, growth patterns, and predictors of growth suggest that there is extensive variability in tumor behaviors across patient populations and that there are few indicators that can accurately predict tumor growth. Given the dearth of consistent evidence, it is difficult to provide specific recommendations on who should undergo conservative management and how they should be monitored. However, a few protocols have been suggested to determine which patients should be conservatively monitored.[3,29,30,32] One of the most cited studies used the following criteria to select patients for conservative management: advanced patient age (> 60 years), poor health or significant medical risks for surgery, risk of further hearing loss (American Academy of Otolaryngology–Head and Neck Surgery Level A or Level B), small tumor size (Koos Grade I or 2), minimal or no incapacitating symptoms, and patient preference.[3] Other protocols have generally used similar selection criteria, including symptoms, patient age, and size of the tumor.

By definition, incidental VSs tend to be associated with few if any symptoms, with rare exceptions.[1,14] Additionally, these lesions tend to be smaller than symptomatic VSs, with one study showing a significant size difference of 1.09 versus 1.50 cm between asymptomatic and symptomatic lesions, respectively.[11] Furthermore, it has been suggested that small and medium-sized incidental VSs tend to have a more benign course and ultimately require less intervention (47% vs 76%, small and medium versus larger lesions).[11] Therefore, the majority of incidental VSs will likely qualify for conservative management according to the above criteria. As mentioned, a few exceptions do exist, including instances in which symptoms are elucidated with further history taking after a positive imaging study[14] or a large high-risk tumor is discovered. Asymptomatic VSs have been reported to be as large as 5 cm.[11]

Specific strategies for follow-up have also been recommended. Martin et al.[16] suggested an initial rescan with MRI at 6 months posttreatment followed by scans annually for 2 years and then every 5 years for the remainder of the patient's life. They recommended reassessment if growth > 2 mm occurs at any interval. The rationale behind their protocol was based on their findings that 90% of the patients demonstrating tumor growth did so before 3 years elapsed and the remaining 10% manifested growth within 6 years. Furthermore, the necessity of MRI as opposed to other screening options was emphasized. Such screening options included hearing tests, which were reported to be equivocal between lesions that grew and those that did not.[9,17,31] Interestingly, in line with the findings of Nikolopoulos et al.,[22] Martin and colleagues[16] also suggested that cystic tumors should be followed more closely, as they represented a disproportionate percentage of the tumors that grew, especially in cases in which a solid tumor converted into a cystic one.

The protocol established by Martin et al.[16] contrasts with the one proposed by Strangerup et al.,[32] who suggested annual scans for 5 years after treatment and then follow-up scans at 7, 9, and 14 years after treatment. Smouha et al.[29] proposed a different strategy, suggesting that those selected for conservative management should undergo MRI studies at 6 months after treatment and ev-

ery year thereafter. Regardless of the specific regimen, all of the studies agree that most tumors showing growth will do so early in the follow-up period. However, the lack of predictability for long-term growth or changes in growth patterns throughout the natural history of individual tumors requires extended follow-up of patients via serial imaging, and practitioners should anticipate growth at any interval.

While we do not specifically endorse one protocol over another or propose our own protocol for conservative management, we do believe that patients should be monitored at regular intervals throughout the course of their lives. Additionally, we support the notions of Nikolopoulos et al.,[22] who emphasized the significance of educating and counseling patients with incidental VSs about the lack of predictability in tumor growth and the importance of patient compliance. A thorough discussion about the method and frequency of follow-up should take place, and any barriers in the patient's life that can lead to poor compliance should be thoroughly explored. The insufficient follow-up of patients undergoing conservative management to monitor growth can result in catastrophic consequences and increase the risks associated with secondary intervention.[8]

Outcomes of Conservative Management

It is postulated that all symptomatic VSs probably have a silent asymptomatic period before presentation to a physician, providing the conceptual rationale for the existence of asymptomatic patients who present with incidental findings.[11] The evolution of symptoms and neurological deficits in initially asymptomatic or minimally symptomatic patients with conservatively managed VSs, while varying significantly from patient to patient, tends to follow certain patterns that correlate with certain stages of increasing lesion size as well as with the location of the lesion.[27] Given that most patients with incidental VSs probably qualify for conservative management, an understanding of how these tumors progress, the effect that this form of management has on the later development of symptoms, and how conservative management affects long-term quality of life is paramount to the successful long-term management of these lesions.

One of the most heavily studied outcomes in regard to conservative management is its effect on long-term hearing preservation. Hearing loss has been suggested to be an integral part of the natural history of VSs, possibly as a result of ischemia of the inner ear or protein shedding from the tumor.[9] Prior studies have shown that hearing may be preserved in up to 49% of individuals undergoing conservative management,[29] while hearing preservation rates vary between 17% and 68%[3] in patients undergoing primary microsurgery. Whitmore et al.[39] suggested that conservative management is associated with a significantly higher incidence of functional hearing loss; however, at this point, the comparison of hearing outcomes between conservative management and other forms of therapy is largely inconclusive.[3] The latest prospective study on conservative management by Breivik et al.[4] suggested that hearing deteriorates in patients over time, regardless of treatment or tumor growth.

Multiple attempts have been made to elucidate the risks for hearing loss based on various tumor and patient characteristics. Sughrue et al.[35] reported an overall hearing preservation rate of 54% in patients undergoing conservative management, showing that tumors with a slower growth rate (≤ 2.5 mm/year) were associated with a significantly higher rate of hearing preservation as compared with lesions that grew faster (75% vs 32%, respectively). However, other studies have suggested that tumor growth is not a reliable indicator of hearing loss.[3,6,24] Furthermore, Bakkouri et al.[3] attempted to classify risk factors based on initial tumor size. These authors reported that patients with preserved hearing tended to have larger initial tumor sizes (11.5 vs 9.3 mm). Taken together, these studies suggest that the impact of specific tumor factors, such as initial size and growth rates, is highly variable.

The relationship between hearing loss and quality of life in the context of other complications is also an important parameter requiring discussion. Breivik et al.[4] showed that hearing deterioration actually had little impact on quality of life, except in terms of social functioning in those with complete unilateral hearing loss. In contrast, previous retrospective analyses suggested that the negative impact of hearing loss was greater than all other complications, including weakness, numbness, spasm, pain, tinnitus, or even hydrocephalus.[39] However, these findings do not necessarily mean that the effect of hearing loss on the quality of life was a significant change from the baseline status of the patient since the study was conducted retrospectively. Again, these data show conclusions in this regard are highly variable. In general, we suggest taking a case-based approach to patients with incidental VSs, focusing on both the hearing status at various intervals and how it affects patients' quality of life and perception of their illness, ultimately using their subjective hearing experience combined with radiological indicators to guide further management.

Other symptoms besides hearing loss have also been discussed in terms of long-term outcomes of conservative management and patient quality of life. Of these symptoms, tinnitus, vertigo, and balance disturbances have been the most heavily studied. Whitmore et al.[39] suggested that the incidence of tinnitus and vertigo is significantly higher in patients undergoing conservative management as opposed to primary intervention. Tinnitus was also shown to be significantly associated with tumor size and type of hearing loss.[2] Interestingly, however, Breivik et al.[4] suggested that, after a period of conservative management, tinnitus was not significantly associated with a reduced quality of life as compared with baseline conditions. Additionally, they showed a significant reduction in the number of patients experiencing vertigo after a period of conservative management. However, the presence of vertigo was also associated with a significant reduction in quality of life. While it is clearly important to follow hearing loss and tinnitus as possible indicators of worsening clinical progression, the presence of vertigo may have the greatest impact on patient quality of life since it is often a debilitating symptom. Thus, the discussion of vertigo in addition to tinnitus and hearing loss is a vital part of patient follow-up in conservative management.

Conversion to Active Treatment

Understanding the frequency of treatment failure as well as when to intervene is an integral aspect of adopting a conservative management strategy. The overall failure rate, defined as conversion to an active treatment, has been found to be 15%–50%.[5,21,29,39,40] Smouha et al.[29] reported that 43% of patients undergoing conservative management ultimately demonstrate positive tumor growth. However, only 20% of the patients undergoing a conservative regimen required definitive treatment, suggesting that treatment failure is not directly linked to tumor growth. When analyzed prospectively, the likelihood of requiring treatment is 13.3% at 2 years and 41.3% at 5 years, suggesting that during a longer follow-up period more patients will require secondary intervention.[4] Fortunately, secondary intervention has been shown to be relatively safe. Numerous studies have also demonstrated that the incidence of postoperative complications is similar between patients undergoing primary surgery and those undergoing surgery due to failure of conservative management.[3] Note, however, that there was an increased risk of hearing deterioration with delayed surgery.[3] Furthermore, up to 83% of patients in whom conservative therapy fails will disproportionately undergo microsurgery.[39] This finding is interesting, suggesting that further investigation into factors determining the selection of microsurgery versus radiotherapy in secondary management, as well as the outcomes that these two strategies have on these patients, would be useful in guiding future management decisions.

Numerous indicators suggest the failure of conservative management and the commencement of secondary intervention. Overt clinical deterioration is a clear indicator to cease conservative management and implement secondary treatment. However, other less obvious indicators can be used as well. Currently, tumor growth rate is a commonly used parameter to indicate the failure of conservative management. Most studies suggest that in patients with small conservatively managed tumors, growth rates > 2–3 mm/year or significant worsening of symptoms should signal the need for treatment.[3,16,35] Bakkouri et al.[3] found that conservative management failed in 23.7% of their patients. They characterized treatment failure by a tumor growth rate ≥ 3 mm between two consecutive MRI studies, disabling vertigo, hearing deterioration, patient choice, or seeking a second opinion from another provider. When utilizing hearing loss as a parameter of failure, it has been suggested that a growth rate > 2.5 mm/year is a strong predictor of failed conservative management at the 3-year follow-up in patients with tumors < 25 mm on presentation.[35] However, it was shown that regardless of the presentation, more than half of the patients managed with observation did not show tumor enlargement (52%–57%), and only 16%–21% required therapy within a period of 3 years.[41] In general, secondary intervention was required in only a minority of patients managed conservatively and tended to be successful in most of these patients. Thus, we suggest that conservative management with careful follow-up is an appropriate therapy in almost all cases of incidental VSs.

Conclusions

With the advent of advanced imaging technologies, an increasing number of patients have been diagnosed with VSs as incidental findings on studies conducted for alternate reasons. These patients probably represent a significant fraction of all patients presenting for the management of VSs. Because these lesions are smaller, produce minimal symptoms, and are relatively indolent, a trial of conservative management is probably appropriate in the majority of cases. And although current data indicate that tumor growth occurs in a minority of patients, long-term growth is largely unpredictable and may follow a wide variety of specific patterns. Furthermore, patients undergoing conservative management can later present with symptoms that significantly affect their quality of life and everyday functioning. Numerous protocols for the selection of and follow-up in conservative management have been proposed, but no large-scale multiinstitutional prospective studies have compared the various options or addressed specific risk factors for tumor growth, symptom onset, or overall treatment failure. Thus, we recommend that practitioners take an individualized approach with early initial follow-up and continued lifelong management, emphasizing the importance of continued contact and observation for both clinical and radiological deterioration.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Liu, Schmidt, Boghani. Acquisition of data: Schmidt. Analysis and interpretation of data: Schmidt, Boghani, Choudhry. Drafting the article: Schmidt, Boghani. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Administrative/technical/material support: Liu, Choudhry, Eloy. Study supervision: Liu.

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Pitfalls in intraoperative nerve monitoring during vestibular schwannoma surgery

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Despite the widespread acceptance of intraoperative neurophysiological monitoring in skull base surgery over the last 2 decades, surgeon training in the technical and interpretive aspects of nerve monitoring has been conspicuously lacking. Inadequate fundamental knowledge of neurophysiological monitoring may lead to misinterpretations and an inability to troubleshoot system errors. Some surgeons perform both the technical and interpretive aspects of monitoring themselves while others enjoin coworkers (surgical residents, nurses, anesthetists, or a separate monitoring service) to perform the technical portion. Regardless, the surgeon must have a thorough understanding to avoid potential medical and legal pitfalls because poor monitoring is worse than no monitoring. A structured curriculum and protocol in both the technical and interpretive aspects of monitoring is recommended for all personnel involved in the monitoring process. This paper details the technical, interpretive, and surgical correlates necessary for optimal intraoperative nerve monitoring during vestibular schwannoma surgery with an emphasis on electromyographic monitoring for facial and recurrent laryngeal nerves. Just as the American Society of Anesthesiologists' 1986 "Standards for Basic Anesthetic Monitoring" became a useful tool for both patients and anesthesiologists, impending guidelines in intraoperative neurophysiological monitoring should likewise become an important instrument for optimizing intraoperative neurophysiological monitoring.
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Key Words • **acoustic neuroma** • **electromyography** • **facial paralysis** • **intraoperative neurophysiological monitoring** • **nerve monitor** • **vestibular schwannoma**

Intraoperative neurophysiological monitoring during VS surgery has become common practice among skull base surgical teams. While numerous modalities are available including auditory brainstem recording, motor and sensory evoked potentials, and vagus nerve monitoring via the RLN, it is facial nerve monitoring that is the simplest and most effective modality to improve outcomes following VS surgery. Despite the burgeoning use of nerve monitoring in skull base surgery, surgeon training in the techniques and interpretation of nerve monitoring has typically been marginal. This lack of knowledge and an inability to troubleshoot system errors can lead to poor monitoring and places the patient at risk for complications. A thorough understanding of the technology is vital to avoid medical and legal pitfalls.

Poor nerve monitoring is worse than no monitoring (J. Kartush, presentation to the American Society of Neurophysiological Monitoring, 1989). Poor nerve monitoring creates a false sense of security akin to walking in a minefield with a dysfunctional minesweeper. In this situation, the unprepared surgeon may have false confidence that the nerve is not being traumatized during dissection. With a thorough understanding of the principles and practice of nerve monitoring, the physician can have confidence that the integrity of the monitoring system is intact and be able to troubleshoot system errors when they occur. Optimal nerve monitoring in combination with sound surgical technique will provide the greatest chance for functional facial nerve preservation during VS surgery.

Abbreviations used in this paper: CMAP = compound muscle action potential; EMG = electromyography; RLN = recurrent laryngeal nerve; VS = vestibular schwannoma.

The Facial Nerve

Avoidance of facial nerve injury during resection of a VS is a primary concern of the skull base surgeon. Facial paralysis has potentially devastating functional, emotional, and social consequences for the patient. The inherent complexity of facial nerve anatomy, combined with tumor distortion of the nerve and adjacent structures, has prompted methods to minimize intraoperative facial nerve injury. While the introduction of the operating microscope and the development of transtemporal microsurgical techniques greatly improved skull base

surgical morbidity and mortality, it was the addition of intraoperative electromyographic facial nerve monitoring that resulted in dramatic reductions in postoperative facial palsy. Facial monitoring is helpful in localizing the nerve within a tumor distorted field and provides real-time neurophysiological status feedback. This feedback allows the surgeon to detect and avoid stretch or ischemic nerve injury that may not otherwise be apparent with a visualized intact facial nerve.

Many studies have demonstrated improved postoperative facial nerve outcomes with the use of intraoperative monitoring.[2,4,10,11,16] This dramatic improvement has led to the routine use of monitoring during VS surgery and has also been advised by the NIH.[13]

Neurophysiology

Electromyography is used intraoperatively to monitor for nerve injury during VS surgery. This test relies on measurements of the CMAP generated by the muscles of facial expression. Depolarization of the facial nerve leads to distal propagation of nerve action potential to the motor endplate, where it is translated into motor unit potentials emanating from the corresponding muscle fibers. These motor unit potentials when summed together result in the measured CMAP, which reflects the activity of the muscles of facial expression. Changes in CMAP reflect changes in the functional status of the facial nerve. Monitoring the CMAP allows an order of magnitude larger response than if a nerve action potential was recorded due to the amplifying effect of the muscle response.

An accurate assessment of nerve conduction with EMG requires stimulation proximal to the potential site of injury. When an electrical stimulus is applied distal to injury in the acute setting, a seemingly normal response may be obtained, because distal Wallerian axonal degeneration following severe nerve injury takes 48–72 hours to reach the motor endplate. Nerves suffering from mild to moderate trauma will exhibit reductions in amplitude and prolonged latency. Increasing injury requires an increasing amount of current to elicit a response. Often a combination of physiological conduction block (neurapraxia) and physically injured neural elements (axonotmesis or neurotmesis) will be evident after significant surgical trauma. These injuries will be variably represented on EMG by reduction in amplitude, increase in latency, and increase in threshold stimulation as the level of nerve trauma increases.

Intraoperative Facial Nerve Monitoring

Intraoperative facial nerve monitoring aids in localization of the nerve displaced by tumor distortion, detects nerve injury during dissection, and provides a means for assessing nerve function after dissection is complete. While many multimodality monitoring devices can be employed, when only EMG recording is required, many centers use dedicated EMG monitoring systems, which are simpler and typically provide direct auditory feedback of responses to the surgeon. Common systems we have employed include the Medtronic Nerve Integrity Monitor and the Neurosign system. The nerve monitoring system is an adjunct, not a replacement, for surgical skill and judgment in the assessment and preservation of neural structures. False-positive and false-negative errors can occur with monitoring; therefore, a knowledgeable surgeon and monitoring team are essential to troubleshoot system errors. If monitoring results contradict the surgeon's assessment of anatomy, tumor, or status of the nerve, then the surgeon should proceed with caution and may choose to reject the monitoring information until proper functioning of the monitoring system can be assessed.

It is important to be aware that EMG monitoring is disabled during the use of electrocautery. Electrocautery generates high intensity electrical artifact that typically overpowers the ability of the monitor to record low amplitude EMG activity. In fact, to reduce the disruptive noise artifact through the loudspeaker, most monitors defeat the loudspeaker during electrocautery. Thus, thermal nerve injury due to electrocautery may not be detected until after the injury has occurred and then only if the nerve has been stimulated to assess its function. Baseline stimulation should be performed as early and often as feasible using moderate-level mapping currents prior to initiating tumor dissection. Stimulation is especially important before and after any particularly risky surgical maneuver to ensure appropriate function of the monitoring system and to detect nerve injury at the earliest time possible.

Training

The successful use of intraoperative facial nerve and RLN monitoring requires the surgeon's interpretation of neurophysiological responses.[7] Furthermore, if the surgeon also takes on the responsibility of the electrode and device setup, specific training in the technical aspects of monitoring is required. Meticulous attention to detail must be paid to anesthesia, the monitoring device, and the electrodes to ensure accurate results. At our institution, the surgeon performs intraoperative monitoring in conjunction with a technologist who has received special training and certification (Certification in Neurophysiological Intraoperative Monitoring).

At many institutions, however, surgeons performing the technical setup have had little or no formal training. At academic institutions, residents who have had only cursory instruction by a coresident may perform the monitoring setup. Many staff surgeons may have had only a brief introduction to the technology through a product representative. Alternatively, some centers contract monitoring to either a service company or an in-house department such as Neurology or Anesthesia. Ultimately, it is incumbent on the operating surgeon to learn the technical and interpretive aspects of monitoring and to incorporate this knowledge into real-time surgical modification. It is suggested that an intraoperative nerve monitoring protocol be established at each institution in conjunction with a core curriculum and competency testing. Many institutions have similar requirements for new or high-risk procedures, such as the use of lasers, endoscopy, and others.

Technical Factors During Setup

To reliably perform EMG-based facial nerve moni-

toring, an intraoperative checklist is beneficial, similar to the orderly stepwise process performed preflight by commercial airline pilots before takeoff. Established protocols (Table 1) can diminish the chance for error by creating obligations that must be completed before progressing to the next step. In this manner, intraoperative monitoring errors may be promptly identified and remedied.

The first step is to ensure that the anesthesiologist avoids the use of long-acting muscle relaxants. Short-acting muscle relaxants such as succinylcholine are acceptable for anesthesia induction, except in the rare patient with pseudocholinesterase deficiency, in which prolonged paralysis is experienced. When in doubt, train-of-four EMG testing should be performed prior to the nerves being exposed.

The second step is the judicious use of local anesthetic (such as lidocaine or bupivacaine), which can chemically induce temporary paresis, rendering monitoring useless. Anesthetic injections near the stylomastoid foramen are to be avoided, especially in children for whom the mastoid tip may be poorly developed. In addition, local anesthetic may track into the middle ear with the potential to come into contact with the tympanic segment of the facial nerve, which may be dehiscent in approximately 20% of patients.[1]

The third step is careful electrode placement. For facial nerve monitoring, intramuscular needle electrodes are inserted in a closely paired manner at the nasolabial groove (orbicularis oris) and near the eyebrow (orbicularis oculi) on the side to be monitored (Fig. 1). Care must be taken to direct the electrodes away from the orbit to avoid inadvertent trauma to the eye. Two additional electrodes are placed subcutaneously over the sternum, one serving as a ground for the recording channels and the other as a return for the monopolar nerve stimulator. We routinely color code electrodes: blue for eyes, red for lips, green for ground, white for anode, and black for cathode. A reliable mnemonic for this set-up is "blue eyes, red lips, green ground." These electrodes are then connected to their corresponding nerve monitor headboards with the cathode (typically colored black) used for stimulation rather than the anode (white), because cathodal stimulation is more effective.

The fourth step is to check the impedances of the different electrode pairs. The independent electrode impedance should be less than 5 kOhm, while interelectrode impedance should be less than 2 kOhm. If impedance is too high, this suggests poor needle position or faulty electrodes; the electrodes should be repositioned or replaced and then retested.

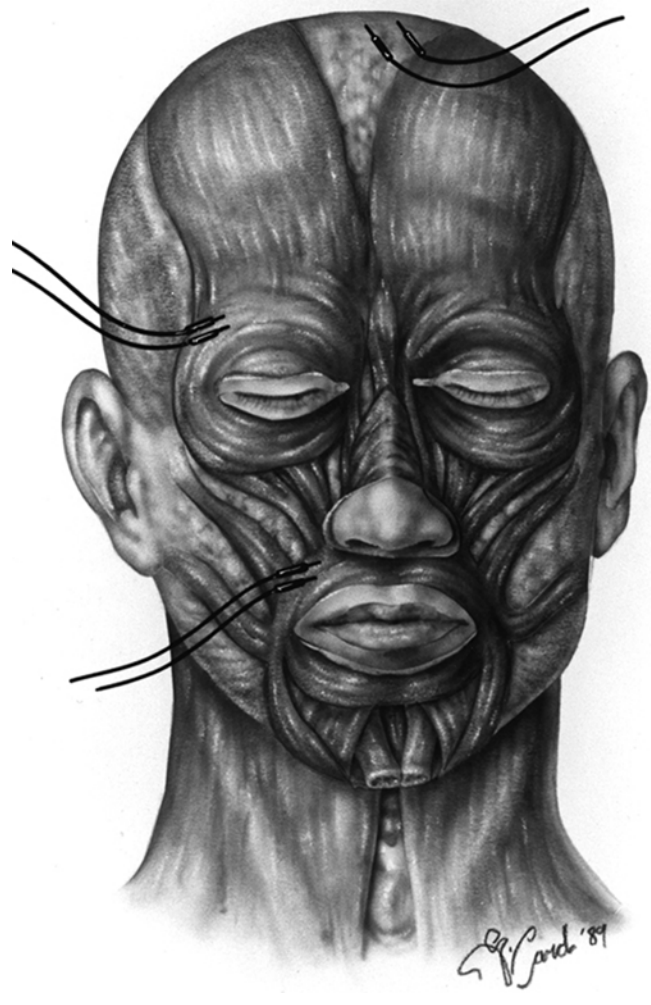

Fig. 1. Illustration of the EMG recording sites used for 2-channel facial nerve monitoring. Paired electrodes are placed subcutaneously at the nasolabial groove (orbicularis oris) and near the eyebrow (orbicularis oculi) on the side to be monitored. Ground and anodal return electrodes can be placed high on the forehead as shown, or more commonly today, near the sternum (not shown). Reproduced with permission from Kartush: **Otolaryngol Head Neck Surg 101:**496–503, 1989.

The fifth step is to perform a tap test to check the integrity of the connection from the electrode to the recording device. Tapping the skin over each pair of facial recording electrodes generates an electrical artifact. Most monitors will display this artifact as a visual signal on the oscilloscope, as well as a concurrent acoustic signal from the loudspeaker. This design allows those surgeons

TABLE 1: Facial nerve monitoring setup protocol

1. Ensure the anesthesiologist avoids use of long-acting muscle relaxants
2. Be wary of local anesthetic (lidocaine or bupivacaine), which can chemically induce a temporary facial paresis, rendering monitoring useless
3. Place electrodes carefully
4. After electrodes are connected to the nerve monitor, check impedances of different electrode pairs
5. Perform a tap test to check integrity from electrode to recording device
6. After incision and soft tissue exposure, check for current flow using nerve stimulator
7. Stimulate the nerve at an early point in surgery before any significant nerve manipulation is performed

who are monitoring without the help of a monitorist to receive real-time audio feedback. It must be remembered that the tap test only represents an intact connection from electrode to monitoring device. Performing a tap test on a paralyzed face will also create the same electrical artifact present in the case of a nonparalyzed facial nerve; a true CMAP is not obtained. Therefore, no information is obtained regarding the functional status of the facial nerve with the tap test.

Pairing of audio and visual feedback allows the monitoring team to be as vigilant as possible in regard to a change in nerve status. For example, for those surgeons relying on the technician watching the screen without audio feedback, an important response may be missed if the technician looks away for a moment. Conversely, devices that only have an audible response with no oscilloscope lose the opportunity to differentiate artifact response from true response. Multimodality monitoring including RLN monitoring, auditory brainstem recording, and somatosensory and transcranial motor evoked potentials increases the complexity of monitoring, which typically therefore mandates the need for a technical assistant.

The sixth step is to check for current flow using the nerve stimulator. After incision and soft tissue exposure, touching the stimulator to soft tissue or wet bone should result in near 100% conduction of current from the stimulator to the monitor. Most devices will display the returned current visually. Others may present an audible warble tone that is distinct from a true response beep tone. At this time, the volume of the nerve monitor should be adjusted to assure the surgeon can hear it over ambient operating room sounds (such as a drill or suction).

The seventh step is to obtain a baseline response by stimulating near the nerve at sufficient current to elicit a response. Doing so at an early point in the surgery before any significant nerve manipulation is performed establishes that all aspects of monitoring and anesthesia are optimized. The distance from the nerve and the amount of intervening tissue will determine the current setting needed to elicit nerve depolarization. A normal facial nerve will respond to as little as 0.05-mA stimulation when the probe is placed directly on the nerve in the cerebellopontine angle. However, with increasing distance, as well as intervening soft tissue, bone, CSF, or blood, a current of 1–2 mA may be required to obtain a baseline "far-field" response by volume conduction of current through tissue. Once a confirmatory baseline response is obtained, current intensity is reduced to the lowest possible stimulating levels based on nerve proximity. In the senior author's experience (J.M.K.), 30 years of using such mapping techniques with modern current settings (such as pulse durations of approximately 100 μsec at approximately 5 Hz) have resulted in no detrimental nerve effects.

The more difficult the tumor dissection, the more often electric stimulation should be performed to constantly assess both the location and integrity of the nerve. Using insulated stimulating surgical instruments such as Kartush Stimulating Instruments (Neurosign) allows simultaneous surgical dissection with frequent electrical stimulation. Once the tumor has been resected, a final threshold response to stimulation should be obtained for comparison with the baseline amplitude.

Intraoperative RLN Monitoring

When VSs are large and extend significantly toward the jugular foramen, consideration should be given to monitoring the vagus nerve by way of its RLN. Similar to facial nerve monitoring, RLN monitoring is also based on EMG. Therefore, many of the principles and potential pitfalls discussed with respect to facial nerve monitoring also apply to RLN monitoring, with a few technical considerations. However, all monitoring modalities have their particular differences, which should be clearly understood to avoid error.

Today, the most commonly used method of intraoperative RLN monitoring utilizes surface electrodes along an endotracheal tube. Direct intramuscular needle placement can be effective but had practical challenges that limited its use, such as performing direct laryngoscopy to insert needles into the vocal cords. Consequently, while not ideal, RLN monitoring is most commonly performed today using laryngeal surface recording electrodes placed on endotracheal tubes. These may be premanufactured and attached to an endotracheal tube (the Medtronic Nerve Integrity Monitor EMG endotracheal tube), or they may be applied externally to the endotracheal tube using stick-on surface electrodes (Neurosign, IOM Solutions). This surface technique requires precise placement of the tube-electrode array during intubation such that electrodes directly contact the true vocal cords to allow recording of laryngeal musculature CMAP. By "hitchhiking" along with the endotracheal tube, the anesthesiologist becomes a key player in the procedure and must understand the proper method to optimize placement under direct laryngoscopic vision, often aided by devices such as the Glidescope (Verathon). Intraoperative head rotation or extension may displace the tube; therefore, consideration should be given to laryngoscopic reassessment of tube position if there is any doubt in tube-electrode positioning. Topical laryngeal anesthetic (such as viscous lidocaine) should be avoided during intubation as well, because this could temporarily cause vocal cord paralysis preventing effective monitoring.

It must be remembered that laryngeal surface electrode contact must be precise with an optimal electrode-to-vocal cord fit. Endotracheal tube size must be considered. A tube with a small outer diameter will result in poor electrode contact, whereas too large of a tube may cause trauma during and following intubation. While the stick-on type laryngeal electrodes allow the anesthesiologist to choose whichever endotracheal tube he or she prefers, unfortunately, as of this writing, Medtronic only offers their Nerve Integrity Monitor endotracheal tubes in sizes 6, 7, and 8; there are no half sizes or pediatric sizes available in the US, which reduces options for glottic sizing. Stick-on electrodes should be taped 1–2 cm above the endotracheal cuff and can be used with any size endotracheal tube to optimize fit. Lubricants should be avoided. Additionally, with longer procedures, the moist environment of the airway can cause these adhesive electrodes to become displaced. New adhesives are being tested.

In regards to RLN stimulation, the surgeon should

remember a few points when mapping for the lower cranial nerves in the posterior fossa. Vagal stimulation at the pars nervosa appears to be safe at stimulation levels of approximately 0.5–1 mA; in our experience, no bradycardia or other untoward vagal effect has been encountered at these levels. However, it is wise to avoid high-level stimulation of the nearby spinal accessory cranial nerve, as this can cause a sudden vigorous muscle contraction of the ipsilateral trapezius and sternocleidomastoid. This contraction can cause a sudden shift that can startle the surgeon; therefore, begin with 0.5 mA and increase amplitude slowly as needed.

Another caveat clinicians should be aware of concerns the Nerve Integrity Monitor EMG endotracheal tube wire reinforcement within the tube. While this reinforcement provides a generally desired increase in rigidity during intubation, the wire can become kinked or unraveled, leading to airway obstruction. To date, the reinforcement wires have been secured by a thin layer of silicone within the lumen of the endotracheal tube, rather than being integrated into the harder outer Silastic, as is common in other "reinforced" or "armored" tubes. Instrumentation of the tube (such as with a tube exchanger or catheter suctioning) can lead to unraveling of the wire within the tube, leading to buildup of clot/mucus with resultant airway obstruction (Fig. 2).[3] Similarly, the patient may inadvertently bite on the tube, resulting in intraluminal wire kinking and obstruction of the endotracheal tube. Thus, to minimize these airway risks, surgeons, technologists, and anesthesiologists should be familiar with the Medtronic package insert caveats: a bite block should be used, and intraluminal manipulations of the Nerve Integrity Monitor tube should be avoided.

None of the described laryngeal electrodes/tubes are intended for use postoperatively, nor are they MRI compatible. When prolonged postoperative intubation is anticipated, the electrode-equipped endotracheal tube should be replaced with a standard endotracheal tube.

Monitoring Interpretation

The prepared surgeon must have a clear understanding of the monitoring technology to interpret data received and adjust surgical manipulation accordingly. The team should be aware that EMG responses can result from many causes such as trauma, ischemia, electric stimulation, thermal changes, patient movement, artifact, and others. Responses may be detected during continuous free-running EMG or during intended electrical stimulation. Prass and Lüders[14] described 2 types of CMAP activity depending on the type of nerve irritation. The first type is the "burst" potential, which consists of a single polyphasic response due to near simultaneous activation of multiple motor units (Fig. 3A). Burst potentials are observed after direct contact of the nerve with surgical instrumentation and are fatiguable with repeated nerve contact. To provide real-time surgeon feedback, these responses will also be represented by a synchronous EMG "click" or "beep" sound produced by the neuromonitor loudspeaker.

The second type of continuous free-running CMAP is a "train" potential. This potential lasts seconds to minutes and is generated by multiple asynchronous responses from different motor units (Fig. 3B). This potential will be represented by a sound from the loudspeaker described as that of popping corn or an airplane engine. The train potential may be caused by mechanical injury (pressure or stretch) or thermal changes to the nerve. With increasing nerve injury, a greater intensity and longer duration of nerve potential will be evident. These findings should prompt the surgeon to adjust or suspend dissection to minimize nerve trauma. With nerve irritation, train potentials may last for several minutes and occasionally, with facial monitoring, 1 channel has prolonged train activity and the other channel does not. Some devices allow 1 channel to be silenced under these circumstances, but one should frequently return to check the silenced channel and unmute as soon as possible to avoid missing relevant feedback.

Thermal nerve injury after the use of laser or electrocautery deserves a special note because this injury may only become evident on EMG in a delayed manner. It may be represented as a gradual increase in baseline, but may also be associated with electrical silence. This finding is important, particularly with electrocautery, because the monitor is disabled during electrical artifact silencing. In this situation, electrocautery nerve injury may result under electrical silence without train EMG potentials. Therefore, if electrocautery injury is suspected, the nerve should be electrically stimulated as soon as possible to determine whether the nerve has been injured and where the injury may have occurred along the nerve's course.

Other nontraumatic causes of train potentials to consider include drill vibration energy transmission to the nerve, temperature irritation from hot or cold irrigation, irritation from osmotic hypertonic saline irrigation, and lightening of general anesthesia resulting in facial muscle fasciculations. Simply aspirating CSF in the cerebellopontine angle may result in a sudden drop in temperature immediately adjacent to the nerve, triggering a train potential. Therefore, some train potentials are not indicative of trauma, but instead represent a transient change in nerve physiology homeostasis and must be kept in mind when interpreting monitor feedback.

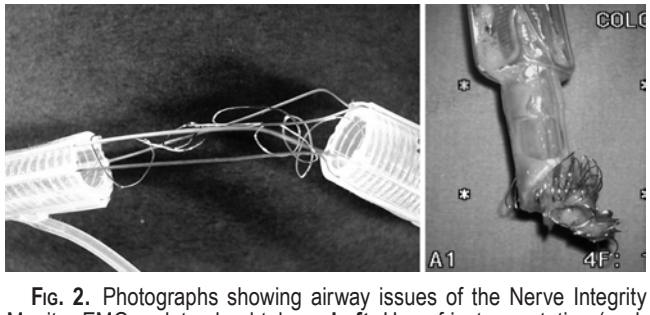

Fig. 2. Photographs showing airway issues of the Nerve Integrity Monitor EMG endotracheal tube. **Left:** Use of instrumentation (such as a suction catheter) can lead to unraveling of the wires within the endotracheal tube and potential airway compromise. Reproduced with permission from Evanina and Hanisak: **AANA J 73:**111–113, 2005. **Right:** A second example of airway obstruction with the Nerve Integrity Monitor endotracheal tube, secondary to fibrin clot/mucus buildup on the exposed wires within the lumen of tube.

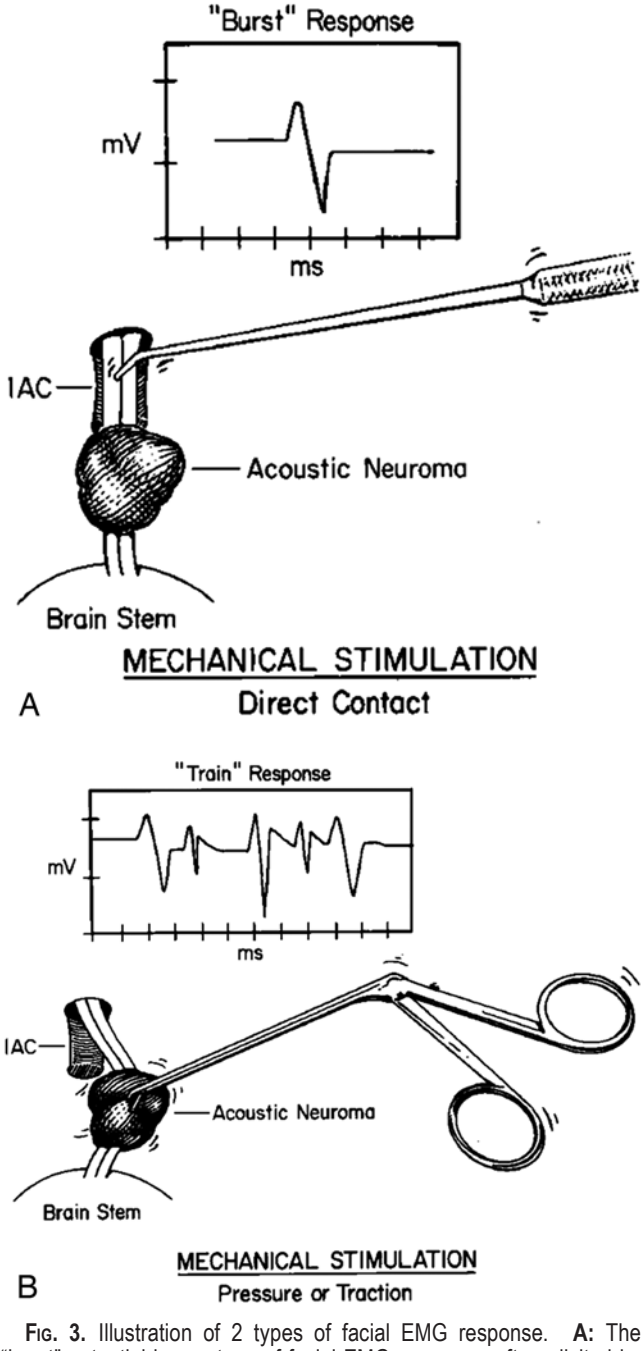

Fig. 3. Illustration of 2 types of facial EMG response. **A:** The "burst" potential is one type of facial EMG response, often elicited by mechanical contact of surgical instruments with the facial nerve. **B:** The "train" potential is a second type of facial EMG response, often due to mechanical/ischemic injury (pressure or stretch) to the nerve from dissection or retraction. IAC = internal auditory canal. Reproduced with permission from Kartush: **Otolaryngol Head Neck Surg 101:**496–503, 1989.

The second manner in which intraoperative nerve monitoring is used is via nerve stimulator–generated EMG. A stimulating probe instrument is touched to or near the nerve, which generates a true stimulus-triggered EMG response. This potential is represented by a "machine gun" sound on the loudspeaker composed of precisely timed potentials coincident with the stimulus frequency, typically set to 3–5 Hz. Stimulating current levels, ranging from 0.05 to 2 mA, are adjusted accordingly in mapping out the rough location of the nerve.

There are numerous commercially available monopolar and bipolar stimulating probes, each with their own distinct advantages. Bipolar stimulators are superior when discrete differentiation of nerve from adjacent tissue is required (such as when differentiating the facial from the vestibular nerve), whereas monopolar stimulation is optimal when mapping the general location of the nerve. However, monopolar stimulators can be made to be quite selective by progressively decreasing the current intensity as the nerve is approached. As noted, the Kartush Stimulating Instruments have added function in that they allow the surgeon to stimulate and dissect concurrently, because they are shaped liked conventional microsurgical instruments.[9] Conventional probes can be used intermittently to stimulate the nerve but sharp transection of the nerve with cold instrumentation may not elicit an EMG response until after the transection has already occurred. Stimulating instruments, on the other hand, will elicit an electrically evoked CMAP during the dissection to provide the surgeon ongoing feedback on nerve location and integrity.

False-positive errors can occur with stimulus-triggered EMG by a phenomenon known as "current jump." This happens when volume conduction of current through nearby tissues leads to stimulation of the nerve (Fig. 4 upper). An inexperienced surgeon may incorrectly identify the stimulated structure as the nerve of interest, leading to surgical error. This error can be minimized by using bipolar stimulating instruments and/or the lowest monopolar current mapping levels possible to obtain a response.

False-negative errors can also occur with stimulated EMG. Due to the presence of CSF and blood in the field, electrical insulation along the stimulator's shaft is required to reduce "current shunting" away from the nerve of interest (Fig. 4 lower). Also, high quality insulation is needed to avoid insulation cracking and inadvertent loss of fragments intracranially. Due to this occurrence being precipitated by repeated autoclaving of reuseable probes, there is now a trend to single use probes, including Kartush Stimulating Instruments.

In regard to RLN monitoring interpretation, there are several limitations to consider. First, laryngeal surface electrodes lack the sensitivity and stability of intramuscular needle electrodes.[5] Second, suboptimal positioning of surface electrodes may markedly impair the ability to detect laryngeal responses (false-negative error), and may also lead to erroneous recording of EMG activity from the adjacent pharyngeal musculature (false-positives), particularly at high levels of stimulation.

Last, failure to respond to stimulation can be due to an anatomically disrupted nerve, but can also be due to lesser levels of trauma. Other possibilities, such as anesthetic paralysis of the nerve and incorrect setup of the nerve monitor, must always be considered. Thus, failure to respond to stimulation cannot, by itself, be used as a determinant to resect and graft an unresponsive nerve segment. Instead, this measure is typically only taken with apparent physical disruption of the nerve.

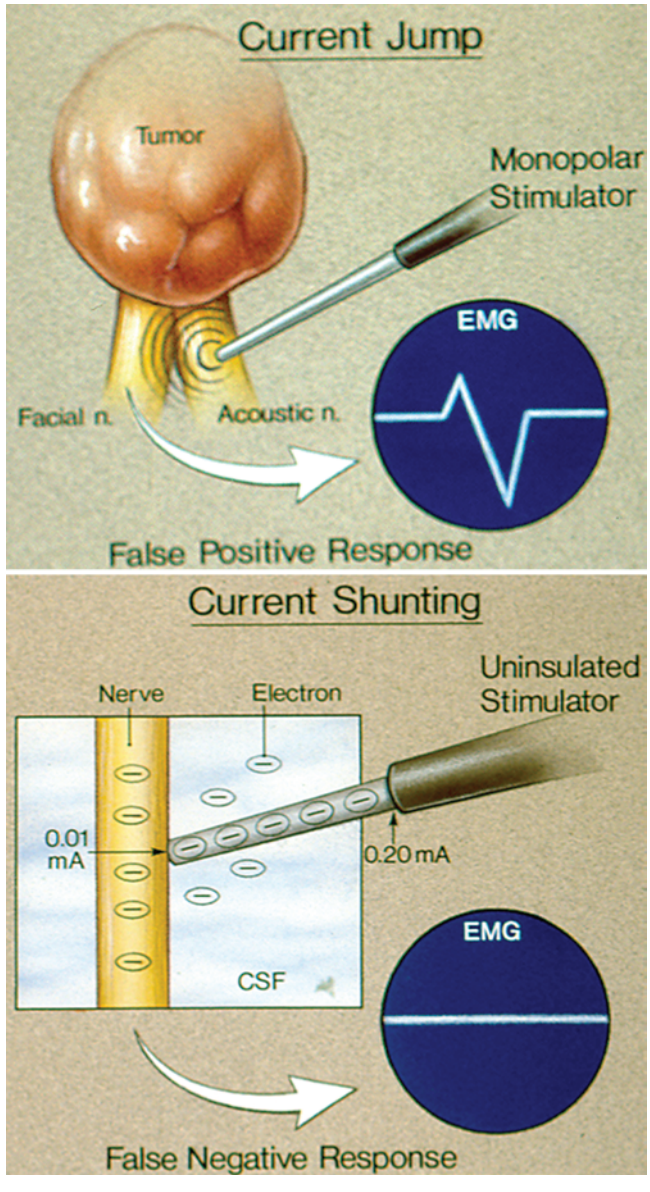

Fig. 4. Illustration of current jump **(upper)** and current shunting **(lower)**. **Upper:** Monopolar stimulation of a structure (such as the acoustic nerve) near the facial nerve can result in a false-positive response if the current jumps via volume conduction to the nearby facial nerve. This event tends to occur at higher current levels and can be minimized by using lower current levels or bipolar stimulators. n. = nerve. **Lower:** False-negative responses can occur with current shunting, if CSF, blood, or soft tissue shunts current from the nerve stimulator away from the facial nerve. Reproduced with permission from Kartush: **Otolaryngol Head Neck Surg 101:**496–503, 1989.

Facial Nerve Outcomes

The use of intraoperative monitoring for VS surgery has resulted in improved facial nerve outcomes.[8] A number of studies have specifically examined the correlation between intraoperative facial nerve monitoring findings and postoperative facial nerve outcomes. Lower final stimulus thresholds, higher CMAP response amplitudes, and lower ratios of proximal to distal stimulation thresholds have been correlated with better postoperative facial nerve outcomes.[12,18] Recently, Prell et al.[15] found that a real-time CMAP measure of train potential time (the A train) is relevant to facial nerve outcomes. Specifically, they found that if the total accumulated A train potential time was greater than 2.5 seconds, consideration should be given to limiting further dissection, because postoperative facial nerve weakness was more likely.

Vagal EMG monitoring has also been shown to reduce nerve morbidity following skull base surgery.[17] Authors in this study found that intraoperative monitoring allowed for improved nerve localization in situations of severely distorted anatomy and that the immediate intraoperative feedback affected the extent and manner of surgical manipulation. These authors concluded that these factors facilitated gross-total tumor resection with cranial nerve preservation.

The real-time neurophysiological feedback provided by intraoperative nerve monitoring benefits the surgeon in always knowing the functional status of the nerve. Monitoring has shown the benefits of sharp over blunt dissection when the nerve is clearly delineated visually or by electric stimulation. Past surgical techniques of "rough and rapid" debulking have been modified due to the neurophysiological feedback provided to surgeons, such as EMG train potentials associated with overzealous blunt debulking and retraction.

Implementation of Intraoperative Checklist

To improve the quality of nerve monitoring, there has been a national movement to establish standards in monitoring techniques and to offer credentialing to those who complete this training. In fact, future malpractice claims are likely to be based on these evolving standards. As a corollary, in 1986, there was great controversy among anesthesiologists as to whether a document titled "Standards for Basic Anesthetic Monitoring" (http://www.asahq.org/For-Members/Standards-Guidelines-and-Statements.aspx) should be adopted by the American Society of Anesthesiologists. Fortunately, it became a useful tool for both patients and anesthesiologists. Likewise, we suggest that impending national guidelines already being developed by the American Society of Neurophysiological Monitoring be adopted and followed by surgeons and their teams to optimize intraoperative neurophysiological monitoring.

Conclusions

Surgeons and their ancillary personnel must be cognizant of the potential limitations and pitfalls of intraoperative neurophysiological monitoring. Ultimately, it is incumbent upon the surgeon to understand both the technical and interpretive aspects of intraoperative nerve monitoring to minimize errors and optimize patient safety.

Disclosure

Dr. Kartush is a consultant to Magstim, a manufacturer of intraoperative monitoring devices. He was a past consultant to Medtronic and was the founding president of the American Society of Neurophysiologic Monitoring.

Author contributions to the study and manuscript preparation include the following. Conception and design: Kartush. Analysis and interpretation of data: Kircher. Drafting the article: both authors. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors. Approved the final version of the manuscript on behalf of both authors: Kartush.

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Intraoperative neuromonitoring techniques in the surgical management of acoustic neuromas

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Unfavorable outcomes such as facial paralysis and deafness were once unfortunate probable complications following resection of acoustic neuromas. However, the implementation of intraoperative neuromonitoring during acoustic neuroma surgery has demonstrated placing more emphasis on quality of life and preserving neurological function. A modern review demonstrates a great degree of recent success in this regard. In facial nerve monitoring, the use of modern electromyography along with improvements in microneurosurgery has significantly improved preservation. Recent studies have evaluated the use of video monitoring as an adjunctive tool to further improve outcomes for patients undergoing surgery. Vestibulocochlear nerve monitoring has also been extensively studied, with the most popular techniques including brainstem auditory evoked potential monitoring, electrocochleography, and direct compound nerve action potential monitoring. Among them, direct recording remains the most promising and preferred monitoring method for functional acoustic preservation. However, when compared with postoperative facial nerve function, the hearing preservation is only maintained at a lower rate. Here, the authors analyze the major intraoperative neuromonitoring techniques available for acoustic neuroma resection.
(http://thejns.org/doi/abs/10.3171/2012.6.FOCUS12194)

Key Words • **acoustic neuroma** • **vestibular schwannoma** • **intraoperative neuromonitoring** • **microneurosurgery** • **tumor resection** • **hearing preservation** • **facial nerve preservation**

Acoustic neuromas (vestibular schwannomas) are categorized as benign, extraaxial brain tumors (Fig. 1) developing near the internal auditory canal, typically with involvement of the cerebellopontine angle.[32,53,60,130,143] Advances in treatment modalities have popularized the application of less invasive management methods such as radiotherapy and radiosurgery,[100,138] which carry high efficacy and low morbidity.[31,53,57,86,87,100,101,107] However, many acoustic neuromas, particularly those that are large in size, necessitate surgical intervention.[33,36,62,101,107,110]

The primary operative goals are gross tumor debulking while safeguarding the adjacent cranial nerves (Fig. 1).[4,9,36,49,103,115,118,124,130] Neural preservation is particularly imperative in the contemporary management of acoustic neuromas.[5,68,103] By virtue of their location, these tumors are close to the facial and vestibulocochlear cranial nerves (Fig. 1), and can thus severely impair the nerve function at the time of initial presentation.[60,62,68,79,143] The neuroma can directly impinge, tightly adhere to, or overtly damage the nerves.[13,22,38,60] These tumors often present as operative challenges, as resection may cause nerve irritation or injury leading to neurapraxia, axonotmesis, or neurotmesis.[38,117]

The various options of surgical approaches (translabyrinthine vs middle fossa vs retrosigmoid) for acoustic neuromas and their respective patterns of postoperative cranial nerve preservation have been described.[3–6,15,23,25,45,49,56,58,61,65,67,112,115,117,118,124,130,131] However, IONM may demonstrate improvements in structural and functional preservation of the cranial nerves during these operations.[139,148] Several IONM techniques have been developed and evaluated with particular focus on CN VII and VIII preservation. Among these methods, the most frequently

Abbreviations used in this paper: BAEP = brainstem auditory evoked potential; CN = cranial nerve; CNAP = compound nerve action potential; ECOG = electrocochleography; EMG = electromyography; IONM = intraoperative neuromonitoring; IOVM = intraoperative video monitoring; MUP = motor unit potential.

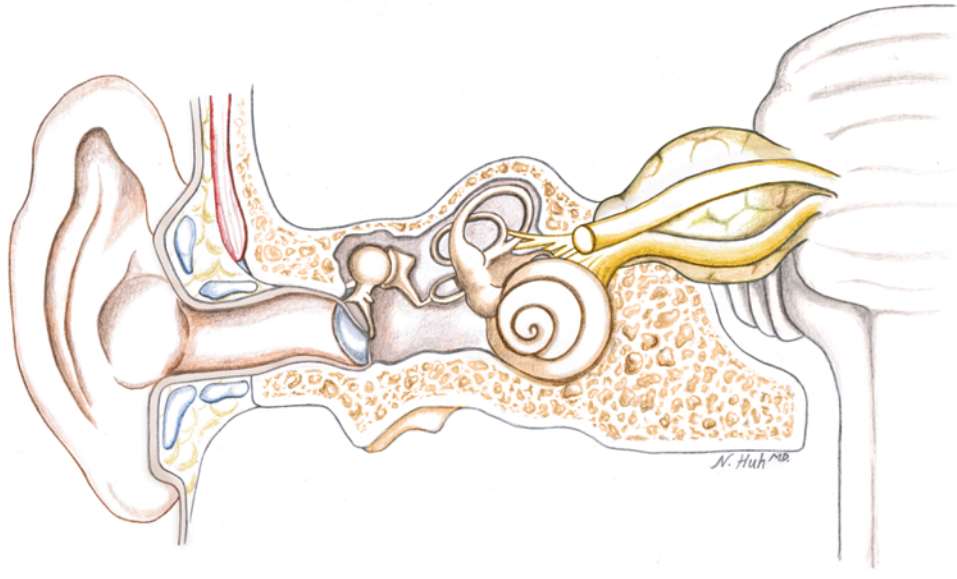

Fig. 1. Illustration showing an acoustic neuroma with displaced facial and cochlear nerves, the nerves we are trying to preserve. Printed with permission from Dr. Nancy Huh, M.D., Illustrations.

used are EMG for the facial nerve and BAEP monitoring for the vestibulocochlear nerve.[41,42,79,142,146,148] Here, we assess the fundamental characteristics underlying the major techniques available in IONM, emphasizing specific advantages and limitations of their utilization for optimal patient management.

Intraoperative Monitoring of the Facial Nerve (CN VII)

Cranial nerve VII plays a critical role in facial muscle function and one's cosmetic appearance, and its weakness can have severe and profound implications on a patient's quality of life.[72,92,140,148] For instance, loss of facial nerve function can ultimately result in an inability to blink, secrete tears, or speak properly, thus imposing a significant burden on the patient.[6,85] Such significant outcomes were once considered a probable morbidity.[78,92,117] However, with the advent of facial neuromonitoring, the morbidity once associated with acoustic neuroma resection has been drastically reduced. The House-Brackmann Grading Scale,[43] which ranges in increasing severity of deficits from Grade I through Grade VI, serves as a standardized method for analyzing postoperative outcomes of facial nerve function. As a result of advances in microneurosurgery and facial nerve IONM, many patients with smaller tumors have minimal functional loss of the nerve, as indicated by low House-Brackmann grades.[3–7,12,27,36,46,62,110,115,117,124,130,146] In patients with larger tumors, the outcomes are not as optimistic, as these patients are at an increased risk of postoperative facial nerve deficits.[4,27,36,56,60,62,117,125]

Electromyography

The use of EMG to monitor facial nerve function has been well documented, leading to its widespread application in modern practice.[9,22,39,40,46,54,82,94,125,126] The operative EMG device consists of a stimulator probe and a "sensor" that detects contractions of the facial muscles. Most operations use a minimum of 2 channels to observe the activity of the orbicularis oris and orbicularis oculi muscles,[12,46,77,82,90,115] although the use of additional channels to observe other facial muscles may provide further benefit.[37,38,85,134] When considering a 2-channel system, a pair of needle electrodes are usually planted in the orbicularis oris and orbicularis oculi muscles while another is placed on the forehead or shoulder for grounding.[9,12,54,82] Prior to the operation, the baseline electrical parameters, including MUPs and insertional activity, of these muscles are measured and recorded for future comparisons.[22,78]

The stimulator probe is applied to determine the location of the facial nerve.[21,40,126,148] During an operation, the ideal location for applying the probe on the facial nerve is near the brainstem[13,37] because it is proximal to the area of resection. Distal stimulation, while possible, yields limited data, as stimulation is being directed on the portion of the nerve that is virtually unaffected by resection.[35] However, distal stimulation is not to be ignored, as several studies have found that higher proximal-to-distal EMG amplitude ratios successfully predict postoperative facial nerve function.[35,46,47,137] When delivering the stimulus, the amount of current that is administered by the probe can be adjusted.[21,22,38,70,90,126] Once the amount of current applied exceeds the action potential threshold of the patient's facial nerve,[111] an action potential is fired that causes twitching of the facial muscles.[22,126] The sensor detects these facial movements and emits a sound alarm, thereby providing direct, immediate, and real-time feedback.[9,34,66,111,126] The facial muscle MUPs corresponding to this stimulation are also projected onto an oscilloscope to facilitate visualization.[39,42,82]

The electrical morphology, frequency, and characteristics of the MUPs vary greatly, and such divergences offer insights into possible abnormal nerve activity.[42,77] As demonstrated in Fig. 2,[111] multiple types of MUP sig-

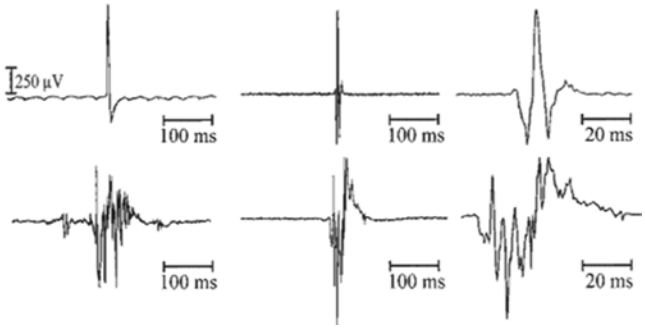

Fig. 2. Electromyography MUP morphologies demonstrating spikes *(upper)* and bursts *(lower)*. Reprinted with permission from Romstöck et al: *J Neurosurg 93*:586–593, 2000.

nals can be observed on an intraoperative EMG study. A single MUP wave is referred to as a "spike," while a short chain of MUPs is classified as a "burst." When a sustained streak of MUPs is distinguished, it is designated as a "train,"[77] which is shown in Fig. 3.[111] Train MUPs possessing a particularly high frequency (greater than 30 Hz) are termed "neurotonic."[41,42,77]

Neurotonic train activity typically serves as an indicator of intense nerve stimulation, as robust nerve stimulation correlates with greater MUP activation.[77] During an operation, neurotonic discharges can occur in the context of nerve stimulation, irritation, or damage.[41,42,148] However, not all neurotonic train waves carry equal clinical significance. The A-train pattern has been most substantially affiliated with postoperative facial nerve deficits (Fig. 3).[111] The A trains are characterized as a high-frequency train pattern with the following features: a duration lasting milliseconds to seconds, an amplitude in the range of 100–200 μV, and a short onset and offset.[104,105,111,148] The duration of "train time," as quantified by the seconds of A-train activity, has been shown to translate to worse postoperative facial nerve paresis.[21,104,105] Other train patterns that may be encountered on an EMG include the B and C trains, although they have not been shown to carry significant value in predicting postoperative nerve function.[111] As described by Romstöck et al.,[111] B trains manifest either in a spike or burst pattern and are distinguished by their gradual onset, low amplitudes, and average duration lasting minutes to hours. C trains, on the other hand, are irregular waveforms of varying amplitudes that bear resemblance to interference. Aside from train time and activity, other electrical EMG findings bear clinical importance as well. Mandpe et al.[66] reported that low immediate postoperative stimulation thresholds in combination with a response amplitude appeared to reliably foretell excellent postoperative facial nerve function. Neff et al.[90] reached similar conclusions but with stimulation thresholds of 0.05 mA or lower and amplitudes greater than 240 μV.

Electromyography provides several benefits. One of its main functions is determining the anatomical location of the facial nerve.[22,126] Direct, pinpoint visualization of the nerve may often prove difficult, as the tumor, its capsule, and bone may interject along the nerve's trajectory. By adjusting and determining the current required for muscle stimulation, however, the relative proximity of the nerve to the probe can be deduced. If there is very little tumor, tissue, or bone covering the nerve, the facial nerve will be more prone to stimulation at lower currents, such as lower than 0.2 mA, thereby implying that the nerve is highly exposed, close to the probe, and in danger of being manipulated.[22,126] Conversely, stimulation at higher currents, such as greater than 0.5 mA ,suggests the presence of a sizable tissue or bone barrier between the nerve from the probe.[126] Highly adherent tumors have a tendency for creating thicker barriers from the probe, thus resulting in higher mean stimulation thresholds.[38]

In addition, EMG helps prevent unplanned manipulation of the facial nerve by emitting a warning noise whenever muscle stimulation is detected. This can warn the surgeon of impending danger and thus advise cessation of current actions or recommend extreme caution. By doing so, EMG directly influences surgical planning and strategy, as the surgeon can appropriately alter the surgical ap-

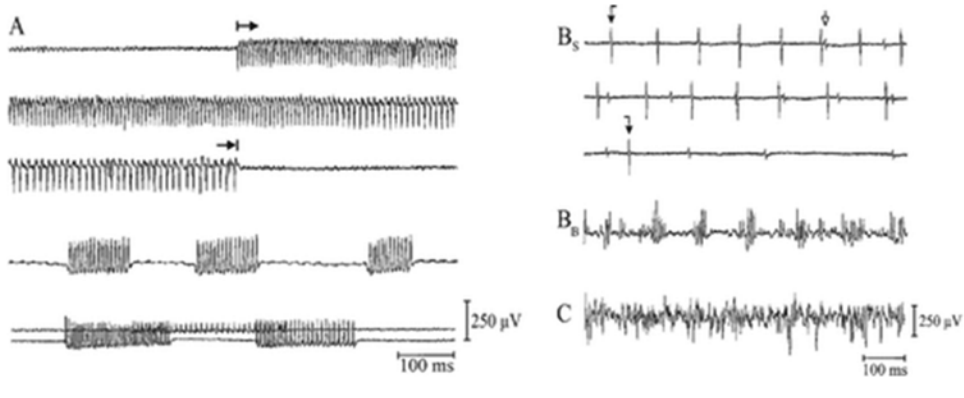

Fig. 3. Electromyographic train activity. **Left:** Examples of A trains of various durations and frequencies. The *upper* tracings show the abrupt onset and termination *(arrows)* of this sinusoidal waveform pattern, which lasted 1600 msec. The fourth train from the top of the figure shows repeated short-term periods of activity, ranging in duration from 100 to 120 msec each. The *lower* A train, which was simultaneously recorded from 2 facial muscle groups, gives an impression of frequency variability between 120 and 190 Hz. **Right:** Waveforms defined as B trains with spikes (B_S) and B trains with bursts (B_B) as predominant single components. The *black* and *white arrows* mark 2 individual B trains with spikes of higher and lower amplitudes recorded in the same channel. The *lowest* tracing represents irregular EMG activity, called a C train. Reprinted with permission from Romstöck et al: *J Neurosurg 93*:586–593, 2000.

proach to avoid causing damage to the nerve.[9,25,46,82,111,125] A-train activity or other abnormal EMG patterns may also encourage caution,[39] although they must be placed in context: neurotonic discharges can sometimes fire even in the presence of a transected nerve.[42] In addition to perioperative nerve preservation, EMG can help clarify the residual function of the nerve postoperatively.[46,54] When comparing postoperative and baseline stimulation thresholds, patients who require high or higher postoperative currents may have endured some degree of nerve injury.[54,70,93,123,126,128,150]

Despite its benefits, EMG is not an infallible monitoring system. During resection, the facial nerve may appear grossly intact; however, this finding does not necessarily convert to true nerve functionality.[11,22,60,70,92] One possible explanation for this phenomenon is that EMG can sometimes receive poor data input. This issue is particularly salient with the application of microinstruments to cauterize tissue or tumor surrounding the facial nerve.[41,85] The generated electrical signal may create artifact, signal interference, and distortion.

Electromyography also runs the added risk of instigating electrical injury from overstimulation. As general principle, application of the stimulator probe should be done conservatively to avoid inducing iatrogenic injury to the facial nerve. Intense or prolonged stimulation theoretically increases the risk of causing irreparable nerve injury.[106,127] To that end, several techniques are encouraged to diminish the risk of injury. Pulsed stimulation, for example, appears to have a lower injury risk than constant stimulation.[26,106] In addition, monopolar stimulation with constant voltage may be superior to bipolar constant-current stimulation.[82] However, the majority of experimental studies done to examine the potential for overstimulation have been conducted in animal models. Through these studies, one of the emerging general principles has been the greater influence of stimulus frequency on the degree of nerve injury. In rats, Sapmaz et al.[119] investigated the respective effects of stimulus amplitude (mA) and frequency on histological axonal degeneration. The authors' results demonstrated that frequency, but not amplitude, was statistically significant in causing greater axonal degeneration. In other words, rats with 20 stimulations had more degeneration than those that underwent 10 stimulations ($p < 0.05$), while rats with 30 stimulations had more degeneration than those that received 20 stimulations ($p < 0.05$).[119] In a cat model, McCreery et al.[76] obtained similar results: stimulation at 100 Hz versus 50 Hz caused greater axonal degeneration, while stimulus amplitude did not appear to have much effect. Another important principle is the superiority of pulsed stimulation when compared with constant stimulation. In mice, pulsed stimulation was associated with less myelin and axonal degeneration.[44] In a cat study, investigators found that extended periods of high frequency stimulation caused greater injury and that pulsed stimulations can reduce the risk of damage.[2] Interestingly, Kartush et al.[55] found that constant current stimulation can be safely applied in guinea pig models as long as the electrode is properly insulated to preclude shunting. In summary, low stimulus frequencies and pulsed stimulations can be applied clinically to minimize the risk of injury from overstimulation.

Direct Observation/Video Monitoring

To increase the sensitivity of facial nerve IONM, recent studies have proposed implementing direct observation of facial muscle movement or intraoperative video monitoring (IOVM).[21,28,29,85] Theoretically, IOVM would supplement EMG by allowing better visualization of facial muscle contractions, thus providing an additional aid in the operating room. During IOVM, an anesthesia mask containing several infrared cameras is fastened to the patient's face, and the infrared properties of these cameras allow video recording under the operative drapes.[28,85] The camera view can be magnified such that even minute movements may be detected by the naked eye.[85] The images are projected on a 4-way split screen: 2 focus on movements of the facial muscles, another displays the microscopic operating field, and the remaining screen projects the EMG tracings.[28,29] These simultaneously derived images are thus juxtaposed next to each other, with a sound alarm triggered by facial muscle contractions.[28,29]

Although IOVM may prove useful, the full utility of this tool remains to be characterized. In a study comparing EMG with IOVM, the use of EMG alone exhibited higher sensitivity in detecting facial nerve activation: EMG detected facial muscle movement at a stimulation of 0.3 mA, whereas IOVM required a minimum of 0.5 mA.[29] De Seta et al.[21] obtained similar results, finding EMG alone to be more sensitive than IOVM. Thus, EMG appears more effective than IOVM based on current data. However, further studies must evaluate the validity of IOVM as a supplementary tool in the operating room.

Intraoperative Monitoring of the Vestibulocochlear Nerve (CN VIII)

Even with modern IONM, current vestibulocochlear nerve retention rates do not compare favorably with the excellent outcomes seen with the facial nerve.[5,12,16,36,49,52,62,75,88,94,99,103,112,115,116,120,130,132,139,147] Although this discrepancy may highlight the need for improvement in IONM of CN VIII,[99] it may also reflect the inherent difficulty in preserving auditory function, as large tumors are more highly associated with postoperative deficits,[1,11,20,23,88,89,103,116] and tumors with extensive infiltration into the cerebellopontine angle render acoustic preservation an arduous task.[23,89,147]

Operative damage to the vestibulocochlear nerve can be induced in various ways.[63,64,149] Direct operative trauma is a potential avenue, with the nerve most prone to exposure during maneuvers, such as drilling into the internal auditory canal, operative traction, or subsequent tumor resection.[1,17,19,51,79,96,129] Cranial nerves are inherently more susceptible to trauma because they are ensheathed in central myelin, thus lacking the extra protective layers, such as the perineurium, that are more prevalent in peripheral myelin.[8,17,19,64,79,123] Ischemic damage also presents further risk of injury. More specifically, vascular changes to the internal auditory artery, such as occlusion, rupture, or vasospasm, are believed to induce postoperative hearing deficits.[17,19,79,84,88,89,122] Strauss et al.[135] found that applying medical therapy to preclude such vasospasms produced

preservation rates that were more than twice as high when compared with controls.

Brainstem Auditory Evoked Potentials

Brainstem auditory evoked potentials are defined as the bioelectric neural activity that materializes in response to stimulation of the vestibulocochlear nerve.[63,97,129] In comparison with the background electrical brain activity,[54,79,142] these BAEP waves are diminutive and difficult to detect.[18,149] To facilitate distinction between BAEPs and background "noise," several thousand samples of the electrical stimulus must be acquired and subsequently averaged to create a distinct auditory evoked potential.[1,50,64,79,97,129,149] On BAEP recordings, the auditory response is extracted from several locations in the entire vestibular nerve pathway, as it travels peripherally to centrally.[97] The peaks of the evoked electrical potentials are classified as Wave I through Wave V, which correspond to the peripheral cochlear nerve and the inferior colliculus, respectively.[42,52,63,64,71,97,98,129] These waves can be seen in Fig. 4.[80]

In BAEP monitoring, scalp and earlobe electrodes are placed, and an auditory stimulator discharges acoustic clicks to the operated ear through an earphone-transducer apparatus.[59,64,79,97,129,149] The electrical pulse rate is set at a range of 20–50 clicks per second.[16,54,79,97,99,148] Before commencing with the operation, the stimulus intensity, as measured in decibels, is adjusted until the patient can hear the click; the stimulus is eventually delivered at several decibels higher than the measured threshold.[71,97] Upon delivery of the stimulus, the ears are stimulated bilaterally so white noise is applied at an intensity several decibels lower to obscure the response of the contralateral ear.[50,54,64,71,96,97,121,129]

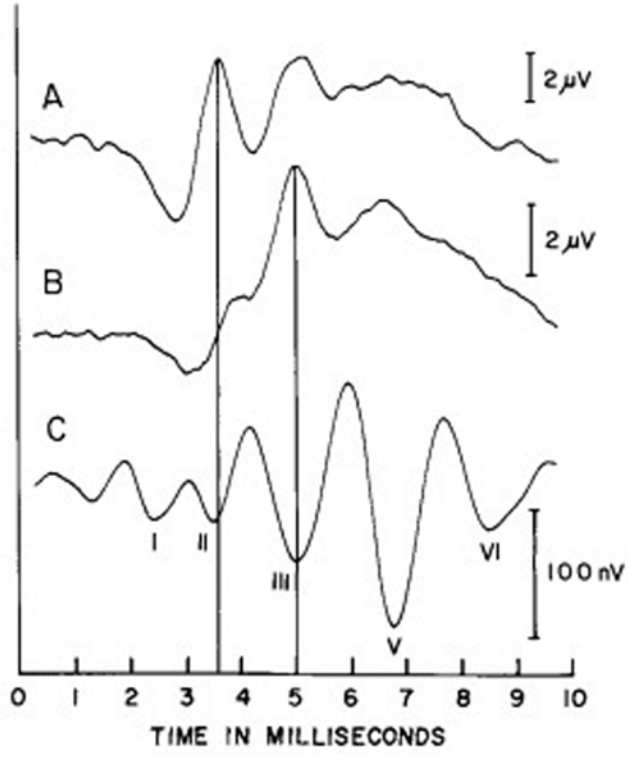

Fig. 4. Figure demonstrating direct CNAPs *(A)*, direct recording from the lateral recess of the fourth ventricle *(B)*, and BAEPs *(C)*. Note the 2 negative peaks seen on direct CNAPs and the relative coincidence of the first negative peak on direct CNAP to Wave I on BAEP monitoring. *Roman numerals* indicate the waves. Reproduced with permission from Møller and Jannetta: *J Neurosurg 59:*1013–1018, 1983.

When considering BAEP waveform shifts, Waves I, III, and V carry the most clinical significance.[54,71,74,97,129] Changes in their amplitude, peak latency, or presence of the peak are heavily scrutinized and compared with baseline BAEPs.[23,50,54,63,71,97,98,121,129,141] More specifically, increased peak latencies of Waves I, III, and V,[10,23,71,97,102] high interaural latency differences,[10,23,71,97] decreased amplitudes of Waves I and V,[63,71,97,121] and increased interpeak latencies between Waves I–III, III–V, and I–V[71,97] are examples of potentially concerning wave changes. Between peak latencies and interpeak latencies, the latter is the more clinically useful marker because peak latencies are more susceptible to influence from external factors such as age, thus rendering them less reliable.[71] However, the majority of these parameters are, at best, warning signs that alert the surgeon; among them, only maintenance of Waves I and V has been consistently shown to correlate with better postoperative hearing preservation rates,[34,50,88,91,98,121,136,141,146] although others have found poor hearing outcomes despite wave preservation.[30,59] The prognostic power of BAEPs is based solely on the preservation of the waves; in other words, when actual changes are seen on BAEPs, the severity or presence of postoperative deficits cannot be predicted reliably.[18,96] Regardless, detecting such BAEP waveform irregularities can still alert the surgeon to potential cranial nerve damage and encourage redirection of the operative plan of action.[63]

The use of BAEPs comes with several limitations. Because the stimulus response must be summed and averaged to obtain a wave of sufficiently high amplitude, the tradeoff to this process is a significant time delay that can last up to several seconds to minutes.[1,18,19,49,50,83] Naturally, such a delay can negatively influence the course of surgery, as BAEPs effectively provide data that were applicable several seconds or minutes prior.[14,18,108] Matthies and Samii[73] reported that direct BAEP monitoring was able to reduce the lag time to 5–15 seconds, suggesting that considerable improvements may be possible. In addition, BAEP recordings are prone to presenting false-positive results. Trauma is not the only causative agent of BAEP waveform shifts, with other physiological or intraoperative processes such as anesthesia, hypothermia, and irrigation all capable of inducing waveform changes.[24,54,63,64,69,71,129,133] Such a wide range of artifact sources can create great difficulty with respect to surgical decision making.[145] The utility of BAEPs may also be patient dependent, as some do not have detectable BAEPs while others have abnormal baseline BAEPs.[18,48,71,132,141] Without a clear starting point, BAEP monitoring may prove too difficult a task to complete. Measuring CNAPs may be more beneficial in such cases, as patients occasionally have BAEP waveform normalization postoperatively despite preoperative absence.[109]

Electrocochleography and Direct CNAPs

Brain auditory evoked potential monitoring is considered a "far-field" technique because the auditory response is measured on the scalp, which is distal from the neural auditory response.[54,98,129,149] In contrast, ECOG and direct CNAPs are "near-field" techniques because the stimulation evokes and records an electrical response close to its origin on the auditory nerve.[48,149] Because these techniques record from the nerve itself, near-field IONM bypasses the noise and artifact created in far-field IONM, which translates to reducing the number of stimuli averages required in addition to affording a larger amplitude for facile visualization.[17,54,79,149] Ultimately, this leads to a much quicker assessment of nerve function.[114,142,149]

In principle, both ECOG and direct CNAPs use electrodes to measure potentials generated from the auditory nerve, with some minor differences in operative setup. For ECOG, electrodes are typically positioned transtympanically on the middle ear promontory of the pathological ear.[45,53,65,80,95,96,101,123] Reference and ground electrodes are placed on the ipsilateral earlobe and on the forehead, respectively.[64,113,114,149] A foam ear plug not only holds the electrode firmly in place but also impedes foreign substances from breaching into the ear canal.[42,99] Similar to the BAEP, the stimulating electrode administers click impulses, and multiple responses must be averaged for a distinct wave pattern to emerge.[18,19,113,114,120,129]

As its name suggests, in direct CNAPs, the action potential is measured directly from the acoustic nerve itself.[64,81,129] The recording electrode is placed directly on the acoustic nerve, the negative electrode is attached to the mastoid of the contralateral ear, and a reference electrode is placed on the scalp.[16,54,79,99,108,148] It is common practice to place the recording electrode proximal to the tumor being resected,[14,17,79,99,129] with adhesive such as Gelfoam applied between the electrode and nerve to reinforce the placement.[54,99,148] Like the BAEP and ECOG, a click stimulus is applied through an earphone, and the resulting compound action potential is measured.[18,48,79,99,108,145]

Electrocochleography and direct CNAP monitoring are both techniques that rely on deducing the compound action potential, which represents a summation of all the action potentials, from the vestibulocochlear nerve.[42,108,113,129] These CNAPs, as they are known, are visualized as negative peaks distinguishable by their high amplitudes (Fig. 4).[80,120,129] In ECOGs, they consist of 2 action potential peaks designated "N1" and "N2,"[113,120,142,149] and in direct CNAP monitoring, comparable peaks are obtained.[81,108,142,149] Because ECOG involves peripheral nerve stimulation, the N1 waveform seen on ECOG is congruent with Wave I on BAEP monitoring.[8,42,64,71,79,113,120,129] The absolute loss of N1 on ECOG[48,88,96,114,120,139,149] or on direct recording[8,108,129,132,145,146,149] is frequently associated with postoperative hearing deficiency. Changes to the latency or amplitude of N1 on either ECOG or direct recording are also electrophysiological signs suggestive of injury.[8,14,17,19,48,54,79,120,131,142,145,146] Further electrical waveforms are seen in ECOG, thus differentiating it from a direct CNAP reading. The cochlear microphonics and summation potential are both electrical responses generated from the organ of Corti,[54,94,113,120] and lower cochlear microphonic detection thresholds may be involved in prognosticating postoperative hearing function.[94] In the overall context of ECOG monitoring, however, cochlear microphonics and summation potentials are generally considered less important than the N1 peak.[64,120]

The primary advantages of ECOG and direct CNAPs are derived from their near-field designation. With shorter latency periods, they reflect pertinent information much faster than BAEPs and provide immediate feedback on the state of the auditory system.[14,48,64,108,129,142,149] Changes seen on compound action potentials also tend to occur immediately, a helpful trait when considering vascular etiologies of dysfunction: vascular changes cause immediate effects that may not be detected quickly enough on BAEP monitoring.[54] The quick response time in conjunction with larger amplitudes than BAEPs[19,48,79,129] has strengthened the reputation of measuring CNAPs as the most preferred monitoring method of choice.[14,16,17,19,20,48,99,146] When comparing direct CNAPs with ECOG, direct CNAPs possess higher predictive value of postoperative functionality, with lower false-positive and higher true-positive rates.[18,146]

Electrocochleography and direct CNAPs have unique disadvantages. Because the recording electrode is placed peripherally, ECOG is unable to provide information about the entirety of the auditory nerve, particularly the more central portions of the auditory pathway.[18,41,83,98,129] As a result, it is possible to completely transect the nerve centrally without observing any credible change on the ECOG study.[48,79] Due to its rather invasive nature, ECOG also presents an increased risk of CSF otorrhea due to tympanic membrane perforation during electrode placement.[49,96,120,149] To circumvent this issue, alternative but viable options include tympanic or extratympanic electrode placement.[95,113,144] Electrocochleography can prove technically challenging as well. The electrode must be held securely in place; moving it manually or unintentionally can induce changes in the baseline amplitude and latency, thus exacerbating the difficulty of making subsequent comparisons.[149]

The disadvantage of using direct CNAPs is mainly practical. In larger tumors, there is very little operating space to place the recording electrode without sacrificing visibility of the surgical field.[49,54,79,99,120,129,149] As a result, direct CNAPs are generally reserved for patients presenting with smaller tumors.[42,83]

Conclusions

Implementation of facial and vestibulocochlear nerve IONM, in combination with the development of improved modern microneurosurgical techniques, has led to a dramatic reduction in the morbidity once associated with acoustic neuroma surgery. The facial nerve, in particular, has shown higher rates of preservation with the use of EMGs. The vestibulocochlear nerve, on the other hand, may be important to investigate as an avenue for further improvement. Despite the combined techniques of BAEPs, ECOG, and direct CNAPs, auditory preservation rates do not yet approximate those of facial nerve preservation. Further efforts and investigations are needed to study and incorporate other adjunctive IONM techniques in an attempt to improve preservation of auditory function.

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Neuroanatomical correlation of the House-Brackmann grading system in the microsurgical treatment of vestibular schwannoma

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Avoidance of facial nerve injury is one of the major goals of vestibular schwannoma (VS) surgery because functional deficits of the facial nerve can lead to physical, cosmetic, and psychological consequences for patients. Clinically, facial nerve function is assessed using the House-Brackmann grading scale, which also allows physicians to track the progress of a patient's facial nerve recovery. Because the facial nerve is a peripheral nerve, it has the ability to regenerate, and the extent of its functional recovery depends largely on the location and nature of its injury. In this report, the authors first describe the facial nerve anatomy, the House-Brackmann grading system, and factors known to be predictors of postoperative facial nerve outcome. The mechanisms and pathophysiology of facial nerve injury during VS surgery are then discussed, as well as factors affecting facial nerve regeneration after surgery.
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Key Words • **facial nerve** • **House-Brackmann scale** • **vestibular schwannoma** • **neuroanatomy** • **acoustic neuroma**

Vestibular schwannoma is a benign CNS tumor arising from one or more constituent nerves of the eighth cranial nerve complex.[87] Management options include observation, radiation, radiosurgery, microsurgery, and a combination of these modalities. Overall, microsurgical resection remains the best cytoreductive therapy and has been shown to be most effective for treating large lesions that cause mass effect and obstructive hydrocephalus. However, microsurgery cannot achieve the facial and cochlear nerve outcomes of radiosurgery.[78,95] Because functional deficits of the facial nerve lead to physical, cosmetic, and psychological consequences for patients, avoidance of facial nerve palsy is a major goal of VS surgery.[3,31,59] Therefore, it is critical to compare the pre- and postoperative facial nerve function in patients with VS, not only for effective care of these patients, but also to assess the likelihood of functional recovery. The House-Brackmann grading system was initially proposed in 1983 as a universal standard system for the assessment of facial nerve function, and it has since become a part of the standard of care for all patients with VS. This review will include discussions of the facial nerve anatomy, the House-Brackmann grading system, the factors known to predict postoperative facial nerve outcome, the pathophysiology of facial nerve injury during surgery, and finally, the mechanisms affecting facial nerve regeneration.

Abbreviations used in this paper: CPA = cerebellopontine angle; IAC = internal auditory canal; VS = vestibular schwannoma.

Facial Nerve Anatomy

The facial nerve has the longest and most tortuous course in the skull of any cranial nerve. The efferent component of the facial nerve mainly innervates the muscles of facial expression; in addition, it carries secretomotor fibers for the submandibular and sublingual salivary glands and the lacrimal glands. The afferent component of the facial nerve carries taste sensation from the tongue and the palate, as well as general sensation from the external ear.[23]

The upper motor neurons of the facial nerve arise from the primary motor cortex anterior to the central fissure in the precentral gyrus (Fig. 1). The majority of the upper motor neuron axons descend through the corticobulbar tracts, cross the midline, and synapse in the contralateral motor nucleus in the pons; others descend without crossing to innervate the ipsilateral subnuclei responsible for the periorbital and frontalis muscles (Fig. 1). Thus, these subnuclei receive bilateral cortical innervation, whereas the subnuclei for the lower half of the face are only innervated by contralateral corticobulbar

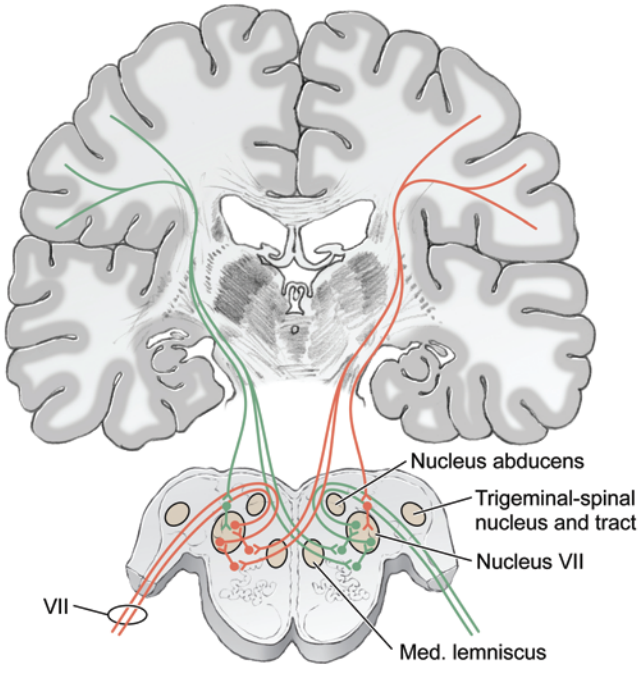

Fig. 1. Upper motor neuron connection from the cerebral cortex to the facial nuclei. The facial nuclei corresponding to the upper face are bilaterally innervated, whereas the facial nuclei targeting the lower face are only unilaterally innervated by the contralateral motor cortex. VII = cranial nerve VII; med. lemniscus = medial lemniscus.

fibers. It is interesting to note that the motor pathways for emotion-induced facial movements, such as smiling and pursing the lips, contain additional synaptic junctions that have been found in the hypothalamus, basal ganglia, and midbrain tegmentum.[23] Within the motor nuclei, subnuclei exist for each corresponding individual nerve branch, and those innervating the upper half of the face are ventral to those innervating the lower half. The fibers emerging from the facial motor nucleus initially loop around the abducens nucleus in a segment often referred to as the internal genu, in contrast to the external genu in the temporal bone.[63]

The facial nerve consists of 2 roots: a larger medial motor root and a smaller lateral sensory root, also called the nervus intermedius, which contains fibers arising from both the sensory nucleus and the salivatory nuclei located in the rostral ventrolateral medulla. Both roots exit the lower lateral aspect of the pons between the inferior cerebellar peduncle and the olive. The average distance that the facial nerve travels from its point of exit in the brainstem at the CPA to its entrance at the IAC (the cisternal portion) is 15.8 mm, and this segment is covered by pia mater and bathed in CSF.[63] Emerging from the brainstem, the facial nerve is accompanied by the nervus intermedius, and it runs anteriorly and superiorly adjacent to the vestibulocochlear nerves. Knowledge of the orientation of the nerves is critical during VS resection to preserve facial nerve function (Fig. 2).[63,93] The anatomical proximity of the facial nerve with the nervus intermedius and the vestibulocochlear nerves at the level of the CPA and in the IAC increases the likelihood that lesions of the CPA can cause concurrent deficits of these nerves.

Transtemporal Bone Portion of the Facial Nerve

The facial nerve traverses the entire temporal bone, and due to variation in the anatomy of the temporal bone, the course of the facial nerve within the bone is also variable (Fig. 3). Because the cisternal (intracranial) segment of the facial nerve from the brainstem to the fundus of the IAC is covered only by a thin layer of glia, it is quite vulnerable to surgical manipulation. However, it has been shown to be somewhat resistant to a slow process of stretching or compression, such as by a slow-growing VS.[63] In the IAC, which is 7 mm long on average in adults, the motor root of the facial nerve lies in a groove on the anterior and superior surface of the auditory nerve, with the sensory root (nervus intermedius) between them (Fig. 3).[23] Although the layer of meninges covering both the facial and auditory nerves usually ends at the fundus as the facial nerve pierces the dura, in some cases the dural coverings extend beyond the canal to as far as the geniculate ganglion.[23]

At the fundus of the IAC, the crista falciformis (transverse crest), a transverse ridge, divides the auditory canal into superior and inferior compartments, and the facial nerve passes across the top of this ledge and is separated from the superior vestibular nerve by a vertical bony structure called the Bill's bar.[63] After passing through the fundus, the facial nerve enters the fallopian canal and makes a Z-shaped course until it reaches the stylomastoid foramen. Its Z-shaped course within the fallopian canal is usually described in 3 anatomical sections that are nearly perpendicular to one another on 3 different 2D planes: the labyrinthine section, the tympanic or horizontal section, and the mastoid or vertical section (Fig. 3).

The labyrinthine segment of the facial nerve is its first segment in the fallopian canal and is also the shortest and the thinnest segment within the canal. It begins at the opening of the fallopian canal and runs laterally above the vestibule, passing between the vestibule and the cochlea until it reaches the medial wall of the middle ear cleft.[23] The end point of this nerve segment is expanded by the geniculate ganglion containing the unipolar cells of the sensory root. Because the nerve fibers in the labyrinthine segment are loosely arranged, without epineurial covering, and pressed into the lateral aspect of the canal where it is narrowest at its entrance, the facial nerve can be easily damaged by any process that decreases this already narrow course. In addition, the blood supply to the nerve in this region is uniquely without anastomosing arterial arcades.[63]

The tympanic portion begins at the geniculate ganglion where the nerve abruptly turns 90° to run posteriorly and inferiorly to form the external genu.[69] The length of this section of the nerve is approximately 12 mm and is above the oval window and below the prominence formed by the horizontal semicircular canal.[23]

The final segment of the facial nerve in the fallopian canal is the mastoid or vertical portion, which begins with a less abrupt bend, called the pyramidal turn, in which the nerve passes below the semicircular canal to descend behind the pyramid.[100] This segment is roughly 15 mm in length and ends when it reaches the stylomastoid foramen.[23] The mastoid segment lies immediately anterior

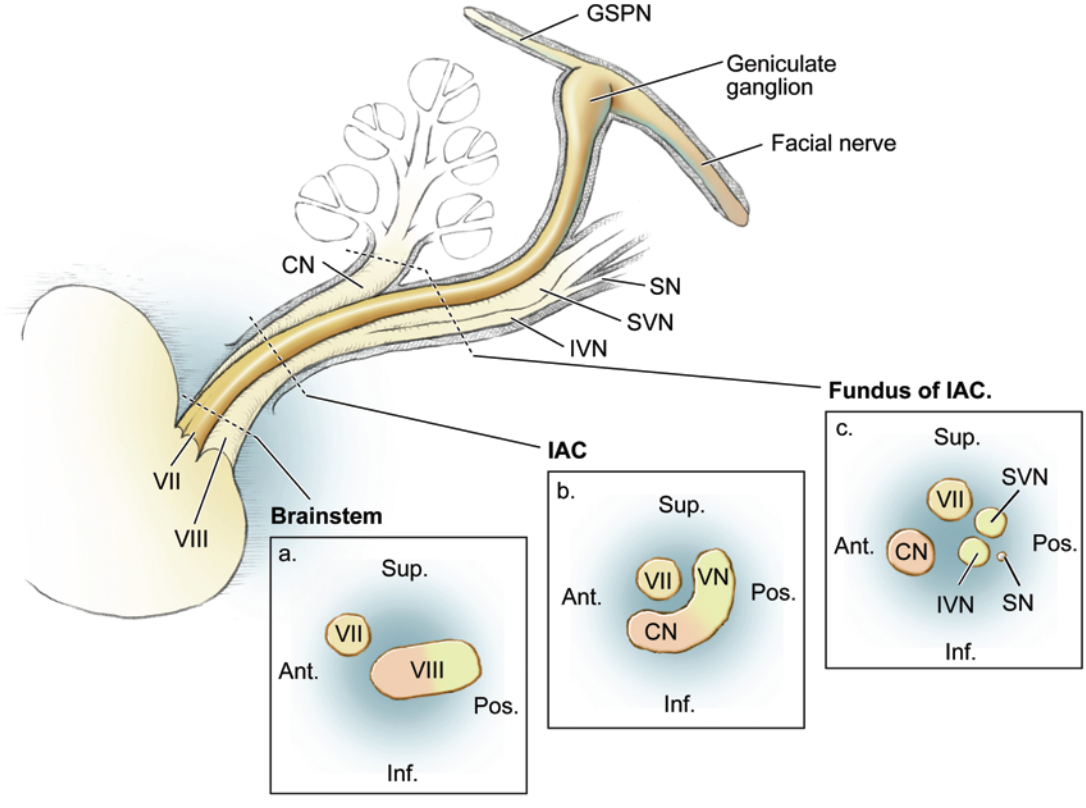

Fig. 2. Anatomy of the cisternal and meatal segments of the facial nerve (cranial nerve [CN] VII). The facial (CN VII) and vestibulocochlear (CN VIII) nerves exit the brainstem at the CPA (cisternal segment) and enter the IAC together. The facial nerve is anterior and superior to the vestibular nerves in the IAC. The orientation of the facial nerve with respect to the superior vestibular nerve (SVN), inferior vestibular nerve (IVN), the cochlear nerve (CN), and the singular nerve (SN) is depicted. Ant. = anterior; GSPN = greater (superficial) petrosal nerve; Inf. = inferior; Pos. = posterior; Sup. = superior; VN = vestibular nerve.

and medial to the lateral semicircular canal.[80] On average, the facial nerve occupies approximately 25%–50% of the cross-sectional area of the fallopian canal.[91]

After the facial nerve emerges from the stylomastoid foramen, it continues its course downward and forward, crosses the lateral surface of the styloid process and the external carotid artery, and reaches the parotid gland. Then, the nerve divides behind the neck of the mandible into 2 main branches, the temporofacial and cervicofacial trunks, which further subdivide to form the 5 terminal branches called the temporal, zygomatic, buccal, marginal mandibular, and cervical branches (Fig. 4).[98]

Branches of the Facial Nerve

The facial nerve has numerous branches throughout its course, and they can be categorized as either branches of communication with other nerves or as branches of distribution to target tissues. The branches of the facial nerve can also be divided anatomically into 3 categories: branches within the temporal bone, branches in the neck, and branches in the face.

There are 2 branches of the facial nerve within the temporal bone: the nerve to the stapedius muscle and the chorda tympani nerve. The nerve to the stapedius muscle innervates the stapedius muscle; the chorda tympani nerve mainly carries fibers for taste sensation of the anterior two-thirds of the tongue, and also contains efferent

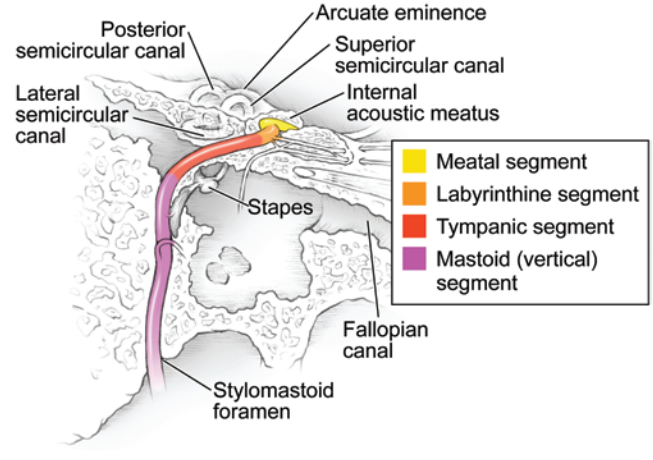

Fig. 3. Facial nerve anatomy within the fallopian canal. The course of the facial nerve within the fallopian canal is usually described in 3 anatomical sections that are nearly perpendicular to one another on 3 different 2D planes: the labyrinthine section, the tympanic or horizontal section, and the mastoid or vertical segments. The labyrinthine segment runs laterally above the vestibule, passing between the vestibule and the cochlea until it reaches the medial wall of the middle ear cleft. The tympanic portion runs lateral to the superior semicircular canal, and the mastoid section runs immediately anterior and medial to the lateral semicircular canal.

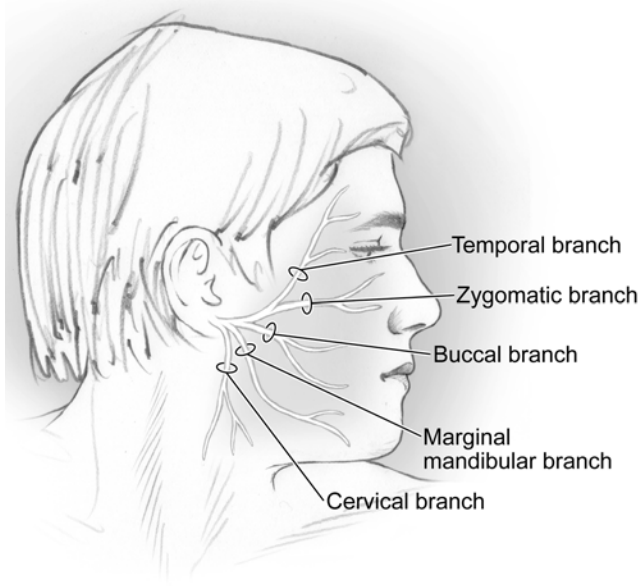

Fig. 4. The House-Brackmann scale measures the function of the following 5 branches of the facial nerve: temporal, zygomatic, buccal, marginal mandibular, and cervical branches. The temporal branch supplies anterior and superior auricular muscles, frontal belly of the occipitofrontalis muscle, and the orbicularis oculi and corrugator muscles. The zygomatic branch innervates the orbicularis oculi principally. The buccal branch supplies procerus, zygomaticus major and minor, levator labii superioris, levator anguli oris, levator labii superioris alaeque nasi, orbicularis oris, and buccinator muscle. The marginal mandibular branch innervates risorius, depressor anguli oris, depressor labii inferioris, and mentalis muscle. The cervical branch mainly supplies the platysma muscle.

secretomotor parasympathetic fibers for the submandibular and sublingual salivary glands.[23]

There are 3 branches of the facial nerve in the neck: the posterior auricular, digastric, and stylohyoid branches. The posterior auricular branch has an auricular division to the posterior auricular muscle and an occipital division to the occipital belly of the occipitofrontalis muscle. The digastric branch supplies the posterior belly of digastric muscle; the stylohyoid branch supplies the stylohyoid muscle.[23]

There are 5 main branches of the facial nerve in the face with direct clinical relevance to the House-Brackmann scale (Fig. 4): temporal, zygomatic, buccal, marginal mandibular, and cervical branches.[98] The functionality of these 5 branches is the main concern in the context of grading facial nerve recovery after surgery for VS using the House-Brackmann scale.[48] The temporal branch supplies anterior and superior auricular muscles, frontal belly of occipitofrontalis muscle, and orbicularis oculi and corrugator muscle. The zygomatic branch innervates the orbicularis oculi principally. The buccal branch supplies procerus, zygomaticus major and minor, levator labii superioris, levator anguli oris, levator labii superioris alaeque nasi, orbicularis oris, and buccinator muscle. The marginal mandibular branch innervates risorius, depressor anguli oris, depressor labii inferioris, and mentalis muscle. The cervical branch mainly supplies the platysma muscle. In addition to the 5 branches in the face, the cutaneous fibers of the facial nerve accompany the auricular branch of the vagus, and these fibers are believed to innervate the skin over the conchal cartilage.[23] It is worth noting that what has been described only represents a general pattern of innervation, and different patterns of innervation are possible because the terminal branches of the facial nerve anastomose in a plexus. Therefore, patients with different innervation patterns of facial nerve branches may recover from VS surgery with variable speed and pattern, resulting in differential outcome.

Blood Supply of the Facial Nerve

The facial nerve is supplied in the temporal bone by 3 arteries with overlapping territories, such that any 1 portion of the nerve has at least 2 supplying vessels, with the only exception being the labyrinthine segment (Fig. 5).[8,23,63] The anterior inferior cerebellar artery feeds the nerve in the posterior fossa, and the internal auditory artery, which is a branch of the anterior inferior cerebellar artery, specifically supplies the nerve in the IAC. The petrosal branch of the middle meningeal artery mainly supplies the nerve in the fallopian canal. Within the canal the artery divides at the geniculate ganglion into a descending branch that runs distally with the nerve to the stylomastoid foramen and an ascending branch that supplies the region proximal to the geniculate ganglion. The stylomastoid branch of the posterior auricular artery enters the facial canal through the stylomastoid foramen, with an ascending branch running up with the nerve to the geniculate ganglion and a descending branch supplying the nerve down to the stylomastoid foramen, while running with the posterior auricular nerve.[23] These arteries and their associated veins course within the loose connective tissue of the epineurium, between the periosteum of the canal wall and the nerve sheath proper. Within the temporal bone, not only do the branches of these arteries anastomose with each other and with the vascular plexus of the middle ear mucosa, but they also anastomose within the marrow spaces of surrounding bone. Finally, the extracranial portion of the facial nerve receives its blood supply from the branches of the stylomastoid, posterior auricular, superficial temporal, and transverse facial arteries, which send anastomosing branches to the nerve.[23]

House-Brackmann Facial Nerve Grading System

Various systems for assessing facial nerve function have been proposed since the 1950s, but none were universally adopted until House and Brackmann in 1983 systematically reviewed most that were in existence at the time. They proposed a new grading system intended to be an international standard that could be widely accepted and had sufficient reliability and validity.[47] The grading system they proposed includes a 6-point scale, with Grade I representing normal and Grade VI representing total, flaccid paralysis (Fig. 6; Table 1).[47,48] While they initially suggested that a grading score can be correlated with an 8-point scale from direct measurements of the movement of the eyebrow and corner of the mouth and comparing the results with those on the unaffected side, it was considered much easier to assign a grade based on the 6-point scale from simple clinical observation.[48]

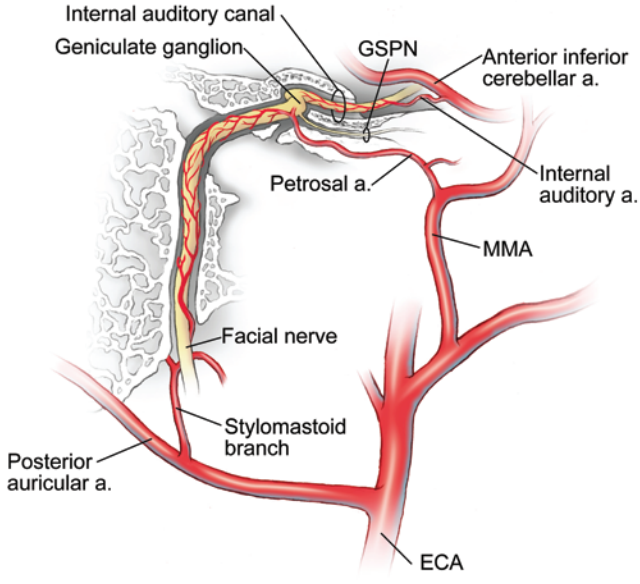

Fig. 5. The blood supply of the facial nerve within the temporal bone. Three arteries supply the facial nerve, which are the internal auditory artery (a branch of the anterior inferior cerebellar artery), the petrosal artery (a branch of the middle meningeal artery [MMA]), and the stylomastoid artery (a branch of the posterior auricular artery). a. = artery; ECA = external carotid artery.

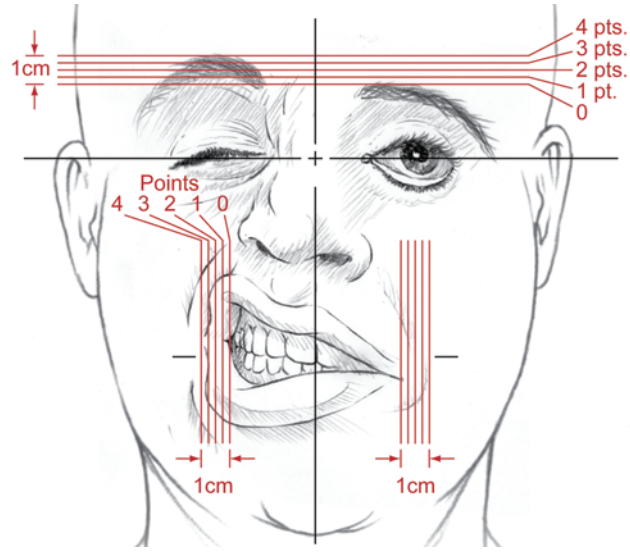

Fig. 6. Dr. Brackmann developed an easy method for measuring facial movement, specifically at the eyebrow and corner of the mouth, and compared the results with measurements on the unaffected side as depicted here. A scale of 0.25-cm divisions is used for the measurements, with a total possible score of 8 (4, or 1 cm, for the mouth; 4, or 1 cm, for the eyebrow) that can be converted to the 6-point House-Brackmann scale.

The implementation of the House-Brackmann grading system involves assessment of 3 components that contribute to the assignment of each grade: observations grossly, the face at rest, and motions of the facial muscles.[48] When the 3 components of the grade do not fall into the same grade level, the usual clinical practice is to assign the most severe grade. Grade I represents normal facial movement in all areas with no weakness or synkinesis, which is the involuntary movement of a part of the face during voluntary movement of another part of the face. Grade II indicates mild dysfunction; one can observe slight asymmetry of facial movements with a possible slight synkinesis, but normal symmetry and tone at rest. Grossly, there is slight weakness and asymmetry on close inspection, but the eye can achieve complete closure with minimum effort.[48] In the published literature, a Grade II or less is often considered to indicate preserved facial nerve function.

In Grade III there is moderate dysfunction, with obvious but not disfiguring differences between the 2 sides; there can be noticeable but not severe synkinesis, contracture, and/or hemifacial spasm. In Grade IV, there is moderately severe dysfunction with obvious weakness and/or disfiguring asymmetry. Importantly, there is normal symmetry and tone at rest from Grades II to IV, but not Grades V or VI. In Grade III slight to moderate forehead movement remains evident, but in Grades IV and higher the forehead has no movement. In Grade III the eye can completely close with effort, but in Grade IV there is incomplete closure of the eye. The mouth is slightly weak with maximum effort in Grade III and is asymmetrical with maximum effort in Grade IV. In Grade V there is severe dysfunction, and grossly there is only barely perceptible motion, and one can observe asymmetry even at rest. The forehead has no motion, the eyelids cannot completely close, and the mouth can move only slightly with maximum effort. Finally, in Grade VI, there is total paralysis, and no movement of any kind is observed.

Vestibular Schwannoma Surgery

Surgical exposure of the facial nerve may be necessary for decompression, grafting, rerouting, or removal of lesions such as VSs, meningiomas, facial nerve neuromas, and cholesteatomas.[12] Current microsurgical techniques make it possible to expose the entire course of the facial nerve without injuring the nerve and without disturbing hearing or vestibular functions.[37]

Currently there are 3 established surgical approaches for VS resection (Fig. 7): the retrosigmoid, translabyrinthine, and middle fossa approaches.[80] The rationale for the translabyrinthine approach is to gain lateral access to the IAC and the CPA lesions with no cerebellar retraction (Fig. 7D–F).[80] The translabyrinthine approach also allows exposure of the neurovascular structures present in the CPA and thus enables removal of a VS of any size.[80] However, unlike the middle fossa and retrosigmoid approaches, the translabyrinthine approach does not allow for hearing preservation.[42] The retrosigmoid approach is a modified suboccipital approach that is performed more anterolaterally, just posterior to the sigmoid sinus (Fig. 7A and B). Unlike the translabyrinthine approach, this approach provides access to the CPA without sacrificing the labyrinthine, and the IAC is exposed by drilling its posterior wall (Fig. 7B and C).[80] The middle fossa approach allows complete exposure of the IAC from the porus to the fundus and limited exposure of the CPA through the superior surface of the temporal bone (Fig. 7H and I);

TABLE 1: The House-Brackmann grading scale

Grade	Description	Characteristics			Estimated Function (%)
		Gross	At Rest	Motion	
I	normal	normal	normal	normal	100
II	mild dysfunction	slight weakness noticeable on close inspection, may have very slight synkinesis	normal symmetry & tone	forehead: moderate to good function; eye: complete closure w/ minimum effort; mouth: slight asymmetry	80
III	moderate dysfunction	obvious but not disfiguring difference between 2 sides; noticeable but not severe synkinesis, contracture, and/or hemifacial spasm	normal symmetry & tone	forehead: slight to moderate movement; eye: complete closure w/ effort; mouth: slightly weak w/ maximum effort	60
IV	moderately severe dysfunction	obvious weakness and/or disfiguring asymmetry	normal symmetry & tone	forehead: none; eye: incomplete closure; mouth: asymmetric w/ maximum effort	40
V	severe dysfunction	only barely perceptible motion	asymmetry	forehead: none; eye: incomplete closure; mouth: slight movement	20
VI	total paralysis	no movement	asymmetry	no movement	0

thus, this approach allows for hearing preservation, while the translabyrinthine approach does not.[80]

For small tumors, typically smaller than 20 mm,[53,86] several studies have found that the preservation of facial nerve function was better achieved with translabyrinthine and retrosigmoid approaches compared with the middle fossa approach: preserved function was noted in more than 90% of cases for the translabyrinthine and retrosigmoid approaches, compared with 80% for the middle fossa approach.[27,72] The worse outcome associated with the use of the middle fossa approach could be explained by the fact that the position of the facial nerve relative to the tumor, which most often arises from the inferior vestibular nerve, makes the nerve particularly vulnerable during the approach. Hillman et al.[42] recently found similar results from comparing postoperative facial nerve outcomes of patients treated surgically using either the middle fossa or retrosigmoid approach; not only was the retrosigmoid approach associated with a higher percentage of preservation (80% for the middle fossa approach vs 90% for the retrosigmoid approach), but facial function recovered faster with the retrosigmoid approach, and there were more long-term House-Brackmann Grade I function results in the retrosigmoid group. This finding was confirmed by Rabelo de Freitas et al.,[72] but they also found that the difference in facial nerve outcome appeared only for extrameatal tumors when they compared size-matched tumors (58.3% preservation in the middle fossa approach vs 98% in the retrosigmoid approach; $p = 0.0006$). Additionally, they found no difference in hearing outcome between the 2 approaches.[72] However, Hillman et al.[42] observed better hearing preservation in the middle fossa group and found that there were more recurrent and residual tumors in the retrosigmoid group.

The rates of CSF leaks between the 3 approaches were not significantly different as reported by Mangus et al.[58] Previously, Brennan et al.[14] reported that there was no difference in the leakage rate between translabyrinthine and retrosigmoid approaches (7.9% vs 10%, respectively; $p = 0.46$), although there were differences in the site of the leak: 56% of translabyrinthine leaks occurred through the wound, compared with only 8% of retrosigmoid leaks ($p = 0.007$), whereas otorrhea accounted for 9% of translabyrinthine leaks compared with 42% of retrosigmoid leaks ($p = 0.02$). Tumor size (maximum extracanalicular diameter) had a significant effect on the leakage rate overall: tumors in cases complicated by CSF leak were significantly larger than in those without a leak (mean diameter 21 vs 15 mm, respectively; $p = 0.001$), although significantly larger tumors were removed via the translabyrinthine approach compared with the retrosigmoid approach (22 vs 6 mm, respectively; $p < 0.001$). On subgroup analysis, it was found that this association of leak with tumor size was only significant for the retrosigmoid, but not the translabyrinthine procedures. However, translabyrinthine-associated leaks (especially rhinorrhea) required surgical repair significantly more often than retrosigmoid-associated leaks.[14]

Performing recurrence-free survival analysis on a prospectively collected database, Sughrue et al.[85] found that there was no significant relation between the extent of resection and the rate of tumor recurrence, but the extent of resection was highly correlated with patient age, tumor size, and surgical approach. However, using Cox regression analysis, the authors found that the approach used did not significantly affect tumor control when the extent of resection was controlled for.[85]

Prognosticators of Facial Nerve Outcome After VS Surgery

Prognostic factors such as age, tumor size, extent of

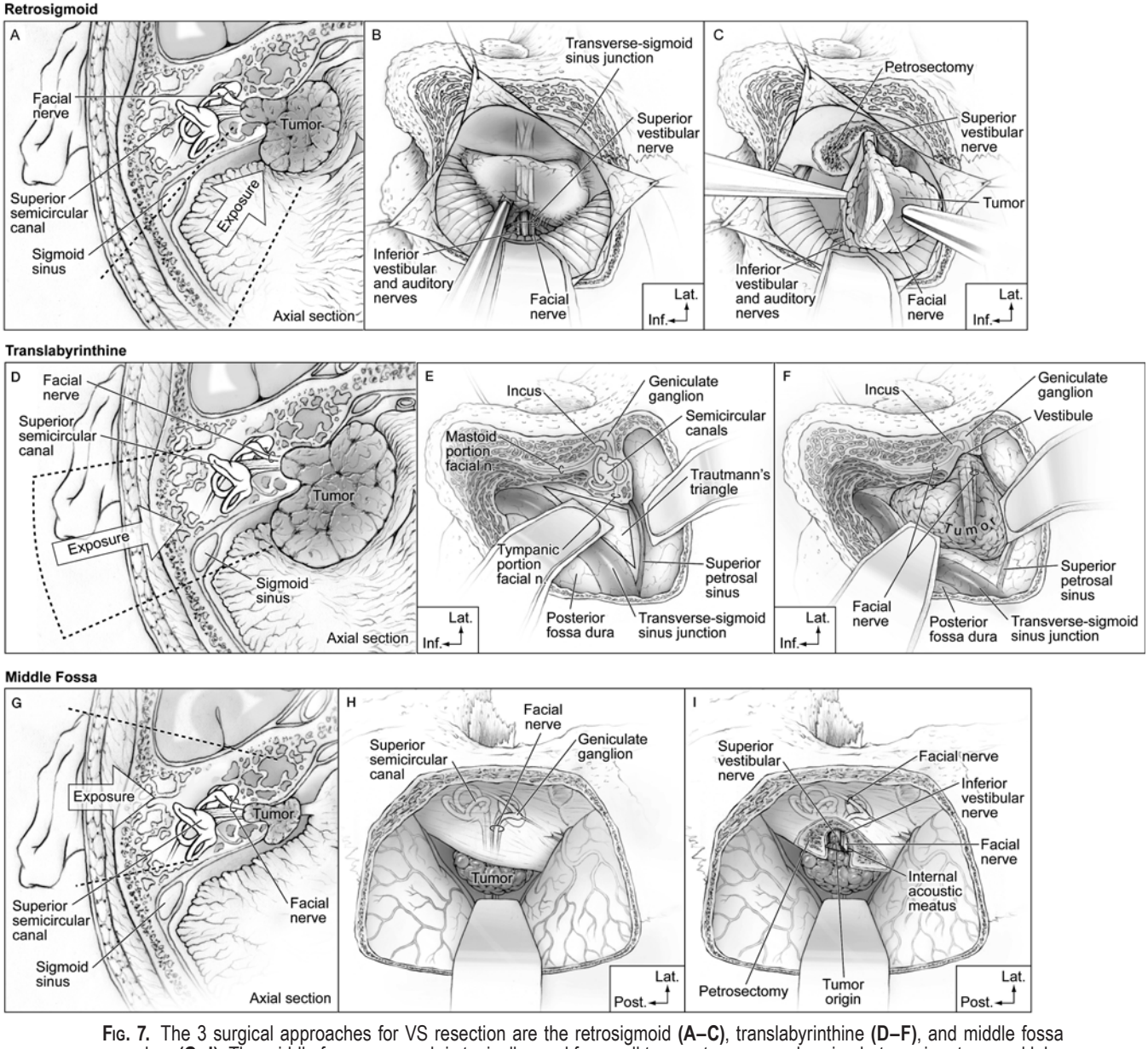

Fig. 7. The 3 surgical approaches for VS resection are the retrosigmoid **(A–C)**, translabyrinthine **(D–F)**, and middle fossa approaches **(G–I)**. The middle fossa approach is typically used for small tumors to preserve hearing but requires temporal lobe retraction and results in poor exposure of the posterior fossa. The translabyrinthine approach is often used for tumors with IAC extension with no serviceable hearing. The retrosigmoid approach is used primarily for cisternal tumor, but can be used for different sizes of tumors. It is also the most familiar approach to many neurosurgeons. The retrosigmoid approach is performed posterior to the sigmoid sinus and provides access to the CPA without sacrificing the labyrinth. The IAC is exposed by drilling its posterior wall. The translabyrinthine approach allows lateral access to the IAC and the CPA lesions with no cerebellar retraction. This approach, however, will sacrifice the labyrinth, and thus hearing. The Trautmann's triangle is entered via this approach (E and F). It is demarcated by the bony labyrinth, sigmoid sinus, and superior petrosal sinus or dura. The middle fossa approach allows complete exposure of the IAC from the porus to the fundus with a limited exposure of the CPA through the superior surface of the temporal bone; thus, this approach allows for hearing preservation. The approach to Kawase's triangle can be seen (H and I), and is demarcated by the greater petrosal nerve, trigeminal nerve (V3), the arcuate eminence, and the medial edge of the petrous ridge (or superior petrosal sinus). Lat. = lateral; n. = nerve; Post. = posterior.

resection, and surgical approach have been implicated for predicting facial nerve function outcome after surgical removal of VS.[20,55,59,102] In a meta-analysis of 296 studies involving more than 25,000 patients that included outcome data for facial nerve function of surgically treated VS patients, Sughrue et al.[86] found that tumor size of less than 20 mm, the use of the middle fossa approach, and the use of neuromonitoring during surgery were associated with facial nerve preservation. However, others have found through retrospective review of patients presenting at single institutions that for small tumors, specifically those smaller than 20 mm, the preservation of facial

nerve function was better achieved with translabyrinthine and retrosigmoid approaches compared with the middle fossa approach.[27,72] Additionally, Brackmann and Barrs[13] and Sanna and Caylan[80] had previously suggested that the enlarged translabyrinthine approach produces the best facial nerve functional outcome; the lowest frequency of postoperative neurological sequelae was achieved, likely due to the absence of cerebellar retraction. Thus, there is a lack of consensus on the effect of surgical approach on preservation of facial nerve function.

In a recent multivariate logistic regression analysis of patients with VS from a prospectively collected database at the University of California San Francisco, Bloch et al.[10] examined the effect of variables such as surgical approach, tumor size, patient age, and extent of resection on rates of facial nerve dysfunction after surgery. Only preoperative tumor size significantly predicted poorer facial nerve outcome for patients followed up for at least 6 months, as well as those followed up for at least 12 months. These investigators found no significant relationship between facial nerve function and any of the other factors they examined, such as extent of resection, surgical approach, and age.[10]

Intraoperative continuous monitoring of evoked electromyography activity has been used by surgeons for more than a decade in preserving cranial nerve functions during VS resection. The usefulness of this procedure has been the subject of many recent studies. In a single institutional report of 477 surgically treated patients with VS, Sughrue et al.[84] reported that elevated stimulation threshold exceeding > 0.05 mA is a highly specific (90%), but very insensitive (29%) finding in their cohort.[84] The positive and negative predictive values of facial nerve electromyography for detection of permanent facial palsy reported were 68% and 63%, respectively.[84] Additionally, they found that the negative predictive value decreased with increasing tumor size (72% vs 64% vs 53%) due to the increasing prevalence of postoperative facial nerve palsy in these patients.[84] Whereas the findings by Sughrue et al. showed that the predictive value for facial nerve function remained to be determined, Amano et al.[2] reported that the postoperative course of facial nerve function appears predictable using intraoperative monitoring. These investigators found that the amplitude preservation ratio correlated significantly with facial nerve function both immediately and 1 year after surgery.[2]

In summary, while there is a lack of consensus on the effect of surgical approach on preservation of facial nerve function, large tumor size and elevated stimulation threshold during intraoperative monitoring have been associated with poor preservation of facial nerve functional outcome. Conversely, small tumor size and the use of neuromonitoring have been associated with good facial nerve functional outcome, likely from the lower risk of nerve injury intraoperatively.

House-Brackmann Correlation of Early Recovery Patterns in Facial Nerve Function After VS Surgery

One of the important functions of the House-Brackmann grading scale is its ability to allow physicians to conveniently and precisely track the recovery of a patient's facial nerve function after injury. Substantial evidence suggests that the most important determinant of successful clinical outcome of the recovery of a peripheral nerve is time to reinnervation.[46,88] The faster the end organ is reinnervated, the less likely it is to undergo atrophy and permanent denervation.[9,88] The facial nerve's recovery after VS surgery can be similarly dependent upon the speed of immediate postoperative recovery.

Anecdotally, we have observed that VS patients with postoperative facial nerve dysfunction more often recover their upper face and especially eyelid function before their mouth or lower face function. House and Brackmann also explained that recovery of forehead movement indicates that there has not been total degeneration of the nerve, implying that the forehead branches are either more resistant to permanent injuries or are more likely to recover immediately after surgery.[48] Thus, if frontalis motion is absent, facial nerve may have been sacrificed.[13] Moreover, Brackmann and Barrs[13] noted that if mouth movement is preserved, then the facial nerve is likely preserved, further indicating that the lower branches are more easily damaged.

Based on self-reported questionnaires, Brackmann and Barr's analysis[13] revealed that although patients' ability to raise an eyebrow on the affected side postoperatively correlated well with their estimated extent of final recovery, their ability to move the corner of their mouth did not. This finding appears to suggest that the recovery of upper facial function directly correlates with the physiology of facial nerve recovery and regeneration, and it further suggests the existence of a potential underlying physiological explanation for our experience of observing patients' clinical course of postoperative facial nerve function.

Physiological Basis of Facial Nerve Dysfunction

The facial nerve is often distorted by VS, both in shape and in relationship to other anatomical landmarks.[7] Thus, most surgeons use intraoperative electrical stimulation for both positive identification and as proof of preserved function even when the normal anatomy is preserved.

Several sources have been reported in the literature regarding the pressure-induced motor neuropraxia of the facial nerve, using physiological studies: primary pressure effects on saltatory nerve conduction, blockage of bulk and rapid axonal transport, and regional ischemia.[6] It has been known that gentle pressure applied to the trunk of a peripheral nerve even for long periods of time, such as one produced by a slow-growing small VS, produces only minor anatomical change and no alteration of conduction; on the other hand, severe pressure, such as that of a surgical clamp, usually results in Wallerian degeneration and prolonged conduction block.[6] Between the two ends of this pressure spectrum is a range of transient or reversible neuropraxias in which the onset, magnitude, and duration of the block after release of the nerve are roughly proportional to the magnitude and duration of the applied pressure.[6] In real-world scenarios, however, the magnitude of force applied to the nerve can be more

important than the duration of the force, given the often observed differential functional outcome of surgical- versus tumor-derived pressure on the nerve.

Because the nerve root central to the geniculate ganglion lacks the perineurium and epineurium, it is more vulnerable to compression injury. This central portion of the nerve also lacks the tensile strength of its peripheral counterpart and is more sensitive to traction injury than its peripheral counterpart.[91] It is therefore possible that due to the mechanical nature of such injuries, the location of the injury site likely predicts functional outcome. During surgery, manipulation of the facial nerve proximal to the geniculate ganglion is thus more likely to injury the nerve fibers and produce functional deficits than if the nerve is manipulated in a similar fashion in the distal portions of the facial nerve. Moreover, the more central the injury site, the longer it takes the nerve fibers to regenerate to reach its innervating target.

Somatotopic Basis of Physiological Manifestation of Partial Facial Nerve Injury: Microscopic Examination of Facial Nerve Anatomy and Spatial Orientation of Fibers in the Temporal Bone

At the levels of the somatomotor cortex and the facial nucleus, the existence of somatotopic organization of neurons and their processes has been known for some time.[34,73,94] The idea of a similar organization at the level of the facial nerve was suggested nearly a century ago,[22,61] but it has not been definitively proven. There is substantial evidence, however, supporting a somatotopic organization of the facial nerve trunk.[34,96] It is currently an accepted theory that somatotopic organization exists within at least some parts of the facial nerve and that the maintenance of this organization during regeneration is crucial for reinnervating correct targets (Fig. 8).[18,19] While evidence suggesting the lack of such an organization also exists, the quality and amount of evidence is insufficient to disprove the existence of a somatotopic organization at this time.

Many of the findings demonstrating such an organization of the facial nerve were obtained initially using clinical observations, scalpel hemisections, and radiofrequency lesions; and as more modern techniques were developed, evoked electromyography activity, microdissection techniques, crush injuries, tease avulsions, and in many instances histopathological correlations have been used. Canuyt first reported that branches supplying the upper region of the face were surrounded superficially by those supplying the lower region.[22,61] Canuyt's initial findings were later confirmed by Eyries and Chouard.[26]

Hofmann[44] proposed a different arrangement with a superior ramus and an inferior ramus, consisting of fibers innervating the upper and lower part of the face, respectively, that rotated slightly as the nerve exited the stylomastoid foramen. May[61,62] confirmed Hofmann's findings and reported that the upper ramus was located posterolaterally and the lower ramus anteromedially, although May pointed out that the anatomical dissection methods Hofmann relied upon could not adequately permit the conclusions he proposed. Additionally, Pollmann and Miehlke both independently reported that fibers within the temporal bone innervating the mouth are located closest to the middle ear.[22,61,65]

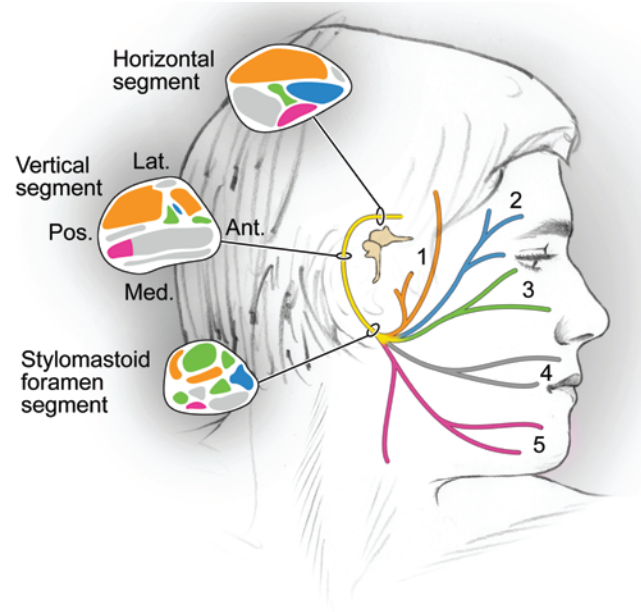

Fig. 8. Somatotopic organization of the facial nerve. Based on several previously published studies using evoked electromyography activity, retrograde axon labeling, and correlating a histopathologically confirmed partial lesion site with peripheral distribution of partial paralysis, a model of the somatotopic organization of the facial nerve was constructed. The spatial distribution of fibers conformed to a roughly clockwise arrangement most strongly in the horizontal segment as depicted in the cross-sections corresponding to fibers innervating the top of the forehead to the lower chin. In the horizontal segment, the branches to the ear and chorda tympani nerve were lateral (1); the branches to the forehead and eye were anterior (2); the branches of the upper midface near the center of the nerve were posterior to fibers innervating the forehead and eye (3); the lower midface branches were further posterior (4); and the lower face, chin, and upper neck branches were anteriormedial to the lower midface (5). Med. = medial.

Obtaining evoked electromyography activity in 48 adult cats, May[61] described a detailed topographical anatomy of the facial nerve in a landmark study. In particular, he clearly mapped out the spatial orientation of the facial nerve within the tympanomastoid segment: the branches to the ear and chorda tympani nerve were lateral (toward the mastoid cortex); the forehead and eye branches were lateroanterior (toward the middle ear); the lower lip branch was anteromedial (toward the floor of the middle ear); the upper midface branch was posterior (toward the horizontal canal); and the lower midface branch was just anterior to the upper midface.[61]

While there was increasing evidence supporting a somatotopic organization of the facial nerve, a group of researchers around the same time period produced results that they argued had suggested the motor fibers may be diffusely distributed in the facial nerve trunk. Sunderland and Cossar[90] originally described repeated alternating plexus formation and reanastomosis in the distal half of the vertical segment and in the extratemporal segment of the facial nerve, relying upon diagrammatic reconstruction of the funicular pattern of the facial nerve from serially sectioned human cadaver material. From these data

they concluded that this plexus formation and interbranching of funiculi disallowed spatial arrangement, although they admitted that it was possible for each peripheral branch to have a predominant representation in a particular sector of the facial nerve because they stipulated that they had only observed a gross anatomical intermingling and funicular redistribution.[92] Additional evidence from Harris'[40] and Scoville's[82] attempts at partial transections of the facial nerve trunk in cats, resulting in no sparing of any of the peripheral branches, was interpreted by those authors as suggestive of a random arrangement of the facial nerve. However, the technical challenges of these experiments have caused others in the field to question the validity of a true hemisection of a nerve, and the lack of definitive data proving the randomization of fibers has allowed many investigators and clinicians to continue to believe the spatial arrangement concept. One critical flaw with Harris' and Scoville's reports, however, was that they did not document the extent of these lesions histopathologically. Thus, it is possible that they actually created near-total lesions. In contrast to the reports of Harris and Scoville, May's[61] landmark study demonstrated that the sparing of selective branches of the facial nerve was, in fact, a direct result of partial lesions as confirmed by histopathological changes specifically at the lesion site.

After May's landmark study, subsequent investigations beginning in the late 1970s and early 1980s using retrograde nerve fiber labeling methods have again produced conflicting data. Using the horseradish peroxidase technique to retrogradely trace axons of the facial motor nerve in the rat and cat, Thomander et al.[96] found that the intratemporal portion of the facial nerve was diffusely distributed. However, Crumley[22] found that a definite spatial orientation was retained at least in the extratemporal portion of the nerve, also using the horseradish peroxidase labeling method. Subsequently, he found that the orbicularis oculi was represented in the posterolateral aspect of the facial nerve near the stylomastoid foramen.[21] It may appear from these studies that the facial nerve could be both diffusely distributed as well as spatially organized, depending on its location, but because few studies used multiple experimental modalities such as evoked electromyography activity and retrograde labeling concurrently to confirm the results obtained from each modality, it becomes difficult to make conclusive statements about the true state of somatotopic organization of the facial nerve at this point. Interestingly, few purely anatomical studies focused on facial nerve somatotopy have been performed since Crumley's reports; therefore, more studies are needed to verify the existence of a somatotopic organization of the facial nerve. Nevertheless, given that the facial nucleus has been proven to be somatotopically organized, it has been generally accepted that there are at least some aspects of somatotopic organization within at least most segments of the facial nerve trunk.

One of the potential concerns about the direct clinical relevance of these basic physiological studies conducted on animals is whether their findings, either supporting or refuting the existence of a spatial organization of the facial nerve, can be applied to the human facial nerve anatomy. Although many studies of the facial nerve anatomy were performed in cats, there is evidence to suggest that the spatial anatomy of the tympanomastoid portion of the facial nerve proposed for the cat model systems may be generally applied to man, a general assumption held in the field supported by the close resemblance of the spatial anatomy of the facial nerve in the motor cortex, pontine nucleus, and facial muscle distribution between the human and the cat.[61]

Pathophysiology of Facial Nerve Lesion During VS Surgery

It has been well established that peripheral nerves can regenerate after nerve injury, and recovery of motor function following nerve damage is due to the ability of peripheral nerves to sprout and reinnervate denervated targets.[17,28,57,75,89,99] The postoperative facial nerve function may thus be determined by the nature of the injury from the tumor growth, damage incurred during tumor resection, or the various factors affecting the recovery of the individual nerve branches. Compared with any potential damage incurred during a resection of a VS, the destructive effects that a slow-growing tumor has on the facial nerve are relatively minimal. In the case of attempted total resection of the tumor, there may be a danger of severing functioning nerve fibers that are microscopically embedded within the tumor.

Tumor-Nerve Interface During VS Surgery

During resection of a VS, the surgeon must find a cleavage plane between the facial nerve and the tumor, which can usually be achieved by fine dissection with the aid of the operating microscope. However, evidence from histological studies of the interface between a VS and the cochlear nerve from en bloc–resected VS tissue suggests that no well-defined connective tissue structure exists between the cochlear nerve and tumor tissue.[60,67,68,104,105] The histological data on the facial nerve and tumor interface are much more scarce, however, due to efforts to preserve facial nerve function among surgeons. Nevertheless, studies exist that examine the interface between facial nerve and tumor, and all demonstrate lack of such interface in at least parts of the tumor, if not in all observable parts of the tumor histologically.[51,56] Jääskeläinen et al.[51] found that where the facial nerve trunk is attached to the surface of the tumor, nerve fibers in the contact area are either abutted directly against tumor cells or penetrated into the tumor tissue. Because these studies examined the histology of large VSs almost exclusively, they are particularly relevant to the current management paradigm for VS because the larger the tumor, the more likely it becomes a surgical candidate. Although there is no definitive evidence that the fibers embedded in the tumor are functional, immunostaining confirmed the existence of axoplasm with neurofilaments.[51] Additionally, where the nerve fibers appeared intact, bundles of axons were sheathed by only a thin endoneurium that could be easily disrupted by the infiltrating tumor, often eliminating the distinct boundary between nerve fibers and tumor tissue, making it difficult to fully visualize the histological

relationship of nerve fibers and tumor tissue through the operating microscope.[51]

The clinical implications of the lack of a tumor-nerve boundary are currently not fully understood, however. Data from a large series of VS resections published by Thomsen et al. suggest that during surgery not all fiber damage as noted by the surgeons leads to functional dysfunction, especially when taking into account the tumor size.[51] In particular, in small- to medium-sized tumors, the postoperative outcome was much better than predicted based on nerve damage noted during surgery, but in large and giant tumors the postoperative outcome was much worse then predicted.[51] Therefore, regarding the small- to medium-sized tumors, the histological questions about the nerve-tumor interface may appear less clinically relevant, because the current microsurgical technique in combination with intraoperative facial nerve monitoring can almost always achieve a de facto cleavage plane and a satisfactory facial nerve outcome. However, in the case of large tumors, unnoticed severing of these fibers can occur during surgery and result more often in immediate postoperative complications, perhaps due to embedding of nerve fibers within the tumor. Nevertheless, recovery of facial nerve function to normal (House-Brackmann Grade I) was shown to be achieved in 53.4% of large tumors and 31.8% of giant tumors, suggesting that not all large or giant tumors engulf all facial nerve fibers or that severing some portion of the fibers does not cause irreversible functional deficits.[51]

It is currently no longer the goal of many VS surgeries to remove every last fragment of tumor that is adherent to the facial nerve, especially in large tumors, mainly to preserve facial nerve function.[95] Moreover, these tumors are generally slow growing, and other adjunctive therapies such as Gamma Knife surgery are available for the residual tumors. In a comparison of 15 patients who underwent subtotal resection of tumors that exceeded 3 cm in diameter with the published results of patients with similarly larger tumors, Raftopoulos et al.[74] found that facial nerve dysfunction was 0% in their patients compared with 20%–35% in patients from other published series. Following their suggestion that planned subtotal resections may be appropriate for large tumors, there is now a multicenter prospective trial to assess subtotal resection as treatment for tumors that are larger than 2.5 cm in diameter (www.clinicaltrials.gov; trial no. NCT01129687). Moreover, Esquia-Medina et al.[25] found not only the expected association between degree of tumor adhesion and facial nerve dysfunction, but also that tumor displacement of the facial nerve predicted worse outcomes. Because both displacement and degree of adhesion were also associated with tumor size and location, they demonstrated that the combination of tumor stage, adhesion, and nerve displacement in a logistic regression model was highly predictive of postoperative facial function.

Based on the current evidence on tumor-nerve interface as well as the slow growth rate of VS, it is recommended that surgeons leave a fragment of the tumor behind if necessary and take the risk that the remnant may eventually grow large enough to cause symptoms. This risk, however, can often be reduced by postoperative Gamma Knife surgery, and it has been reported to be quite effective in the literature.[95]

Ischemia of the Nerve Fiber During VS Surgery

It has been shown that during VS resection, preserving the blood supply of the cochlear nerve is crucial for hearing preservation.[39] Similarly, it has been believed that preserving integrity of the vascular supply of the facial nerve is also crucial for preventing facial nerve dysfunction.[6,8] Anecdotally, surgeons recognize that some blood vessels shared between tumor and the nerve may become damaged when the tumors are removed, and they have observed that local microvascular damage during surgery has been associated with facial nerve dysfunction postoperatively. Recent data suggest that ischemic injury of the facial nerve incurred during VS resection caused by disturbance of the microcirculation of the nerve can cause facial nerve paralysis.[81] Moreover, it has been shown that the use of vasoactive treatments postoperatively can mask the onset of facial nerve dysfunction, and the termination of these treatments results in delayed onset of dysfunction.[81]

Factors Affecting Regeneration of Facial Nerve After VS Surgery

During VS surgery, microscopic damage of nerve fibers can occur due to the nature of a tumor's adherence to the fibers of the nerve. It has been known that there are inherent limitations of nerve regeneration limiting the speed and extent of regeneration. Additionally, several lines of evidence suggest that factors such as misguided axonal regeneration, excessive axonal branching, and lack of specificity of axonal guidance all contribute to the failure of precise regeneration.[19] Recent evidence even suggests that it is possible for the regenerating axons to become aberrant throughout the length of the facial nerve, not only at the site of the lesion.[19] Thus, successful peripheral nerve repair depends not only upon targeting of axons to the periphery, but also upon reestablishment of appropriate connections between the periphery and the CNS.

The consequences of nonspecific regeneration are usually clinically obvious. Incorrect localization of sensory stimuli has long been recognized as a hallmark of nerve regeneration.[66] Misdirected motor axon regeneration can present with gross distortion of the face as the patient attempts to smile.[54] Based on various retrograde labeling techniques, evidence has also emerged in recent years that suggests somatotopic reorganization of the facial nucleus after lesioning, surgical repair, and regeneration of the facial nerve.[4,18,19,29,50] This somatotopic reorganization as a result of axon misguidance during regrowth is believed to underlie involuntary movement of a part of the face during voluntary movement of another part of the face (synkinesis).[11,30] The results of these studies demonstrate the failure of nerve axons to make correct connections with their distal targets during regeneration. While the more recent findings of facial nucleus reorganization after facial nerve regeneration do not directly

demonstrate any facial nerve organization, they certainly have not contradicted previous findings of facial nerve somatotopic organization.[34,70,96]

Inherent Limitations of Peripheral Nerve Regeneration

The speed of peripheral nerve regeneration in mammalian nervous systems is largely limited by the intrinsic rate of axonal outgrowth, which is fairly constant across species. The axonal outgrowth is, in turn, limited by the rate of slow axonal transport, which is 1–4 mm/day.[36,43] This rate, however, declines with aging and contributes to poor recovery in older adults.[101] Given the restricted speed of axonal outgrowth, the time to full recovery clearly depends on the distance that the axons need to travel to reach their targets. Because the different branches of the facial nerve travel different lengths to reach their targets after the nerve exits the stylomastoid foramen, it is very likely that muscles innervated by shorter branches will recover faster than muscles innervated by longer branches. This mechanism may underlie the difference in the rate of recovery between orbicularis oculi and orbicularis oris, which we have observed anecdotally.

Another rate-limiting step impeding functional recovery is that at the site of injury, regenerating axons have to overcome the growth-inhibitory environment of the scar tissue that forms. Unlike the CNS axons, peripheral axons can overcome this inhibitory environment, and axonal regeneration can probably be enhanced by modifying the inhibitory environment.[38,45]

Misguided Axonal Regeneration and the Need for Axonal Guidance

Successful recovery of facial nerve function from VS surgery not only depends on the speed of recovery but also on full and correct restoration of facial nerve function to prevent synkinesis caused by axon misguidance. The establishment of topographic and end organ specificity is necessary to prevent misguided axonal regeneration. Topographic specificity allows axons to return to the muscle or area of skin they served initially; end organ specificity can then match regenerating axons with end organs of the sensory modality or muscle type to which they were connected originally.[46] Presuming the somatotopic organization of the facial nerve, the phenomenon of synkinesis, which can occur after nerve repair, can be easily explained by an injury-induced disorganization of the original somatotopy of the facial nerve.

To achieve full facial nerve recovery without synkinesis in adults, the facial nerve needs to reestablish somatotopy. Evidence suggests that the ability of neonatal rats to reestablish somatotopy of the facial motor nucleus after nerve lesion and repair is likely due to the influence of target-derived trophic factors in the neonate.[1,5] Target-derived factors also regulate collateral axon sprouting,[41,77] and diffusible inhibitory factors may be produced by nontarget regions to repel developing axons away from incorrect paths.[32,71] Additionally, extracellular molecules such as laminin and soluble isoforms of cell surface adhesion molecules such as neural cell adhesion molecule and N-cadherin can act as chemoattractants for axon growth cones and may be produced by targets to create a trophic gradient in the extracellular matrix.[52]

It has been suggested that excessive axonal branching is a major factor contributing to the poor functional results of facial nerve repair due to aberrant projection within several nerve fascicles, such that the branches of 1 axon often synchronously reinnervate muscles with antagonistic functions and impair any coordinated activity.[24] Additionally, often at the site of injury, there are multiple branches with growth cones that are trying to regenerate, and many neurotrophic factors appear to enhance this multiple branching behavior (such as nerve growth factor). However, directed longitudinal elongation of a main axonal branch is necessary for enhanced speed and success of regeneration.

The specificity of axon regeneration is determined primarily at the site of nerve repair; once a regenerating axon is confined to a Schwann cell tube in the distal nerve stump, it usually follows that tube to its peripheral termination.[15,16] Neurotropism, neurotrophism, and mechanical alignment are factors that may influence the specificity of distal Schwann cell tube reinnervation.

Effects of Facial Nerve Injury on the Primary Motor Cortex

Poor recovery of facial function after VS surgery of individual facial muscles may also occur if the organization of higher motor centers is changed after facial nerve lesion.[79,97] This phenomenon has been demonstrated in patients with facial palsy,[76] in a rat model,[33] and in other peripheral nerve lesions.[49] Recent studies in humans comparing ipsilateral and contralateral facial motor cortices showed that, while in healthy patients motor evoked potentials of perioral muscles elicited by transcranial magnetic stimulation of the contralateral hemisphere were always higher in amplitude than that of the ipsilateral cortical transcranial magnetic stimulation, in patients with unilateral peripheral facial paralysis a significant increase was found in the amplitudes of intact perioral motor evoked potentials to hemisphere stimulation contralateral to the paretic side, but not to stimulation ipsilateral to the paretic side.[103] This finding suggests that patients with peripheral facial paralysis can more strongly activate their intact perioral muscles with their ipsilateral cortices.[103] Moreover, it has been suggested that in acute and chronic nerve injuries in humans and in primates, cortical changes are accompanied by subcortical alterations at all levels of the somatosensory core.[103]

Another factor affecting the recovery of facial nerve function that may explain the observed differences in the recovery rate of upper versus lower facial function may be attributable to the bilateral innervation of the upper face that can result in differential disruption of cortical input after facial nerve injury. Neurons in the cortex that project to the facial nucleus are CNS neurons and behave differently compared with peripheral neurons in the facial nerve after nerve injury. It has been shown that the survival of CNS neurons, unlike those in the peripheral nervous system, depends not only on the presence of trophic factors, but also on neural activity in the form of depolarization or increased levels of cyclic adenosine

monophosphate.[35,64,83] Thus, perhaps the lack of signaling from facial muscle motor feedback loops after facial nerve injury alters the behavior and even survival of the CNS neurons in the facial area of the primary motor cortex, at least until function is restored via peripheral regeneration of the facial nerve. Given that the lower face is unilaterally innervated compared with the upper face, which is bilaterally innervated, the probability that lower face dysfunction affects corresponding ipsilateral cortical neurons is greater than the probability that upper face dysfunction affects bihemispheric cortical neurons to the same extent. In other words, in upper face dysfunction, both sides of the cortex must be affected to disrupt cortical output, whereas for the lower face, ipsilateral lesions alone can interfere with cortical output. There is currently no conclusive evidence on the effect of peripheral neuropathy on CNS cortical neuron activity or survival, but given the difference in the anatomy of cortical output to the facial nucleus corresponding to the upper versus lower face, we are likely to discover yet unknown mechanisms regulating peripheral recovery.

Conclusions

Given the importance of preserving facial nerve function in VS surgeries, improved management algorithms regarding surgical approach, extent of resection, intraoperative monitoring, and postoperative rehabilitation can be implemented based on a combination of tumor size, location, growth rate, and intraoperative observations of tumor adhesiveness. For neurosurgeons planning VS surgeries, it is important to remember that attempted gross-total resection of the tumor may pose significant risks of injuring the facial nerve. For resection of large tumors, it may be harder to correctly discern the tumor-nerve interface, and thus care must be taken such as using neuromonitoring to avoid injuring the facial nerve. Anatomically, the portion of the facial nerve central to the geniculate ganglion is more susceptible to injury due to the lack of epineurium. Additionally, because the blood supply to the geniculate ganglion is mostly supplied by 1 artery, in comparison with the existence of overlapping supply from at least 2 arteries in the more distal facial nerve trunk, that region is also more prone to ischemic injury during surgery. Favorable prognostic factors for facial nerve outcome after VS surgery include tumor size of less than 20 mm and the use of neuromonitoring during surgery. Currently there remains a lack of consensus on the use of any particular surgical approach that reduces the risk of facial nerve injury.

The differential recovery speed of the upper face compared with the lower face after surgery can be explained by the presence of bilateral cortical innervation of the upper face, the somatotopic organization of the facial nerve, the distance needed to travel by regenerating axons, and intrinsic differences in regenerating capacity of different facial nerve fibers. Diligent follow-up postoperatively with frequent functional assessments using the House-Brackmann scale is needed to allow physicians to adjust treatment plans for individual patients to deliver optimal care. The recent increase in the use of stereotactic radiosurgery as an alternative to VS microsurgery presents a viable alternative for treating select types of VS, but its efficacy in tumor control and complication profile have yet to be established.

The landscape of VS treatment is changing. As we gain a deeper understanding of the biological behavior of VS and the effect of its treatment on facial nerve function, we may further optimize our management of this disease to provide the best clinical outcome in tumor control and facial nerve preservation.

Disclosure

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The newly diagnosed vestibular schwannoma: radiosurgery, resection, or observation?

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Object. Management recommendations for patients with smaller-volume or newly diagnosed vestibular schwannomas (< 4 cm^3) need to be based on an understanding of the anticipated natural history of the tumor and the side effects it produces. The natural history can then be compared with the risks and benefits of therapeutic intervention using a minimally invasive strategy such as stereotactic radiosurgery (SRS).

Methods. The authors reviewed the emerging literature stemming from recent recommendations to "wait and scan" (observation) and compared this strategy with published outcomes after early intervention using SRS or results from matched cohort studies of resection and SRS.

Results. Various retrospective studies indicate that vestibular schwannomas grow at a rate of 0–3.9 mm per year and double in volume between 1.65 and 4.4 years. Stereotactic radiosurgery arrests growth in up to 98% of patients when studied at intervals of 10–15 years. Most patients who select "wait and scan" note gradually decreasing hearing function leading to the loss of useful hearing by 5 years. In contrast, current studies indicate that 3–5 years after Gamma Knife surgery, 61%–80% of patients maintain useful hearing (speech discrimination score > 50%, pure tone average < 50).

Conclusions. Based on published data on both volume and hearing preservation for both strategies, the authors devised a management recommendation for patients with small vestibular schwannomas. When resection is not chosen by the patient, the authors believe that early SRS intervention, in contrast to observation, results in long-term tumor control and improved rates of hearing preservation.
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Key Words • **vestibular schwannoma** • **acoustic neuroma** • **outcome** • **radiosurgery** • **observation** • **hearing**

Vestibular schwannomas (acoustic neuromas) are generally slow-growing, intracranial extraaxial benign tumors that usually develop from the vestibular portion of the eighth cranial nerve. The incidence is thought be 1 per 100,000. Although some patients note disequilibrium, vertigo, or tinnitus, progressive unilateral hearing decline is the most common symptom that leads to the diagnosis of a vestibular schwannoma.[22] Because of the earlier use of higher-quality MRI, an increasing number of vestibular schwannomas are diagnosed at a time when patients still have useful or even normal hearing. The anticipated growth pattern (using average diameters) of newly diagnosed vestibular schwannomas has been estimated to be one of the following 3 types: 1) no or very slow growth; 2) slow growth (2 mm/year linear growth on imaging studies); or 3) fast growth (> 8 mm/year). In certain cases a doubling of tumor volume within 12 months has been reported. In fact, the tumor volume doubling time may be a better measure of tumor growth than average tumor diameter.[63] Cystic vestibular schwannomas occasionally demonstrate early enlargement of the cystic component of the tumor. Rarely, intratumoral bleeding may lead to rapid enlargement of the mass.[58]

Since the era of both Cushing and Dandy in the early 20th century, almost all patients with newly diagnosed vestibular schwannomas have undergone attempts at surgical removal of their tumors. Although outcomes in the era of microsurgery greatly improved the quality of life of patients after surgery, the earlier diagnosis of these tumors prompted a comprehensive evaluation of less invasive management strategies. In 1969 SRS was first advocated by Leksell and Norén as a potential alternative surgical procedure.[28] Since then, more than 50,000 patients worldwide have undergone SRS using the Leksell Gamma Knife (AB Elekta). This incision-free procedure, as well as other linear accelerator–based technolo-

Abbreviations used in this paper: GKS = Gamma Knife surgery; LINAC = linear accelerator; SRS = stereotactic radiosurgery; SRT = stereotactic radiotherapy.

gies, has greatly expanded the management options for patients with vestibular schwannomas. Patients no longer need to choose simply between craniotomy or observation, a strategy that only makes sense if such tumors cease to grow after initial recognition and cease to cause additional neurological dysfunction.

Increasingly, patients with small tumors are choosing not to undergo resection in favor of a less invasive approach, either observation or irradiation. We believe that early diagnosis and early SRS provide the highest likelihood of achieving the twin goals of successful management: tumor control and maintenance of existing neurological function. Referring physicians and affected patients need to know the long-term risk/benefit ratio of initial observation versus initial SRS.

Observation: the "Wait and Scan" Option

The basic premise of this hypothesis is 2-fold: 1) that the vestibular schwannomas grew but will not grow further after recognition; and 2) that even if some growth is confirmed over time, generally thought to be many years, the patient will maintain a higher level of function than if early treatment is performed.[3,64] The observation strategy was first proposed for elderly patients or those with significant medical comorbidities with an estimated lifespan of less than the growth/symptom progression rate of the vestibular tumor.[15] The patient is evaluated periodically for symptom assessment, and follow-up MRI scans are obtained to monitor the tumor for signs of growth.[59] The ostensible goal of serial observation is to obviate treatment until growth (or perhaps symptom worsening) is confirmed.[15,48] In the elderly, the goal may be to avoid any treatment during the remaining years of life. In the younger patient, it may be to defer potential complications associated with treatment for as long as possible. In our combined 52-year experience in the management of vestibular schwannomas referred to our center, we have observed that 70% patients have measurable growth within 5 years, increasing to more than 95% by the time 10 years has elapsed. Recent reports continue to define annual tumor growth rates of 1–3 mm/year in at least 1 plane. Extracanalicular tumors may progress at an even faster rate,[12] perhaps related to the easier determination of volumetric changes in larger tumors.

Simple linear tumor measurements are associated with a number of problems in the volumetric assessment of tumor growth.[23] First, measurements are dependent on image type, quality, slice thickness, and contrast administration. During the last 20 years high-definition multiplanar MRI has evolved. Axial T2-weighted 1-mm-slice MRI and T1-weighted contrast-enhanced axial and coronal MRI are the only current methods that provide a reasonable way to measure tumor volumes. Computed tomography scanning is insufficient to allow such volumetric measurements. Second, high-resolution MRI must be performed at annual intervals to plot out a reliable tumor growth rate. Third, measurements must be made in the same planes on each scan to maintain consistency. Most screening MRI scans use a 256 × 256 grid, which indicates a pixel size of 1 mm. Since measurements are made by "eyeballing" the 3 tumor diameters (x, y, and z), it has proven almost impossible to differentiate 1- to 2-mm changes in a single plane. Ideally, it is tumor volume that we truly want to know. The lack of simple volumetric tools such as summated region of interest areas in the MRI scanner software makes tumor volume estimates unreliable.

Varughese et al.[63] described their experience with conservative management of patients who had a vestibular schwannoma between 2000 and 2006. The authors evaluated both linear diameter measurements and provided volumetric calculations. The duration of follow-up was not reported. Volume changes were reported according to the "volume doubling time," which they concluded best described the growth rate of untreated tumors. In that report the tumor volume doubling time was 4.40 years. Other authors have reported that tumor volume doubling times range from 1.65 to 2.3 years. During this observation period, many patients will experience deterioration in hearing or may suddenly lose their hearing even without imaging-defined growth. Many patients will also note the development of symptoms such as tinnitus, vertigo, or disequilibrium.

Varughese et al.[63] concluded that "wait and scan" was a realistic option for patients with small vestibular schwannomas. With more widespread MRI screening of patients who present with unilateral hearing dysfunction, tinnitus, or vestibular disorders, early intervention with a minimally invasive procedure offers an option that improves hearing preservation rates compared with observation.[49,54] The patient and referring physicians should have access to available data to devise a balanced initial management strategy.

Régis et al.[49] performed a study to compare a "wait and scan" strategy with GKS in patients with intracanalicular vestibular schwannomas. Forty-seven patients were in the observation arm, and 34 underwent early GKS (median dose 12 Gy to the 50% isodose). The median follow-up was 34.7 months. Conservative management failed in 35 patients (74%), indicated by documented tumor growth or worsening of hearing. During the observation period, 10 patients (21%) had no change in tumor size, 36 (77%) had tumor growth, and 1 (2%) had a slight decrease in tumor size. The authors also studied the tumor volume doubling time in 35 patients. The doubling time was less than 1 year in 11 patients (31%), 1–3 years in 18 (51%), and longer than 3 years in 6 (17%). In the radiosurgery group, 31 patients (66%) had useful hearing at the time of diagnosis. Twenty-one patients (68%) retained useful hearing, but 10 (32%) lost useful hearing during follow-up. Régis et al. confirmed that tumor control and functional hearing preservation rates were higher in patients who underwent early GKS (88%, 79%, and 60% at 1, 2, and 5 years, respectively). In contrast, in patients who underwent observation, hearing preservation rates were 78%, 43%, and 14% at 1, 2, and 5 years, respectively. In this study the useful hearing preservation rate also was better in patients who underwent SRS (77%, 70%, and 64% at 3, 4, and 5 years, respectively) than in those who underwent "wait and scan" (75%, 52%, and 41% at 3, 4, and 5 years, respectively). This study supports our belief

that observation results in tumor growth and hearing deterioration at a much greater degree than does early SRS, at least early GKS.

Shirato et al.[54] compared observation with fractionated SRT in patients with vestibular schwannomas. Twenty-seven patients were observed, and 50 underwent SRT with mean follow-up periods of 35 and 31 months, respectively. In the SRT group, 37 patients underwent the procedure as primary tumor management and 13 had SRS after a prior resection (all 13 were deaf). In the SRT group, 34 had measureable hearing before treatment compared with 23 of 27 in the observation group. The mean tumor diameter was 18 mm. Eight patients were noted to have tumor enlargement (transient in 6) greater than 2 mm after SRT within the first 2 years. One patient underwent a later resection (2%). Eleven patients (41%) in the observation group required salvage therapy at 21 months or later; 7 (26%) underwent resection; and 4 (15%) had SRT. The mean tumor growth rate was 3.87 mm/year for the observation group and −0.75 mm/year for the SRT group. Preservation of Gardner-Robertson class hearing rates at 3 and 5 years' follow-up were 61% and 31%, respectively, for the observation group compared with 53% at both 3 and 5 years in the SRT group.

In a recent report, Rasmussen et al.[47] compared outcomes of 42 patients following fractionated SRT using mask localization (54 Gy in 27–30 fractions) with a cohort of 409 control individuals who were observed. They noted that fractionated radiotherapy accelerated hearing loss and that cochlear dose was relevant. In the observation group, hearing deterioration was not dependent of tumor growth. By 5 years, half of these patients had lost hearing. The authors believed that their results showed that fractionated radiotherapy was not superior to radiosurgery and that it appeared to accelerate hearing loss rather than prevent it. The use of 54 Gy may indeed be excessive. Mask localization rather than stereotactic frame–based localization may also have contributed to their results.

Between 1990 and 2005, Bakkouri et al.[2] evaluated 386 patients who harbored unilateral vestibular schwannomas. At 1 year, 61 patients were lost to follow-up, and the strategy was discontinued for another 77 patients (24%) due to tumor growth (> 3 mm for 43%). Neurological symptoms that developed included disabling vertigo (in 11 patients [14%]) and hearing deterioration (in 29 [38%]) Six patients requested surgery. The annual tumor growth rate was less than 1 mm/year in 59%, 1–3 mm/year in 29%, and greater than 3 mm/year in 12%. Despite the absence of long-term data, the authors continued to advocate observation for patients whose tumors initially showed a "slow growth rate."

Martin et al.[34] analyzed 320 patients who underwent observation after an intracanalicular tumor or a tumor smaller than 2 cm in diameter was found. The mean follow-up was 43 months, and 276 patients had at least 1 follow-up image. Sixty-two patients (22%) exhibited tumor growth, a rate that increased to 90% within 3 years. The average annual growth rate was 4 mm/year. The authors believed that 65% of the tumors grew slowly (0.5–5 mm/year) and 35% grew rapidly (> 5–17 mm/year). This study also indicated that cystic tumors were more likely to enlarge and to do so at a faster rate.

Hajioff et al.[12] studied 72 patients with unilateral tumors with an extended median follow-up of 121 months. The median tumor diameter growth rate at 10 years was 1 mm/year. The median tumor size at the time of diagnosis was 9.8 mm. The authors found that extracanalicular tumors tended to grow faster than intracanalicular tumors. During the follow-up period, conservative management failed in 25 patients (35%). During the first 5-year follow-up period after diagnosis, conservative treatment failed in 75% of these patients at an average of 37 months, and 1 patient died. During follow-up, 29 patients (40%) exhibited growth of greater than 1 mm/year (50% of cerebellopontine angle tumors and 6% of intracanalicular tumors). Twenty-seven patients (38%) experienced a growth rate of less than 1 mm/year, and 16 (22%) remained unchanged. Audiometric follow-up in 40 patients at 80 months showed that most patients had significant hearing loss even in the absence of measurable tumor growth. Hearing loss was worse in the presence of measurable tumor growth (the mean speech discrimination score deteriorated by 40%).

A review of 47 patients with unilateral intracanalicular schwannomas was performed by Pennings et al.[42] to evaluate hearing function during a period of observation. The mean follow-up was 3.6 years. Nineteen patients (40%) had tumor growth greater than 2 mm (8 patients underwent treatment), 24 (51%) had stable tumors, and 4 (9%) had slight tumor regression. All patients showed hearing degradation during follow-up. The mean pure tone average at the first audiogram was 37.5 dB, which diminished to 50.9 dB at the time of the last audiogram. The speech discrimination scores decreased from 66.2% to 54.5%. Despite the documentation of both tumor progression and hearing deterioration in many patients, the authors continued to recommend observation rather than intervention.

Sughrue et al.[57] performed a literature analysis that combined data of 982 patients from 34 studies. The follow-up period varied from 26 to 52 months. The authors found a mean growth rate of 2.9 mm/year. Patients with slower-growing tumors (< 2.5 mm/year) had higher hearing preservation rates. The authors concluded that the growth rate was a more important predictor of hearing loss than the initial tumor diameter. Figure 1 indicates a decision analysis tree for patients after initial diagnosis.

Stereotactic Radiosurgery: the Minimally Invasive Treatment Option

Stereotactic radiosurgery for vestibular schwannoma using the Gamma Knife has been practiced for more than 40 years. Long-term outcome results have established SRS as an important, minimally invasive alternative to resection. Stereotactic radiosurgery is likely the most common procedure performed for smaller vestibular tumors, although the case volume of patients receiving fractionated radiotherapy is not known. Advanced dose planning software, intraoperative high-resolution MRI, dose optimization, and robotic delivery reflect the evolution of this technology. To reduce risk, various image-guided lin-

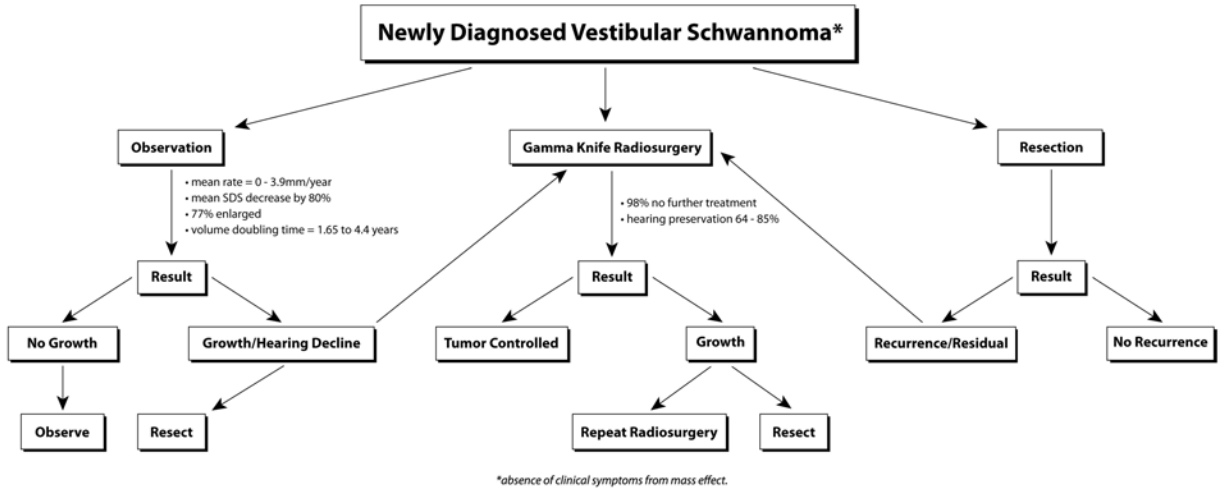

Fig. 1. Decision chart for vestibular schwannoma management. SDS = speech discrimination score.

ear accelerator devices (such as Trilogy, Synergy S, Novalis, and CyberKnife) are often used under fractionation schedules of 3–30 sessions. Proton beam technology is also used to deliver fractionated radiation therapy. The goals of SRS for vestibular schwannoma include prevention of further tumor growth and preservation of existing neurological function.

Optimizing Radiosurgical Dose Planning

Image interpretation, dose planning, and dose delivery are 3 critical components of successful radiosurgery. Complete volumetric conformal and selective tumor radiosurgery improves the rates of facial, cochlear and trigeminal nerve preservation.[29] Reduction of the dose delivered to the brainstem is especially relevant during treatment of larger tumors. Specific GKS techniques include accurate MRI-based definition (or CT-based definition in patients ineligible for MRI) of the tumor volume, use of multiple isocenters, beam weighting, and selective use of beam-blocking patterns to reduce the dose to adjacent critical structures. This degree of conformality can be achieved through multiple isocenter planning, typically by using small beam diameters. A series of 4 mm isocenters are used to create a tapered isodose plan to conform to the intracanalicular portion of the tumor.

Dose Selection

After optimizing the computer dose plan, a maximum and marginal tumor edge dose is prescribed. In GKS a dose of 12–13 Gy is typically prescribed to the 50% (or other) isodose line that conforms to the 3D tumor margin. The most common dose is 12.5 Gy and is most often prescribed to maximize hearing preservation in patients with smaller tumors. We prescribe 12 Gy to the tumor margin of larger tumors. For patients with deafness related to prior resection, we often prescribe 13 Gy to the tumor margin. These marginal doses are associated with a low complication rate and yet maintain a high rate of tumor control. Although experienced centers including the Gamma Knife group from the Hopital Timone in Marseille often use marginal doses of 11 Gy, we suspect that further dose reduction is unlikely to improve hearing preservation rates and may lead to higher rates of tumor progression after many years.[61] Doses in the range of 12–13 Gy at the margin are also used for patients with bilateral (neurofibromatosis Type 2–related) vestibular schwannomas and for patients with contralateral deafness from other causes, for whom hearing preservation is highly desirable.

After prescribing the tumor margin dose, we use computer software to outline adjacent critical structures and then measure the mean dose to the cochlea, semicircular canals, and brainstem. Long-relaxation time (T2) 1-mm axial plane volumetric MRI is necessary to identify the cochlea for dose planning. A mean cochlear dose less than 4.2 Gy may be important for hearing preservation,[19] a finding confirmed by others.[13,61] The majority of the tumor volume receives a radiobiological dose up to 4 times the biologically equivalent dose delivered by fractionated image-guided radiation therapy. The maximum radiosurgical dose of 25 Gy may be radiobiologically equivalent to 100 Gy of fractionated radiation. The SRS technology must also be able to restrict the dose to adjacent structures by having a very sharp dose gradient at the tumor edge. While many radiosurgical centers have evolved toward similar dose selection parameters, the doses and regimens chosen for fractionated radiotherapy continue to vary.

Gamma Knife Surgery: Clinical Results

Long-term results of GKS for vestibular schwannomas have been documented.[5,8,14,24,27,33] Recent reports suggest a tumor control rate of 93%–100% after radiosurgery.[5–11,14,16–18,21,24–27,29–33,38,41,43,45,46] Kondziolka et al.[26] studied 5- to 10-year outcomes in 162 patients with vestibular schwannomas who had undergone radiosurgery at the University of Pittsburgh. In this study a long-term 98% tumor control rate was reported. In further analysis of this cohort, the median follow-up for the 136 patients still living at the time of the study was 10.2 years. Serial imaging studies obtained after radiosurgery in 157

patients showed a decrease in tumor size in 114 patients (73%), no change in 40 patients (25.5%), and an increase in 3 patients who later underwent resection (1.9%).[26,27] No patient developed a radiation-associated malignant or benign tumor (defined as a histologically confirmed and distinct neoplasm arising in the initial radiation field after at least 2 years have passed). In patients younger than 40 years with minimum 4-year follow-up, all remained employed and active.[32] Only 2% of patients required tumor resection after radiosurgery. Norén,[41] in his 28-year experience with radiosurgery for vestibular schwannomas, reported a 95% long-term tumor control rate. Niranjan et al.[40] analyzed the outcome of intracanalicular tumor radiosurgery performed at the University of Pittsburgh. All patients had imaging-documented tumor growth control.

Hearing Preservation

Preradiosurgery hearing can now be preserved in 60%–90% of patients. The best hearing preservation rates are found in patients with smaller tumors. In a long-term (5- to 10-year follow-up) study conducted at the University of Pittsburgh, 51% of patients had no change in hearing.[7,26] All patients who were treated with a margin dose of 14 Gy or less maintained a serviceable level of hearing after intracanalicular tumor radiosurgery.[40] Among patients treated after 1992, the 5-year actuarial rates of hearing level preservation and speech preservation were 75.2% and 89.2%, respectively, for 89 patients treated with a 13-Gy tumor margin dose.

In a longer-term assessment of hearing at a median of 6 years, the same Gardner-Robertson level was preserved in 71%, serviceable hearing was confirmed in 74%, and any testable hearing was present in 95%. For intracanalicular tumors, these rates were 84%, 92%, and 100%, respectively. Our recent research has shown that the mean cochlear dose is important for hearing preservation. Seventy-seven patients with serviceable hearing (Gardner-Robertson Classes I and II) underwent GKS between 2004 and 2007.[19] This interval reflects a period when newer dose planning systems facilitate measurements of dose delivered to critical structures such as the cochlea, trigeminal nerve, and brainstem. The median tumor volume was 0.75 cm^3 (range 0.07–7.7 cm^3), and the median marginal dose was 12.5 Gy (range 12–13 Gy). At diagnosis, a longer distance from the lateral tumor to the end of the internal auditory canal correlated with better hearing. At a median of 20 months, no patient required any additional management. Serviceable hearing was preserved in 71% of patients but in 89% of patients who had Class I hearing (46 patients). Significant prognostic factors for serviceable hearing preservation were Gardner-Robertson Class I, pre-SRS speech discrimination scores of 80% or more, pre-SRS pure tone averages of less than 20 dB, patient age younger than 60 years, intracanalicular tumor location, and tumor volumes less than 0.75 cm^3. All 12 patients younger than 60 years old with a cochlear dose of less than 4.2 Gy maintained serviceable hearing at 2 years. An average cochlea dose of less than 4.2 Gy was associated with better hearing, a finding similar to the dose of 4 Gy noted from Marseille. Younger age is also important for hearing preservation with age under 60 (Pittsburgh group[19]) or 50 (Marseille group[61]) being relevant.

Recently, Hasegawa et al.[13] provided data on 117 patients who underwent GKS and had a median follow-up of 7 years. The tumor control rate was 97.5%, which is similar to that reported by other centers. The cochlear dose again proved important. In a subset of patients with Grade I hearing who were treated using current techniques, the 3- and 5-year hearing preservation rates were 80% and 70%, respectively.

In 2010, Yang et al.[66] performed a systematic literature review of the results of GKS hearing preservation. Forty-five articles that included 4234 patients provided the data. The mean follow-up was 44 months. Overall, the hearing preservation rate was 61% with a dose of 13 Gy or lower, and 50.4% at more than 13 Gy. Neither patient age nor tumor volume correlated with hearing preservation.

Facial Nerve and Trigeminal Nerve Preservation

Facial and trigeminal nerve function can now be preserved in the majority of patients (> 95%). In a study using MRI-based dose planning, a 13-Gy tumor margin dose was associated with a 0% risk of new facial weakness and a 3.1% risk of trigeminal sensory loss (5-year actuarial rates). A margin dose of less than 14 Gy was associated with a 2.5% risk of new facial weakness and a 3.9% risk of trigeminal sensory loss.[8] No patient who underwent radiosurgery for an intracanalicular tumor developed new facial or trigeminal neuropathies. In the current 12- to 13-Gy dose range, any degree of facial weakness is exceedingly rare.

Linear Accelerator Radiosurgery: Clinical Results

Suh et al.[59] evaluated 29 patients treated with a modified linear accelerator stereotactic radiosurgery system. The median margin dose was 1600 cGy. The 5-year local disease control rate was 94%. Long-term complications included new or progressive trigeminal and facial nerve deficits (estimated 5-year incidence) of 15% and 32%, respectively. Subjective hearing reduction or loss occurred in 14 (74%) of the 19 patients who had useful hearing prior to treatment. Since there was a high risk of cranial neuropathy, these authors did not recommend using only CT-based planning and high prescription doses. Spiegelmann et al.[56] reported their results of LINAC radiosurgery for 44 patients with vestibular schwannomas. After a mean follow-up period of 32 months (range 12–60 months), 98% of the tumors were controlled. The actuarial hearing preservation rate was 71%. New transient facial neuropathy developed in 24% of the patients and persisted to a mild degree in 8%. The University of Florida group published clinical outcomes in a series of 390 patients, with a high control rate and a facial neuropathy rate of 0.7% using current techniques and dose.[10]

Stereotactic Radiation Therapy: Clinical Results

Stereotactic radiation therapy or fractionated SRT refers to the delivery of a standard fractionation scheme of

radiation, used with rigidly applied or relocatable stereotactic-guiding devices. Many LINAC-based radiosurgery centers (driven by the desire to reduce complication rates) use dose fractionation for vestibular schwannomas.[18,36,44,52–55,59,60] Ishihara et al.[18] reported a 94% tumor control rate at a median follow-up of 31.9 months in a series of 38 patients who underwent CyberKnife radiosurgery for vestibular schwannoma. One patient developed transient facial paresis (2.6%) and another developed trigeminal nerve neuropathy (2.6%). Fuss et al.[11] described 51 patients with vestibular schwannomas who were treated with SRT. The mean follow-up period was 42 months, and the actuarial 5-year tumor control rate was 95%. One patient developed a transient facial nerve paresis, and 2 noted new trigeminal dysesthesias. Chung et al.,[4] using SRT for 25 patients with useful hearing, reported 57% hearing preservation at 2 years. The mean pre- and post-SRT speech recognition threshold was 20 and 38 dB, respectively. The mean proportion of pre- and post-SRT speech discrimination was 91% and 59%, respectively.

Sawamura et al.[53] treated 101 patients with vestibular schwannomas using fractionated SRT to a total dose of 40–50 Gy, administered in 20–25 fractions over a 5- to 6-week period. The median follow-up period was 45 months, and the actuarial 5-year tumor control rate was 91.4%. The actuarial 5-year rate of useful hearing preservation (Gardner-Robertson Class I or II) was 71%. The complications of fractionated SRT included transient facial nerve palsy (4%), trigeminal neuropathy (14%), and balance disturbance (17%). Eleven patients (11%) developed progressive communicating hydrocephalus after SRT and required a shunt.

Meijer et al.[35] performed a single-institution trial to study whether fractionated stereotactic radiation therapy is superior to single-session LINAC-based radiosurgery. They assessed treatment-related toxicity and local tumor control in patients with vestibular schwannomas. These authors analyzed 129 patients with vestibular schwannomas who were treated at an LINAC-based radiosurgery facility. Stereotactic radiation therapy was performed in 80 patients with a relocatable guidance device using 5 sessions that delivered either 4 or 5 Gy to the tumor margin at the 80% isodose. Forty-nine patients had SRS of 1 × 10 Gy and later 1 × 12.5 Gy at the 80% isodose using a stereotactic frame. There was no statistically significant difference between the single-fraction group and the fractionated group with respect to mean tumor diameter (2.6 vs 2.5 cm) or mean follow-up time (both 33 months). Outcome differences between the single-session group and the fractionated treatment group with respect to 5-year local control probability (100% vs 94%), 5-year facial nerve preservation probability (93% vs 97%), and 5-year hearing preservation probability (75% vs 61%) were not statistically significant.

Andrews et al.[1] published the Thomas Jefferson University experience using stereotactic radiotherapy at a total dose of 50.4 or 46.8 Gy. In patients with Class I or II hearing, the median follow-up was 65 weeks. Although no patient had later tumor growth, the hearing preservation rates were better at the lower dose. At 3 years, the hearing preservation rate was 55%–60%, and no patient with Class II hearing maintained hearing if they received the 50-Gy dose.[1] Based on these findings, the group reported the use of even lower doses to try to improve hearing outcomes (D. W. Andrews, personal communication, meeting of the Acoustic Neuroma Association, 2011). As noted above, Rasmussen et al.[47] concluded that fractionated radiotherapy at a dose of 54 Gy (higher than used in the Thomas Jefferson University report), appeared to accelerate hearing loss.

Kapoor et al.[20] published outcomes after fractionated SRT from Johns Hopkins Hospital in 496 patients, of whom 385 had follow-up. Radiation was administered in five 5-Gy fractions or ten 3-Gy fractions. Resection was later performed in 3%. Attempted hearing preservation is often given as a reason why some centers choose to use fractionated radiotherapy, but hearing results were not provided.

The Risk of Delayed Malignancy

The risk of a benign or malignant secondary tumor development after SRS has been suggested as a reason to continue observation rather than to perform early radiosurgery. After fractionated external-beam radiation therapy, this risk may be as high as 2%, as has been reported many years after such radiation therapy for pituitary tumors.[39] Delayed oncogenesis following radiosurgery is rare because the target and regional tissue volume irradiated are small, the procedure results in only a single radiation exposure, and the high central dose more likely leads to cell death rather than cell transmutation. There are case reports after radiosurgery or radiotherapy.[62,65] Although we quote to our radiosurgery patients a less than 1:1000 risk of secondary tumor formation over a 5- to 30-year follow-up period, this figure is almost certainly too high.[39] Neither the incidence nor the prevalence of secondary radiation-related tumors is known despite the more than 40 years of radiosurgery experience using the Gamma Knife. Rowe et al.[51] reviewed their experience in 5000 patients treated with SRS and 30,000 patient-years of follow-up. More than 1200 patients had delayed assessments beyond 10 years. The authors detected a single new brain astrocytoma but anticipated 2.47 cases based on population incidence statistics.

Comparison of GKS and Resection: Level 2 Studies

Patients with small vestibular schwannomas may choose resection as their initial form of care or after a period of observation when growth or new symptoms develop. Results after surgery are dependent on surgeon experience. There can be strong opinions about the different treatment choices. Thus, we reviewed the available comparative literature. Despite these available reports, patient selection bias, personal choice, physician skill, and quality of data collection all remain important variables that can affect outcome. There is a large case-series literature on outcomes after resection that continues to evolve. Individual outcomes are dependent on the factors noted earlier, including surgeon goals for each patient.

Unfortunately, it is likely that a randomized clinical trial will probably never be completed to compare resection with radiosurgery for vestibular schwannomas. However, there are several well-matched (Level 2) cohort studies that compare outcomes for patients with tumors smaller than 2.5 cm in extracanalicular diameter. Karpinos et al.[21] analyzed 96 patients with unilateral vestibular schwannomas treated using the Leksell Gamma Knife or microsurgery and concluded that radiosurgery was associated with a lower rate of immediate and long-term development of facial and trigeminal neuropathy, postoperative complications, and hospital stay. Radiosurgery yielded better measurable hearing preservation than microsurgery and equivalent serviceable hearing preservation rate and tumor growth control.

Between 1990 and 1991, Pollock et al.[46] studied 87 patients who were treated treated at the University of Pittsburgh and had unilateral, previously unoperated vestibular schwannomas with an average diameter of less than 3 cm. In this matched cohort trial preoperative patient characteristics and average tumor size were similar between the treatment groups. Microsurgical or radiosurgical techniques were used by experienced surgeons in both treatment groups. The treatment groups were compared based on cranial nerve preservation, tumor control, postoperative complications, patient symptoms, length of hospital stay, total management charges, effect on employment status, and overall patient satisfaction. Stereotactic radiosurgery was more effective in preserving normal postoperative facial function and hearing preservation with less treatment associated morbidity. Effect on preoperative symptoms was similar between the treatment groups. Postoperative functional outcomes and patients' satisfaction were greater after radiosurgery when compared with microsurgery. Patients returned to independent functioning sooner after radiosurgery. Hospital length of stay and total management charges were less in the radiosurgical group.

In a similar study of patients with vestibular schwannomas, Régis et al.[50] used objective results and questionnaire answers to compare the results of radiosurgery (97 consecutive patients) with a microsurgery group (110 patients who fulfilled the inclusion criteria). Questionnaire answers indicated that 100% of patients who underwent GKS compared with 63% of patients who underwent microsurgery had no new facial motor disturbance. The mean hospitalization stay was 3 days after radiosurgery and 23 days after microsurgery. All working patients who underwent SRS kept the same professional activity, compared with 56% in the microsurgery arm. The mean time away from work was 7 days for the SRS group compared with 130 days for the microsurgery group. Among patients whose preoperative hearing level was Class I according to the Gardner-Robertson scale, 70% had preserved functional hearing after radiosurgery (Class I or II), compared with only 37.5% in the microsurgery group. At 4 years of follow-up, GKS provided better functional outcomes than microsurgery. It was concluded that radiosurgery was an effective and less costly management strategy for unilateral vestibular schwannomas smaller than 3 cm in diameter, and it should be considered a primary management option.

In another study, Myrseth et al.[38] compared the quality of life outcomes for 189 patients treated with either microsurgery or radiosurgery, who harbored vestibular schwannomas that were less than 30 mm in diameter. The outcome analysis included assessments of tumor control, cranial nerve preservation rates, and complications. The results showed that cranial nerve function and overall patient outcomes were better in the radiosurgery group. The results reveal that from the patients' perspective, radiosurgery provides a more desirable outcome than microsurgery. A second 2009 report confirmed these findings.[37] Pollock et al.[45] prospectively collected data on patients undergoing either resection or GKS at the Mayo Clinic and found similar or better outcomes after radiosurgery, including quality of life measures.

Conclusions

Numerous studies show that vestibular schwannomas have variable growth rates. Tumor volume in some patients may be linear; in others it may be stepwise. By 10 years most clinical experience demonstrates that the vast majority of patients will have tumor growth. As the tumor grows, cranial nerve function, especially hearing, is likely to deteriorate. Hearing loss may progress even without imaging-defined growth. Linear growth measurements are not sensitive to 3D changes in tumor volume, which may be better understood according to the tumor volume doubling time calculation. Stereotactic radiosurgery arrests the growth of almost all vestibular schwannomas. When performed at experienced centers, cranial nerve function is preserved and quality of life is enhanced. When applied early after tumor diagnosis, useful hearing is much more likely to be preserved. Fractionated radiotherapy techniques have shown less consistent outcomes. Matched cohort studies show that radiosurgery has either better or similar outcomes to resection, depending on the outcome measured. The "wait and scan" option has been advocated in recent years, especially since the minimally invasive strategy of radiosurgery emerged. We believe that "wait and scan" only makes sense in patients whose medical comorbidities indicate a high likelihood of death from other causes in the next 5 years of life.

Disclosure

Dr. Kondziolka is a consultant for AB Elekta. Dr. Lunsford owns stock in and is a consultant for AB Elekta.

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Surgical approaches for resection of vestibular schwannomas: translabyrinthine, retrosigmoid, and middle fossa approaches

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Surgical removal remains one of the key treatment modalities for vestibular schwannomas. A team approach between a neurotologist and a neurosurgeon offers the patient the expertise of both specialties and maximizes the chances for an optimal outcome. Vestibular schwannomas can typically be resected through 1 of 3 main surgical approaches: the translabyrinthine, the retrosigmoid, or the middle fossa approaches. In this report and videos, the authors describe and illustrate the indications and surgical techniques for the removal of these tumors.
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Key Words • **vestibular schwannoma** • **surgical technique** • **translabyrinthine approach** • **retrosigmoid approach** • **middle fossa approach**

Surgical removal remains one of the key treatment modalities for VSs.[3] A team approach between a neurotologist and a neurosurgeon offers the patient the expertise of both specialties and maximizes safety and the chances for an optimal outcome. Vestibular schwannomas can typically be resected through 1 of 3 main surgical approaches: the translabyrinthine,[2] the retrosigmoid,[4] or the middle fossa approaches.[1] In this article, we describe and illustrate our indications and surgical technique for the removal of these tumors.

The Translabyrinthine Approach

The translabyrinthine approach (Fig. 1, Video 1) provides excellent access to the IAC.

Video 1. Clip showing the translabyrinthine approach for resection of a VS. Click here to view with Media Player. Click here to view with Quicktime.

The translabyrinthine approach is chosen for patients with poor preoperative hearing and for patients with large tumors who have a low probability of hearing preservation. The size of the tumor is typically not a limiting factor for this approach. The approach has several advantages, including 1) it offers early identification of the facial nerve in the auditory canal, and 2) there is absolutely no need for cerebellar retraction.

The patient is positioned supine, and the head is rotated to the contralateral side. Somatosensory evoked potentials, motor evoked potentials, and facial nerve monitoring are used. A hockey stick–shaped retroauricular skin incision extending behind the mastoid tip is made. The mastoidectomy is performed with the high-speed drill, and the sigmoid sinus, presigmoid dura mater, and middle fossa dura are exposed. The mastoid segment of the facial nerve is skeletonized. The eustachian tube is packed with bone wax and Surgicel to prevent CSF leakage.

Next, the labyrinthectomy is performed to access the IAC. The translabyrinthine approach exposes 270° of the circumference of the IAC. The dura of the IAC is then opened. We prefer to open along the axis of the IAC and use a Y-shaped incision to open the dura in the posterior fossa. The facial nerve is located and confirmed using stimulation. The vestibular nerves are cut, and the tumor is dissected from the facial nerve. The presigmoid dura is then opened. The tumor is dissected from the cerebellum and the brainstem and is debulked using the ultrasonic aspirator.

It should be noted that the operative corridor is smaller with this approach and that debulking of the tumor is necessary prior to dissection of the tumor capsule from the brainstem and nerves. Once the tumor has been debulked, the extracapsular dissection proceeds more easily. The root entry zone of the facial nerve at the brainstem is identified and confirmed with stimulation. The flocculus and choroid plexus emerging from the lateral foramen are useful landmarks to identify the facial nerve origin just superior to the pontomedullary junction. The origin of the vestibulocochlear nerve at the brainstem is identified just ventral and superior to the facial nerve origin, where it is then divided. This allows the tumor to be gently reflected

Abbreviations used in this paper: CPA = cerebellopontine angle; IAC = internal auditory canal; VS = vestibular schwannoma.

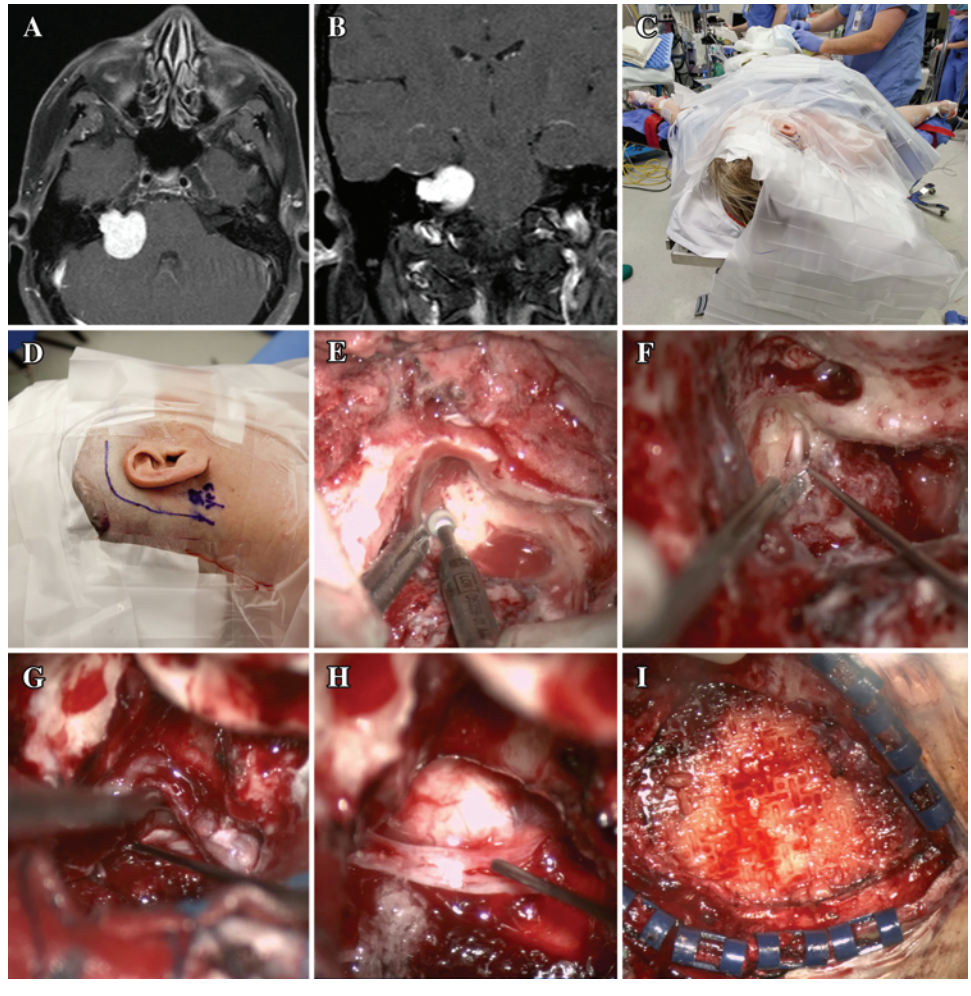

Fig. 1. Axial **(A)** and coronal **(B)** T1-weighted MRI studies obtained with contrast enhancement of the brain showing a VS occupying the right CPA and extending into the IAC of a 42-year-old woman who presented with a history of hearing loss in the right ear. **C–I:** Intraoperative photographs obtained during the translabyrinthine approach to the tumor. **C:** The patient is positioned supine, and her head is rotated to the contralateral side. **D:** A hockey stick–shaped retroauricular skin incision extending behind the mastoid tip is made. **E:** The mastoidectomy is performed with the high-speed drill. **F:** After the dura of the IAC is opened, the facial nerve is located and confirmed using stimulation. The vestibular nerves are cut, and the tumor is dissected from the facial nerve. **G:** The tumor is debulked with extracapsular dissection. The root entry zone of the facial nerve at the brainstem is identified and confirmed with stimulation, and then the tumor is gently reflected laterally and dissected from the facial nerve under direct visualization. **H:** After tumor removal, the facial nerve is stimulated at the root entry zone. **I:** Free abdominal fat is used to close the dural defect and occlude the mastoidectomy cavity. The bone defect is covered with a mesh implant.

laterally and superiorly to dissect it away from the facial nerve under direct vision.

After tumor removal and hemostasis, free abdominal fat is used to close the dural defect and occlude the mastoidectomy cavity. The bone defect is covered with a mesh implant. The rest of the closure is done using 2-0 Vicryl interrupted sutures for the galea and 4-0 Vicryl in a subcuticular fashion for the skin. This offers the advantage that the stitches do not have to be removed.

The Retrosigmoid Approach

The retrosigmoid approach (Fig. 2, Video 2) provides a trajectory that is parallel to the petrous bone.

Video 2. Clip showing the right retrosigmoid approach for resection of a VS. Click here to view with Media Player. Click here to view with Quicktime.

It allows removal of tumors of different sizes and offers the possibility of hearing preservation.[5,7] The senior authors prefer this approach for tumors with significant mass in the cistern and in patients with serviceable hearing, in whom hearing preservation is the goal. The advantage is that the approach offers the surgeon a wide view of the cisternal component of the tumor and thus good access to the root entry zone of the acoustic nerve. The disadvantages include the necessity for cerebellar retraction and less access to the facial and cochlear nerves in the distal IAC, which increases the potential to leave a residual tumor fragment behind.

Patients with tumors that do not involve the lateral one-third of the IAC and do not impinge on the brainstem are the best candidates for hearing preservation. For a medium-to-large tumor (> 2.5–3 cm) that extends far lateral to the fundus of the IAC in a patient with good hearing,

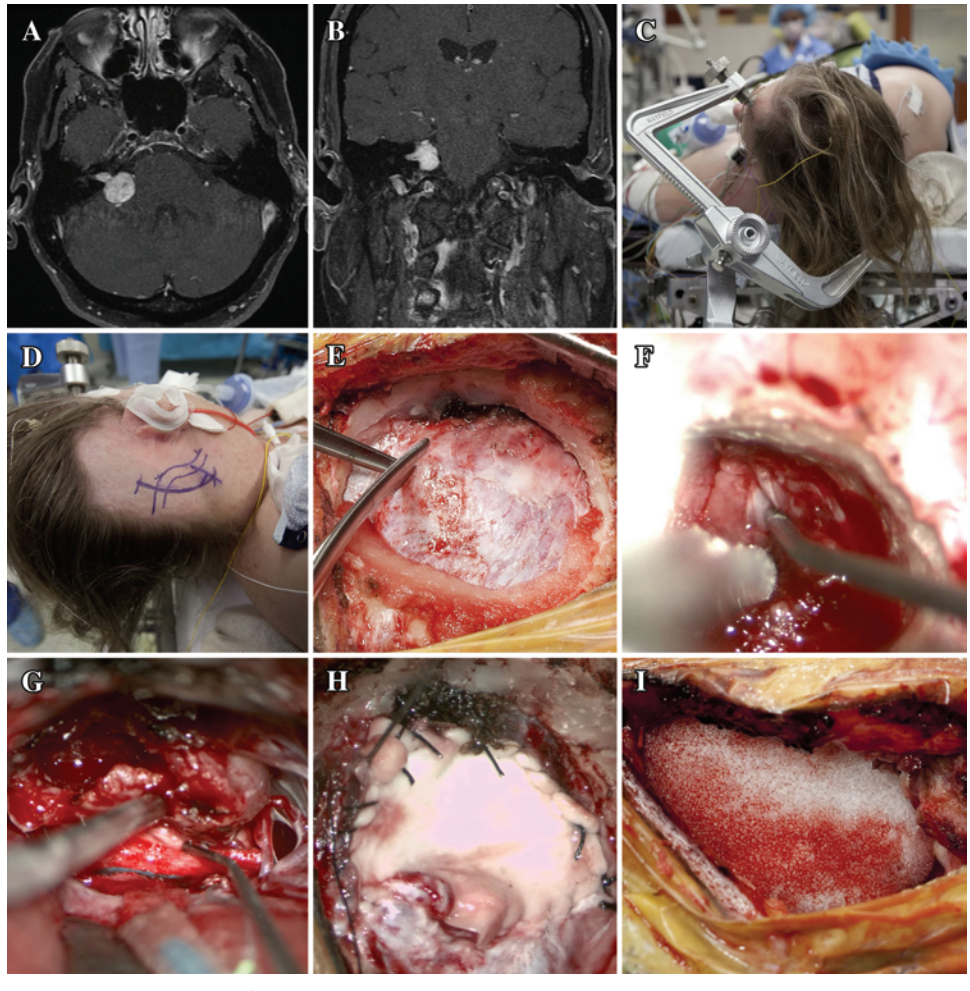

Fig. 2. Axial **(A)** and coronal **(B)** T1-weighted MRI studies with contrast enhancement showing a VS occupying the right CPA and extending into the IAC of a 59-year-old woman. She presented with a history of tinnitus and decreased hearing in the right ear, and her audiogram showed that she had serviceable hearing on the right side. **C–I:** Intraoperative photographs obtained during the retrosigmoid approach to the tumor. **C:** The patient is placed supine or in the lateral position, and the head is fixed in a Mayfield 3-point head holder. **D:** The incision is located approximately 2 fingerbreadths behind the pinna of the ear. The planned suboccipital craniectomy exposes the edges of the transverse and sigmoid sinuses. **E:** The dura is opened parallel to the sinuses and then retracted with 4-0 silk sutures. **F:** The posterior lip of the IAC is drilled, the vestibular nerves are divided, and the facial nerve is identified anterior to them and confirmed with stimulation. The cochlear nerve is identified just inferolateral to the facial nerve. **G:** The tumor is then dissected off the cerebellum and brainstem and debulked using the ultrasonic aspirator. The root entry zones of the facial and cochlear nerves at the brainstem are identified, and the facial nerve is confirmed with stimulation. The tumor is dissected off these 2 nerves very carefully and under direct vision. **H:** After tumor removal, the dura is closed in a watertight fashion by using an AlloDerm patch. **I:** The bone defect is covered with Medpor cranioplasty (porous polyethylene implant).

we believe that hearing preservation is not likely and that the retrosigmoid approach does not adequately expose the lateral end of the IAC. We believe that such cases are best managed via the translabyrinthine approach.

The patient is placed in the lateral position, and the head is fixed in a Mayfield 3-point head holder. Somatosensory evoked potentials, motor evoked potentials, and facial nerve monitoring are used. Brainstem auditory evoked responses are also used when hearing preservation is attempted.

The incision is located approximately 2 fingerbreadths behind the pinna of the ear. A suboccipital craniectomy is performed, and the edges of the transverse and sigmoid sinuses are exposed. The air cells are carefully waxed to prevent CSF leakage. The dura mater is then opened in a cruciate fashion such that there is a flap superiorly and laterally to allow flat trajectory along the axis of the sigmoid sinus, and the cerebellum is gently retracted to expose the tumor in the CPA. The posterior lip of the IAC is drilled until the transverse crest can be palpated with a hook. The drilling is performed using a diamond drill bit and with copious irrigation to prevent thermal injury. The retrosigmoid approach exposes 180° of the circumference of the IAC. During drilling, extensive irrigation is used to avoid any thermal damage to the nerves in the canal. Air cells at this level can be opened during the drilling and should also be waxed to prevent CSF leakage.

Nuance for Nerve Dissection

The vestibular nerves are divided, and the facial nerve is identified anterior to them and confirmed with stimulation. The cochlear nerve is identified just inferolateral to the facial nerve. Specific techniques for preserving the facial and vestibular nerves when dissecting from the tumor include gentle dissection using fine otology instruments, such as the right-angled hook used in the video. The tumor is dissected from medial to lateral to avoid traction on the cochlear nerve as it exits the perforated bone at the end of the IAC. Careful dissection is performed under direct visualization, and the interface of the nerve with the tumor is dissected under high magnification. Both facial nerve and brainstem auditory evoked response monitoring are used during this dissection.

The senior authors use automated irrigation systems during the removal of VSs to avoid desiccation of the nerves. The labyrinthine artery is carefully preserved, and up to 1 ml of papaverine diluted in 10 ml of saline at room temperature administered locally is used to treat vasospasm. The rationale for using diluted solution is to avoid any potential for cranial nerve injury. The tumor is then dissected off the cerebellum and brainstem and is debulked using the ultrasonic aspirator, which makes the extracapsular dissection proceed more easily. The root entry zones of the facial and acoustic nerves at the brainstem are identified, and the facial nerve is confirmed with stimulation. The tumor is dissected off these 2 nerves very carefully and under direct vision. Once the tumor is removed and hemostasis is obtained, the dura is closed in a watertight fashion by using an AlloDerm patch (if necessary). The bone defect is covered with Medpor cranioplasty (porous polyethylene implant). The muscular layer and the galea are then closed using 2-0 Vicryl interrupted sutures. The skin is closed in a subcuticular fashion by using 4-0 Vicryl.

The Middle Fossa Approach

The middle fossa approach (Fig. 3, Video 3) exposes the IAC and its contents from a superior trajectory.

Video 3. Clip showing the middle fossa approach for resection of a VS. Click here to view with Media Player. Click here to view with Quicktime.

It is chosen for small tumors located primarily within the IAC and offers the possibility of hearing preservation.[6,7] It is an excellent approach for small tumors that predominately occupy the IAC with a minor component in the cistern (usually < 10 mm); it provides exceptional access to the lateral end of the canal.

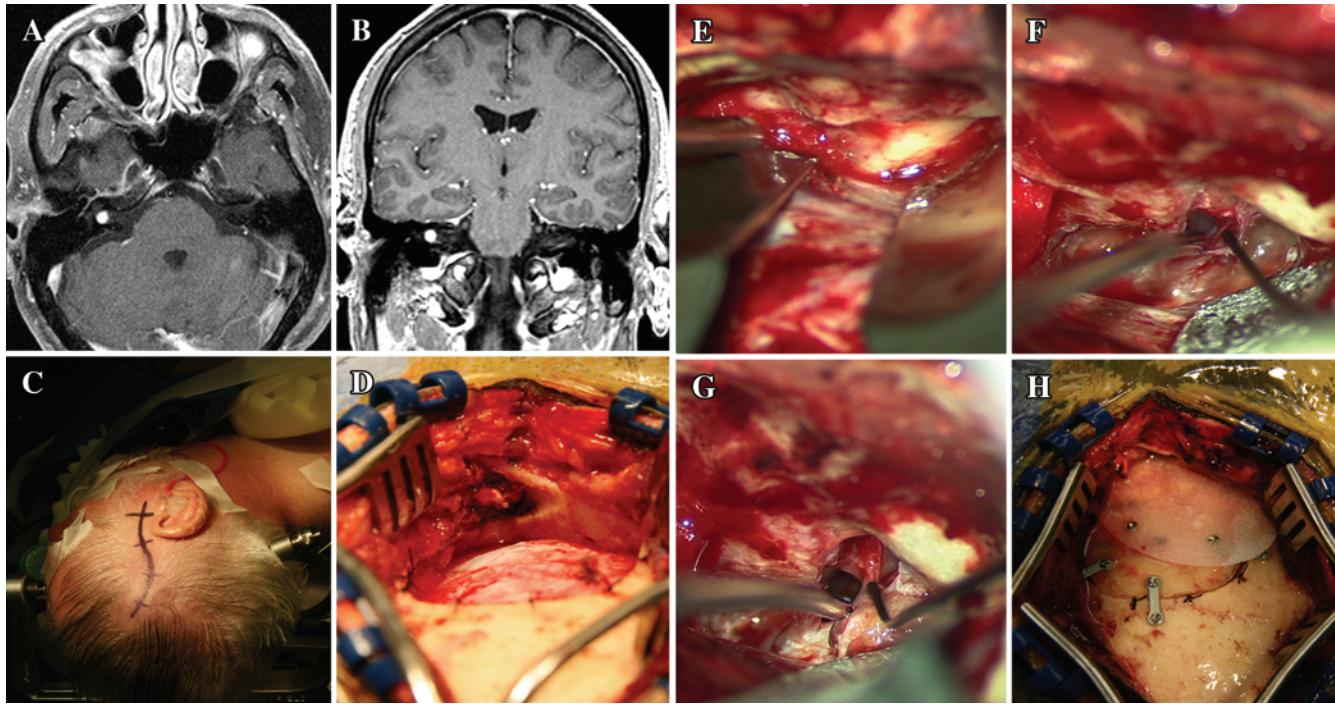

Fig. 3. Axial **(A)** and coronal **(B)** T1-weighted MRI studies of the brain showing a VS occupying the distal aspect of the right IAC of a 60-year-old man who presented with a history of dizziness. **C–H:** Intraoperative photographs obtained during a middle fossa approach to the tumor. **C:** The head of the patient is fixed in the Mayfield 3-point head holder and positioned parallel to the floor. A lazy S-shaped skin incision is made anterior to the tragus; it extends from the zygomatic arch inferiorly to the temporal line superiorly. **D:** The craniotomy is positioned with one-third posterior and two-thirds anterior to the external auditory canal. The craniotomy has to be as low as possible, flush with the cranial base, and additional drilling is performed if the craniotomy flap is not low enough. **E:** The dura is then elevated from posterior to anterior, the edge of the petrous ridge is exposed, and then drilling is performed to expose the IAC. **F:** The dura of the IAC is opened. **G:** The vestibular nerves are divided; the facial nerve is identified anterior to them and confirmed with stimulation. The cochlear nerve is identified inferior to the facial nerve. The tumor is then dissected off the facial and cochlear nerves while preserving all the blood vessels. **H:** The craniotomy flap is then replaced. A Medpor cranioplasty is used to cover the bone defect in case of additional drilling during the opening.

The patient's head is fixed in the Mayfield 3-point head holder and positioned parallel to the floor. Facial nerve monitoring and brainstem auditory evoked responses are used. A lazy S-shaped skin incision is made anterior to the tragus; it extends from the zygomatic arch inferiorly to the temporal line superiorly. The temporalis muscle and fascia are cut vertically and retracted with a self-retaining retractor. The craniotomy is positioned with one-third posterior and two-thirds anterior to the external auditory canal. The craniotomy has to be as low as possible, flush with the cranial base; additional drilling is performed if the craniotomy flap is not low enough. The dura is then elevated from posterior to anterior to prevent injury to the greater superficial petrosal nerve.

After the edge of the petrous ridge is exposed, drilling is performed to identify the superior semicircular canal; this is followed anteriorly to the geniculate ganglion, and then drilling is continued medially to expose the IAC. The basal turn of the cochlea lies very close to the labyrinthine segment of the facial nerve. When dissecting the lateral end of the IAC, it is imperative to be aware of this relationship and keep that dissection very tight to the facial nerve course in the labyrinthine segment. Typically, it is possible to dissect this area with a 2-mm diamond bur, which allows for adequate exposure of the facial nerve but without violating the cochlea.

Once the roof of the IAC is drilled, the dura of the IAC is opened along its posterior aspect along the axis of the IAC, away from the course of the facial nerve. The vestibular nerves are divided; the facial nerve is identified anterior to them and confirmed with stimulation. The cochlear nerve is identified inferior to the facial nerve. The tumor is then dissected off the facial and cochlear nerves while preserving all the blood vessels. The tumor may be located ventral to the facial nerve. In this case, the facial nerve is carefully dissected from the superior aspect of the tumor.

After tumor removal and hemostasis, fat graft is placed into the IAC to prevent CSF leakage. The craniotomy flap is then replaced. A Medpor cranioplasty is used to cover the bone defect if additional drilling is needed during the opening. The rest of the closure is done layer by layer, with closure of the muscle, fascia, galea, and skin. The temporalis muscle and fascia are closed using interrupted 2-0 Vicryl sutures; the galea is closed with 3-0 Vicryl, and the skin is closed in a subcuticular fashion by using 4-0 Vicryl.

Conclusions

The contemporary surgical management of VS is facilitated by familiarity with all 3 approaches described above. Indications for approaches depend on the size of the tumor, its location, the quality of preoperative hearing, and the desire for attempts at hearing preservation. Meticulous microsurgical technique for dissection of the tumor from the adjacent facial and cochlear nerves, electrophysiological adjuncts such as intraoperative cranial nerve monitoring, and careful closure to prevent CSF leakage are critical technical aspects of these surgical approaches.

Disclosure

Dr. Shelton received support from the Cochlear Corporation for a non–study-related clinical or research effort that he oversaw. The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Couldwell. Acquisition of data: Couldwell. Drafting the article: Chamoun. Critically revising the article: Couldwell, MacDonald, Shelton. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Couldwell.

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Video 1: http://mfile.akamai.com/21490/wmv/digitalwbc.download.akamai.com/21492/wm.digitalsource-na-regional/FOCUS12-190_video_1a.asx (Media Player).
http://mfile.akamai.com/21488/mov/digitalwbc.download.akamai.com/21492/qt.digitalsource-global/FOCUS12-190_video_1a.mov (Quicktime).
Video 2: http://mfile.akamai.com/21490/wmv/digitalwbc.download.akamai.com/21492/wm.digitalsource-na-regional/FOCUS12-190_video_2a.asx (Media Player).
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Hearing preservation surgery for vestibular schwannoma: experience with the middle fossa approach

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Object. In the early 1960s William F. House developed the middle fossa approach for the removal of small vestibular schwannomas (VSs) with the preservation of hearing. It is the best approach for tumors that extend laterally to the fundus of the internal auditory canal, although it does have the potential disadvantage of increased facial nerve manipulation, especially for tumors arising from the inferior vestibular nerve. The aim of this study was to monitor the hearing preservation and facial nerve outcomes of this approach.

Methods. A prospective database was constructed, and data were retrospectively reviewed.

Results. Between December 2004 and January 2012, 30 patients with small VSs underwent surgery via a middle fossa approach for hearing preservation. The patients consisted of 13 men and 17 women with a mean age of 46 years. Tumor size ranged from 7 to 19 mm. Gross-total resection was accomplished in 25 of 30 patients. Preoperative hearing was American Academy of Otolaryngology–Head and Neck Surgery (AAO-HNS) Class A in 21 patients, Class B in 5, Class C in 3, and undocumented in 1. Postoperatively, hearing was graded as AAO-HNS Class A in 15 patients, Class B in 7, Class C in 1, Class D in 2, and undocumented in 5. Facial nerve function was House-Brackmann (HB) Grade I in all patients preoperatively. Postoperatively, facial nerve function was HB Grade I in 28 patients, Grade III in 1, and Grade IV in 1. There were 3 complications: CSF leakage in 1 patient, superficial wound infection in 1, and extradural hematoma (asymptomatic) in 1. The overall hearing preservation rate of at least 73% and HB Grade I facial nerve outcome of 93% in this cohort are in keeping with other contemporary reports.

Conclusions. The middle fossa approach for the resection of small VSs with hearing preservation is a viable and relatively safe option. It should be considered among the various options available for the management of small, growing VSs.

(http://thejns.org/doi/abs/10.3171/2012.7.FOCUS12172)

Key Words • **acoustic neuroma** • **middle fossa approach** • **hearing preservation** • **vestibular schwannoma**

In the early 1960s William F. House developed the middle fossa approach. It was originally designed to decompress the auditory nerve in cases of far advanced otosclerosis.[12,13] This procedure was later adapted to VS surgery in 1968.[14] The main indications for the middle fossa approach include the removal of small laterally placed VSs, exposure of the labyrinthine and upper tympanic segments of the facial nerve for decompression, vestibular nerve section, and repair of superior semicircular canal dehiscence. As with all surgical approaches for VS resection, there are both advantages and disadvantages to the middle fossa approach. Advantages include the highest reported hearing preservation rates, a very low incidence of postoperative headache, improved exposure of the lateral IAC as compared with that in the retrosigmoid approach, and the completion of bone removal prior to dural opening. Its disadvantages are limited access to the posterior fossa, a tumor size limitation, and a higher risk of postoperative facial nerve weakness.

Abbreviations used in this paper: AAO-HNS = American Academy of Otolaryngology–Head and Neck Surgery; HB = House-Brackmann; IAC = internal auditory canal; VS = vestibular schwannoma.

Methods

Institutional review board approval was obtained for this study.

Patient Selection

The optimal patient for middle fossa surgery for VS excision and hearing preservation has a tumor that extends 1 cm or less into the cerebellopontine angle, a tumor that involves the distal end of the IAC, hearing loss no greater than 40 dB of pure tone loss with speech discrimination of at least 80% (AAO-HNS Class A and upper Class B hearing[4]), and an age under 65 years since dural elevation becomes more difficult in older patients.

Surgical Techniques and Landmarks

After general endotracheal anesthesia is established, the patient is positioned in a Mayfield headrest with the head turned so that the affected ear is as nearly parallel

to the floor as possible. Monitoring for facial nerve electromyography and auditory brainstem response is set up.

A variety of skin incisions can be used to access the lateral temporal area for craniotomy to expose the middle fossa. We choose a curvilinear incision, which begins just behind the ear and follows along the hairline to create a posteriorly based 5-cm wide scalp flap held with a self-retaining retractor. A temporalis fascia graft is harvested and set aside for later use. An anteriorly based temporalis muscle flap is then elevated off the calvaria and held with a self-retaining retractor. This arrangement of flaps offsets the incision and is helpful in creating a watertight closure at the end of the procedure.

The root of the zygomatic arch is an external landmark for the IAC and should be in the central portion of the exposure. We then create a 4.5 × 4.5–cm craniotomy. Placement of this craniotomy has been described as extending one-third in front and two-thirds behind the external auditory canal. Exact localization of the craniotomy can be improved by using frameless stereotactic navigation. The use of image guidance ensures that the craniotomy is centered directly over the IAC. The temporal squamous bone is then removed using a high-speed drill and rongeurs to create a flush approach to the floor of the middle cranial fossa. The patient is hyperventilated to a PCO_2 of 28 mm Hg, and 0.25 g/kg of mannitol is given. The dura mater is detached from the underside of the calvaria anteriorly and posteriorly, which helps to relax the dura and allows easier elevation of the temporal dura. Under magnification, the dura is elevated off the floor of the temporal fossa beginning posteriorly at the petrous ridge and continuing anteriorly to identify the greater superficial petrosal nerve. The posterior to anterior dissection avoids the risk of dissecting below the greater superficial petrosal nerve and inadvertently avulsing it from the facial hiatus. Venous bleeding is commonly encountered in the region of the foramen spinosum. We avoid coagulation in this location to minimize the risk of both injury to the greater superficial petrosal nerve and interruption of any blood supply to the facial nerve that could arise from the region of the foramen spinosum. Generally, this bleeding is easily managed with gentle tamponading with one of a variety of hemostatic materials. Once the greater superficial petrosal nerve is identified, dural elevation continues to the true petrous ridge. We then place a House-Urban retractor with its edge over the petrous ridge to maintain visualization of the floor of the middle cranial fossa. The superior semicircular canal is positively identified either by direct visualization or by blue-lining of the canal. Typically, the IAC bisects the angle between the course of the greater superficial petrosal nerve and the orientation of the superior semicircular canal.[25] This angle tends to be approximately 60° from the course of the greater superficial petrosal nerve, but the anatomical relationship varies (Fig. 1). Bone anterior and medial to the superior semicircular canal is removed to identify the dura around the porus acousticus. Bone removal continues laterally along the course of the IAC until Bill's bar, or separation of the facial nerve from the vestibular nerve, comes into view. The cochlea lies just anterior to the lateral IAC, and no attempt is made to expose it. The bone between the labyrinthine facial nerve and the cochlea is less than 1 mm thick.

Bone is removed to expose the IAC and extends both anteriorly and posteriorly along the petrous ridge to create at least a 200°–270° exposure of the internal auditory canal (Fig. 2). This exposure allows greater manipulation of the structures of the IAC in a safe way and is particularly relevant for tumors of inferior vestibular nerve origin. Maximized bone removal improves the ability to mobilize the facial nerve anteriorly and allows a better angle of dissection beneath the facial nerve as viewed from a posteroanterior direction. As the bone removal extends laterally, great care is taken to avoid entry into the cochlea anteriorly or into the labyrinth posteriorly.

Once the bone has been removed, exposing the contents of the IAC, the dura is opened sharply posteriorly to avoid injury to the underlying facial nerve (Fig. 3). Cerebrospinal fluid is allowed to egress. A stimulating dissecting probe is liberally used to identify the facial nerve and separate the facial nerve from the superior vestibular nerve (Fig. 2). At this point it is usually evident whether the tumor is arising from the superior or the inferior vestibular nerve. The distal superior vestibular nerve is divided, allowing greater access to the tumor. In the unusual circumstance that the tumor is in the facial nerve, the nerve is decompressed, and no additional tumor is removed to avoid producing facial paralysis.

Depending on the size and consistency of the tumor, internal debulking is performed with the cup forceps and occasionally the fine tip on the ultrasonic aspirator. Dissection of the IAC is generally more difficult on the right side for a right-handed surgeon because of the "height" of the superior semicircular canal. This bony prominence tends to inhibit dissection of the posterior pole of the tumor and visualization below the facial nerve. The tumor is gently dissected from the posterior edge of the facial nerve and from the underside of the facial nerve. The tumor is progressively dissected posteriorly, allowing identification of the cochlear nerve just inferior to the facial nerve. Tumor dissection is generally performed in a medial to lateral direction to avoid avulsing the auditory nerve from the lamina cribrosa laterally. The inferior vestibular nerve is also divided. The tumor is then gently rolled out of the IAC, and its proximal attachments are divided (Fig. 4). Bipolar cautery within the IAC is avoided, and dissection is generally done under constant irrigation to allow optimal visualization. We believe these techniques are crucial to facial nerve and hearing outcomes.

Following tumor resection a small piece of temporalis muscle is placed over the canal to close off the IAC. Any opened air cells are carefully occluded with bone wax. The middle fossa floor is then covered with the temporalis fascial graft, and a small corner of the bone flap is used to create a bony reconstruction of the middle cranial fossa floor. Careful epidural hemostasis is then achieved with both bipolar cautery and gentle tamponading with hemostatic agents. The self-retaining retractor is removed, and the bone plate is replaced in its anatomical position and rigidly fixed. The temporalis muscle is returned to its anatomical position, and the temporalis fascia is carefully approximated to achieve a watertight closure. The

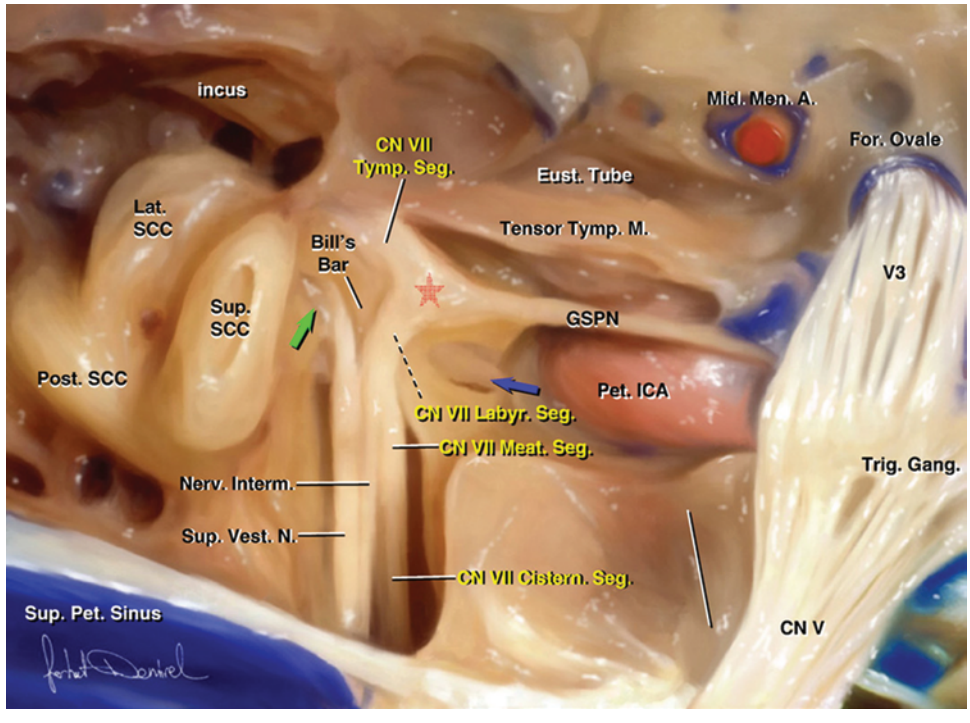

Fig. 1. Anatomical illustration of the floor of the middle cranial fossa. A line drawn along the course of the greater superficial petrosal nerve (GSPN) creates an approximately 120° angle with a similar line drawn along the orientation of the superior semicircular canal (Sup. SCC). A line bisecting this angle approximates the location of the IAC. The *blue arrow* points to the opened cochlea, and the *green arrow* points to the opened vestibule of the labyrinth. The *red star* indicates the geniculate ganglion. Cistern. Seg. = cisternal segment; CN = cranial nerve; Eust. Tube = Eustachian tube; For. Ovale = foramen ovale; Labyr. Seg. = labyrinthine segment; Lat. SCC = lateral semicircular canal; Meat. Seg. = meatal segment; Mid. Men. A. = middle meningeal artery; Nerv. Interm. = nervus intermedius; Pet. ICA = petrous internal carotid artery; Post. SCC = posterior semicircular canal; Sup. Pet. Sinus = superior petrosal sinus; Sup. Vest. N. = superior vestibular nerve; Tensor Tymp. M. = tensor tympani muscle; Tymp. Seg. = tympanic segment; Trig. Gang. = trigeminal ganglion. Reprinted from *Surgical Neurology,* 71/5, Tanriover et al., Middle fossa approach: microsurgical anatomy and surgical technique from the neurosurgical perspective, pp 586–596, 2009, with permission from Elsevier.

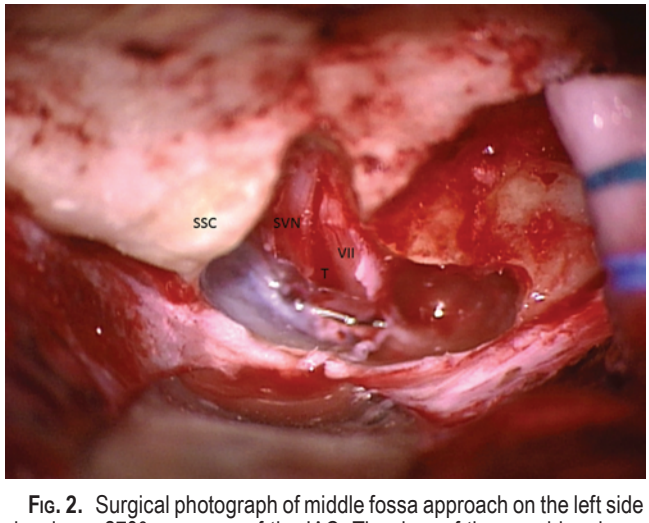

Fig. 2. Surgical photograph of middle fossa approach on the left side showing a 270° exposure of the IAC. The dura of the canal has been opened, and the superior vestibular nerve has been separated from the anteriorly located facial nerve. The tumor is visible between the two nerves and has an inferior vestibular nerve origin. SSC = superior semicircular canal; SVN = superior vestibular nerve; T = tumor; VII = facial nerve.

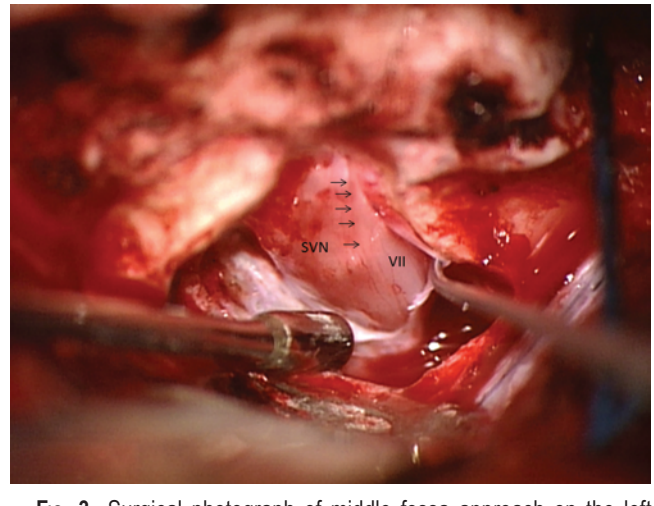

Fig. 3. Surgical photograph of middle fossa approach on the left side, featuring the opening of the dura of the IAC. The cleft between the anteriorly located facial nerve (VII) and the superior vestibular nerve (SVN) is faintly visible *(arrows).* This cleft is developed by dissection with the stimulating dissecting probe.

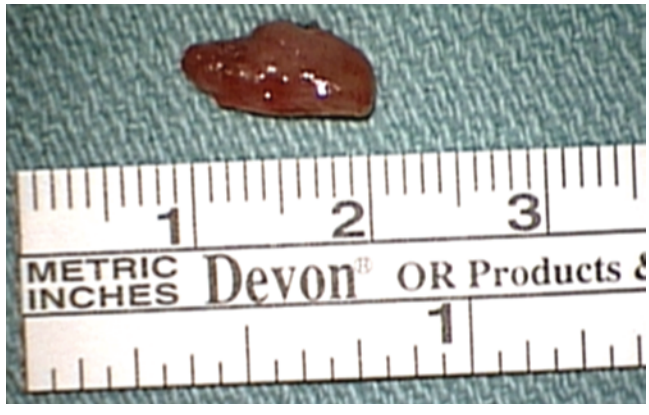

Fig. 4. Photograph of tumor whose consistency and size made en bloc removal technically possible.

scalp is returned to its anatomical position, and the galea aponeurotica is closed. The skin edges are approximated and closed with a nonabsorbable nylon suture. A mastoid dressing is applied.

Results

All patients undergoing a middle fossa approach for VS resection with attempted hearing preservation were included in this study extending from December 2004 to January 2012. Thirty patients, 13 men and 17 women, made up the study cohort. The mean patient age was 46 years (range 21–64 years). The mean tumor size was 12 mm, and the median tumor size was 11 mm (range 7–19 mm). Twenty-five of the 30 patients underwent gross-total resection, as determined by intraoperative observation and postoperative MRI (Fig. 5). Three of 30 patients underwent near-total resection in which microscopically visible tumor was left in place for functional reasons. Note that postoperative MRI may or may not have shown the residual lesion. Two patients underwent subtotal resection, and residual tumor was evident on postoperative MRI. Both of these patients had tumors on the right side with an inferior vestibular nerve origin. Superior displacement of the facial nerve and the constricted anatomy did not allow adequate visualization into the anterior portion of the IAC.

Hearing was scored according to the AAO-HNS classification of hearing (Table 1). Twenty-nine of 30 patients had preoperative audiograms available for review. Twenty-one patients had AAO-HNS Class A hearing, 5 had Class B hearing, and 3 had Class C hearing. Postoperative audiograms were available in 25 of the 30 patients. Postoperative hearing was Class A in 15 patients, Class B in 7, and Class C in 1. Two patients had profound hearing loss. Among the 5 patients without postoperative audiograms, 3 probably had Class B hearing since they could use the telephone on the surgically treated side, although this fact could not be definitively documented. Among the 21 patients with Class A hearing preoperatively, that level of hearing remained in 14; hearing diminished to Class B in 4 patients, all hearing was lost in 1, and no postoperative audiograms were available in 2 (Fig. 6). Of the 5 patients with Class B hearing preoperatively, hearing improved to Class A in 1 patient, remained stable (Class B) in 2 patients, and diminished to Class C in 1 patient; a postoperative audiogram was not available in 1 patient (Fig. 7). Among the 3 patients with preoperative Class C, hearing improved to Class B in 1 patient and was lost in 1 patient; 1 patient did not have an audiogram. Twenty-two of 30 patients had Class A or B hearing postoperatively. This result is consistent with a 73% rate of functional hearing preservation.

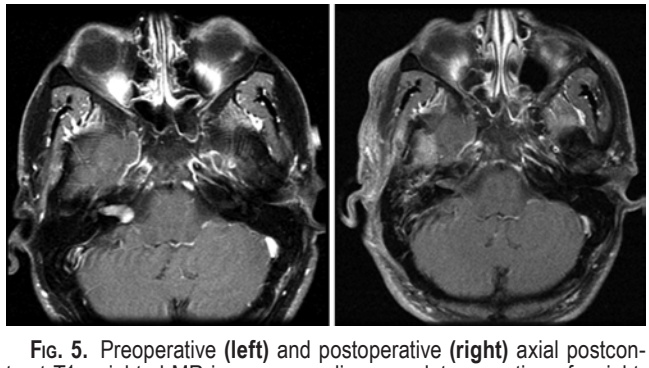

Fig. 5. Preoperative **(left)** and postoperative **(right)** axial postcontrast T1-weighted MR images revealing complete resection of a right-sided VS via the middle fossa approach.

TABLE 1: The AAO-HNS classification of hearing*

Hearing Class	PTA (dB)	SDS (%)
A	≤30	≥70
B	>30 & ≤50	≥50
C	>50	≥50
D	any level	≤50

* "Serviceable hearing" was defined as Class A or B hearing. Abbreviations: PTA = pure tone average; SDS = speech discrimination score.

Of those patients with complete pre- and postoperative data, 11 had Class A or B hearing preoperatively with tumors that were 10 mm or smaller. Nine of these 11 patients remained in hearing Class A or B, and thus an 82% rate of hearing preservation was achieved in this subgroup. Twelve patients with Class A or B hearing preoperatively had tumors larger than 10 mm. All 12 of these patients remained in hearing Class A or B. This finding is consistent with a 100% rate of hearing preservation. There was no significant difference in hearing preservation between groups ($p = 0.22$, Fisher exact test).

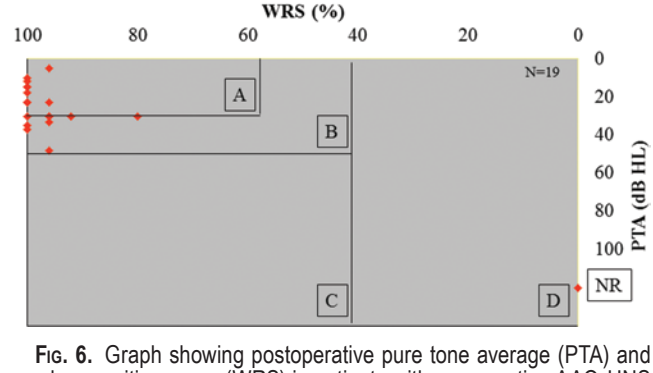

Fig. 6. Graph showing postoperative pure tone average (PTA) and word recognition score (WRS) in patients with preoperative AAO-HNS Class A hearing. The letters A, B, C, and D represent the postoperative AAO-HNS hearing class. HL = hearing level; N = number of patients; NR = no response.

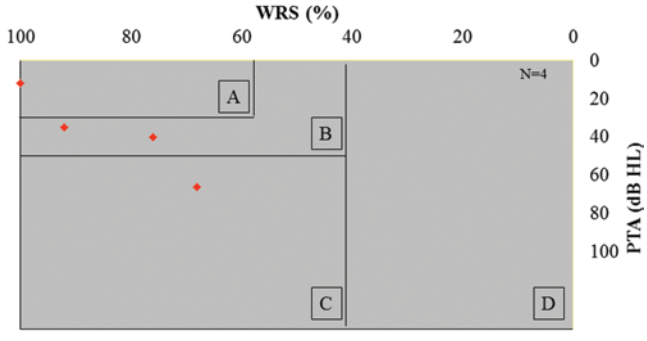

Fig. 7. Graph showing postoperative PTA and WRS in patients with preoperative AAO-HNS Class B hearing. The letters A, B, C, and D represent the postoperative AAO-HNS hearing class.

Facial nerve function was graded using the HB grading scale. All 30 patients had an HB Grade I level of facial function preoperatively. Postoperatively, 28 (93%) of 30 patients maintained an HB Grade I level of facial function. One patient had facial weakness of HB Grade III, and 1 patient with only 1 week of postoperative follow-up had an HB grade of IV.

Three patients suffered postoperative complications. One patient required reoperation for a CSF leak through a highly aerated temporal bone; the defect was repaired directly with complete resolution. One patient had an asymptomatic but significantly sized extradural hematoma, which was surgically evacuated. Unbeknownst to the surgical team, this patient was a chronic aspirin user. There was 1 superficial wound infection, which resolved with oral antibiotics.

Discussion

Several options exist for the management of small VSs in patients with serviceable hearing. Interestingly, in a recent report, Di Maio et al.[6] could not identify a difference in quality of life among the therapeutic options of observation, radiation, or surgery. A commonly used management strategy is careful interval clinical and MRI evaluation without intervention, or the "wait-and-scan" option. In a natural history study from France, Bakkouri et al.[2] identified a growth rate of < 1 mm/year in 58.6% of patients. Only 12.2% of patients had a growth rate of > 3 mm/year. Over time, however, many patients will suffer a deterioration in hearing.[23] Good high-frequency hearing and speech discrimination at diagnosis are positive predictors of good hearing after several years of observation, although about 15% of patients will have worsened hearing after a year of observation.[23] Radiosurgery has assumed a prominent role in the management of patients with small VSs. The short-term tumor growth control rates are in excess of 95%, and long-term control remains in over 85% of patients. Short-term hearing preservation rates are also quite good and range from 60% to 75%;[5,7,18,27] however, long-term results reveal a marked tendency to deteriorate over time. Hearing preservation rates at 5 years postradiosurgery range from 43% to 57%, whereas those at 10 years range from 34% to 45%.[3,9,20] The middle fossa approach for VSs with attempted hearing preservation has been described by several groups over the years. Data from recent publications reveal the maintenance of postoperative serviceable hearing in 37%–77% of patients (Table 2).[1,15–17,19,22,24] These rates are typically superior to hearing preservation outcomes reported for the retrosigmoid approach.[10,21] In contrast to rates in radiosurgical series, the long-term hearing preservation rates following surgical tumor removal typically remain stable over time.[8,11,24] Friedman et al.[8] reported hearing preservation rates of 70% more than 5 years after surgery, and Hilton et al.[11] reported a 10-year hearing preservation rate of 72%. Ninety-six percent of patients in the study by Woodson et al.[26] maintained their immediate postoperative hearing levels with > 5 years of follow-up. Thus, despite the inherent variability and selection biases among treatment groups, it seems that microsurgical removal of VSs results in better long-term hearing preservation rates than radiosurgery.

A disadvantage of the middle fossa approach is the need for facial nerve manipulation. Nonetheless, most centers report postoperative HB Grade I and II facial function outcomes in 89%–100% of patients. Facial nerve outcomes have typically been superior in patients undergoing a retrosigmoid approach, with functional outcomes of HB Grades I and II of 90%–100% (Table 3). We chose to report only

TABLE 2: Published hearing preservation rates

Authors & Year	Serviceable Hearing Maintained (% of patients)*
Satar et al., 2002	53
Friedman et al., 2003	61
Arts et al., 2006	73
Meyer et al., 2006	56
Jacob et al., 2007	37
Phillips et al., 2010	71
Hillman et al., 2010	59
Sameshima et al., 2010	77
Kutz et al., 2012	63
current study	73

* "Serviceable hearing" was defined as PTA ≤ 50 dB and SDS ≥ 50%, or AAO-HNS Class A or B.

TABLE 3: Published facial nerve outcomes

Authors & Year	HB Grade I–II (% of patients)
Satar et al., 2002	90.8
Friedman et al., 2003	94
Arts et al., 2006	96
Meyer et al., 2006	97
Jacob et al., 2007	94
Hillman et al., 2010	88
Sameshima et al., 2010	100
Kutz et al., 2012	89
current study	93*

* House-Brackmann Grade I only.

those patients with HB Grade I function, and this level of facial nerve function was achieved in 93% of patients.

Conclusions

The middle fossa approach for the resection of VSs with hearing preservation is a viable alternative to both a retrosigmoid approach and stereotactic radiosurgery. Hearing preservation rates in excess of 70% can be achieved in well-selected patients. Facial nerve outcomes are typically good despite the need for nerve manipulation.

Disclosure

Dr. DeMonte is a yearly lecturer for a Medtronic skull base course.

Author contributions to the study and manuscript preparation include the following. Conception and design: both authors. Acquisition of data: both authors. Analysis and interpretation of data: both authors. Drafting the article: DeMonte. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors. Approved the final version of the manuscript on behalf of both authors: DeMonte. Statistical analysis: DeMonte. Study supervision: both authors.

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A stepwise illustration of the translabyrinthine approach to a large cystic vestibular schwannoma

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Of the presigmoid approaches, the translabyrinthine approach is often used when a large exposure is needed to gain access to the cerebellopontine angle but when hearing preservation is not a concern. At the authors' institution, this approach is done with the aid of ENT/otolaryngology for temporal bone drilling and exposure.

In the present article and video, the authors demonstrate the use of the translabyrinthine approach for resection of a large cystic vestibular schwannoma, delineating the steps of positioning, opening, temporal bone drilling, tumor resection, and closure. Gross-total resection was achieved in the featured case. The patient's postoperative facial function was House-Brackmann Grade II on the side ipsilateral to the tumor, although function improved with time.

The translabyrinthine route to the cerebellopontine angle is an excellent approach for masses that extend toward the midline or anterior to the pons. Although hearing is sacrificed, facial nerve function is generally spared.
(http://thejns.org/doi/abs/10.3171/2012.7.FOCUS12208)

Key Words • **translabyrinthine approach** • **vestibular schwannoma** • **surgical technique** • **video**

Although the translabyrinthine approach was described by Panse in 1904 and first used to resect a cerebellopontine angle tumor by Quix in 1912, it was not until House reported on 47 resections with no deaths in 1964 that the approach was truly popularized.[1] Since that time it has been well described in the literature as a useful approach for resecting vestibular schwannomas in cases in which hearing preservation is not a concern. Additionally, a modified use of this approach has been described in combination with a transtentorial component for the resection of vestibular schwannomas and other lesions of the cerebellopontine angle and proximate anatomy.[4]

Surgical series of translabyrinthine resections often include meningiomas of the cerebellopontine angle and the internal acoustic meatus, schwannomas of the facial and trigeminal nerves, and cholesteatomas, neurinomas, and chordomas, illustrating the multiple uses of this approach.[3] In the present report, the translabyrinthine approach for resection of a large cystic vestibular schwannoma (Fig. 1) is described in a stepwise manner using text, a narrated video (Video 1), and still images obtained in a single patient.

Abbreviation used in this paper: IAC = internal auditory canal.

Video 1. Narrated video describing the steps of the translabyrinthine approach for vestibular schwannomas from positioning and opening through closure. In this example, the tumor was left-sided, large, and cystic. Click here to view with Media Player. Click here to view with Quicktime.

Surgical Indications

Although case series exist describing hearing preservation in the setting of partial labyrinthectomy, the main indication for the approach remains resection of vestibular schwannomas in patients in whom preoperative hearing is absent or nonserviceable.[2] When hearing preservation is a goal of surgery, or when a smaller lesion is being resected, a retrosigmoid craniotomy may be appropriate. In the case of the largest of vestibular schwannomas, a combined translabyrinthine-transtentorial approach may be indicated.[4]

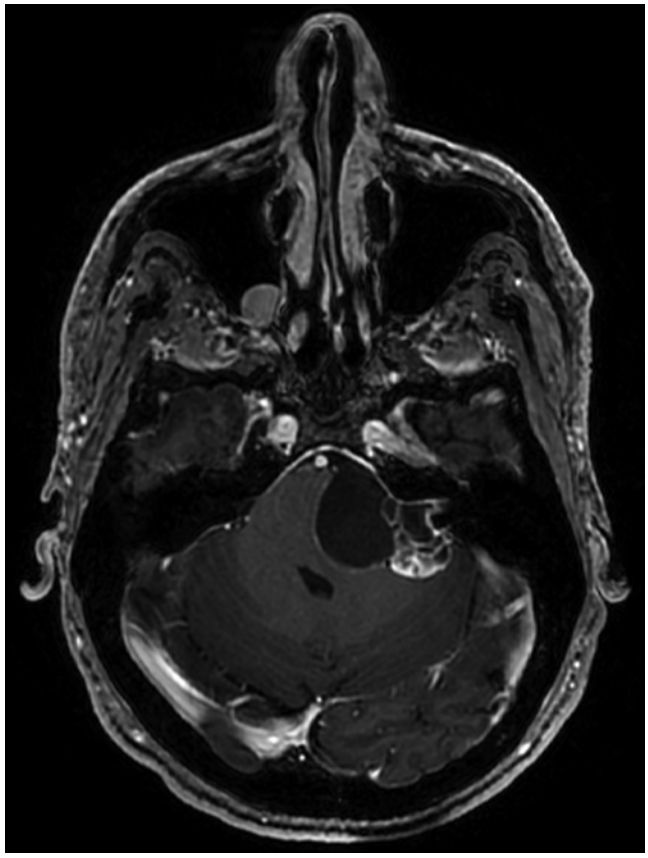

Fig. 1. Preoperative axial contrast-enhanced T1-weighted MR image obtained at the level of the IAC and demonstrating a large left cystic vestibular schwannoma.

Positioning and Incision

The patient is positioned supine with the head turned contralateral to the side of the lesion. This must be done to the point that the zygomatic arch is almost parallel to the floor. If the patient's neck is inflexible, a shoulder roll may be required for positioning. The chin must also be assessed, and should be 2 fingerbreadths away from the sternum; flexion should not be severe enough to compromise venous return from the head. Distance should be kept between the planned incision and the patient's ipsilateral shoulder, which will otherwise limit the surgeon's range of motion later in the procedure.

The head is placed in pins in the Mayfield skull clamp to which the navigational frame is anchored. The neuronavigation system is registered and used for incision planning and, later, to aid in tumor resection. Facial electromyography needles are placed for seventh cranial nerve monitoring to be performed later in the procedure. At our institution, the otolaryngology team prefers to position the patient facing away from the anesthesiologist, with the surgeon sitting between the patient and the anesthesiologist.

A C-shaped incision is planned behind the ear, as needed, to expose the mastoid tip, sigmoid sinus, and transverse-sigmoid junction, and to be as high as the floor of the middle cranial fossa (Fig. 2). The floor of the middle fossa is marked by the level of the zygomatic arch,

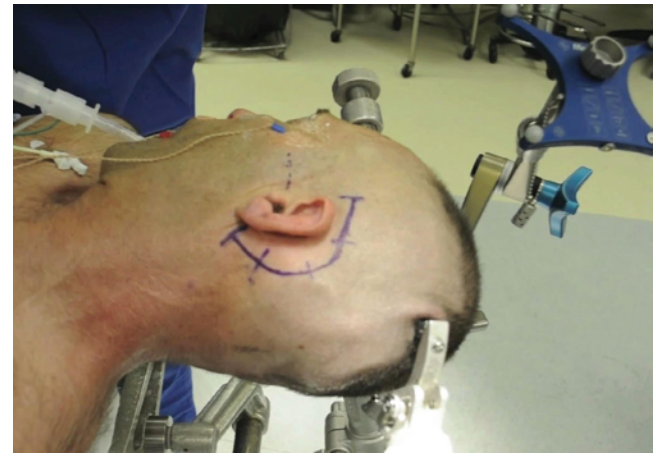

Fig. 2. Photograph depicting the patient's head positioned and secured in pins, with electromyography needles in place and a navigational frame attached. The zygomatic arch and planned incision are marked.

but neuronavigation can assist in determining the position as well as the location of the junction of the transverse and sigmoid sinuses. As a final check before prepping and draping, the table should be air-planed toward and away from the anesthesiologist. This ensures that the patient will be stable later in the procedure when the table will need to be manipulated to facilitate dissection distally along the seventh cranial nerve and in the internal acoustic meatus. When prepping, one must keep in mind that an abdominal field and potentially a lateral thigh field should be prepared for fat graft harvest and potential fascial harvest.

Opening and Approach

The skin is divided sharply and electrocautery is used to divide the muscle, leaving the periosteum intact for the time being. The periosteal layer will be used as an additional layer and protection against CSF leakage at the time of closure. Skin edges are undermined anteriorly and posteriorly, both to benefit exposure and to enable the surgeon to harvest a fascial graft from the temporalis muscle. In the event that this is not possible, a fascial graft can be harvested either from the rectus muscle when the abdominal fat graft is taken or from the tensor fascia lata.

The periosteum is then elevated. As emissary veins are encountered, the periosteum around them should be cleared completely from the bone, and the veins waxed. As the periosteum is elevated, the limits of the exposure are as follows: the mastoid tip inferiorly, 1–2 cm behind the sigmoid sinus posteriorly, and the spine of Henle anteriorly. Once this is accomplished, mastoid drilling may begin.

At the our institution, drilling starts with a large cutting bur, as well as the suction irrigator. A standard mastoidectomy begins with skeletonization of the tegmen, sigmoid sinus, and posterior border of the external auditory canal. The drill is then used to enter the mastoid antrum and identify the incus and the lateral semicircular canal. A diamond bit can then be used to remove the remainder

of bone off of the sigmoid sinus and 1–2 cm of the posterior fossa dura. The tegmen is removed to expose middle fossa dura, and the presigmoid dura is also exposed by finishing the bony removal there (the endolymphatic duct will likely be transected during this process). This boney removal is accomplished using a small rongeur once the drill has sufficiently thinned the bone. The rongeur will also be used at this point to remove the petrous ridge over the superior petrosal sinus heading medially (Fig. 3).

At this time, further drilling with the diamond bit allows the surgeon to identify the vertical segment of the facial nerve in the proximal mastoid. This is followed toward the stylomastoid foramen, leaving a thin shell of bone over the nerve in the fallopian canal. The labyrinthectomy is begun by entering the lumen of the lateral semicircular canal and then the posterior semicircular canal. The vestibule is exposed and the lateral semicircular canal is removed. The subarcuate artery will often be encountered at this point in the foramen of the superior semicircular canal.

The sigmoid sinus can now be traced toward the jugular bulb, which is defined with the drill. Variations in this anatomy may limit exposure in the presence of a high-riding jugular bulb, which can be directly inferior to the IAC in some patients. This limits the trough that will later be drilled below the IAC, because the inferior limit of this trough is the jugular bulb. Once the jugular bulb is identified, drilling can commence to uncover the dura of the IAC. The troughs are then drilled superior and inferior to the IAC with the goal being to expose the IAC 270° around its circumference. The superior trough is delimited by the superior edge of the IAC and the middle fossa dura. At this stage, the only bone on the IAC dura should be along its anterior aspect (Fig. 4).

To identify the superior vestibular nerve, which sits superior to the falciform crest, the falciform crest should now be located. This is the nerve from which most vestibular schwannomas arise, and it can be transected while causing little morbidity. Most frequently its transection results in transient vertiginous symptoms. The dura of the IAC is then opened, proceeding medially, and a plane can be developed between the tumor and the facial nerve. The dura is opened further laterally and then inferiorly and superiorly along the presigmoid posterior fossa dura.

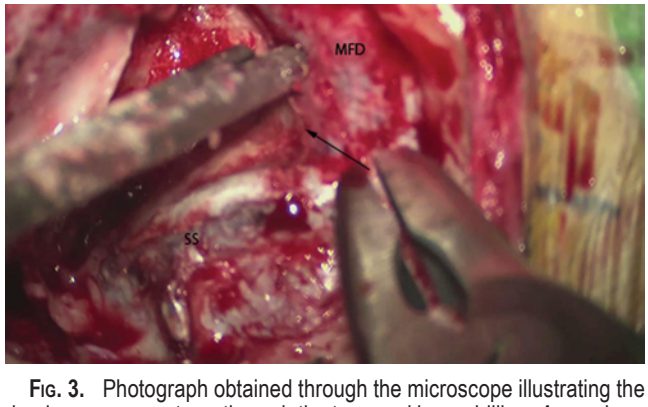

Fig. 3. Photograph obtained through the microscope illustrating the dural exposure partway through the temporal bone drilling. *Arrow* demonstrates the direction of the superior petrosal sinus along the petrous ridge. MFD = middle fossa dura; SS = sigmoid sinus.

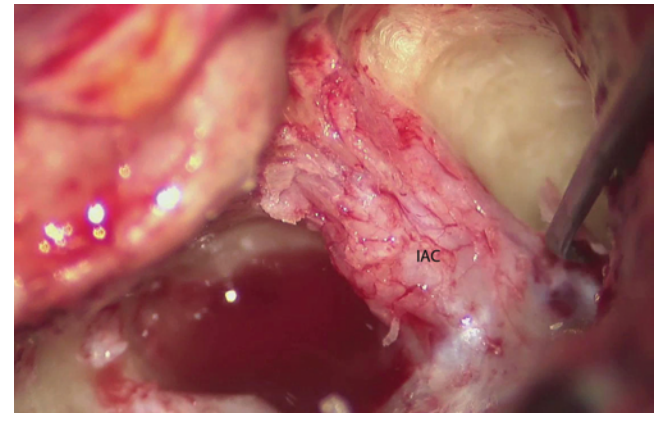

Fig. 4. Photograph obtained through the microscope showing the dura of the IAC exposed with troughs drilled superiorly and inferiorly.

This is best done using a microsurgical blade followed by scissors.

Tumor Removal

While the first portion of the tumor in the IAC may be dissected partially off of the facial nerve, the majority of this portion of the dissection is left for the end of the resection. It is the cisternal portion of the tumor that actually is resected first. Once enough general dissection has been achieved to visualize the tumor, the lesion is debulked internally to improve its mobility. This process allows the surgeon to dissect the mass partially away from the brainstem and begin attempting to identify cranial nerves. The goal is ultimately to achieve circumferential dissection around the tumor. To debulk the tumor internally, its surface is coagulated and cut with scissors. This allows the surgeon access to the center of the tumor, which can be resected with pituitary rongeurs or an ultrasonic aspirator. If the course of the facial nerve has not already been established, the surface of the tumor must be stimulated to determine that the facial nerve does not have twigs running through the area that is about to be coagulated and cut. The facial nerve most commonly runs along the anterior surface of the tumor and is also commonly found along the anterosuperior or anteroinferior surface. Although the surface of the tumor is coagulated with the bipolar electrocautery, care is taken not to use electrocautery near the brainstem or any cranial nerves.

In the standard translabyrinthine approach, in which hearing is sacrificed, the eighth cranial nerve can be identified at the brainstem, coagulated, and transected (Fig. 5). The nerve is first stimulated to verify that it is not the facial nerve. In cases in which the facial nerve has been damaged by the tumor, the surgeon must establish a threshold for stimulation of the facial nerve before concluding that a nerve does not stimulate, and therefore must not be, the facial nerve. One should note that the cochlear nerve in the IAC is not removed at this time. Instead it is left for the end of the resection, so that the facial nerve is given some stability to handle the dissection of the adherent tumor. Thus, the cochlear nerve in the IAC is taken only as the last portions of the tumor are resected (Fig. 6).

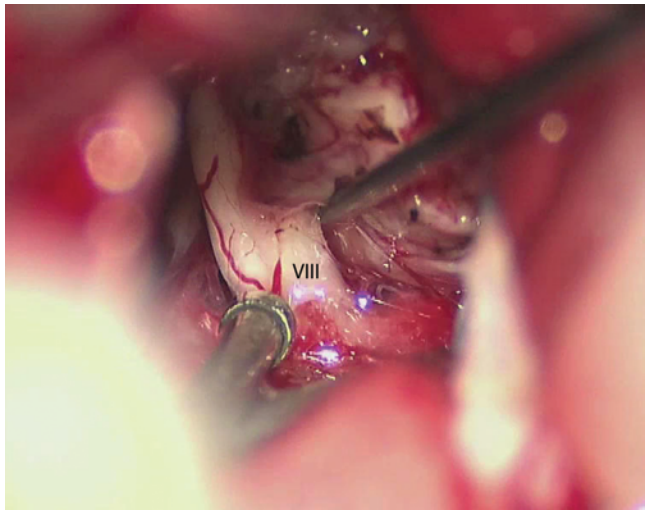

Fig. 5. Photograph obtained through the microscope. Cranial nerve VIII is identified along the posterior edge of the tumor at its entry zone along the brainstem.

Transection of the eighth cranial nerve allows for further dissection of the mass off of the brainstem and therefore further debulking of the mass. As this back-and-forth process of debulking and dissection continues, the tumor will eventually be a husk that has been separated from the brainstem. At this time, the shell of the tumor is resected with curved microscissors, leaving only the outer portion of the tumor, which is adherent to the facial nerve. This last fragment is dissected from the facial nerve using sharp dissection techniques and the diamond and arachnoid knives.

In large masses, the anatomy is quite distorted and a systematic approach to identification of landmarks is needed. A helpful strategy in this case is first to identify the inferior pole of the tumor. From this point, cranial nerves can be identified as the tumor is rolled from inferior to superior, starting with cranial nerves IX, X, and XI. From here, the flocculus is identified along with choroid plexus in the foramen of Luschka. The surgeon should find the eighth and seventh cranial nerves just ventral to it. The anterior inferior cerebellar artery should have a loop between these 2 nerves at the brainstem. Whichever strategy the surgeon takes, the arteries and veins of the posterior fossa should be respected, and every effort should be made not to take the superior petrosal veins.

Closure

Once the tumor resection is completed and hemostasis has been achieved, the wound is packed closed. The dura that has been opened cannot be closed primarily, and a fat graft harvested from the abdomen is placed. One large piece of fat may be harvested, but strips will be cut and inserted into the surgical cavity to the level of the dura. It is important not to allow these fat strips to advance past the level of the dura into the cerebellopontine angle or to pack them too aggressively because cranial nerve deficits may result. A fascial graft, as mentioned, may be obtained from the temporalis muscle during the opening.

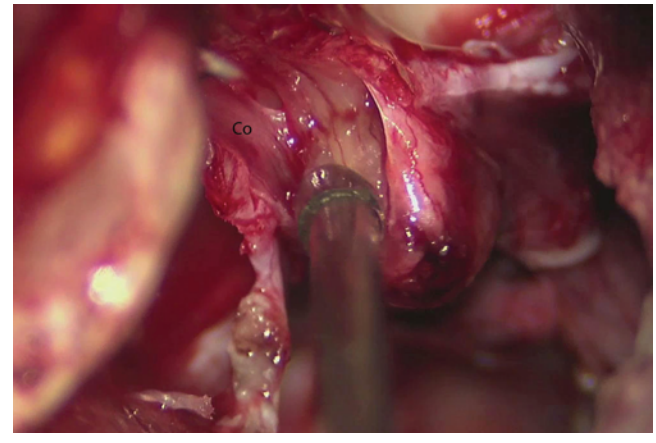

Fig. 6. Photograph obtained through the microscope revealing a small remnant of tumor adherent to the facial nerve, which is not well visualized. At this stage in the resection, the cochlear nerve (Co) is about to be divided in the IAC.

Alternatively, it may be harvested from the rectus muscle via the fat graft incision or from the tensor fascia lata of the thigh. This fascial graft is simply draped over the mastoid antrum, mastoid and tympanic facial nerve, and all air cells of the facial recess and zygomatic groove. At this point in the procedure, synthetic fibrin glue or other suitable epoxy can be interspersed among the fat strips and fascial graft to guard against CSF leakage.

The periosteum, muscle, and skin are closed in layers. The skin is often closed with a running locked stitch; in redo cases or those in which poor wound healing is a concern due to radiation or other causes, interrupted vertical mattress sutures are used. A mastoid dressing is then applied and is left in place for several days.

Postoperative Management

If there is evidence of hydrocephalus or if there is some other reason to be concerned about wound healing, a lumbar drain is placed at the outset of surgery, although this is uncommon. The patient is admitted to the neurosurgical ICU, and drainage is undertaken hourly. At the authors' institution, drainage is typically begun at a rate of 5 ml/hour and increased as long as the patient does not suffer intolerable headaches and nausea. It is often increased to a rate of 15 ml/hour, and occasionally higher. The duration of drainage depends on the surgeon's level of concern for CSF leak. Usually the lumbar drain can be clamped for 24 hours at the conclusion of the healing process to ensure that no CSF leak is present, and drainage is then discontinued.

In the majority of cases, however, no such CSF drainage is required. The patients spend their first postoperative night in the ICU and undergo MRI on postoperative Day 1 (Fig. 7). After this, and barring any specific concerns, the patient can be transferred to the floor. Although particular attention is paid to the posterior fossa on the MRI, these patients will often have a small filling defect in the sigmoid sinus, representing a clot. The clot is generally treated by keeping the patient well hydrated; specific anticoagulation therapy is not performed. As long as

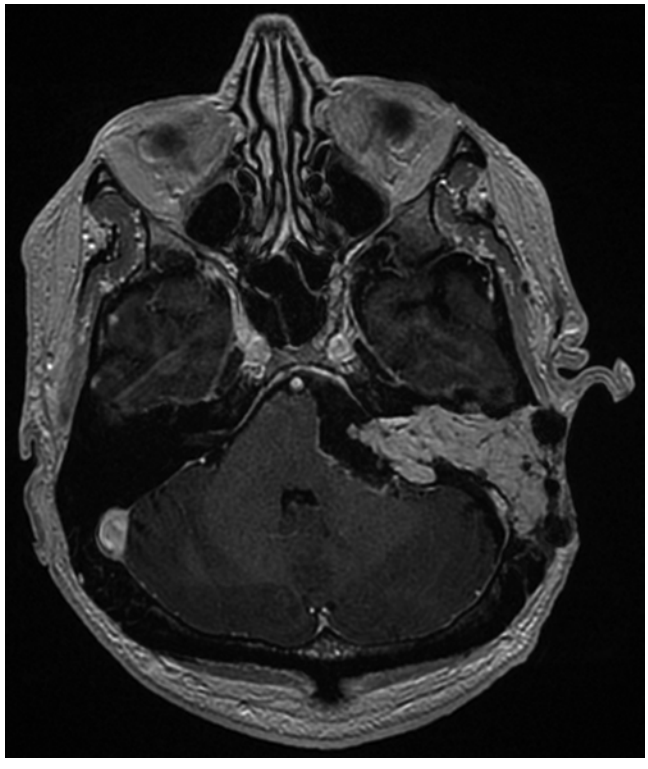

Fig. 7. Postoperative axial contrast-enhanced T1-weighted MR image demonstrating the extent of resection of the tumor and fat strips placed in the surgical cavity to the level of the dural defect.

there is no significant edema, the patient can be weaned quickly from Decadron. Other postoperative care is not specific to this procedure.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Acquisition of data: Nickele, Akture. Analysis and interpretation of data: Nickele, Akture, Gubbels. Drafting the article: Nickele, Gubbels. Critically revising the article: Başkaya, Nickele, Gubbels. Reviewed submitted version of manuscript: Nickele. Administrative/technical/material support: Nickele. Study supervision: Başkaya.

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Do cystic vestibular schwannomas have worse surgical outcomes? Systematic analysis of the literature

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Object. The goal of this study was to perform a systematic quantitative comparison of the surgical outcomes between cystic vestibular schwannomas (CVSs) and solid vestibular schwannomas (SVSs).

Methods. A review of English-language literature published between 1990 and 2011 was performed using various search engines including PubMed, Google Scholar, and the Cochrane database. Only studies that reported surgical results of CVSs in comparison with SVSs were included in the analysis. The primary end point of this study was surgical outcomes, defined by the following: 1) facial nerve outcomes at latest follow-up; 2) mortality rates; or 3) non–facial nerve complication index. Secondary end points included extent of resection and brainstem adherence.

Results. Nine studies comprising 428 CVSs and 1287 SVSs were included in the study. The mean age of patients undergoing surgery was 48.3 ± 6.75 and 47.1 ± 9 years for CVSs and SVSs, respectively ($p = 0.8$). The mean tumor diameter for CVSs was 3.9 ± 0.84 cm and that for SVSs was 3.7 ± 1.2 cm ($p = 0.7$). There was no significant difference in the extent of resection among CVSs and SVSs (81.2% vs 80.7%, $p = 0.87$) Facial nerve outcomes were significantly better in the cohort of patients with SVSs than in those with CVSs (52.1% vs 39%, $p = 0.0001$). The perioperative mortality rates for CVSs and SVSs were not significantly different (3% and 3.8%, respectively; $p = 0.6$). No significant difference was noted between the cumulative non–facial nerve complication rate (including mortality) among patients with CVSs and SVSs (24.5% and 25.6%, respectively; $p = 0.75$)

Conclusions. Facial nerve outcomes are worse in patients undergoing resection for CVSs than in patients undergoing resection for SVSs. There were no significant differences in the extent of resection or postoperative morbidity and mortality rates between the cohorts of patients with vestibular schwannomas.
(http://thejns.org/doi/abs/10.3171/2012.6.FOCUS12200)

Key Words • **cystic vestibular schwannoma** • **acoustic neuroma** • **solid vestibular schwannoma**

Vestibular schwannomas are the most common tumors occupying the cerebellopontine angle.[7,19] Based on their consistency, they can be broadly categorized into SVSs (homogeneous or heterogeneous) and CVSs.[13,25]

It has been generally stated that CVSs are more aggressive, unpredictable variants of VSs, and patients present to their physicians after shorter symptomatic periods.[4,9,13,25] Due to the improved understanding of CVSs, it was recommended in a 2003 consensus meeting that a VS with a cystic component (especially multicystic VSs and those with the cyst on the surface) should be categorized as a different group within VSs and should be appropriately analyzed and reported separately.[13]

It is widely believed that CVSs have less favorable surgical outcomes than SVSs with regard to facial nerve outcomes, surgery-related complications, and mortality.[1,5,6,26,29,32] On the contrary, recently there have been reports that have shown no significant differences in the surgical outcomes among the 2 cohorts of VSs.[9,10,25] To our knowledge, currently there is no systematic review in

Abbreviations used in this paper: CVS = cystic vestibular schwannoma; GTR = gross-total resection; STR = subtotal resection; SVS = solid vestibular schwannoma; VS = vestibular schwannoma.

the literature comparing the surgical outcomes between CVS and SVS. To address this question and summarize the evidence, we performed a quantitative analysis of the literature to elucidate if there was any difference in the surgical outcome among CVSs and SVSs. Additionally, this article will provide a brief synopsis of the literature to highlight the possible pathophysiology behind the development of the cystic component in a VS and operative considerations pertinent to CVS.

Methods

A review of the English-language literature published between 1990 and 2011 was done using various search engines including PubMed, Google Scholar, and the Cochrane database. Initially, the articles were identified using a combination of the following key words: "cystic vestibular schwannoma," "solid vestibular schwannoma," "vestibular schwannoma," "acoustic neuroma," "cyst," "facial nerve," and "surgical outcome."

The abstracts of the various selected studies were screened, and only those studies that reported the surgical results of CVS in comparison with SVS were included in our analysis. The studies examining only 1 of the cohorts were excluded from the analysis.[12,33] Furthermore, references from the selected studies were also screened to identify any missed studies.

The primary end point in our study was surgical outcomes defined by the following: 1) facial nerve outcomes at latest follow-up; 2) mortality rates; or 3) non–facial nerve complication index. Secondary end points included the extent of resection and brainstem adherence. As with most tumor studies, patient age and tumor size are important in determining surgical outcome. Thus, whenever possible, this pertinent information was noted, and a statistical comparison was performed to see if the difference in tumor size and age was significant between the cohorts.

Relative rates of facial nerve outcomes at the last follow-up were compared. Good facial nerve outcome was defined as House-Brackmann Grade I or II. The studies that reported good facial nerve outcomes as Grades I–III were excluded from the main analysis; their results were analyzed and reported separately.[1,29]

Since there was a possibility that a patient could have more than 1 type of non-audiofacial complication, we analyze the non-audiofacial complications, such as CSF leak, lower cranial nerve deficits, and stroke, as the total cumulative complication percentage per cohort (non-audiofacial complication index).

Statistical Analysis

The Fisher exact test was used to analyze the binary variables, while an independent sample t-test was used to determine the statistical difference among the continuous variables. The result was considered statistically significant at $p < 0.05$.

Results

After the initial screening, 35 unduplicated studies were reviewed, from which 10 studies comparing the results of CVS versus SVS were extracted. Two studies comparing the results of fractionated stereotactic radiotherapy between the groups were excluded from the analysis of the surgical outcomes.[24,28]

Overall, 9 studies comprising 428 CVSs and 1287 SVSs were included in our review (Table 1). The mean age of patients undergoing surgery for CVSs and SVSs was 48.3 ± 6.75 and 47.1 ± 9 years, respectively; this difference was not significant ($p = 0.8$). The mean tumor diameter of the CVSs was 3.9 ± 0.84 cm and that of the SVSs was 3.7 ± 1.2 cm; this difference was also not significant ($p = 0.7$). The 2 studies that did not report their mean tumor sizes did mention in their methods section that the patients were recruited only after matching the tumor sizes.[6,10]

Extent of Resection

To analyze the extent of resection, data were available for 341 patients with CVSs and 1210 with SVSs (Table 2). The extent of resection was classified into GTR and STR. Authors of 1 study divided their results into 3 categories that included GTR, near-total resection, and STR.[9] For this paper, we included the patients undergoing near-total resection in the STR group. There was no significant difference with regard to the extent of resection among surgery for CVSs and SVSs (81.2% and 80.7%, respectively; $p = 0.87$) (Fig. 1).

Facial Nerve Preservation Rate

For analysis of the facial nerve function, good outcome was defined as House-Brackmann Grade I or II at the latest follow-up (Table 3). A total of 302 patients with CVSs and 959 patients with SVSs were analyzed. Patients from 2 studies were excluded from this analysis (analyzed separately) since they reported good outcome as House-Brackmann Grades I–III, and no detailed table was available from which information pertaining to House-Brackmann Grade I or II could be extracted. Facial nerve outcomes at the last follow-up were significantly better in the cohort of patients with SVSs than for those with CVSs (52.1% and 39%, respectively; $p = 0.0001$) (Fig. 2). Facial nerve preservation rates in the studies that reported good outcomes as House-Brackmann Grades I–III were also significantly better for patients with SVSs than for those with CVSs (78.3% and 65.2%, respectively; $p = 0.03$).[1,29]

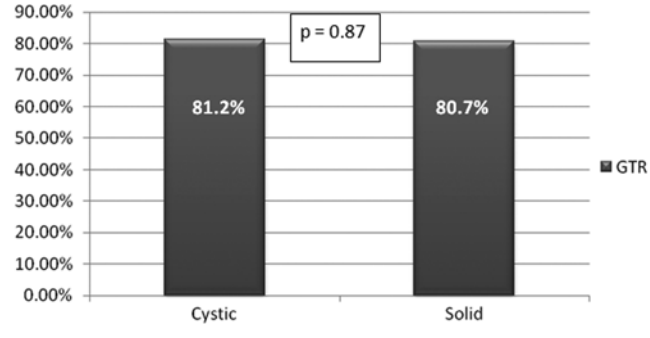

Fig. 1. Bar graph showing a comparison of the extent of resection between CVSs and SVSs.

TABLE 1: Study type and basic demographic details of the available comparative studies in the literature*

Authors & Year	Level of Evidence	Study Setting	No. of Patients		Incidence of CVS (%)	Mean Age (yrs)		Tumor Size (cm)		Tumor Size Matched	Mean Duration of Symptoms	
			CVS	SVS		CVS	SVS	CVS	SVS		CVS	SVS
Jian et al., 2011	II	prospective comparative	58	410	12	52	49	2.7	1.7	no	NR	NR
Piccirillo et al., 2009	III	retrospective comparative	96	96	6.7	51	49	2.8	2.7	yes	NR	NR
Mehrotra et al., 2008	III	retrospective comparative	22	40	21.2	42.1	34.5	4.7	4.6	yes	26.2 mos	21.1 mos
Sinha & Sharma, 2008	III	retrospective comparative	58	226	20.4	39.5	NR	mean NR	mean NR	no	16 mos	22.9 mos
Jones et al., 2007	III	retrospective comparative (prospective database)	77	77	9.6	matched but NR	matched but NR	matched but NR	matched but NR	yes	NR	NR
Moon et al., 2007	III	retrospective comparative	24	82	22.6	40.5	NR	4.38/3.42	3.42	no	14 mos	26.1 mos
Benech et al., 2005	III	retrospective comparative	26	54	20	58	56	4.5	4.8	yes	shorter in CVS ($p < 0.001$)	
Fundová et al., 2000	III	retrospective comparative	44	151	5.7	52.7	NR	3.9	NR	yes	4.2 yrs	NR
Charabi et al., 1994[5]	II	prospective comparative	23	151	4	51.2	NR	4.5	4.98	yes	no difference ($p > 0.05$)	

* NR = not reported.

TABLE 2: Extent of resection of the VSs*

Cohort	Extent of Resection		
	GTR (%)	STR	Total
CVS	277 (81.2)	64	341
SVS	977 (80.7)	233	1210
total	1254	297	1551

* There was no significant difference in the extent of resection between CVSs and SVSs ($p = 0.87$).

Mortality and Non–Facial Nerve Morbidity

To analyze the perioperative mortality and morbidity rates, data were available for 293 patients with CVSs and 800 patients with SVSs (Table 4). The mortality rate was not significantly different between the groups (3% and 3.8%, respectively; $p = 0.6$). Furthermore, no significant difference was noted between the cumulative non-audiofacial complication rate (including mortality) among the CVS and SVS cohorts (24.5% and 25.6%, respectively; $p = 0.75$) (Fig. 3).

Brainstem Adherence

Only 3 studies had tabulated the comparative frequencies of tumor-brainstem adherence.[1,5,6] To analyze the same, data were available for 94 patients with CVSs and 356 patients with SVSs. Patients with SVSs had a significantly higher rate of brainstem adherence than those with CVSs (86.8% and 77.6%, $p = 0.03$ [Table 5]).

Discussion

The incidence of CVSs based on the current review of comparative studies range from 4% to 23% (mean 13.5% [Table 1]). Previous contemporary studies noted the incidence of CVSs as ranging from 5.7% to 48%.[6,25] Generally, CVS is considered to be a more aggressive and less predictable tumor than SVS. Patients with CVSs also tend to have shorter symptomatic periods.[4,9,13,25] In our review of the literature, the mean duration of symptoms was noted in 3 studies (Table 1), and a trend of a shorter duration of symptoms was noted in patients with CVSs, although the difference was not significant (18.7 vs 23.3 months, $p = 0.3$).

Fundová et al.[6] pointed out that the heterogeneity in the incidence of CVS will likely remain until an effective

TABLE 3: Analysis of facial nerve function*

Cohort	Facial Nerve Function		
	Good Outcome (%)	Worse Outcome	Total
CVS	118 (39)	184	302
SVS	500 (52.1)	459	959
total	618	643	1261

* A good outcome was defined as House-Brackmann Grade I or II at the latest follow-up. Facial nerve outcomes at the last follow-up were significantly better in patients with SVS than in those with CVS ($p = 0.0001$).

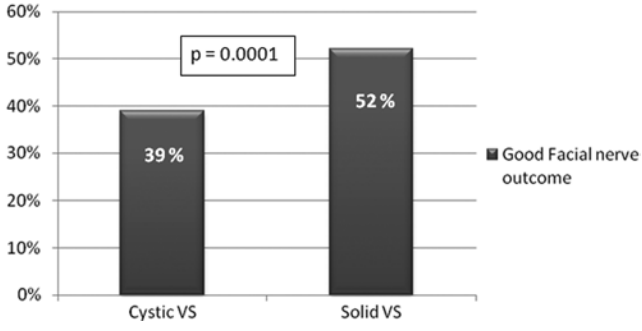

Fig. 2. Bar graph showing a comparison of the facial nerve function between CVSs and SVSs. Good outcome was defined by a House-Brackmann grade of I or II.

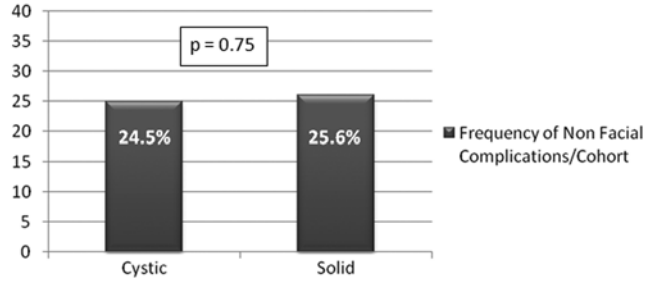

Fig. 3. Bar graph showing a comparison of the non–facial nerve complications per cohort between CVS and SVS.

standard classification system is universally used. There have been numerous attempts in the literature to propose a standard classification (Fig. 4).[6,12,13,24,25,31,33] Most of the studies considered VSs to be cystic if the mean diameter of the cyst was more than two-thirds the diameter of the tumor on MRI imaging. Among the available studies, the most extensive classification system has been proposed by Piccirillo et al.[25] (Table 6 and Figs. 5 and 6); nevertheless, the need to standardize a criterion for CVS remains.

Pathophysiology of Cyst Formation

Vestibular schwannomas are composed of 2 types of tissue that were originally described by Antoni in 1920.[30] Type A tissue is formed by a compact interwoven bundle of long bipolar spindle cells that have a tendency to palisade. Type B tissue is characterized by the presence of loosely organized tissue with small round satellite cells and tumor cell polymorphism.[20]

While the exact pathogenesis of CVS remains unclear, various observations and hypotheses have been put forth. Originally, it was thought that the cyst formation was due to the increased cell growth rate.[11,18] However, later studies did not find any difference in the proliferative index of CVSs compared with SVSs.[20,23] Now it is believed that the unpredictable and rapid growth in CVSs is due to the expansion of the cyst itself.[20,23]

Charabi et al.[3] concluded that the formation of cysts was due to the degeneration of tumor tissue. The authors observed the production a myxomatous material in small cystic areas by Antoni Type B tissue and hypothesized that the former may eventually coalesce into larger cysts and compress the surrounding Antoni Type A cells.[3,4] Their finding that the cyst walls consisted of Type A tissue and that the inner tissue was made up of Antoni Type B tissue has drawn general agreement. Some investigators have explained the presence of cysts by the difference in the distribution of Antoni cells and the degeneration of tumor tissues.[11,16,22] However, recent studies have not found any relationship between the distribution of cysts and the 2 types of tissues.[20,23]

An impairment of the blood-tumor barrier leading to an extravasation of serum proteins has also been proposed as a reason for the increase in cyst size.[27] Protein secretion by the tumor cells, due to an osmotic effect, augmenting the accumulation of fluid within the cyst is also thought to contribute to an increase in size.[14,17] While these mechanisms may explain the expansion of already present cysts, they do not explain their genesis.

While massive intratumoral hemorrhage has been classically attributed to an increase in the size of the tumor,[2,8,15] the role of microhemorrhages has also come to light as a possible reason for the presence of cysts.[22,24] Park et al.[23] documented the presence of significantly increased histological evidence of microhemorrhage, such as hemosiderin-laden macrophages, hemosiderin deposits, thrombotic vessels, and abnormal vessel proliferation, in CVSs compared with their solid counterparts (Fig. 7).

Another interesting observation is the increased expression of MMP-2 in the cyst fluid and cyst-lining wall of the tumor.[21] The proteolytic enzyme MMP-2 has been shown to be present in renal cystic lesions and ovarian cystic neoplasms and is thought to significantly contribute to the genesis of the cyst as well as to its biological invasiveness. Although it was thought that the presence of MMP-2 may explain the increased adhesion of the tumor to the neighboring neurovascular structures,[1,21] our review of the literature showed that SVSs had a significantly higher probability of adherence to the brainstem (Table 5).

TABLE 4: Frequency of non–facial nerve complications per cohort*

	Complications Present		
Cohort	Yes (%)	No	Total
CVS	72 (24.5)	221	293
SVS	205 (25.6)	595	800
total	277	816	1093

* No significant difference was noted between the cumulative nonfacial complications including death between the groups (p = 0.75).

TABLE 5: Analysis of studies reporting the comparative frequencies of tumor-brainstem adherence*

	Brainstem Adherence		
Cohort	Yes (%)	No	Total
CVS	73 (77.6)	21	94
SVS	309 (86.8)	47	356
total	382	68	450

* Data available from the 3 studies that reported brainstem adherence show that SVSs have a significantly higher rate of adherence than CVSs (p = 0.03).

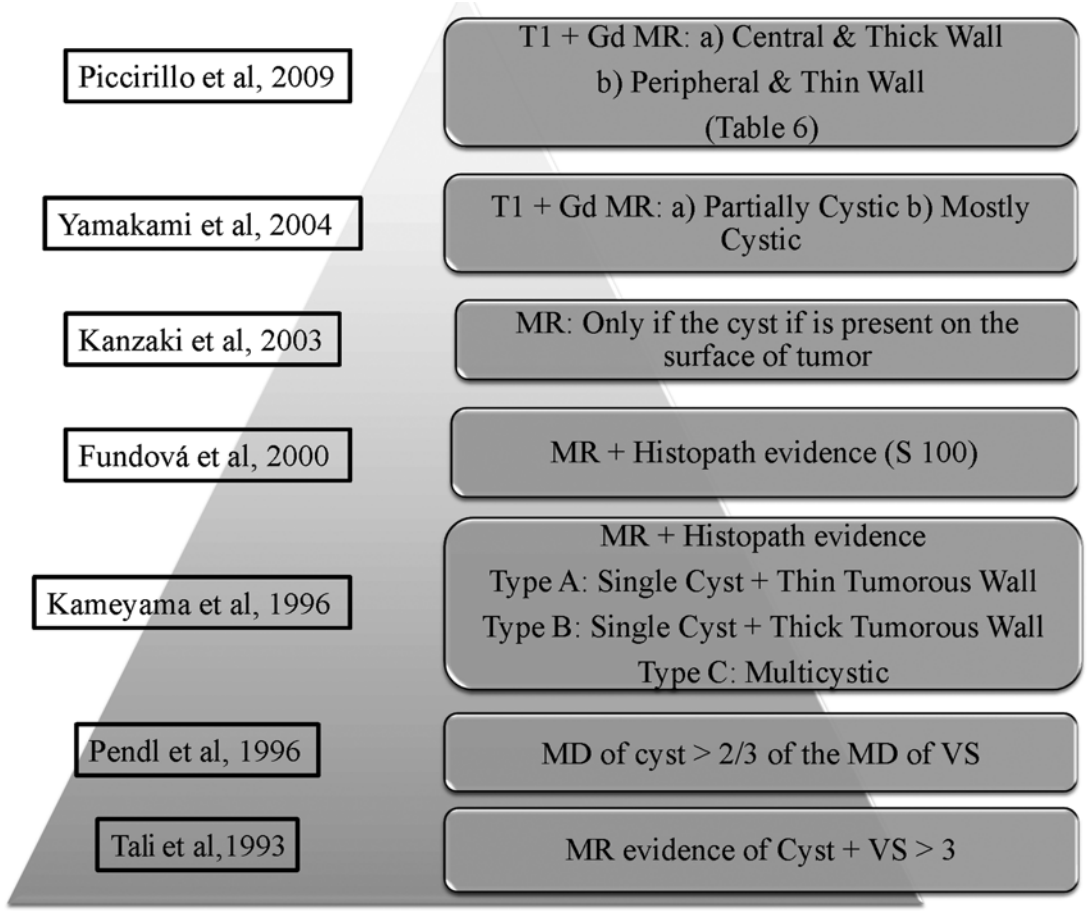

Fig. 4. Schematic illustration showing the evolution of and the variations in the classification system used to define CVS. Histopath = histopathological; MD = mean diameter; T1 = T1-weighted.

Operative Considerations

Surgery for CVSs can be technically challenging. Standard surgical approaches (translabyrinthine, middle fossa, and retrosigmoid) have been used to resect CVSs.[5,9,25,26] The patient's hearing status, size of the tumor with respect to cisternal and metal segments, brainstem compression, and surgeon's preference all play an integral role in determining which surgical approach is used. However, irrespective of which approach is used, there are fundamental differences and pertinent considerations while resecting CVSs versus SVSs.

Generally, the following are important factors that govern the surgical strategy among CVS treatment.

Number and Size of the Cysts. Multiple cysts in a tu-

TABLE 6: Proposed classification for CVS*

Type	Overall Cyst Location/ Cyst Wall Thickness	Subtype	Definition
A	central & thick wall	1	polycystic (multiple small intratumoral cysts w/ a thick cyst wall)
		2	polycystic (multiple moderate size intratumoral cysts w/ a thick cyst wall)
		3	monocystic (single large cyst w/ a thick or thin cyst wall)
B	peripheral & thin wall	1	anterior
		2	medial
		3	posterior
		4	combined

* The proposed classification is first based on overall cyst location (central or peripheral) and cyst wall thickness (thick or thin). Type A lesions are further subdivided by the cyst characteristics (polycystic or monocystic) and size. Type B lesions are further classified according to cyst orientation with respect to the internal auditory meatus (anterior, medial, posterior, or a combination of these locations). Reprinted with permission from Piccirillo et al: *Otol Neurol 30*:826–834, 2009.

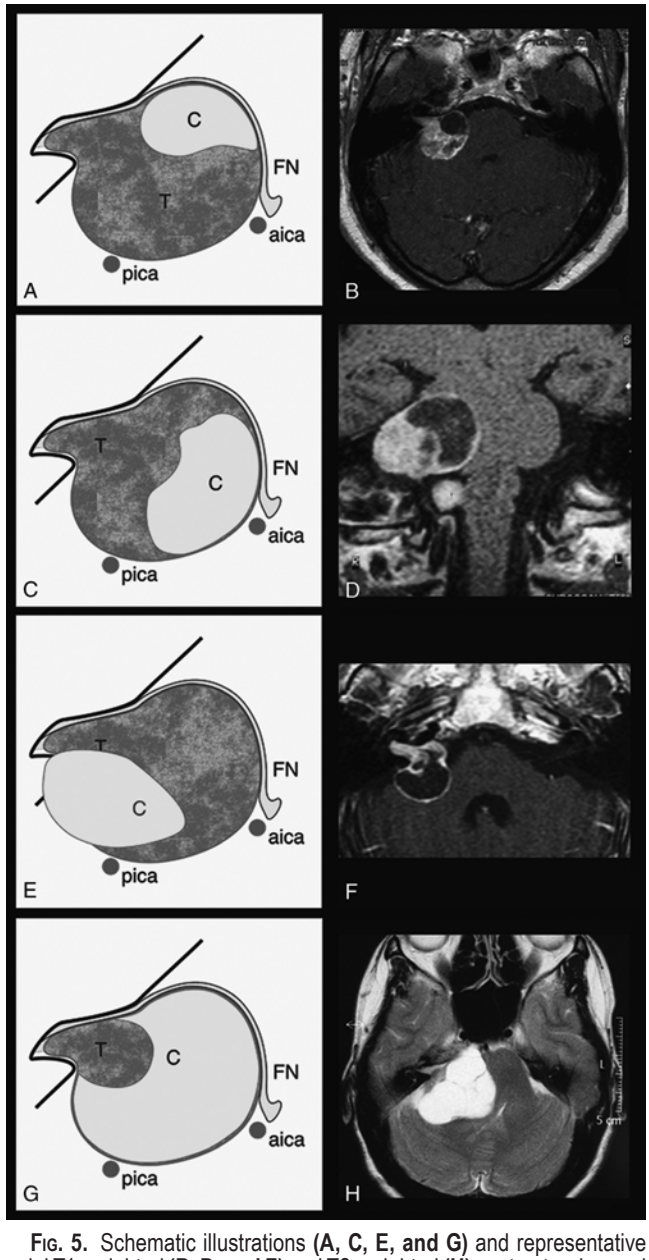

Fig. 5. Schematic illustrations **(A, C, E, and G)** and representative axial T1-weighted **(B, D, and F)** and T2-weighted **(H)** contrast-enhanced MRI studies of each subtype of Type B CVSs according to the proposed classification in Table 6. Type B1 (A and B), Type B2 (C and D), Type B3 (E and F), and Type B4 (G and H). aica = anterior inferior cerebellar artery; C = cyst; FN = facial nerve; pica = posterior inferior cerebellar artery; T = solid tumor. Magnetic resonance images B, D, and F are T1-weighted axial images with contrast. Reproduced with permission from Piccirillo et al: *Otol Neurol 30:*826–834, 2009.

mor can be challenging because of the heterogeneity in the consistency of the tumor. Resection usually proceeds more rapidly through cystic areas.[1,5] Nevertheless, tumors with large cystic components can be difficult to resect. Removal of large cyst contents can cause deformation of the tumor structure and subsequently may change the structural integrity by lowering the internal resistance. Cyst decompression is also likely to alter the existing relationship to adjacent structures, especially the facial nerve.

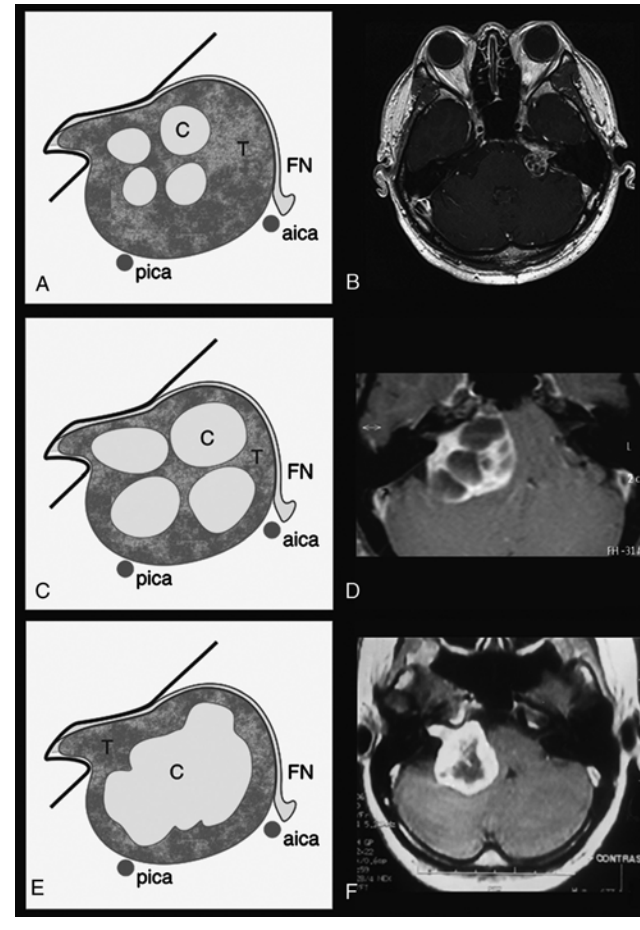

Fig. 6. Schematic illustrations **(A, C,and E)** and representative axial T1-weighted contrast-enhanced MRI studies **(B, D, and F)** of each subtype of Type A CVSs according to the proposed classification in Table 6. Type A1 (A and B), Type A2 (C and D), and Type A3 (E and F). Reproduced with permission from Piccirillo et al: *Otol Neurol 30:*826–834, 2009.

Thickness of the Cyst Wall. The thickness of the cyst wall is an important feature on preoperative imaging.[1,25] Thinner cyst walls provide less of a barrier and can be easily breached during dissection. This can be especially problematic if the cystic component is anterior or adherent to the facial nerve. One can imagine traversing a cyst with a thin wall and injuring the facial nerve due to the minimal barrier afforded by the wall. A thin cyst wall also provides a minimal subarachnoid plane for dissection. Therefore, it is suggested that the tumor should always be sharply dissected from the facial nerve, thus eliminating the traction forces associated with blunt dissection.[1] Also, using higher-voltage stimulation for facial nerve monitoring has been advocated in some studies.[9] This can potentially allow for earlier identification and preservation of the facial nerve while working in the cystic component.

Location of the Cyst Wall. Cystic areas along the tumor periphery can cause loss of the dissection plane, especially when the cyst is in the anterior portion of the tumor in contact with the facial nerve.[5,25] Similar issues may arise when the cystic component is located more

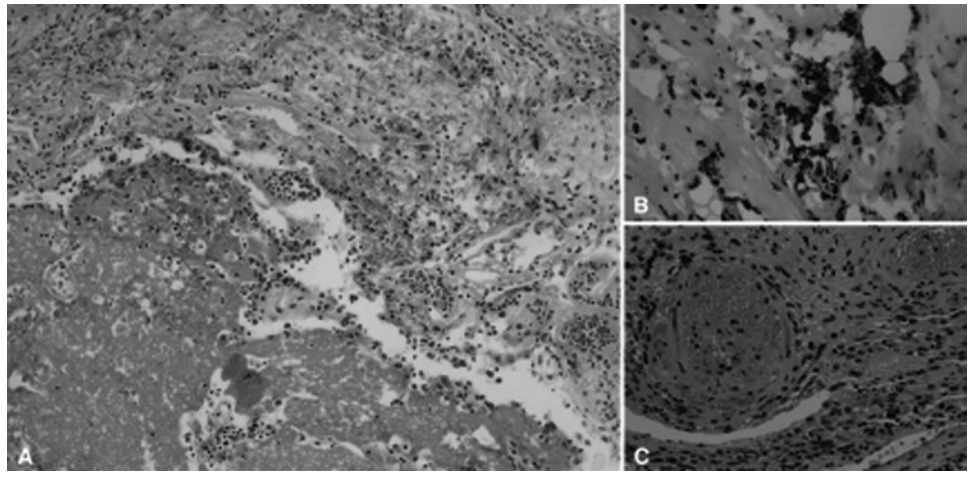

Fig. 7. Photomicrographs showing the histopathological appearance of a CVS. **A:** Note the abnormal vessel proliferation and hemosiderin deposits along the interface between the tumor and organizing hemorrhage. **B and C:** Hemosiderin deposits and hemosiderin-laden macrophages (B) and thrombotic vessels (C) are visible in the tumor area. H & E, original magnification × 45 (A), × 200 (B), and × 100 (C). Reproduced with permission from Park et al: *J Neurosurg 105:*576–580, 2006.

medially against the brainstem. On the other hand, these technical challenges may not be as evident in the centrally located cysts.

Adherence to Neighboring Neurovascular Structures. Individual cystic tumors may be notoriously adherent to neurovascular structures and may make the resection difficult. Historically, it was thought that the presence of a cyst increased the risk of adherence to these neurovascular structures; however, our systematic review found that SVSs were more likely to be adherent to the brainstem than CVSs (Table 5). Hence, even though adherence of CVSs may be a factor in making the resection difficult, this is likely also the case with SVSs.

Study Limitations

Our current quantitative study is based on the results of 2 studies with Level II evidence and 7 studies with Level III evidence; therefore, the results should be interpreted with caution. Having said that, performing a prospective controlled study and comparing the surgical outcomes between SVSs and CVSs may be difficult in terms of accruing enough patients to achieve desirable statistical power. Thus, a systematic review such as ours does evaluate a relatively large patient population and provides some trends for interpretation. A comparison of hearing outcomes was reported by only 2 studies and thus could not be considered as a potential primary end point. Lastly, stereotactic radiosurgery is an important component of the treatment paradigm for all types of VSs. Our study does not incorporate results of this treatment modality since there were only limited comparative series.[24,28]

Conclusions

Facial nerve outcomes are worse in patients undergoing resection for CVSs than in those undergoing resection for SVSs. There were no significant differences in the extent of resection or postoperative morbidity and mortality rates between CVS and SVS treatment. Further prospective comparason studies are required to validate the results of the current study.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Thakur, Guthikonda. Acquisition of data: Thakur, Khan. Analysis and interpretation of data: Thakur, Shorter, Sonig. Drafting the article: Thakur, Khan, Shorter. Critically revising the article: Nanda, Sonig, Gardner, Guthikonda. Approved the final version of the manuscript on behalf of all authors: Nanda. Statistical analysis: Thakur, Khan. Administrative/technical/material support: Nanda. Study supervision: Gardner, Guthikonda.

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Extent of resection and early postoperative outcomes following removal of cystic vestibular schwannomas: surgical experience over a decade and review of the literature

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Object. Vestibular schwannomas (VSs) are benign tumors of the eighth cranial nerve sheath, representing approximately 6%–8% of all newly diagnosed brain tumors, with an annual incidence of 2000–2500 cases in the US. Although most of these lesions are solid, cystic vestibular schwannomas (CVSs) compose 4%–20% of all VSs and are commonly larger at the time of presentation. The authors present their experience with the operative management of CVSs, including surgical approach, extent of resection, and postoperative facial nerve outcomes. The literature pertaining to clinical and histopathological differences between CVSs and their solid counterparts is reviewed.

Methods. The University of Southern California Department of Neurosurgery database was retrospectively reviewed to identify patients who had undergone resection of a VS between 2000 and 2010. One hundred seventy-nine patients with VS were identified. Patients with CVSs were the subject of the present analysis. Diagnosis of a CVS was made based on MRI findings. Clinical and neuroimaging data, including pre- and postoperative assessments and operative notes, were collected and reviewed.

Results. Twenty-three patients, 14 men (61%) and 9 women (39%), underwent 24 operations for CVSs. These patients composed 12.8% of all cases of VS. Patient ages ranged from 28 to 78 years (mean 55 years), and the mean maximal tumor diameter was 3.6 cm (range 2.0–4.0 cm). Patients most frequently presented with headache, hearing loss, vertigo, and dizziness. Preoperative facial numbness was reported in 44% of patients. Among the 24 cases, 13 were treated with retrosigmoid craniotomy and 11 via a translabyrinthine approach. Complete resection was achieved in 11 patients (48%), subtotal resection (STR) in 8 patients (35%), and near-total resection (NTR) in 4 patients (17%). Facial nerve outcomes were available in all except one case. Good facial nerve outcomes (House-Brackmann [HB] Grades I–III) were achieved in 82% of the patients who had undergone either NTR or STR, as compared with 73% of patients who had undergone gross-total resection (GTR; $p > 0.05$, Fisher exact test). In comparison, 83% of patients with solid VSs had a good HB grade ($p = 0.38$, Fisher exact test), although this finding did not reach statistical significance. Complications included wound infection (2 patients), delayed CSF leakage (1 patient), and a delayed temporal encephalocele following a translabyrinthine approach and requiring surgical repair (1 patient).

Conclusions. Cystic vestibular schwannoma represents a clinical and surgical entity separate from its solid counterpart, as demonstrated by its more rapid clinical course and early surgical outcomes. Facial nerve grades may correlate with the degree of tumor resection, trending toward poorer grades with more significant resections. Although GTR is recommended whenever possible, performing an STR when facial nerve preservation is in jeopardy to improve facial nerve outcomes is the preferred strategy at the authors' institution.
(http://thejns.org/doi/abs/10.3171/2012.7.FOCUS12206)

Key Words • **cystic vestibular schwannoma** • **retrosigmoid approach** • **translabyrinthine approach** • **facial nerve**

Vestibular schwannomas are one of the most common benign intracranial tumors, representing 6%–8% of all newly diagnosed brain tumors, with an approximate annual incidence of 2000–2500 cases in the US.[16] Specifically, they compose approximately 80% of the tumors arising within the cerebellopontine angle.[16] Although the majority of VSs are solid, a small proportion are cystic.

This cystic variety has been reported to account for 5.7%–48% of these tumors, although more recent studies have documented an incidence of 10% among all VSs.[6,11,12,23] However, the accuracy of these estimates is

Abbreviations used in this paper: CVS = cystic vestibular schwannoma; GTR = gross-total resection; HB = House-Brackmann; NTR = near-total resection; RS = retrosigmoid; STR = subtotal resection; TL = translabyrinthine; USC = University of Southern California; VS = vestibular schwannoma.

debatable given the variety of definitions of what constitutes a CVS.[3,6,12,13,27] Radiographically, CVSs are best diagnosed on MRI studies, with the presence of fluid-filled hyperintense compartments on T2-weighted sequences and, typically, hypointense compartments on T1-weighted imaging (Fig. 1). Epidermoid and arachnoid cysts, also in the differential diagnosis, can be distinguished from CVSs by the presence of Gd enhancement on MRI, which is generally not noted in the former pathologies.[5]

The cystic variety of tumors can also vary from its solid counterparts in its clinical presentation. Solid tumors have been reported by Selesnick and Johnson[22] to grow at a rate of about 2–6 mm per year. Conversely, cystic tumors can demonstrate a more unpredictable growth pattern, with rapid expansion of the cystic elements. These lesions can distort the brainstem, stretch cranial nerves, and lead to a rapid rise in intracranial pressure, without allowing the brain to gradually compensate for such compressive effects. As a result, patients presenting with CVSs may have a higher incidence and more rapid clinical course of elevated intracranial pressure, papilledema, hydrocephalus, cranial nerve paresis, and facial paresthesias.

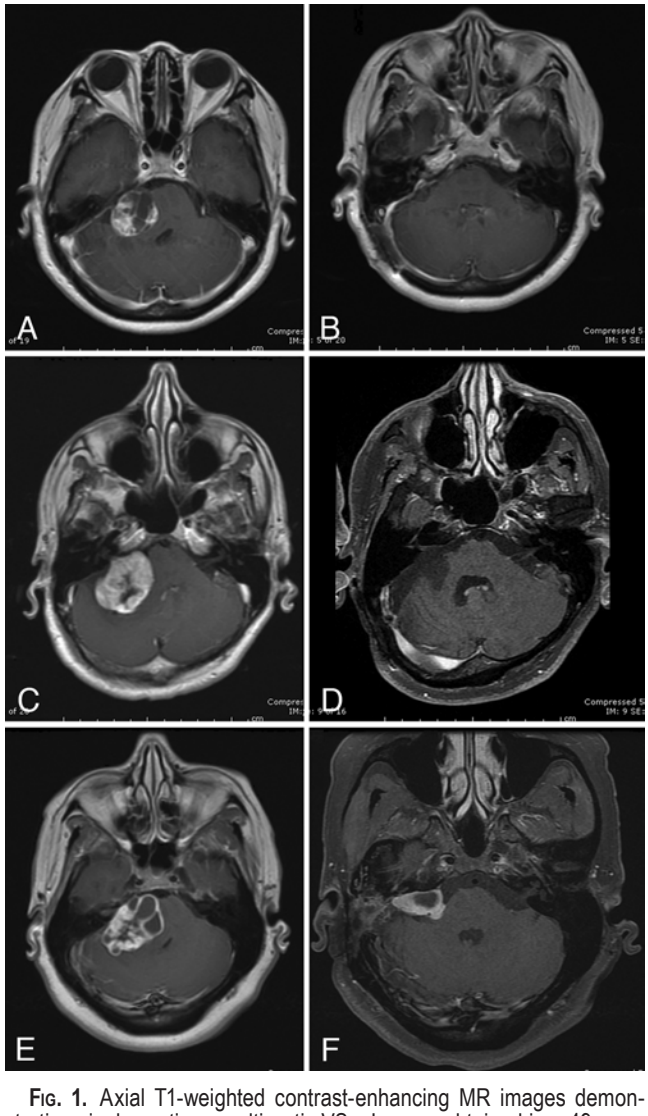

Fig. 1. Axial T1-weighted contrast-enhancing MR images demonstrating single cystic or multicystic VSs. Images obtained in a 49-year-old woman, showing a right-sided multicystic VS preoperatively **(A)** and successful GTR via a retrosigmoid approach at the 1-year follow-up **(B)**. This patient had HB Grade I facial nerve function after treatment. Images obtained in a 50-year-old man, demonstrating a right-sided central single CVS preoperatively **(C)** and successful GTR via a retrosigmoid approach at the 5-year follow-up **(D)**. This patient had HB Grade IV facial nerve function after resection. Images obtained in a 45-year-old woman, showing a right-sided multicystic VS preoperatively **(E)** and successful STR via a translabyrinthine approach at the 4-year follow-up **(F)**. This patient had HB Grade I facial nerve function after resection, although a cystic component not present on the immediate postoperative MR image later recurred (not shown).

Management paradigms for CVSs include clinical observation, radiosurgery, open resection, or a combination of surgery and radiosurgery. When the goal of surgery has been GTR, reported outcomes for facial nerve function have unfortunately been less successful than those for solid VSs, which can be attributed to the adherent nature of these cystic tumors to surrounding structures.[3,6,8,17,20,23]

In this paper we review our experience in the surgical treatment of CVSs, including surgical approached used, extent of resection, and early postoperative facial nerve outcomes. We also briefly review the clinical and histopathological differences between CVSs and their solid counterparts.

Methods

A search of the USC Department of Neurosurgery patient database was conducted to identify patients who had undergone craniotomy for the treatment of VS between the years 2000 and 2010. Only patients with CVSs were included in our analysis, and those treated solely with observation or stereotactic radiosurgery were excluded. Patient charts were retrospectively reviewed, including clinical data and notes, operative reports, pathology reports, and neuroimaging results. Data were reviewed in a confidential manner after obtaining approval from the institutional review board at the Keck Hospital of USC and LAC + USC Medical Center, in accordance with the Health Insurance Portability and Accountability Act.

All patients with CVS underwent a rigorous preoperative study panel. Computed tomography imaging data were available in most patients, whereas all patients underwent Gd-enhanced MRI studies. Audiometry testing was also performed in all patients, most importantly in those for whom a translabyrinthine approach was recommended. Gross-total resection was reported in patients who demonstrated no evidence of macroscopic tumor at the time of surgery. In cases in which a small remnant of tumor was left on the facial nerve or brainstem, NTR was reported. Any larger residual tumor was reported as STR.

Surgical approaches used in our patient population consisted of either retrosigmoid or translabyrinthine craniotomy. The choice depended on the presence or absence of serviceable hearing on preoperative testing, as well as the extent of tumor within the internal acoustic canal, regardless of tumor size. Patients were positioned either supine with the head turned 90° or, when neces-

sary, in a lateral or park bench position. In the operative suite, a neurophysiologist monitored facial nerve function as well as brainstem auditory evoked responses, somatosensory evoked potentials, and motor evoked potentials. Whenever possible, the Cavitron ultrasonic surgical aspirator (CUSA) was used. Postoperative assessment of facial nerve function was performed using the HB grading system.[9] For this study, we reported only early patient follow-up, including significant postoperative events and complications, early clinical follow-up, and neuroimaging study results. The senior author (S.L.G.) performed all operations.

Results

We identified 179 patients who had undergone resection of a histologically verified VS at USC during a 10-year period (2000–2010). Twenty-three patients (12.8%) had a CVS, as determined by neuroimaging characteristics. Fourteen patients (61%) were men, and 9 (39%) were women, with a mean age of 55 years (range 28–78 years). Sixty-seven percent of the tumors were located on the right, and all patients had single (unilateral) tumors on preoperative imaging, with no history of neurofibromatosis Type 2. The most frequent presenting symptom was diminished hearing in the ipsilateral ear (94%), followed by vertigo (69%), facial sensory disturbances (44%), and headaches (31%; Table 1). All patients were treated primarily at our institution, without any prior radiation or surgical therapy. Magnetic resonance imaging was performed in all patients, and the mean maximal tumor diameter was 3.6 cm (range 2.5–4.0 cm).

Surgical and Facial Nerve Outcomes

The two surgical approaches favored by the senior author (S.L.G.) for CVSs are retrosigmoid and translabyrinthine craniotomy. Twelve patients (52%) underwent a primary retrosigmoid approach and the remainder (48%) underwent a translabyrinthine approach. After presenting with symptomatic tumor progression, 1 patient underwent a retrosigmoid craniotomy 21 months after the translabyrinthine approach. Subtotal resection was achieved in 8 patients (35%), NTR in 4 (17%), and GTR was possible in 11 (48%). Subtotal tumor resection was possible in 42% (5 of 12) of the patients who underwent the retrosigmoid approach and 27% (3 of 11) of those who underwent the translabyrinthine approach. Near-total resection was documented in 17% (2 of 12) of those who underwent the retrosigmoid approach and 18% (2 of 11) of those who underwent the translabyrinthine approach. Lastly, in patients with complete tumor resection, 42% (5 of 12) underwent the retrosigmoid approach, and 55% (6 of 11) the translabyrinthine approach. The most common reason for NTR or STR was tumor adherence to or invasion of surrounding vital structures (facial nerve, brainstem, and so forth) or difficulty in accurately identifying the facial nerve.

TABLE 1: Neurological symptoms on presentation with CVS

Symptom	% of Patients
hearing loss	93.9
vertigo/dizziness	68.75
facial sensory disturbance	43.75
headaches	31.25
facial nerve symptoms or weakness	18.75
tinnitus	12.5

Facial nerve outcomes were available in all but 1 patient (this patient underwent STR via a retrosigmoid approach). Postoperative facial nerve outcomes were good (HB Grade I–III) in 77% (17 of 22) of the patients. Among them, we noted HB Grade I in 65% (11 of 17), Grade II in 12% (2 of 17), and Grade III in 24% (4 of 17; Fig. 2). A good HB grade was achieved in 86% (6 of 7) of the patients with STR, 75% (3 of 4) of those with NTR, and 73% (8 of 11) of those with GTR ($p > 0.05$). A poor HB grade (Grades IV–VI) was evident in 23% of the patients overall: A poor HB grade (IV–VI) facial palsy developed in 14% (1 of 7) of patients with STR, 25% (1 of 4) of those with NTR, and 27% (3 of 11) of those with GTR. Good HB outcomes were obtained in 82% (9 of 11) of patients who underwent either NTR or STR, as compared with 73% of those who underwent GTR ($p > 0.05$, Fisher exact test). Anatomical preservation of the facial nerve was noted in all 23 patients.

As a comparison, in patients with solid VSs, 83% had a good HB grade ($p = 0.38$, Fisher exact test). A good HB outcome was obtained in 89% (16 of 18) of those who underwent STR for a solid VS and 83% (114 of 138) of those who underwent GTR of a solid VS. These results were not statistically significant, as compared with those in patients with CVSs.

Patient Complications

No patients undergoing resection of a CVS died. Wound infections without meningitis developed in 2 patients postoperatively, requiring wound revision. One of these patients, who initially underwent a translabyrinthine approach followed by a retrosigmoid approach 21 months later, experienced delayed wound breakdown. One patient presented with delayed CSF leakage 2 years after a retrosigmoid approach, and the leak was surgically

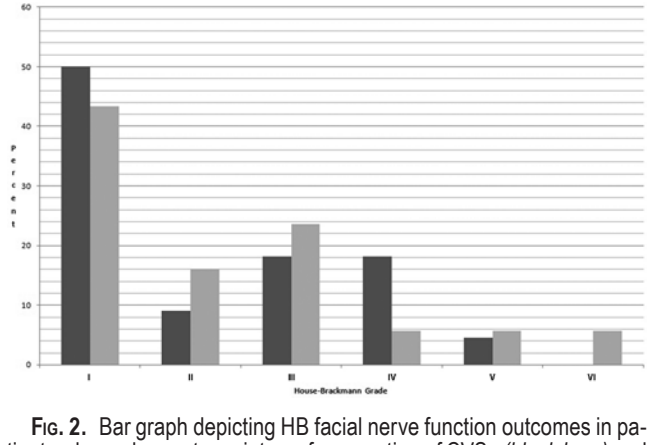

Fig. 2. Bar graph depicting HB facial nerve function outcomes in patients who underwent craniotomy for resection of CVSs *(black bars)* and solid VSs *(gray bars)*. The y axis represents the percentage of patients.

repaired. Another patient presented with seizures 2 years after resection of a CVS via the translabyrinthine approach. Follow-up MRI demonstrated what appeared to be a temporal lobe encephalocele in the translabyrinthine defect. The patient had complete resolution of seizures following surgical repair of this lesion.

Discussion

Vestibular schwannomas are benign tumors believed to arise from Schwann cells of the eighth cranial nerve. The annual incidence in the US is approximately 10–15 cases/million people.[10,25] Although VSs are generally considered slow-growing neoplasms, the cystic subtype can grow quite rapidly. As a result, the presentation for CVS can differ from that for solid VS, including a shorter duration of symptoms prior to presentation, increased risk of cranial nerve palsy, and increased rates of hydrocephalus.[1,6,12,20,23]

Previous reports have suggested that CVSs compose 5.7%–48% of all VSs. More recent studies have favored a proportion closer to 10%.[6,11,12,23] Our series corroborates the findings of more recent studies, demonstrating an incidence of 12.8% among all patients with VS (Table 2). However, it is important to note that our data are based on neuroimaging findings alone. In a recent study by Fundová et al.,[6] in which 2 additional criteria were used for the diagnosis of CVS (intraoperative identification of cystic elements and histopathological staining for S100 protein positivity in addition to radiographic data), an incidence of only 5.7% was reported.

The precise etiology of the cystic component of these tumors remains unknown, although various mechanisms for cystic formation have been proposed. Although not mutually exclusive, these mechanisms have included tumor growth with subsequent central necrosis, coalescence of microcysts within the Antoni B environment, and/or repeated hemorrhage within the tumor.[2,18] Cyst enlargement can subsequently occur because of an osmotic gradient resulting from serum proteins or the production of mucinous material within the cyst.[2,23] In a more recent report by Moon et al.,[17] matrix metalloproteinases—proteolytic enzymes found naturally during embryogenesis and tissue remodeling—were identified within the cyst fluid and walls of CVSs. These authors believe that matrix metalloproteinase-2 may contribute to cyst formation as well as tumor adhesion to the facial nerve by promoting tumor expansion and growth or by stimulating proteolytic degradation at the tumor-nerve interface. Future studies analyzing the genomic and epigenetic substrates for a CVS phenotype may provide more information regarding the etiology of this cystic formation.

TABLE 2: Literature survey of the incidence of CVS

Authors & Year	Total No. of VSs	No. of CVSs (% of total)
Kendall & Symon, 1977	31	3 (9.7)
Robbins & Marshall, 1978	39	8 (20.5)
Wallace et al., 1993	35	7 (20)
Tali et al., 1993	80	15 (18.8)
Charabi et al., 1994	571	23 (4)
Jeng et al., 1995	27	13 (48)
Pendl et al., 1996	148	9 (6.1)
Fundová et al., 2000	773	44 (5.7)
Sinha & Sharma, 2008	284	58 (20.4)
Piccirillo et al., 2009	1416	96 (6.8)
present study	179	23 (12.8)

Cystic vestibular schwannomas have been associated with more rapid tumor growth, a shorter duration of symptoms, and increased involvement of the facial nerve.[3,6] As evident in our cohort of patients, the symptoms most commonly found in patients presenting with CVS include hearing loss, headache, cerebellar signs, trigeminal nerve involvement, and facial nerve involvement. Other signs, such as tinnitus, lower cranial nerve paresis, diplopia, and visual symptoms or loss, can also be seen.[23]

Surgical outcomes after craniotomy for CVS have been reported to be worse than those for solid VSs, specifically when evaluating facial nerve function. In the current study, worse facial nerve outcomes were associated with GTR of a CVS rather than a solid VSs, although these results did not reach statistical significance. A previous report by Fundová et al.[6] demonstrated a poor HB grade (IV–VI) in 66% of patients at 1 year or more of follow-up. In the report by Sinha and Sharma,[23] facial nerve preservation rates were 67.9% and 82.7% in the CVS and solid VS groups, respectively. A good HB grade (I–III) was noted postoperatively in 67.9% of patients with CVS. In a more recent report by Piccirillo et al.,[20] 81% of patients with CVSs who underwent resection had a good HB grade. All of these studies demonstrate improved facial nerve outcomes in solid VS cases as compared with outcomes in CVS cases. Our results correspond with previous findings: 77% of patients with CVSs who underwent treatment demonstrated an HB grade between I and III, as compared with 83% in solid VS cases.

The role of stereotactic radiosurgery has yet to be elucidated in the treatment of CVS. In a report by Pendl et al.,[19] radiosurgery outcomes were suboptimal, with 3 of 6 cystic tumors demonstrating rapid and significant cystic expansion requiring urgent surgery for neurological decline. Ganslandt et al.[7] described an intratumoral hemorrhage leading to death in a patient with CVS 15 months after treatment with stereotactic radiosurgery. Delsanti and Régis[4] described their experience with stereotactic radiosurgery in the treatment of 54 CVS cases. Their failure rate, defined as the need for a second procedure, was 6.4%, almost 3-fold greater than that reported for solid VS.[15] In our series, only 1 patient was treated with radiosurgery (5040 cGy in the form of intensity-modulated radiation therapy). As can be seen in Fig. 3, despite 2 resections and adjuvant radiation therapy, the CVS recurred after each intervention, with ongoing cyst formation. Surprisingly, however, almost 2 years after external radiotherapy, the cystic and solid components of the tumor demonstrated a noticeable decrement in size. Cystic VSs with multiple recurrences, in particular, pose a significant challenge in achieving a surgical cure or tumor control.

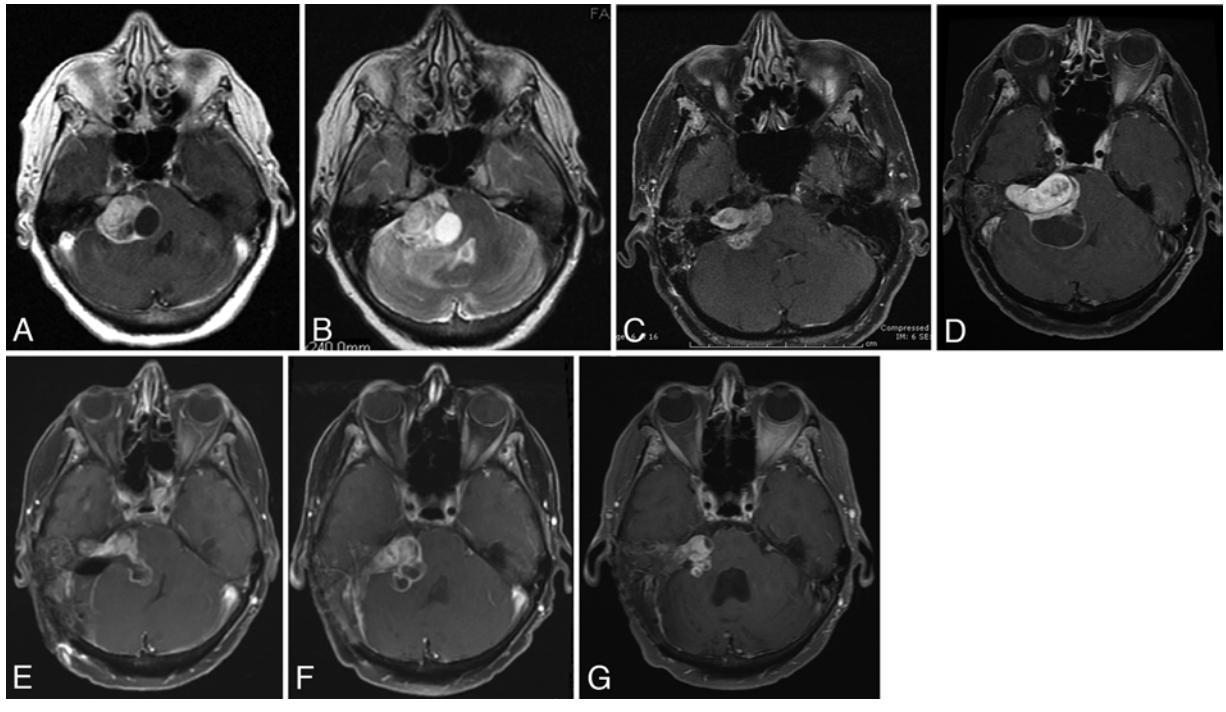

Fig. 3. Images obtained in a 66-year-old woman presenting with hearing loss, right facial sensory disturbance, and vertigo. Preoperative T1-weighted contrast-enhanced MR image **(A)** demonstrating a right VS with multiple small cysts and a single large medial cyst adjacent to the brainstem. Axial T2-weighted MR image **(B)** revealing the classic appearance of CVS with hyperintense signal within the cyst. Postoperative MR image **(C)** obtained after a translabyrinthine approach, showing residual tumor after STR. Axial MR image **(D)** obtained 1 year after the image featured in panel B, demonstrating interval development of a large medial cyst with significant compression of the pons and cerebellum. Postoperative axial MR image **(E)** obtained immediately after a second craniotomy (retrosigmoid approach), demonstrating significant debulking of the tumor, brainstem decompression, and cyst resection. Follow-up MR image **(F)** obtained 8 months later, showing tumor recurrence along with 2 small medial cysts, which prompted adjuvant external radiation treatment with 5040 cGy. Axial MR image **(G)** obtained 2 years after the completion of radiation, showing a mild decrease in the size of the 2 small cysts and the residual tumor.

Conclusions

Cystic vestibular schwannomas represent an entity separate from their solid counterparts, as demonstrated by their greater likelihood for facial numbness and hydrocephalus at presentation, rapid clinical course, adherence to vital neurological structures, and early surgical outcomes. Early facial nerve function appears to correlate with the degree of tumor resection, with poorer grades generally noted with more radical resections. We recommend GTR whenever possible, performing STR when facial nerve preservation is in jeopardy to improve facial nerve outcome and limit complications. Although anecdotal, our experience suggests that "once a CVS, always a CVS," often making the treatment of recurrent CVS a more substantial challenge.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Yashar, Zada, Giannotta. Acquisition of data: Yashar, Harris, Giannotta. Analysis and interpretation of data: Yashar, Harris, Giannotta. Drafting the article: Yashar, Zada, Harris. Critically revising the article: Yashar, Zada, Giannotta. Reviewed submitted version of manuscript: Yashar, Zada, Giannotta. Approved the final version of the manuscript on behalf of all authors: Yashar. Statistical analysis: Yashar. Administrative/technical/material support: Yashar, Zada, Giannotta. Study supervision: Giannotta.

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Surgery for vestibular schwannomas: a systematic review of complications by approach

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Object. Various studies report outcomes of vestibular schwannoma (VS) surgery, but few studies have compared outcomes across the various approaches. The authors conducted a systematic review of the available data on VS surgery, comparing the different approaches and their associated complications.

Methods. MEDLINE searches were conducted to collect studies that reported information on patients undergoing VS surgery. The authors set inclusion criteria for such studies, including the availability of follow-up data for at least 3 months, inclusion of preoperative and postoperative audiometric data, intraoperative monitoring, and reporting of results using established and standardized metrics. Data were collected on hearing loss, facial nerve dysfunction, persistent postoperative headache, CSF leak, operative mortality, residual tumor, tumor recurrence, cranial nerve (CN) dysfunction involving nerves other than CN VII or VIII, and other neurological complications. The authors reviewed data from 35 studies pertaining to 5064 patients who had undergone VS surgery.

Results. The analyses for hearing loss and facial nerve dysfunction were stratified into the following tumor categories: intracanalicular (IC), size (extrameatal diameter) < 1.5 cm, size 1.5–3.0 cm, and size > 3.0 cm. The middle cranial fossa approach was found to be superior to the retrosigmoid approach for hearing preservation in patients with tumors < 1.5 cm (hearing loss in 43.6% vs 64.3%, $p < 0.001$). All other size categories showed no significant difference between middle cranial fossa and retrosigmoid approaches with respect to hearing loss. The retrosigmoid approach was associated with significantly less facial nerve dysfunction in patients with IC tumors than the middle cranial fossa method was; however, neither differed significantly from the translabyrinthine corridor (4%, 16.7%, 0%, respectively, $p < 0.001$). The middle cranial fossa approach differed significantly from the translabyrinthine approach for patients with tumors < 1.5 cm, whereas neither differed from the retrosigmoid approach (3.3%, 11.5%, and 7.2%, respectively, $p = 0.001$). The retrosigmoid approach involved less facial nerve dysfunction than the middle cranial fossa or translabyrinthine approaches for tumors 1.5–3.0 cm (6.1%, 17.3%, and 15.8%, respectively; $p < 0.001$). The retrosigmoid approach was also superior to the translabyrinthine approach for tumors > 3.0 cm (30.2% vs 42.5%, respectively, $p < 0.001$). Postoperative headache was significantly more likely after the retrosigmoid approach than after the translabyrinthine approach, but neither differed significantly from the middle cranial fossa approach (17.3%, 0%, and 8%, respectively; $p < 0.001$). The incidence of CSF leak was significantly greater after the retrosigmoid approach than after either the middle cranial fossa or translabyrinthine approaches (10.3%, 5.3%, 7.1%; $p = 0.001$). The incidences of residual tumor, mortality, major non-CN complications, residual tumor, tumor recurrence, and dysfunction of other cranial nerves were not significantly different across the approaches.

Conclusions. The middle cranial fossa approach seems safest for hearing preservation in patients with smaller tumors. Based on the data, the retrosigmoid approach seems to be the most versatile corridor for facial nerve preservation for most tumor sizes, but it is associated with a higher risk of postoperative pain and CSF fistula. The translabyrinthine approach is associated with complete hearing loss but may be useful for patients with large tumors and poor preoperative hearing.
(http://thejns.org/doi/abs/10.3171/2012.6.FOCUS12163)

Key Words • **vestibular schwannoma** • **neurosurgical procedure** • **complication** • **facial nerve weakness** • **hearing loss**

The first operation for VS removal was performed by Sir Charles Ballance[21] in 1894. At the time, intracranial operations carried mortality rates as high as 84%,[14] so when Harvey Cushing wrote his 1917 monograph on tumors of the acoustic nerve, he advocated a subtotal resection to decrease mortality and morbidity.[14,21] Over the ensuing years, the goal of VS surgery evolved from preservation of life to preservation of facial function,[27] and now to conservation of hearing.[39,61] In modern studies, mortality rates as low as 0.4% have been documented,[16] with rates of anatomical preservation of the facial nerve approaching 100%.[4,5,14,48,57,67] Moving forward, the primary goal in VS therapy will likely become early detection and complete removal or near-complete removal in combination with radiosurgery, with optimal preservation of hearing and facial nerve function when possible.[37,55,97]

Currently, there are 3 options for managing cases of

Abbreviations used in this paper: AAO-HNS = American Academy of Otolaryngology–Head and Neck Surgery; CN = cranial nerve; CPA = cerebellopontine angle; IAC = internal auditory canal; IC = intracanalicular; VS = vestibular schwannoma.

VS: 1) conservative treatment ("wait and see"); 2) resection; and 3) radiotherapy/radiosurgery. Many physicians choose the conservative approach if patients have small tumors,[18] minimal or no symptoms, poor overall health status, or if they are elderly,[24,33,87] but this approach may still result in hearing loss over time.[35,41,65,70,91] Radiosurgery is often chosen for patients with tumors smaller than 2.5 cm or for the elderly. Patients harboring recurrent or residual tumors are also frequently referred for radiosurgery rather than an additional operation.[61,67,68] Outcomes with respect to facial nerve function and hearing have been reported to be more favorable with radiosurgery than with traditional open surgery.[63] This former method has recently been advocated as the primary therapy for larger tumors, but there is a lack of data on outcomes of radiosurgery in patients with larger VSs.[96]

Three different approaches are generally considered when planning VS surgery, and opinions abound about when to use a particular approach. To this end, some criteria have been consistently established in the literature, and factors for consideration have been named: tumor size, patient age and overall health status, anatomy of the vestibule and CPA, involvement of the brainstem and facial nerve, and extent of involvement of the IAC.[19,20,80,93] One group suggested the surgeon's comfort as one of the main determinants of this choice.[45] Anderson et al.[2] suggested a combined translabyrinthine and retrosigmoid approach for very large tumors (diameter > 3 cm), as this allows better proximal and distal identification of the facial nerve. Each approach has associated advantages and disadvantages.

The middle cranial fossa approach offers some of the highest historical hearing preservation rates, but it places the facial nerve between the surgeon and the tumor (which results in the need for blind dissection in some cases[26]), and it places some degree of retraction on the temporal lobe (which entails a risk of postoperative seizures and speech disturbances),[20,43,49,72,73,92] while providing a limited view of the CPA.[47] Satar et al.[81] believe that facial nerve dysfunction following the middle cranial fossa approach is transient and that the long-term outcome is the same as with the other approaches. This approach may be poorly tolerated by the elderly, as extradural dissection of the adherent dura may be difficult.[85] The middle cranial fossa corridor is suggested for younger patients with smaller tumors[7,38,81] in the IAC with less involvement of the CPA, specifically the tumors that involve the fundus of the IAC, a location to which access and visualization are restricted during the retrosigmoid approach.[20] This route is also indicated for patients with useful preoperative hearing (although the definition of "useful" is still open to debate).[25,80,93]

The retrosigmoid approach is well known to neurosurgeons and allows a panoramic visualization of the CPA.[49] Opponents of this approach quote cerebellar retraction as a risk for postoperative ataxia[42,88] and maintain that this approach carries a higher incidence of postoperative headache.[11,31,76,93] The retrosigmoid corridor may also provide a limited visualization of the fundus of the IAC, which may necessitate blind dissection to remove the entire tumor.[43] The retrosigmoid approach is a versatile route for tumors of any size regardless of the preoperative hearing status.[28]

The translabyrinthine approach precludes the possibility of hearing preservation but allows removal of a tumor of almost any size, with early identification of the facial nerve,[38,79,89,93] and some argue that the translabyrinthine approach permits good preservation of all CNs.[54] It also provides very good visualization of the lateral IAC and fundus, whereas the exposure of these in the retrosigmoid approach is limited due to the risk of damaging the vestibule and cochlea; therefore the translabyrinthine approach may allow for a more complete removal of tumor from these areas. Indications for the translabyrinthine approach include larger tumors and the preoperative lack of serviceable hearing.[12,23,49,60,79,95]

Because no single study to date includes a large enough number of patients, and a randomized trial would not be feasible to compare the value of different operative approaches, we attempted to conduct a systematic review of the available outcome data.

Methods

Inclusion Criteria

We defined inclusion criteria for the studies included in this review to ensure a relatively homogeneous patient population while maintaining the largest possible group size. To be included in the final analysis, studies had to fit the following criteria: the authors must have included follow-up data of at least 3 months, provided preoperative and postoperative audiometric data, used intraoperative monitoring for facial and cochlear nerves, and reported facial nerve results using the House-Brackmann scale.[46] In addition, we required reporting on hearing preservation using the AAO-HNS Committee on Hearing and Equilibrium classification[1] (or its equivalent)—studies providing raw audiometric data were also included, as were studies using the Gardner-Robertson classification.[34] Studies that included patients who underwent repeat operations, radiosurgery, or endoscopic surgery were excluded.

Normal facial nerve function was defined as House-Brackmann Grade I or II. Hearing loss was defined as any AAO-HNS classification below B, a pure-tone audiometry score of more than 50 dB, or a speech discrimination score of less than 50%.[34,94]

Literature Search

A literature search was conducted using PubMed and the search terms "acoustic neuroma surgery," "acoustic neuroma outcomes," "acoustic neuroma approach," "vestibular schwannoma surgery," "vestibular schwannoma approach," and "vestibular schwannoma outcomes." This yielded approximately 6800 articles, which were then screened by title and abstract. The initial screening process selected 180 articles, which were again screened for relevance to the inquiry and usable data. Eighty articles were found to offer data helpful to the analysis. Of these, 45 were excluded for reasons including inadequate follow-up and data not reported in standard metrics, as well as the other exclusion criteria mentioned above. Bibliographies of certain articles were also searched for further studies. The final quantitative analysis included 35

articles. See Fig. 1 for a flowchart outlining this selection process.

The search included studies published during January 1992 through December 2010 to allow for review of the cases in which modern microsurgical techniques, including intraoperative CN monitoring, had been used. Finally, this review included data on 5064 patients who underwent VS surgery and who were included in the above-mentioned studies.

Review

Data were summarized across all studies using counts (percentages). Comparisons across groups were performed using Fisher exact tests. Pairwise comparisons between treatment methods were also performed using Fisher exact tests with significance levels adjusted using the Bonferroni correction. Pairwise comparisons were considered significant only if the p value was less than 0.017. Statistical tests were performed using R for Windows (version 2.10.1).

Initially, comparisons were made across all 3 approaches. When significant differences were detected, pairwise comparisons were performed, as needed, to test for differences among individual groups. Pairwise comparisons are notated in Table 1 with superscript letters "a" and "b." Two cells that contain the same superscript letter are not significantly different. Conversely, 2 cells with different superscript letters are significantly different. Data for facial nerve dysfunction and hearing loss were stratified based on tumor size because tumor size has been shown to be the most important predictive factor for outcome related to these variables.[17,26,42,65,66,77] Tumors were stratified as entirely IC (no extrameatal extension) or based on their extrameatal diameter (< 1.5 cm, 1.5–3.0 cm, or > 3.0 cm into the CPA).

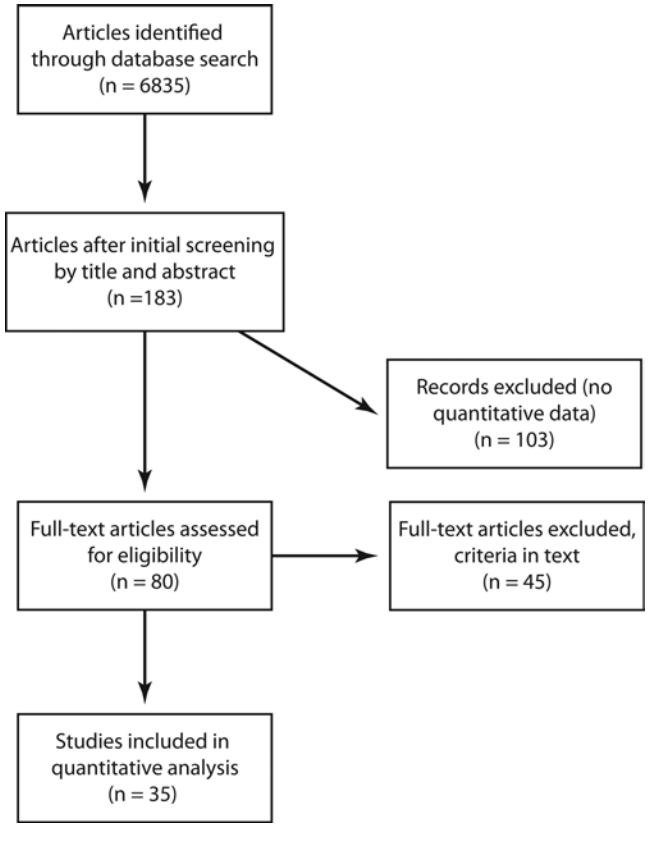

Fig. 1. Flow diagram outlining the search and screening method used in this study.

Results

Hearing Loss

Table 1 summarizes the results for different clinical variables based on surgical approach. For patients harboring tumors less than 1.5 cm in diameter, the proportion of patients experiencing hearing loss was significantly lower among those who underwent surgery via a middle cranial fossa approach than those who were treated via the retrosigmoid route (43.6% vs 64.3%; $p < 0.001$). Among patients with tumors 1.5–3.0 cm in diameter, 82.7% of those treated with a middle cranial fossa approach experienced hearing loss, whereas 71.6% of those treated with a retrosigmoid approach suffered from this complication ($p = 0.051$). Finally, among the patients with intracanalicular tumors, 40.6% and 44.3% of the patients who underwent middle cranial fossa and retrosigmoid surgery, respectively, experienced hearing loss ($p = 0.492$).

Facial Nerve Dysfunction

Considered to be the most important complication of VS surgery,[2,10,18,47] facial nerve dysfunction was defined as House-Brackmann Grade III or higher at last follow-up. In the group of patients with tumors less than 1.5 cm in diameter, 3.3% of those treated with a middle cranial fossa approach, 7.2% of those treated with a retrosigmoid approach, and 11.5% of those treated with a translabyrinthine approach suffered facial nerve dysfunction postoperatively ($p = 0.001$). The middle cranial fossa approach was associated with significantly lower rates of facial nerve dysfunction than the translabyrinthine approach; there was no significant difference between the retrosigmoid approach and either of the other 2 approaches with respect to rates of facial nerve dysfunction.

Among patients with 1.5- to 3.0-cm tumors, 17.3% of those treated with a middle cranial fossa approach, 6.1% of those treated with a retrosigmoid approach, and 15.8% of those treated with a translabyrinthine approach had facial nerve dysfunction ($p < 0.001$). In this analysis, the retrosigmoid approach was associated with significantly less facial nerve dysfunction than either the middle cranial fossa or translabyrinthine approach. Among patients with tumors larger than 3.0 cm in diameter, 30.2% of those treated with a retrosigmoid approach and 42.5% of those treated with a translabyrinthine approach had facial nerve dysfunction ($p < 0.001$), and, expectedly, insufficient data were available on the patients in this tumor size group undergoing middle cranial fossa surgery. Among patients with intracanalicular tumors, 16.7% of those treated with a middle cranial fossa approach, 4.0% of those treated with a retrosigmoid approach, and 0% of those treated with a translabyrinthine approach suffered facial nerve dysfunction ($p < 0.001$).

TABLE 1: Results of systematic review of complications per approach*

Variable	Approach			p Value
	MCF	RS	TL	
hearing loss				
tumor size <1.5 cm	72/165 (43.6)	137/213 (64.3)	NA	<0.001
tumor size 1.5 cm–3.0 cm	62/75 (82.7)	214/299 (71.6)	NA	0.051
tumor size >3.0 cm	NA	91/127 (71.7)	NA	NA
intracanalicular tumors	95/234 (40.6)	86/194 (44.3)	NA	0.492
CN VII dysfunction				
tumor size <1.5 cm	8/240 (3.3)[a]	20/279 (7.2)[a,b]	38/331 (11.5)[b]	0.001
tumor size 1.5 cm–3.0 cm	9/52 (17.3)[a]	28/456 (6.1)[b]	65/412 (15.8)[a]	<0.001
tumor size >3.0 cm	NA	134/444 (30.2)	144/339 (42.5)	<0.001
intracanalicular tumors	41/245 (16.7)[a]	8/200 (4.0)[b]	0/11 (0.0)[a,b]	<0.001
CSF leak	23/436 (5.3)[a]	110/1067 (10.3)[b]	116/1623 (7.1)[a]	0.001
post-op headache	4/50 (8.0)[a,b]	127/732 (17.3)[a]	0/40 (0.0)[b]	<0.001
mortality	0/42 (0.0)	2/772 (0.3)	1/75 (1.3)	0.346
major neurol compl	1/42 (2.4)	12/655 (1.8)	13/500 (2.6)	0.513
residual tumor	6/231 (2.6)	44/735 (6.0)	48/857 (5.6)	0.107
other CN dysfunction	0/35 (0.0)	17/607 (2.8)	2/167 (1.2)	0.457
recurrence	1/91 (1.1)	31/501 (6.2)	NA	0.045

* Values represent numbers of patients (%). Data were summarized across all studies using counts (%). Tumor size refers to extrameatal diameter (extension into the CPA). Comparisons across groups were performed using Fisher exact tests. Pairwise comparisons between treatment methods were also performed using Fisher exact tests with significance levels adjusted using the Bonferroni correction. Pairwise comparisons were considered significant only if $p < 0.017$. Statistical tests were performed using R (version 2.10.1). Abbreviations: compl = complications; MCF = middle cranial fossa; NA = data not available; neurol = neurological; RS = retrosigmoid; TL = translabyrinthine.

Pairwise comparisons are notated by superscript letters ([a] and [b]). Two cells that contain the same letter are not significantly different. Conversely, 2 cells with different letters are significantly different.

Cerebrospinal Fluid Leak

Among patients treated with the middle cranial fossa approach, 5.3% experienced a CSF leak, whereas 10.3% of patients undergoing retrosigmoid surgery and 7.1% of those undergoing translabyrinthine surgery suffered from this complication ($p = 0.001$). Pairwise comparisons indicated that the middle cranial fossa and translabyrinthine approaches were associated with significantly lower rates of CSF leak than the retrosigmoid approach.

Postoperative Headache

Postoperative headache was found to be most prevalent among patients undergoing retrosigmoid surgery (17.3%), with significantly fewer patients undergoing translabyrinthine surgery reported to experience this complication (0%). Notably, the middle cranial fossa approach did not differ significantly from either of the other 2 groups in this regard (8.0%), but the retrosigmoid approach was associated with a significantly higher incidence of postoperative headache ($p < 0.001$). This analysis was likely skewed due to the lack of reporting in the translabyrinthine category. The trend in the data seems to indicate that the incidence of postoperative headache is higher with the retrosigmoid approach than with the middle cranial fossa approach.

Mortality

There was no significant difference detected in mortality rates across the 3 groups. The reported operative mortality rates were 0%, 0.3%, and 1.3% for middle cranial fossa, retrosigmoid, and translabyrinthine approaches, respectively ($p = 0.346$).

Major Neurological Complications

Major neurological complications included strokes (arterial and venous), seizure disorder, and persistent cerebellar dysfunction. Among patients undergoing middle cranial fossa surgery, the rate of major neurological complication was 2.4%, whereas it was 1.8% among those treated with a retrosigmoid approach and 2.6% among those treated with a translabyrinthine approach ($p = 0.513$).

Residual Tumor

Residual tumor was defined as any tumor visible on MRI after surgery. Cases in which subtotal resections had been planned were excluded from this analysis. Among the patients treated with a middle cranial fossa approach, 2.6% harbored residual tumor, whereas 6.0% of the those treated with a retrosigmoid approach and 5.6% of those treated with a translabyrinthine approach had some residual tumor. There was no significant difference across the 3 groups with regard to residual tumor ($p = 0.107$).

Tumor Recurrence

Based on the investigators' reports, tumor recurrence was diagnosed in 1.1% of patients treated with a middle crania fossa approach and 6.2% of those treated with a retrosigmoid approach. Insufficient data were available to analyze this variable among patients treated with a translabyrinthine approach. The difference between the retrosigmoid and middle cranial fossa groups approached significance ($p = 0.045$).

Follow-Up Duration

We found 33 studies with information regarding the duration of follow-up as well as facial nerve function. The mean duration of follow-up was 23 months.

Discussion

Hearing Preservation

Our review suggests that the middle cranial fossa approach is preferable for hearing preservation in patients with smaller tumors (< 1.5 cm extension into the CPA). However, for tumors extending 1.5–3.0 cm into the CPA, the retrosigmoid approach may actually provide better rates of hearing preservation than the middle cranial fossa route. The middle cranial fossa approach provides a limited window into the posterior fossa, and the potentially blind dissection necessitated by the presence of the facial nerve in the surgeon's field of view may limit the resection of larger tumors and allow damage to the cochlear portion of CN VIII and its vasculature during resection.[15] The retrosigmoid approach, on the other hand, provides a more panoramic view of the portion of the tumor in the CPA cistern and its relationship to the surrounding cerebrovascular structures.

The practice of offering patients early surgery upon diagnosis of a small tumor as a method to preserve hearing has been controversial in VS surgery.[6,30,32,50,51,56,62,77,78,86,92] Glasscock and others submitted that no patient should be excluded from surgery based on tumor size alone.[7,36,55,97] Several groups have supported this position, declaring that aggressive tumor removal as early as possible provides the greatest chance of hearing preservation.[41,50,70,76,77] Consistent with previous studies,[17,26,42,66,77] our analysis indicates improved hearing preservation with smaller tumors.

Facial Nerve Function

The best indicator of quality of life following VS surgery is facial nerve function.[2,8,16,44] Facial function at discharge is strongly suggestive of the nerve's final level of function.[8] Delayed facial nerve dysfunction has been documented in many reports,[7,8,11,53] potentially related to delayed ischemia, edema,[7,53] or viral reactivation (Bell palsy).[11] The timing for reanimation of the facial nerve if it is severed or damaged during surgery remains controversial.[3,11] However, it is known that reconstruction of a facial nerve, no matter what method is employed, is not likely to have an excellent outcome (House-Brackmann Grade I or II).[11,75]

Frequently cited predictors of facial nerve function include tumor size, surgical approach, and the use of intraoperative monitoring.[15,22,40,64,75] The length of contact of the tumor with the nerve has also been suggested as a potential prognostic factor.[18] Samii et al.[74] believe that prior surgery or radiosurgery and the presence of an intratumoral cyst are negative predictors of postoperative facial nerve function. Facial nerve function may be best preserved through the retrosigmoid corridor, according to current data.[15]

Our analysis points out that for tumors smaller than 1.5 cm in diameter, the middle cranial fossa approach provides better facial nerve preservation than does the translabyrinthine approach. However, the middle cranial fossa and retrosigmoid approaches do not seem to differ significantly with regard to facial nerve preservation in patients with tumors in this size category. On the other hand, for larger tumors (1.5–3.0 cm), the retrosigmoid approach provides a clear advantage. During the middle cranial fossa approach for larger tumors, the facial nerve is often located between the surgeon and the tumor; this topography places the facial nerve at a greater risk for damage.[81] The retrosigmoid approach also seems to provide better outcomes for patients with intracanalicular lesions. (It should be noted, however, that the 0% incidence of facial nerve dysfunction with the translabyrinthine approach should be interpreted with caution, as it is based on a very small number of patients from a single study).[60]

The middle cranial fossa approach has been suggested as the approach of choice for intracanalicular tumors and for hearing preservation.[41,50,52,90] The increased incidence of facial nerve dysfunction associated with this approach, as demonstrated by our analysis, has also been previously mentioned in the literature.[59,81] The higher incidence of facial nerve dysfunction for all patients with tumors larger than 3.0 cm is likely related to the effect of tumor size.[17,26,42,65,66,77] In summary, our analysis demonstrates the clear benefit of the retrosigmoid approach specifically for facial function among patients with tumors that are intracanalicular and 1.5–3.0 cm in size, with no difference in benefit between the middle cranial fossa and retrosigmoid approaches for patients with extrameatal tumor diameters less than 1.5 cm. For most tumor sizes, the translabyrinthine approach seems to be associated with a higher rate of facial nerve dysfunction. Our close examination of the studies that contributed patients to our review for the translabyrinthine approach (specifically, patients with tumors > 3 cm in diameter) revealed that at least half of the patients harbored tumors larger than 4 cm, which may account for the higher incidence of facial dysfunction in this group.

Postoperative CSF Leakage

The risk of CSF fistula formation has been considered higher for retrosigmoid surgeries due to the difficulty encountered with a watertight dural closure.[13,75] Tumor size is not correlated with the risk of CSF fistula,[69] although larger tumors approached through the translabyrinthine route may be associated with a higher CSF leak rate.[58,75] This could also be related to the approach itself, as meticulous dural closure is difficult, if not impossible, with this technique. The present analysis confirms the belief that the retrosigmoid approach may result in a greater risk of CSF leakage than the middle cranial fossa and translabyrinthine approaches, which we did not find to

differ significantly from each other in this respect. The confounding factor in this analysis is the different ways surgeons handle their closures to prevent a CSF leak. The retrosigmoid approach, which often violates the mastoid air cells during the craniectomy or craniotomy, may indeed place the patient at a higher risk of CSF leak.

Postoperative Headaches

Several causes have been suggested for the persistent postoperative headaches that seem to plague patients undergoing retrosigmoid surgery. Among these are chemical irritation from the bone dust created by drilling the IAC[11,82] and fibrous adhesions formed between the dura mater and suboccipital muscles.[83] Cranioplasty to seal the bony defect can significantly reduce the incidence of postoperative headaches.[31,82,83] Ruckenstein et al.[71] pointed out that although cranioplasty reduces the incidence of headache in the long term, it does not significantly reduce the incidence of headache (compared with the incidence associated with the translabyrinthine approach) in the immediate postoperative period (within 1 year of surgery). Our analysis confirms a significantly higher rate of postoperative headache among patients treated with a retrosigmoid approach as compared with those treated with a translabyrinthine approach.[83]

Although no statistically significant difference was detected between the retrosigmoid and middle cranial fossa approaches with regard to postoperative headache, the raw data suggest that a difference may indeed exist (4 of 50 patients treated with a middle cranial fossa approach vs 127 of 732 treated with a retrosigmoid approach suffered from headache). The translabyrinthine approach seems to be associated with a small risk of postoperative headaches (0% in this analysis). However, this point should be considered carefully, as the data regarding this variable were reported only for a group of 40 patients, all in a single study.[83]

Residual Tumor

Indications for subtotal resection include preservation of facial nerve integrity, development of an arrhythmia during brainstem dissection, advanced age, poor overall medical status, and previous failure of radiation therapy.[9] Subtotal resection may be a reasonable strategy, especially if facial nerve integrity is at risk. A relatively slow rate of regrowth of residual tumor and the effectiveness of radiosurgery for controlling small tumors are justifications for this approach, and we advocate such a strategy to preserve facial nerve function.[67] Unfortunately the studies included in this review did not provide the stratified data needed to conduct an analysis of residual tumor based on approach. This analysis indicates that the rates of residual tumor were similar for all 3 approaches, but if further investigation were carried out into the incidence of residual (and indeed, recurrent) tumor based on size, we believe there would be a clear trend toward higher incidence of residual or recurrent tumor with larger tumors.

Tumor Recurrence

Recurrence of tumor is documented, especially among patients undergoing less than total resection of the tumor. Reducing the volume of residual tumor as much as possible may help minimize this risk.[29] Schmerber et al.,[84] in a long-term study, found that microscopic remnants of tumor within the nerve may retain some growth potential. The present analysis points out that patients undergoing the retrosigmoid approach may have higher rates of recurrence. The higher frequency of tumor recurrence among patients treated with a retrosigmoid approach may indeed be related to the smaller size of tumors selected for the middle cranial fossa approach, as well as poor visualization of the lateral aspect of the IAC offered through the retrosigmoid approach.

Limitations of the Study and Future Directions

In 1993, Glasscock et al.[38] decried the "disarray of data" in the VS literature regarding outcome reporting. Although there are available standard metrics for reporting tumor size, hearing, and facial function, some authors do not adhere to these metrics, providing nonuniform data that are difficult to compare across studies.[93] Additionally, there are multiple standard scales for reporting hearing function, some of which do not reconcile. This fact prevents fair comparisons among institutions.[59] The need for a new hearing classification system has also been suggested.[5] Ideally, a universally accepted system—specifically for VS outcome classification and reporting—that considers tumor size, hearing, and facial function would be desirable and practical.

Limitations of this review include the absence of randomized or controlled studies and bias introduced by the surgeon's preoperative decision regarding the selection of approach. Chronology also potentially confounds our results, because the included studies were retrieved from both older and more recent series. These series have employed different techniques to maximize tumor resection. The introduction of radiosurgery has affected the decision-making process and the outcome of the recent series as compared with the old ones. Furthermore, there is a certain skew in the data, as evidenced in Table 2. Patients treated by means of a middle cranial fossa approach tend to have smaller tumors and thus may have more positive outcomes based on this fact alone. The translabyrinthine approach is well represented among patients with larger tumors, who may have worse outcomes due to the size of their tumor. There is certainly a preponderance of patients in the retrosigmoid approach group overall, but of those patients included in the final size-stratified analyses (hearing and facial nerve function), this difference is minimal, and, in fact, there are approximately 80 more patients in the translabyrinthine approach group. The composition of these populations is unfortunately out of the control of the authors, as surgeons choose approaches based on the potential benefit to each individual patient, and each approach has its advantages, disadvantages, and indications, as described above. Finally, the heterogeneity of data and lack of monitoring necessitated the exclusion of several otherwise large and potentially useful studies.

Conclusions

Much of the data presented here correlate with the findings in prior studies on VS surgery. Our data suggest

TABLE 2: Number of patients per approach and tumor size*

Approach & Tumor Size	No. of Patients
retrosigmoid	2295
intracanalicular	214
tumor size <1.5 cm	525
tumor size 1.5–3.0 cm	487
tumor size >3.0 cm	551
middle cranial fossa	814
intracanalicular	301
tumor size <1.5 cm	285
tumor size 1.5–3.0 cm	78
tumor size >3.0 cm	0
translabyrinthine	1955
intracanalicular	11
tumor size <1.5 cm	518
tumor size 1.5–3.0 cm	637
tumor size >3.0 cm	674

* Not all studies included data stratified by tumor size. The values listed here are based on available data.

that the retrosigmoid corridor remains a versatile approach that may be useful for preserving hearing in the case of larger tumors but carries a higher risk of postoperative CSF leakage and headache. Indeed, the middle cranial fossa approach seems to have a lower incidence of CSF leak and postoperative headache compared with the retrosigmoid approach and carries with it superior hearing preservation in the case of smaller tumors. Finally, the translabyrinthine approach may be associated with poorer facial nerve function, but the analysis was confounded by inclusion of larger tumors approached through the translabyrinthine route.

With more emphasis on hearing preservation, there is a need to standardize the classification system for hearing preservation based on a universally accepted, objective, and reproducible method of measurement. Finally, the treatment plan and surgical approach should be chosen based on careful assessment of the patient and her or his tumor, with individualized consideration and discussion of the risks to maximize the benefit to the patient.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: all authors. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Statistical analysis: Ansari, Terry. Study supervision: Cohen-Gadol.

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Technical nuances of resection of giant (> 5 cm) vestibular schwannomas: pearls for success

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Removal of vestibular schwannomas (VSs, or acoustic neuromas) remains one of the most challenging operations in neurosurgery. Giant or huge tumors (> 5 cm) heighten these challenges, and technical nuances play a special role in maximizing tumor resection while minimizing complications. In this article, the senior author describes his technical experience with microsurgical excision of giant VSs. The accompanying video further illustrates these details.
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Key Words • **acoustic neuroma** • **vestibular schwannoma** • **technical nuance** • **complication** • **giant tumor**

Acoustic neuromas (or VSs) are one of the more technically challenging neurosurgical lesions to remove as exemplified by the well-demonstrated career-long learning curve in VS surgery.[6,23] There is a long-recognized difference in outcome between low- and high-volume surgical centers.[31] Giant VSs, greater than 4–5 cm in greatest extracanalicular diameter,[12] carry their own unique risks for resection. Tumor size has been repeatedly shown to correlate with patient outcome, including postoperative cranial nerve and brainstem function.[11] Higher complication rates[2,28] have been reported with large (> 2 cm) and giant tumors. However, in the hands of experienced surgeons, giant VSs can be removed safely with no deaths, low morbidity, and cranial nerve preservation.[25] In this paper we describe the nuances of the preoperative, intraoperative, and postoperative management of giant VSs framed in the context of the case presented in the accompanying video (Video 1).

Video 1. Clip showing removal of a large VS (> 5 cm). Technical nuances play an important role in maximizing tumor resection while minimizing complications. This video of an operation performed by Aaron A. Cohen-Gadol, M.D., M.Sc., illustrates these details. Click here to view with Media Player. Click here to view with Quicktime.

Abbreviations used in this paper: IAC = internal auditory canal; VS = vestibular schwannoma.

Preoperative Evaluation

All patients should be evaluated clinically with a complete history and neurological examination focusing on cranial nerve and brainstem/cerebellar function, as well as long tract signs: most tumors of this size (> 5 cm) are Hanover Class T4A or T4B with significant brainstem compression. The most common presenting symptom is significantly decreased or absent ipsilateral hearing. In patients with residual useful hearing, formal audiometry should be performed because preservation of hearing is reported to be possible occasionally and may influence the surgical approach. Pure-tone audiograms and speech discrimination testing is the standard, with results classified according to a number of methods, commonly the Gardner-Robertson scale.[9] Auditory brainstem response screening plays little role, given the overt clinical symptomatology at presentation. Other common cranial neuropathies in recent series of giant VSs include tinnitus (34%–100%); extraocular muscle impairment (4%, usually abducent nerve dysfunction); trigeminal sensory dysfunction (4%–17%); facial palsy (1.6%–14%), which should be objectively evaluated using the House-Brackmann scale; and dysphagia/lower cranial nerve dysfunction (4%–5%), which if significant, merits formal swallow and vocal cord mobility studies. Signs of cerebellar or brainstem compression may also be present, including ataxia (also related to vestibular dysfunction, 28%–64%), dysmetria (7%), hyperreflexia (14%),

as well as signs and symptoms of intracranial mass effect or hydrocephalus (7%–33%).[3,25,30]

An important consideration is the management of hydrocephalus. Preoperative untreated hydrocephalus has previously been connected to poorer outcomes and higher complication rates.[18,34] The majority of patients do not require further treatment for their hydrocephalus besides tumor resection; however, a minority do not achieve such a goal and may require postoperative ventriculoperitoneal shunting. As expected, increased tumor size is a risk factor for development of postoperative hydrocephalus.[10] If symptomatic obstructive hydrocephalus is present, we prefer to place an external ventricular drain at the time of craniotomy with the goal of weaning the drain postoperatively, if possible, and shunt placement if necessary. In the particular case described in the accompanying video, due to the patient's gravid state and associated risks, an initial shunting procedure was performed to temporize the patient's symptoms until postpartum to allow for delayed tumor resection.

Imaging Studies

Imaging studies should include CT and MRI. Fine bone detail is important to evaluate the bone anatomy that will be removed during the creation of the surgical corridor. Recognition of the extent of pneumatization of the temporal bones is important to prevent postoperative CSF leak. The position of the venous anatomy, specifically a high-riding jugular bulb or posterior sigmoid sinus, may affect the choice and extent of bone removal.[13] Tumor anatomy and its relation to neurovascular structures is best evaluated using MRI, which allows differentiation from other cerebellopontine angle lesions, such as meningioma or epidermoid.[32] Outgrowth from the posterior fossa is critical to note, and extension of giant tumors around the tentorium may indicate the need to resect a portion of the tentorium for adequate tumor exposure and microdissection. Beyond the local anatomical relationships of the tumor, its intrinsic characteristics such as a cystic component can be identified, which has been correlated with poorer outcomes.[8,26] The relationship of the tumor to the basilar and posterior inferior cerebellar arteries may be appreciated; such vessels should be carefully protected if ultrasonic aspirator devices are used for tumor decompression. Despite extension of these giant tumors through the jugular foramen, the tumor can typically be microsurgically dissected off of the lower cranial nerves with no significant risk, as exemplified by the low rate of postoperative deficit (2%–6%). Presence of edema in the brainstem indicates a high risk of brainstem pial violation during microdissection of the tumor capsule.[19] If significant brainstem edema is evident, staging the surgery may be strongly considered, as the interval between 2 stages would allow the tumor to deliver itself into the resection cavity created during the first operative session.[29]

Representative preoperative and postoperative MR images from the senior author's patient are shown in Fig. 1, demonstrating the presence of minimal brainstem edema, extent of brainstem compression, and the presence of tumor within the jugular foramen.

Operative Preparation

Staging the operative session should be strongly considered in giant tumors to provide the patient with the best outcome. Fatigue of the surgeon and the operative team during the later (and more critical) parts of the operation is an important factor.[29] In a staged procedure, one accepts the risks of a second surgery for the benefits of shorter procedures that are less taxing for the patient and surgeon. Indeed, early series of staged surgery for these giant tumors demonstrated no deaths and acceptable morbidity (65% functional facial nerve preservation).[29] Modern staged series have shown that comparable outcomes with minimal additional risk can be achieved via staged resection for large tumors, and have even suggested superior facial nerve outcomes.[4,17,19] Recent microsurgical and neuroanesthetic techniques weigh in favor of a single-stage surgery: modern series have achieved excellent results (< 1% death, 70%–75% functional facial nerve preservation, 97%–100% excision).[3,25] In a young, otherwise healthy patient such as the one described in the video, a single-stage surgery is a reasonable consideration.

Routine use of intraoperative facial nerve electromyography as well as brainstem auditory evoked response potentials and somatosensory evoked potentials to identify and monitor facial nerve and cochlear nerve/brainstem integrity, respectively, are important. We find these to be an invaluable guide to safe resection and for revising a surgeon's maneuvers intraoperatively to prevent neural injury. It should be noted, however, that the significant attenuation and atrophy of the facial nerve in giant tumors, as apparent by preoperative facial weakness, often complicates mapping the exact location of the nerve splayed over the tumor capsule.

Various skull base approaches and their combinations are described for removal of giant VSs, including retrosigmoid suboccipital and translabyrinthine routes. Numerous experts have advocated for each route, generally based on their personal expertise, achieving excellent outcomes.[3,14,25,30] Those experienced in both denote benefits to each approach and tailor their own approaches to each patient's unique anatomy,[13] while admitting that the best approach is often the one most familiar to the surgeon. Raslan et al.[19] recently described a 2-stage operative session for these tumors and employed both retrosigmoid and translabyrinthine approaches in each session with great results. The decision to stage tumor removal was made based on evidence of cerebellar or brainstem edema, significant tumor adherence to the brainstem and facial nerve, a poorly stimulating facial nerve (partial nerve injury), and attenuated facial nerve.

In the modern era, when the goal of zero deaths and minimal morbidity is achievable, hearing conservation may be considered; in large and giant tumors, this is the exception rather than the rule,[35] and only in the few cases in which serviceable hearing remains preoperatively. Venous anatomy should be considered; a high and/or anteriorly located sigmoid sinus significantly favors selection of a retrosigmoid approach.[13] In our experience, the translabyrinthine approach affords the opportunity for less cerebellar retraction and may be a consideration for

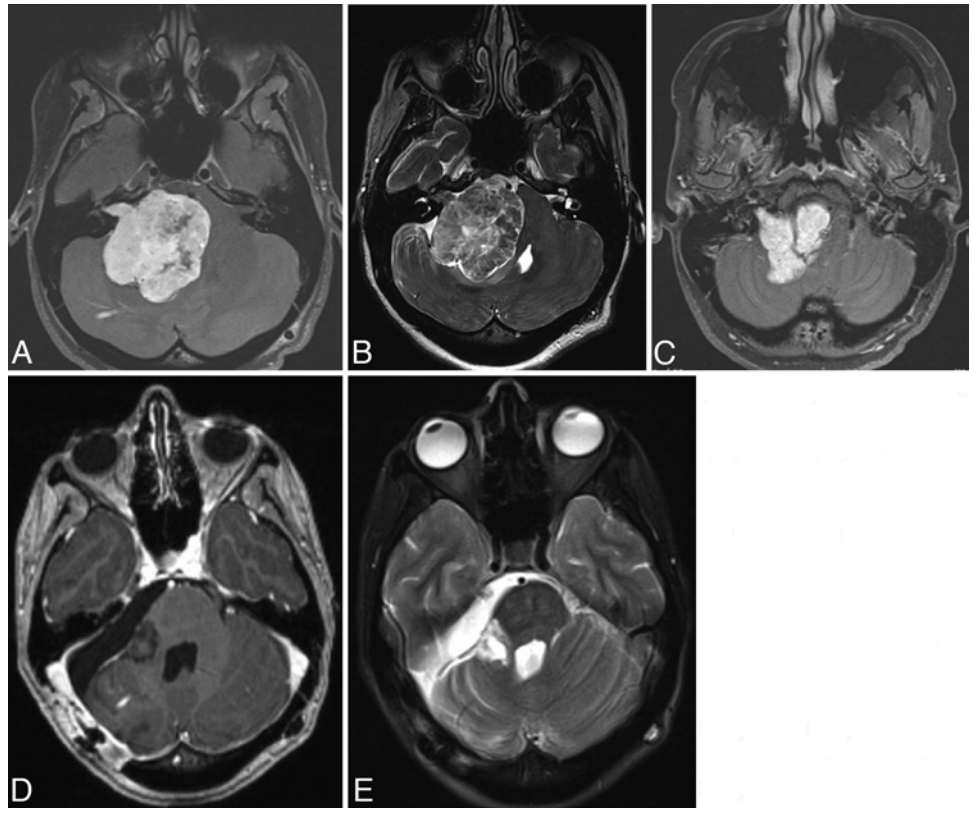

Fig. 1. Representative axial MR images from the senior author's patient, demonstrating the extent of brainstem compression **(A)**, presence of minimal brainstem edema **(B)**, and tumor within the jugular foramen **(C)**. Postoperative images **(D and E)** reveal gross-total removal of the mass. Images A, C, and D are T1-weighted with contrast enhancement, images B and E are T2-weighted.

younger patients with a "full" cerebellum. This route may be used in combination with the retrosigmoid route as it may not provide enough exposure of the brainstem for giant tumors. The retrosigmoid approach provides a more panoramic view of the compressed brainstem, however, and has generally been our preference. This approach allows for a single-stage resection of the tumor.

Patient Positioning

The patient may be positioned in sitting/semisitting or horizontal (supine, oblique, park bench) fashion. The sitting position is classically associated with a risk of venous air embolism; precordial Doppler and/or end-tidal CO_2 monitoring are used. Some operators report less blood loss, less operative time, and lower cranial nerve dysfunction with the sitting position.[1,20] The sitting/semisitting position will facilitate a clear surgical field by using irrigation alone. Bimanual microdissection is a great advantage without the use of bipolar electrocautery, which could place the facial nerve at risk. The sitting position will place the arms of the surgeons at risk for fatigue. Nonetheless, surgeon's preference and familiarity with a particular position is most important.[5] We avoid the supine position due to the risk of neck stiffness associated with such long operative sessions. Our preference has been the park-bench position.

Intraoperative Nuances

For our retrosigmoid approach, a curvilinear incision (Fig. 2A) prevents the scalp flap from interfering with the working zone of the surgeon and decreases the operator's working distance to the tumor (Fig. 2B and C). A large retromastoid craniotomy/craniectomy extending along the edges of the transverse and sigmoid sinuses is desirable. Smaller craniotomy/craniectomy may not provide adequate decompression if intraoperative cerebellar swelling is encountered. Bone removal for giant tumors extends to the posterior fossa floor, but does not involve the opening of the foramen magnum. If significant obstructive hydrocephalus is present preoperatively, CSF drainage through a previously placed external ventricular drain is achieved before dural opening to avoid cerebellar herniation. Dural opening is completed using a curvilinear incision (Fig. 3A). Additional CSF is released by opening the arachnoid membranes caudal to the tumor (Fig. 3B). A small lateral portion of the cerebellum may be excised to allow for adequate tumor exposure without aggressive cerebellar retraction; this may be necessary only in younger patients with a "full" cerebellum, otherwise cisternal opening and drainage may be adequate.[33] The arachnoid membranes over the posterior aspect of the tumor capsule are excised.

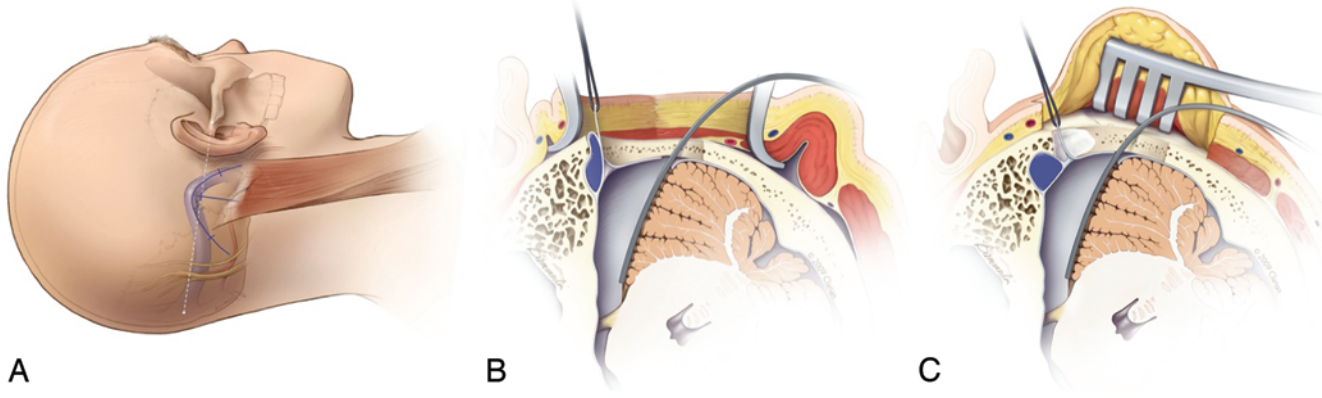

Fig. 2. The curvilinear incision is used and the patient's head is minimally turned away from the surgeon. The landmarks for localizing the incision relative to dural venous sinuses and inion are demonstrated **(A)**. The curvilinear incision **(C)** prevents the scalp flap from interfering with the working zone of the surgeon and decreases the operator's working distance to the tumor, as opposed to the linear incision **(B)**. **A:** From The Neurosurgical Atlas (http://neurosurgicalatlas.com), reprinted with permission from Aaron A. Cohen-Gadol, M.D., M.Sc. **B and C:** Reprinted with permission from IU Health.

Stimulation of the posterior/inferior capsule will exclude an aberrant posterior/inferior displacement of the facial nerve (Fig. 3C). Coagulation of the posterior capsule is followed by aggressive internal tumor debulking (Fig. 4). Maximal internal debulking will tremendously facilitate the later stages of the operation due to enhanced tumor capsule mobilization from the surrounding cerebrovascular structures, as well as decrease neural tissue stretch/retraction.[24] Following devascularization of the tumor by coagulating the feeders from the dura over the porus acusticus, microdissection continues inferiorly as the tumor capsule is mobilized away from the lower cranial nerves and out of the jugular foramen (Fig. 5A). This stage of microdissection should be atraumatic as the lower cranial nerves do not significantly attach to the capsule. Stimulation of the inferior pole of the tumor should exclude the presence of the facial nerve. Subsequently, the tumor is internally debulked along its superior pole and rostral dissection continues, mobilizing the capsule away from the tentorium and fifth cranial nerve.[29] The facial nerve is often adherent along the superior pole of the tumor and adjacent to the trigeminal nerve. This portion of the capsule should be carefully mapped with the stimulator. Movement of the temporalis muscle caused by trigeminal nerve stimulation should not be mistaken for localization of the facial nerve. Mapping of the often attenuated/atrophied facial nerve in giant tumors can be difficult, especially in the presence of preoperative facial weakness. Repetitive mapping at slightly higher stimulation parameters may be necessary to completely exclude the presence of the facial nerve in the region. The capsule is sharply dissected away from the distal trigeminal nerve (Fig. 5B); the trochlear nerve and superior cerebellar artery are preserved. The superior petrosal vein is protected

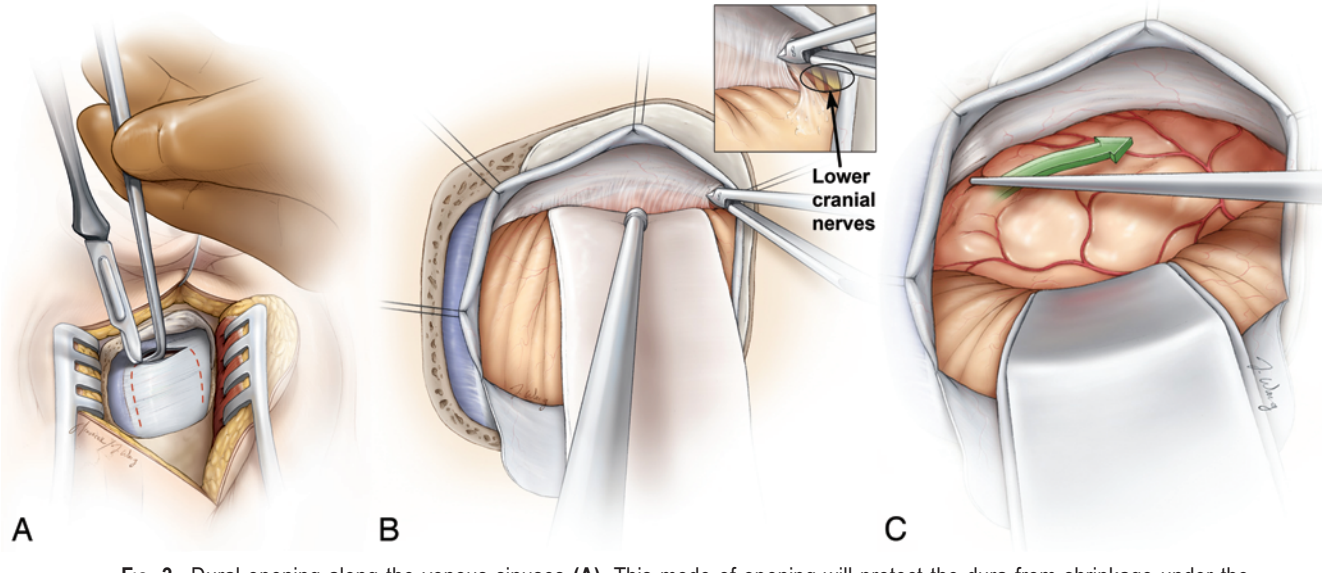

Fig. 3. Dural opening along the venous sinuses **(A)**. This mode of opening will protect the dura from shrinkage under the intense light of the microscope. Cerebrospinal fluid is drained by opening the arachnoid membranes along the inferior pole of the tumor **(B)**. Stimulation of the posterior/inferior capsule will exclude an aberrant posterior/inferior displacement of the facial nerve **(C)**. *Green arrow* indicates direction of the movement of the instrument. From The Neurosurgical Atlas (http://neurosurgicalatlas.com), reprinted with permission from Aaron A. Cohen-Gadol, M.D., M.Sc.

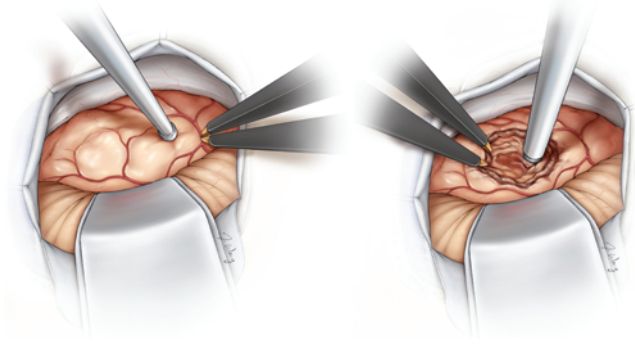

Fig. 4. Coagulation of the posterior capsule *(left)* is followed by aggressive internal tumor debulking *(right)*. From The Neurosurgical Atlas (http://neurosurgicalatlas.com), reprinted with permission from Aaron A. Cohen-Gadol, M.D., M.Sc.

if possible. Venous bleeding from the petrosal-tentorial junction should be carefully controlled.

The tumor capsule is subsequently rolled laterally away from the middle cerebellar peduncle and brainstem toward the porus acousticus. Meticulous hemostasis will allow the operator to appreciate the most important factor in safe resection of these challenging tumors: microdissection along the arachnoid membranes and respecting the brainstem pial membranes. The capsule is pulled on gently as dissection is performed using microforceps to detach the arachnoid membranes from the tumor while periodic irrigation by the assistant clears the field. Suction over the brainstem and cranial nerves is strictly avoided. The veins along the surface of the brainstem are often engorged and prone to avulsion, leading to blood loss and interference with adequate visualization of the dissection planes.[25] Gentle pressure using a cotton ball over the site of the hemorrhage followed by coagulation of the vein along its more proximal segment away from the brainstem is a possible strategy.

Internal debulking using an ultrasonic aspirator followed by tumor mobilization[24] is a safe maneuver to avoid inadvertent injury by undue retraction of the surrounding structures. Further mobilization of the inferior pole of the tumor should protect the posterior inferior cerebellar artery and its branches. En passage vessels are microsurgically mobilized using sharp dissection as these may be crucial vessels,[27] while small tumor-feeding vessels are carefully coagulated and cut. Blunt dissection of the perforators and their subsequent avulsion must be avoided. Cranial nerve VIII is often encountered in the region; its preservation in giant tumors is almost impossible and not advisable if preoperative hearing is nonfunctional.

In the presence of preoperative brainstem edema, violation of the pial membranes is likely in the giant tumors. If such an event occurs, a small piece of cottonoid may be used to mobilize (peel away) the brainstem from the tumor (Fig. 5C) without placing the former at risk for injury by the suction apparatus. Additional cottonoid patties are added and left behind until the end of the operation, when they are irrigated away. The pial membranes may be reidentified along the inferior pole of the tumor. As the tumor is mobilized away from the brainstem along its superior pole and midsection, mapping will localize the facial nerve along the capsule or at its exit zone along the brainstem. The most reliable maneuver to expose the nerve safely is to peel the tumor laterally and identify the nerve along its root exit zone at the brainstem. Removal of the tumor in the deep cerebellopontine cleft may require the most amount of cerebellar retraction; changing the angle of the microscope's view and intermittent dynamic retraction using the suction tip may minimize the required persistent force. The length of the operation, any change in vital signs, or significant violation of the pial membranes or concerning facial nerve recordings may lead the operator to stage the operation.[17]

Localizing the facial nerve along the superior half of the capsule, the surgeon can be aggressive in removal of the inferior pole, which is often not very adherent to the brainstem; the abducent nerve is often adherent to the capsule. Continuing to roll the superior pole laterally, the surgeon will expose the root entry zone of the trigeminal

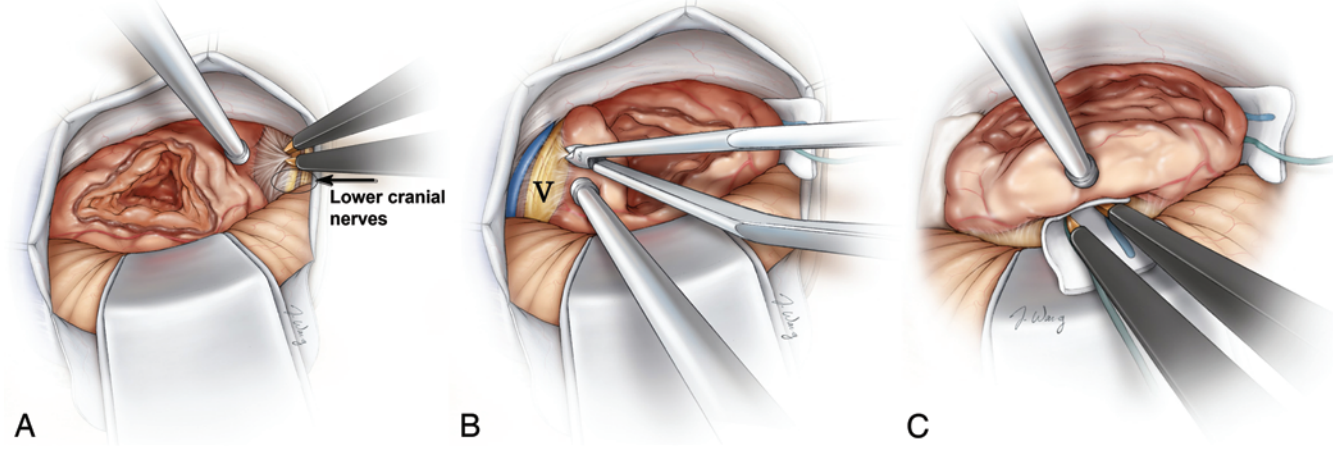

Fig. 5. The arachnoid over the lower cranial nerves is dissected off of the tumor **(A)**. Sharp dissection is used to mobilize the tumor away from the trigeminal nerve (cranial nerve V; **B)**. In the presence of preoperative brainstem edema, violation of the pial membranes is likely. If such an event occurs, a small piece of cottonoid may be used to mobilize (peel away) the brainstem from the tumor without placing the former at risk for injury by the suction apparatus **(C)**. From The Neurosurgical Atlas (http://neurosurgicalatlas.com), reprinted with permission from Aaron A. Cohen-Gadol, M.D., M.Sc.

nerve. This part of the nerve is often very adherent and draped over the tumor, and careful dissection technique will minimize the risk of postoperative trigeminal neuropathy and resultant corneal anesthesia. Attentive internal decompression of the residual tumor will facilitate capsule mobilization using sharp dissection techniques. Using meticulous frequent stimulation, the facial nerve is dissected and peeled away from the tumor; mobilizing the tumor away from the nerve may place the nerve at an increased risk of injury (Fig. 6A–C). Bleeding can be a nuisance, but blind bipolar coagulation should be avoided. At this stage in a cooperative procedure, the neurosurgeon may take a break while the neurotologist removes the tumor within the IAC (Fig. 6D–F).

The inferior wall of the IAC is drilled to the fundus after the dura is dissected from the petrous bone. Cutting and diamond burs may be used to remove the bone under generous irrigation to avoid heat injury to the nerves in the canal; a large piece of soaked soft cotton is used to cover the cerebrovascular structures in the subarachnoid space to protect them from the bone dust during drilling. Extension of bone removal will allow identification of the distal cranial nerve VII/VIII complex free of tumor (Fig. 6E). The mastoid air cells will often be entered and should be carefully waxed at the end of this stage. The dura within the IAC is cut and the tumor is debulked and rolled medially. The vestibular nerve is meticulously identified and the facial nerve is mapped before the vestibular nerve is cut, to allow the tumor to be mobilized out of the canal (Fig. 6F). The facial nerve becomes very attenuated and adherent to the tumor along the junction of its subarachnoid and intracanalicular segments. The operator has to use a careful combination of gentle blunt and sharp microdissection techniques during tumor mobilization at the level of the porus to avoid facial nerve injury.

Removal of the tumor within the IAC will convey additional information regarding the route of the facial nerve over the capsule from its already identified root exit zone to the area of the porus acousticus. If the facial nerve is significantly attenuated, its anatomical preservation may not be possible in giant tumors.[15] Although gross-total tumor resection is attempted, if the tumor is very adherent to the nerve at the level of the porus, a small piece of the tumor may be left behind to optimize facial function. This small piece of the tumor left over the nerve can be managed postoperatively through surveillance imaging and treated with radiosurgery (if enlarging) with good rates of tumor control.[16] The surgeon may use the blunt-tipped stimulator as an instrument to peel away the nerve from the capsule. Significant tension on the nerve is avoided and sharp dissection is used (Fig. 7). Any injury to the nerve may require an increase in stimulation parameters to map the nerve. The basilar artery is evident at the end of the resection. Basilar artery perforators should be preserved. Meticulous hemostasis is followed by a watertight dural closure and generous application of bone wax to the mastoid air cells. If cerebellar swelling is evident, the bone flap should not be replaced and a generous suboccipital decompressive craniectomy is performed. Figure 8 illustrates the typical patterns of facial nerve displacement by large and giant VSs.

Postoperative Considerations

Patients are observed in the intensive care unit for

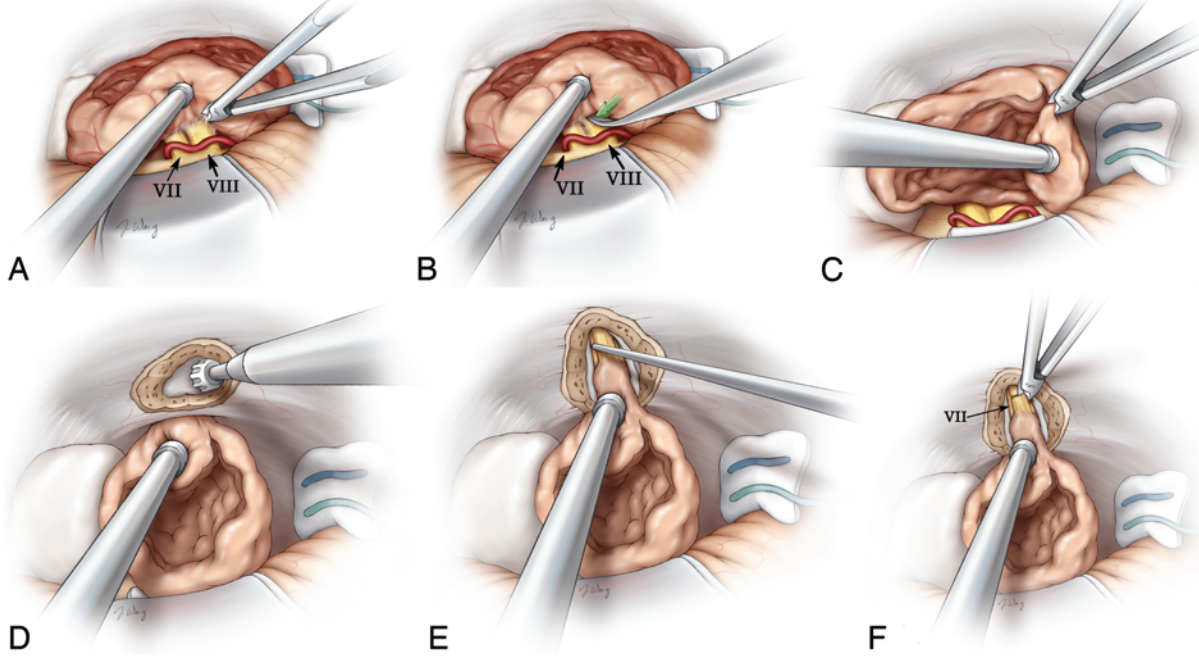

Fig. 6. Using meticulous frequent stimulation, the facial nerve is dissected and peeled away from the tumor (mobilizing the tumor away from the nerve may place the nerve at an increased risk of injury), and further tumor resection is continued **(A–C)**. At this stage, the surgeon may take a break while the otolaryngologist removes the tumor within the IAC **(D–F)**. VII = cranial nerve VII; VIII = cranial nerve VIII. *Green arrow* indicates direction of the movement of the instrument. From The Neurosurgical Atlas (http://neurosurgicalatlas.com), reprinted with permission from Aaron A. Cohen-Gadol, M.D., M.Sc.

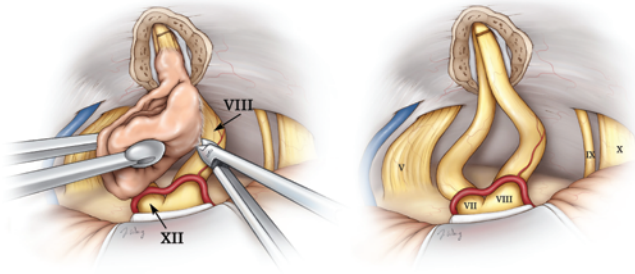

Fig. 7. The surgeon may use the blunt-tipped stimulator as an instrument to peel away the nerve from the capsule. Significant tension on the nerve is avoided and sharp dissection is used. IX, X, XII = cranial nerves IX, X, and XII, respectively. From The Neurosurgical Atlas (http://neurosurgicalatlas.com), reprinted with permission from Aaron A. Cohen-Gadol, M.D., M.Sc.

any signs of neurological deterioration, sudden hypertension, or breathing and swallowing difficulty. Meticulous postoperative care is mandatory to detect and prevent complications.[22] Appropriate eye care is provided: the combination of trigeminal neuropathy and facial nerve paralysis places the eye at risk and appropriate precautions are taken in the presence of one or both. If dysphagia or respiratory insufficiency is encountered, early percutaneous gastrostomy and tracheostomy tubes are placed to avoid any complications such as aspiration pneumonia and hypoxemia; the possibility of these is important to discuss with the patient preoperatively.

If a staged operation is planned, the interval between operations may be 2–4 weeks based on the patient's recovery process from the first operation.[19] Others have advocated for a longer interval to allow further cranial nerve and brainstem recovery.[17] If poor facial function is present from the first operation, the second stage is delayed until the nerve achieves a good functional recovery.

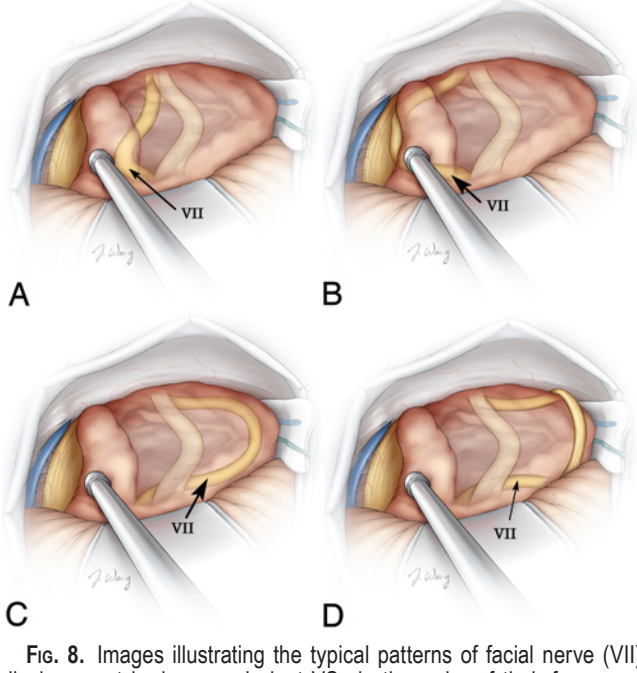

Fig. 8. Images illustrating the typical patterns of facial nerve (VII) displacement by large and giant VSs in the order of their frequency (**A–D**, most to least frequent), based on the experience of the senior author. Note that the location of the cochlear nerve (cranial nerve VIII) is variable and not known in the larger tumors due to its significant attenuation. From The Neurosurgical Atlas (http://neurosurgicalatlas.com), reprinted with permission from Aaron A. Cohen-Gadol, M.D., M.Sc.

Complication Management

The most feared complication with this operation is intraparenchymal hemorrhage and cerebellar edema, which contributes the main source of perioperative morbidity.[3,22] Avoidance of significant retraction on the nervous structures intraoperatively, meticulous microdissection along the arachnoid membranes, and prompt management of postoperative hypertension will minimize these unfortunate events. Watertight dural closure is important to avoid pseudomeningocele formation. If rhinorrhea is encountered, we perform early mastoidectomy and obliterate the air cells with a fat graft.

The main neurological morbidity of VS surgery is facial nerve palsy, which is both functionally and psychologically damaging to the patient. This is especially true with giant tumors, as tumor size is the main factor predicting postoperative facial weakness.[7] Patients with an incomplete eye closure may undergo gold weight placement within the eyelid and/or tarsorrhaphy. Facial reanimation procedures may be considered if facial nerve function does not return within 1 year postoperatively.[21] This maneuver may be considered earlier if the nerve was anatomically noncontinuous at the time of surgery.

The goal of the surgery remains gross-total resection of the tumor and preservation of function. Meticulous microsurgical techniques remain the important factor in operative success.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: both authors. Acquisition of data: both authors. Analysis and interpretation of data: both authors. Drafting the article: both authors. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors. Study supervision: Cohen-Gadol.

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Facial nerve outcomes after surgery for large vestibular schwannomas: do surgical approach and extent of resection matter?

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Object. The object of this study was to evaluate facial nerve outcomes in the surgical treatment of large vestibular schwannomas (VSs; ≥ 2.5 cm maximal or extrameatal cerebellopontine angle diameter) based on both the operative approach and extent of tumor resection.

Methods. A PubMed search was conducted of English language studies on the treatment of large VSs published from 1985 to 2011. Studies were then evaluated and included if they contained data regarding the size of the tumor, surgical approach, extent of resection, and postoperative facial nerve function.

Results. Of the 536 studies initially screened, 59 full-text articles were assessed for eligibility, and 30 studies were included for analysis. A total of 1688 tumor resections were reported. Surgical approach was reported in 1390 patients and was significantly associated with facial nerve outcome ($\phi = 0.29$, $p < 0.0001$). Good facial nerve outcomes (House-Brackmann Grade I or II) were produced in 62.5% of the 555 translabyrinthine approaches, 65.2% of the 601 retrosigmoid approaches, and 27.4% of the 234 extended translabyrinthine approaches. Facial nerve outcomes from translabyrinthine and retrosigmoid approaches were not significantly different from each other, but both showed significantly more good facial nerve outcomes, compared with the extended translabyrinthine approach (OR for translabyrinthine vs extended translabyrinthine = 4.43, 95% CI 3.17–6.19, $p < 0.0001$; OR for retrosigmoid vs extended translabyrinthine = 4.98, 95% CI 3.57–6.95, $p < 0.0001$). There were 471 patients for whom extent of resection was reported. There was a strong and significant association between degree of resection and outcome ($\phi = 0.38$, $p < 0.0001$). Of the 80 patients receiving subtotal resections, 92.5% had good facial nerve outcomes, compared with 74.6% (n = 55) and 47.3% (n = 336) of those who received near-total resections and gross-total resections, respectively. In the 2-way comparison of good versus suboptimal/poor outcomes (House-Brackmann Grade III–VI), subtotal resection was significantly better than near-total resection (OR = 4.21, 95% CI 1.50–11.79; $p = 0.004$), and near-total resection was significantly better than gross-total resection (OR = 3.26, 95% CI 1.71–6.20; $p = 0.0002$) in producing better facial nerve outcomes.

Conclusions. In a pooled patient population from studies evaluating the treatment of large VSs, subtotal and near-total resections were shown to produce better facial nerve outcomes when compared with gross-total resections. The translabyrinthine and retrosigmoid surgical approaches are likely to result in similar rates of good facial nerve outcomes. Both of these approaches show better facial nerve outcomes when compared with the extended translabyrinthine approach, which is typically reserved for especially large tumors. The reported literature on treatment of large VSs is extremely heterogeneous and minimal consistency in reporting outcomes was observed.
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Key Words • **vestibular schwannoma** • **facial nerve outcome** • **subtotal resection** • **translabyrinthine approach** • **retrosigmoid approach** • **near-total resection**

In the era of modern microsurgery, surgical outcomes for the removal of VSs have markedly improved.[14,24,35] Despite advancements in facial nerve monitoring and surgical techniques, functional preservation of the facial nerve in surgery for larger tumors remains a challenge.[4,36] For small and medium-sized tumors, long-term facial nerve preservation rates are reported to be more than 90%, but this rate is substantially lower for large tumors.[20,29]

Series of large VSs frequently have been published as distinct clinical entities, because large tumors present a greater challenge to surgeons regarding total removal, cranial nerve preservation, and other postoperative complications.[32,49] Due to the paucity of data for such large tumors, a systematic literature review of all the available reports would be greatly beneficial in determining opti-

Abbreviations used in this paper: GTR = gross-total resection; NTR = near-total resection; STR = subtotal resection; VS = vestibular schwannoma.

mal treatment strategies. What renders this task nearly insurmountable is the lack of reporting standardization for VS outcomes. In these series, except for the House-Brackmann grading system of facial nerve function, there appears to be no agreement on what is the minimum size of a "large" tumor, measurement of tumor size, and degree of resection.[23] Instead of enumerating the inconsistencies in the reported literature, we will define each variable we measured in the *Methods* section and then indicate specific issues with each included paper in the *Results* and *Discussion* sections.

The ideal therapeutic goal in VS surgery is complete removal of the tumor in a single stage with complete preservation of all cranial nerve function. However, for larger tumors, this has remained an elusive goal. For this reason, many studies have advocated partial resections, 2-stage resections, and combined partial resection with radiation therapy.[3,17,40,42] Moreover, questions remain regarding the surgical approach that provides the optimal facial nerve outcome.

In this study, we systematically reviewed studies reporting postoperative outcomes of large VSs (≥ 2.5 cm of maximal or extrameatal diameter), with special attention to the facial nerve outcomes as a function of surgical approach and degree of tumor resection.

Methods

Search Criteria

Following an Institutional Review Board exemption, we conducted a systematic review using a PubMed search of the English language literature from 1985 to 2011. The date range was chosen to represent the era of more routine use of facial nerve monitoring in VS surgery. The search terms "large acoustic neuroma," "large vestibular schwannoma," "acoustic neuroma surgical resection," "acoustic neuromas retrosigmoid," "acoustic neuromas translabyrinthine," "subtotal resection acoustic neuroma," "vestibular schwannoma," and "acoustic neuroma" were used to identify appropriate papers. Figure 1 provides a flowchart of the number of papers identified, screened, and included in the study. The 59 full-text articles assessed for eligibility were all screened independently by 3 of the authors (R.K.G., S.D., and A.M.).

Studies were screened for data on large VSs defined by a greatest extrameatal diameter of at least 2.5 cm according to the Kanzaki standard, largest diameter, or a Koos classification ≥ 4.[28] We did not exclude papers if they did not measure extrameatal diameter only. The majority of included papers (22/30, 73%) reported extrameatal measurement, while 17% reported sizes at least 2.5 cm in largest dimension (in all but 1 of these studies, the smallest tumor was 3.0 cm in longest dimension), 7% of papers used the Koos classification, and 1 paper did not state how measurements were obtained, although tumors in this paper were still reported as > 2.5 cm.

Studies were included if facial nerve data along with surgical approach and/or extent of resection were provided. Extent of resection as GTR, NTR, or STR was determined as defined by each author. Facial nerve outcomes were determined by the House-Brackmann grading scale.[23] Any anatomically disrupted facial nerves were given a VI/VI grade, even if a subsequent hypoglossal-facial or primary anastomosis of the nerve provided improved facial nerve outcome. Papers that reported surgery without continuous intraoperative facial nerve monitoring were excluded. If papers clearly identified neurofibromatosis Type 2 patients and their outcomes, those patients were excluded from analysis because of the more aggressive nature and neural invasiveness of the disease.[39]

Assessment of Study Quality

After inclusion, each paper was given a relative value score by assigning points for quality of the paper. The grading system included 1 or 0 points for the respective presence or absence of the following: average tumor size reported, inclusion of neurofibromatosis Type 2 patients reported, tumor measurement of largest extrameatal diameter specifically mentioned, facial nerve function reported as function of approach, facial nerve function reported as function of degree of resection, degree of resection, postoperative MRI correlation of extent of resection reported, and postoperative follow-up of at least 12 months. Degree of cranial nerve function was used to measure treatment outcome. Outcome levels of good (House-Brackmann Grade I or II), suboptimal (House-Brackmann Grade III or IV), and poor (House-Brackmann Grade V or VI) were compared.

Surgical Approach and Extent of Resection

Treatment outcomes for the 3 surgical approaches (translabyrinthine, retrosigmoid, and extended translabyrinthine) and 3 degrees of resection (STR, NTR, and GTR) were compared across all studies by summing the number of patients with good, suboptimal, and poor facial nerve outcomes, and using a 3 × 3 chi-square to evaluate the association of treatment outcome with surgical approach and degree of resection. If these analyses were significant, each pair of treatments was compared in a 2 × 2 chi-square examining good versus suboptimal/poor facial nerve outcomes.

The extended translabyrinthine approach is defined as any standard translabyrinthine approach that was modified to provide greater access for extremely large tumors. These modifications included the translabyrinthine-transapical exposure described by Angeli et al.[2] in which the IAC is opened with greater than 300° of exposure, or the combined translabyrinthine-retrosigmoid exposure as described by Anderson et al.[1]

The definition of what constitutes an NTR compared with an STR varies by author, with no universally agreed-upon definition. Some authors have subjectively defined the residual tumor with words such as "minimal," "tiny," "small" amount, or "thin layer" of residual tumor, as defined by the operative surgeon.[33,43,47] Bloch et al.[3] defined an NTR as 25 mm^2 or a 2-mm-thick pad of residual tumor, and an STR as anything less than an NTR. Haque et al.[22] defined an STR as when > 90% of the tumor was removed and GTR as when the entire tumor was microscopically removed. For studies that reported extent of resection, the

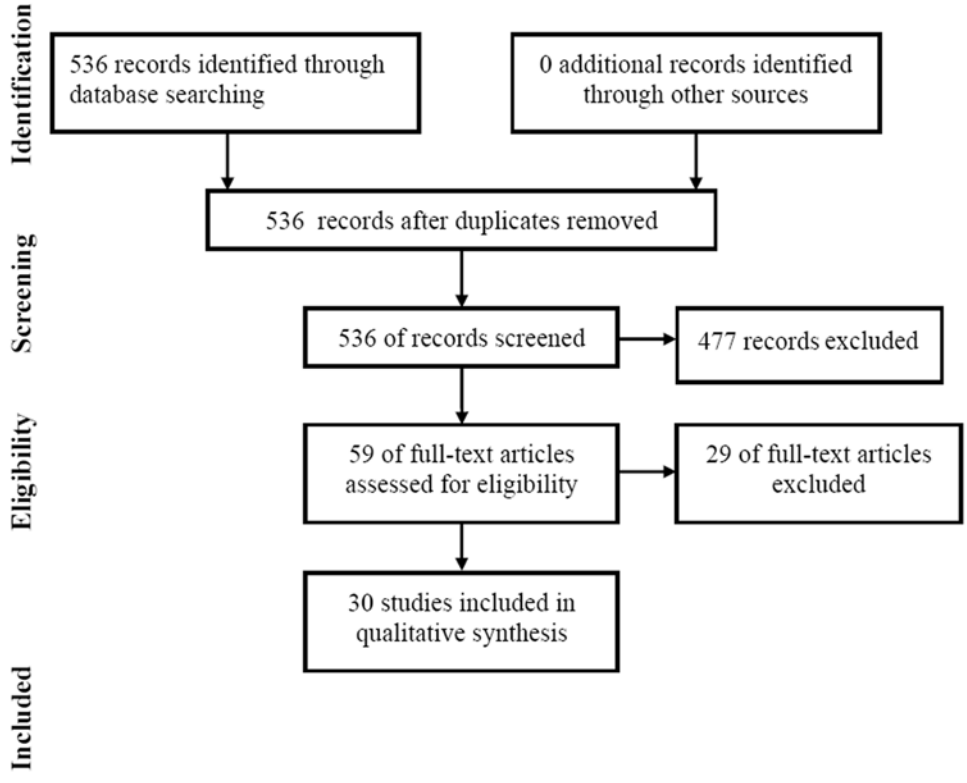

Fig. 1. This flowchart illustrates how many articles were initially identified, screened, and included in the study analysis.

most commonly used definition in this systematic review was that an STR represented > 5% of residual tumor, and an NTR was any residual tumor ≤ 5%.[2,21,31,57]

Statistical Analysis

The percentage of surgeries with good facial nerve outcome (House-Brackmann Grade I or II) was examined across studies. Univariate Pearson correlations were examined across studies, between percentage of functional outcomes, and other study characteristics including date of publication, number of tumors, size cutoff used to define large tumors, and average patient age. Multiple regression analysis was used to test multivariate models predicting functional outcomes, to try to determine whether degree of resection or type of approach had an independent association with outcome, after controlling for other study characteristics.

Statistics were calculated using SAS version 9.2 (SAS Institute, Inc.) and graphs were created using Microsoft Excel.

Results

Of the 536 papers initially identified, 30 fit our criteria, all of which were retrospective case series.[1,2,4,7,8,12,13,18,21,25,27,30–34,37,38,41,43–46,48,50–52,54,55,57] Table 1 summarizes the characteristics of each paper. The retrospective nature of the papers and heterogeneity in reporting tumor size, degree of tumor resection, indications for degree of resection, length of follow-up, and reporting facial nerve outcome for individual groups (degree of resection and surgical approach) precluded a true statistical meta-analysis.

Tumor Size

One thousand six hundred and eighty-eight tumors measuring at least 2.5 cm were reported. The average size of the tumor was 3.9 cm in the 40% of papers that reported an average size. In regard to measuring the dimensions of the tumor, 21 studies reported the longest measurement in the cerebellopontine angle excluding the portion of the tumor in the internal auditory canal, 6 reported the absolute longest dimension, 2 used the Koos classification, and 1 made no mention of measurement criteria.

Surgical Approach

Of the 1636 cases for which a surgical approach was mentioned, 729 underwent translabyrinthine, 644 retrosigmoid, and 263 extended translabyrinthine tumor resection. Ten papers provided a definition for what consisted of less-than-total resection of tumor, and only 4 of those used the Kanzaki standards.[28,45] Two studies contained only patients who underwent less-than-total resection.

Extent of Resection

We were able to decipher the approximate degree of resection for 1158 patients, of whom 938 underwent GTR, 102 NTR, and 147 STR. Only 2 papers correlated their surgical degree of resection with postoperative MR images. Twenty-one papers reported facial nerve outcome after 12 months of follow-up. Fourteen papers reported facial nerve outcome as a function of degree of resection, and 471 patients' treatments were examined. Of these pa-

TABLE 1: Summary of the studies used in the review*

Authors & Year	No. of Pts	Mean Age (yrs)	VS			Surgical Approach			Degree of Resection			Determination of Facial Nerve Outcome	FU (mos)
			Size Cutoff (cm)	Mean Size (cm)	Measurement	TL	RS	EX	GTR	STR	NTR		
Godefroy et al., 2009	50	49.0	2.6	3.2	extrameatal	50	0	0	13	8	29	"post-operative"	mean 29, range 15–51
Roland et al., 2004	56	53	3.0	3.7	larger of either extrameatal only or extrameatal & up to 10 mm of IAC	46	10	0	35	6	5	most recent FU	at least 12, range 17–120
Samii et al., 2010	36	42.1	4.0	4.4	extrameatal	0	36	0	36	0	0	postop	mean 34, range 5–62
Anderson et al., 2005	71	44.7	3.0	3.8	extrameatal	25	22	24	68	3	0	6 mos postop	6
Angeli et al., 2011	88	42.5	4.0	NA	extrameatal	0	0	88	NA	NA	NA	1-yr FU	at least 12 in 88 pts
Charpiot et al., 2010	123	46.6	4.0	NA	extrameatal	123	0	0	119	4	0	1-yr FU	at least 12
Iwai et al., 2003	14	47	3.0	NA	extrameatal	0	13	1	0	14	0	at least 12 mos	at least 12, range 12–72
Chen et al., 2009	85	NA	2.5	NA	extrameatal	0	85	0	83	2	0	1-yr FU	at least 12
Fuentes et al., 2008	8	53.0	3.5	3.9	extrameatal	0	8	0	0	8	0	at least 12 mos	at least 12, range 12–73
Lanman et al., 1999	95	46.1	3.0	3.6	extrameatal	95	0	0	NA	NA	NA	1-yr FU	at least 12
Darrouzet et al., 2004	152	NA	NA	NA	Koos	116	8	28	148	4	2	postop	65.2
Nadol et al., 1992	8	NA	2.5	NA	extrameatal	0	8	0	8	0	0	at least 12 mos	at least 12
Jung et al., 2000	30	45.2	4	4.92	extrameatal	0	30	0	22	8	0	at least 12 mos	at least 12
Mamikoglu et al., 2002	81	47	3	3.7	extrameatal	81	0	0	77	4	0	at least 12 mos	at least 12
Raftopoulos et al., 2005	13	49	3	NA	extrameatal	1	15	0	10	2	1	at least 12 mos	at least 12
Sanna et al., 2004	122	48	3	3.5	extrameatal	0	0	122	NA	NA	NA	at least 12 mos	NA
Torrens et al., 1994	29	54.8	2.5	3.46	extrameatal	0	32	0	20	NA	9	at least 12 mos	12–48
Wu & Sterkers, 2000	40	24.8	3	4.25	extrameatal	40	0	0	39	1	0	1–3 mos	NA
Yamakami et al., 2004	50	52	3	4.1	extrameatal	0	50	0	43	7	0	at least 12 mos	58, range 12–115
Brackmann et al., 2007	141	NA	2.5	NA	NA	141	0	0	NA	NA	NA	at least 12 mos	at least 12
Lalwani et al., 1994	24	NA	3	NA	extrameatal	11	13	NA	NA	NA	NA	at least 12 mos	at least 12
Zhao et al., 2010	89	48.2	4	NA	extrameatal	0	89	0	38	16	35	at least 12 mos	at least 12
Silva et al., 2012	29	NA	4	NA	largest dimension	0	29	0	29	0	0	at least 9 mos	at least 9
van de Langenberg et al., 2011	42	52	3	NA	Koos	NA	NA	0	0	42	0	at least 12 mos	at least 12
Darwish et al., 2005	32	NA	3	NA	largest dimension	0	32	0	32	0	0	at least 12 mos	at least 12
Misra et al., 2009	100	NA	3	NA	largest dimension	0	100	0	86	0	14	at least 2 mos	NA
Lee et al., 2002	36	NA	3	NA	largest dimension	0	36	0	11	18	7	at least 3 mos	at least 3, range 3–58
Veronezi et al., 2008	5	NA	4	NA	extrameatal	0	5	0	5	0	0	at least 18 mos	at least 18
Kirkpatrick et al., 1993	16	NA	2.5	NA	largest dimension	NA	NA	NA	16	0	0	at least 6 mos	at least 6
Uziel et al., 1993	23	NA	3	NA	extrameatal	0	23	0	NA	NA	NA	at least 12 mos	at least 12

* EX = extended translabyrinthine; FU = follow-up; IAC = internal auditory canal; NA = not applicable (not performed or not reported); Pts = patients; RS = retrosigmoid; TL = translabyrinthine.

tients, 274 (58%) had good facial nerve outcomes (House-Brackmann Grade I or II), 104 (22%) were suboptimal (House-Brackmann Grade III or IV), and 93 (20%) were poor (House-Brackmann Grade V or VI). There was a strong and significant association between degree of resection and outcome ($\phi = 0.38$, $p < 0.0001$). Of the 80 patients receiving STR, 92.5% had good facial nerve outcomes, compared with 74.6% (n = 55) and 47.3% (n = 336) of those who received NTR and GTR, respectively (Fig. 2). In the 2-way comparison of good versus suboptimal/poor outcomes (House-Brackmann Grade III–VI), STR was significantly better than NTR (OR = 4.21, 95% CI 1.50–11.79; p = 0.004), and NTR was significantly better than GTR (OR = 3.26, 95% CI 1.71–6.20; p = 0.0002) in producing better facial nerve outcomes. A Forrest plot was created to display the effect sizes for facial nerve outcomes for all studies reporting on degree of resection (Fig. 3).

Facial Nerve Outcome

We were able to identify the facial nerve outcome as the function of surgical approach in 27 of the papers. Studies reported outcomes for 1390 patients, 803 (58%) with good facial nerve outcomes, 365 suboptimal (26%), and 222 (16%) with poor outcomes. Surgical approach was significantly associated with outcome ($\phi = 0.29$, $p < 0.0001$). Good facial nerve outcomes were produced in 62.5% of the 555 translabyrinthine approaches, 65.2% of the 601 treatments using the retrosigmoid approach, and 27.4% of the 234 treatments using the extended translabyrinthine approach (Fig. 4). Facial nerve outcomes from translabyrinthine and retrosigmoid approaches were not significantly different from each other, but both produced significantly more good outcomes compared with the extended translabyrinthine approach (OR for translabyrinthine vs extended translabyrinthine approach = 4.43, 95% CI 3.17–6.19, $p < 0.0001$; OR for retrosigmoid vs extended translabyrinthine approach = 4.98, 95% CI 3.57–6.95, $p < 0.0001$).

The mean percentage of surgeries with good facial nerve outcomes was 0.61 ± 0.24 (95% CI 0.52–0.70). In univariate analysis, patient age, year published, and number of tumors were not significantly related to percentage of functional outcomes, but size cutoff used to define large

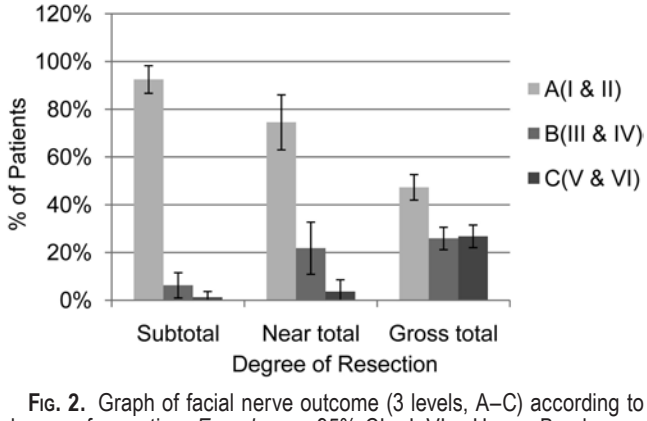

Fig. 2. Graph of facial nerve outcome (3 levels, A–C) according to degree of resection. *Error bars* = 95% CIs. I–VI = House-Brackmann grades.

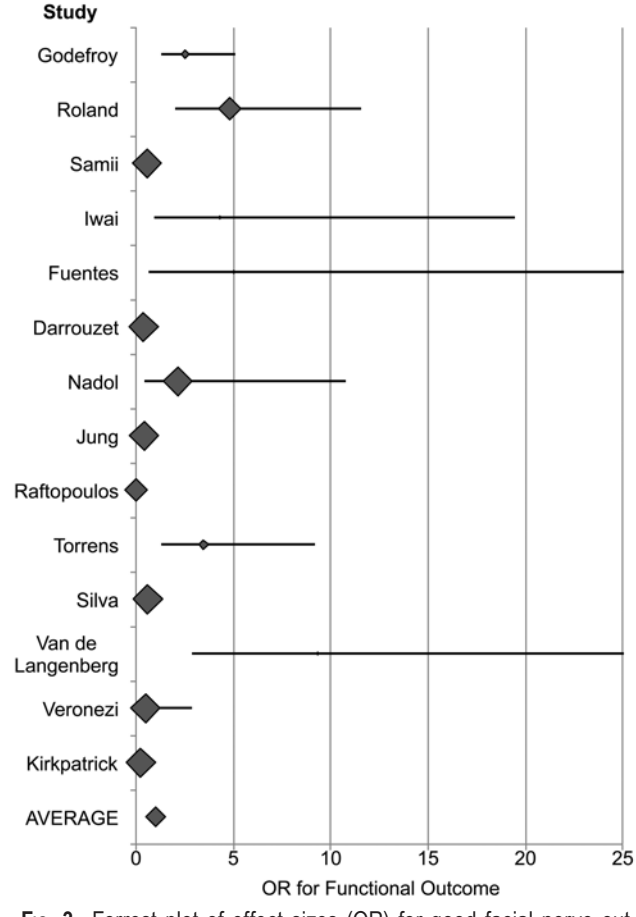

Fig. 3. Forrest plot of effect sizes (OR) for good facial nerve outcomes (House-Brackmann Grade I or II) for all studies reporting on degree of resection. *Horizontal lines* = 95% CIs. *Vertical dashed line* = average effect size. Size of *diamond* is proportional to the percentage of resections that were GTR (*larger diamond* indicates more GTRs, *smaller diamond* indicates more NTRs or STRs).

tumors was significantly associated ($r = -0.43$, $p < 0.02$). This indicated that studies with larger size cutoffs tended to have fewer patients with good facial nerve outcomes. Multiple regression models were tested with predictors including size cutoff, number of treatments using translabyrinthine, retrosigmoid, or extended translabyrinthine approaches, and number using GTR, NTR, and STR. We found a model that was significant ($p < 0.001$) with moderate prediction accuracy ($R^2 = 0.48$), which included the predictors size cutoff, number of subtotal resections, and number of surgeries using the extended translabyrinthine approach. In this model, after accounting for other variables in the model, size cutoff and extended translabyrinthine approach were negatively associated with functional outcomes (p = 0.02 and 0.03, respectively), while STR was positively associated with functional outcomes (p = 0.01). Thus, each of these 3 variables independently predicted the percentage of good facial nerve outcomes across studies.

Discussion

The goal in managing VSs is to control tumor growth

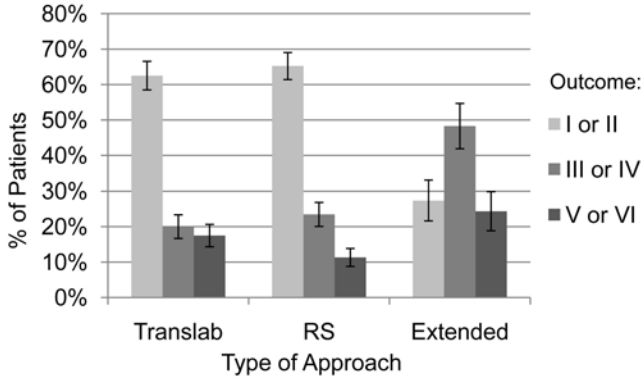

Fig. 4. Graph of facial nerve outcome (3 levels) according to surgical approach. RS = retrosigmoid; Translab = translabyrinthine.

while preserving neurological function. Large VSs pose a particular challenge in attaining this goal. Many reports have cited the high rate of poor facial nerve outcomes following microsurgical resection of VSs.[5,27,32,34,44,53,56] In these studies, facial nerve outcomes of House-Brackmann Grade I or II were reported in only 27%–58% of patients with VSs ≥ 3.0 cm following GTR. Facial nerve preservation is particularly difficult in large tumors because as the tumor slowly enlarges, the facial nerve becomes stretched and often "ribbons" over the surface of the tumor.[3] Moreover, the tumor-arachnoid dissection plane is often obscured as the tumor enlarges.[9] This is particularly true just medial to the porous acusticus, where a pressure "bottleneck" can occur at the entrance of the bony internal auditory canal.

Surgical Approach

The findings of this study suggest that there is no statistically significant difference in facial nerve outcomes between the translabyrinthine and retrosigmoid approaches. However, there was a difference between the extended translabyrinthine and either the translabyrinthine or retrosigmoid approach. As described by Sanna et al.,[45] the extended translabyrinthine approach is a modification of the translabyrinthine approach, which allows for greater exposure for especially large VSs. The finding that the extended translabyrinthine approach resulted in poor facial nerve outcomes is likely due to a selection bias of patients with particularly large VSs, and possibly recurrent tumors operated on at that institution, thus warranting the extended approach.

Extent of Resection

The goal of microsurgical VS resection has traditionally been microscropic GTR. In the era of high-quality imaging modalities and treatment modalities, such as stereotactic radiation with excellent tumor control rates, is total resection warranted for a benign tumor when the facial nerve would be put at high risk for postoperative dysfunction? Answering this question has led to the concept of partial resections of VSs if GTR cannot be achieved without injuring the facial nerve.

The controversy over partial resection dates back to the 2 surgeons who truly revolutionized the art of acoustic neuroma resection, Harvey Cushing and Walter Dandy. Cushing advocated a partial resection, which was vehemently opposed by Dandy, considering the high mortality rate of recurrent tumors.[10,11,15] William House reported in 1968[24] that partial removal was a reasonable alternative to total removal when intraoperative vital signs were labile, or for "the elderly patient, or the patient who is poor surgical risk."

The results of this study show that STR of large VSs results in improved facial nerve outcomes when compared with NTR and GTR. Facial nerve outcomes of NTR are also improved when compared with results of GTR. This result is not surprising given that partial resections likely cause less surgical trauma to the nerve compared with GTRs.

We were unable to evaluate rates of recurrence in the present study due to lack of data. Only 4 papers made any comments on recurrence and most did not have sufficiently long follow-up. However, in general VS literature, there are considerable data to suggest that the rate of recurrence is correlated with the amount of residual tumor following resection. El-Kashlan et al.[16] reported that 43.6% of their 39 patients showed signs of VS regrowth following STR and NTR after a mean follow-up period of 6.2 years (range 3.5–10.2 years). No patient in their series, however, experienced regrowth following tumor resection greater than 98% (n = 8). Residual postoperative tumor thickness of 10.9 ± 4.1 mm versus 5.7 ± 3.0 mm has been shown to be a risk factor for regrowth ($p < 0.001$).[19] Carlson et al.[6] reported that in 350 patients with VSs treated with microsurgical resection, patients receiving STR were more than 9 times more likely to experience recurrence compared with those undergoing NTR or GTR ($p < 0.001$). Whereas GTR is optimal to prevent tumor recurrence, NTR appears to provide similarly low recurrence rates, likely due to a lack of adequate blood supply or critical tumor mass following NTR to allow subsequent growth.[3]

The major limitations of this study include the retrospective and often uncontrolled nature of the studies included. Many surgeons with high volumes of patients only used 1 approach, which makes clear comparison between surgical approaches difficult. Also, certain centers would only perform GTR or STR. The decision as to degree of tumor resection largely depends on the surgeon and intraoperative findings, which is an inherent bias of the literature: the more difficult, aggressive, and adherent tumors probably were resected in a less-than-total fashion. Studies also often included heterogeneous populations; some reported on patients with preoperative facial palsy or included patients with neurofibromatosis Type 2 and those with unilateral, sporadic VS in the same patient population. Neurofibromatosis Type 2 tumors are more infiltrative of the facial nerve than unilateral, sporadic VSs, and will therefore be more likely to cause facial palsy following GTR.[39]

The results of this current review should be viewed considering the limitations of the available literature. As this study sought to pool similar data from many studies, it was imperative that the patients be similar. Despite our best efforts to compare similar populations, there were limitations due to variability in reporting tumor size,

facial nerve outcome, degree of resection, and length of follow-up. In 2003, Kanzaki et al.[28] reported on minimal standards for describing outcomes in VS surgery. Unfortunately, these standards have yet to be universally implemented, but would aid in comparing data for studies such as this.[26] We recommend creating more strict reporting guidelines for surgeons interested in publishing their series of VS resections, similar to the House-Brackmann facial nerve outcome. Also, a prospective, multicenter study would limit some of the biases affecting the literature and is currently underway (http://clinicaltrials.gov/ct2/show/NCT01129687).

Conclusions

The surgical treatment of large VSs often leads to suboptimal and poor facial nerve outcomes. Taking into account the inherent biases in the literature, STR and NTR appear to produce improved facial nerve outcomes when compared with GTR. The retrosigmoid and translabyrinthine surgical approaches both provide similar rates of facial nerve outcomes. Both of these approaches have better facial nerve outcomes when compared with the extended translabyrinthine approach, which is typically reserved for especially large tumors. The reported literature on treatment of large VSs is extremely heterogeneous, and minimal consistency in reporting outcomes was observed.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Monfared, Gurgel. Acquisition of data: Monfared, Gurgel, Dogru. Analysis and interpretation of data: Monfared, Gurgel, Dogru. Drafting the article: Monfared, Gurgel, Dogru. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Monfared. Statistical analysis: Amdur. Administrative/technical/material support: Monfared, Gurgel, Dogru. Study supervision: Monfared.

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Fascial sling technique for dural reconstruction after translabyrinthine resection of acoustic neuroma: technical note

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Reconstruction of presigmoid dural defects after resection of acoustic neuromas via the translabyrinthine approach is paramount to prevent postoperative CSF leakage. However, primary dural reapproximation and achieving a watertight closure of the dural defect in this anatomical region are quite difficult. Standard closure techniques after the translabyrinthine approach often involve packing an abdominal fat graft that plugs the dural defect and mastoidectomy cavity. This technique, however, may pose the risk of direct compression of the fat graft on the facial nerve and brainstem. Nonetheless, even with the evolution in dural repair techniques, postoperative CSF leaks can still occur and provide a route for infection and meningitis. In this report, the authors describe a novel dural "sling" reconstruction technique using autologous fascia lata to repair presigmoid dural defects created after translabyrinthine resection of acoustic neuromas. The fascia lata is sewn to the edges of the presigmoid dural defect to create a sling to suspend the fat graft within the mastoidectomy defect. A titanium mesh plate embedded in porous polyethylene is secured over the mastoidectomy defect to apply pressure to the fat graft. In the authors' experience, this has been a successful technique for dural reconstruction after translabyrinthine removal of acoustic neuromas to prevent postoperative CSF leakage. There were no cases of CSF leakage in the first 8 patients treated using this technique. The operative details and preliminary results of this technique are presented.
(http://thejns.org/doi/abs/10.3171/2012.6.FOCUS12168)

Key Words • **dural sling reconstruction** • **autologous fascia** • **cerebrospinal fluid leak** • **translabyrinthine approach** • **petrosectomy** • **acoustic neuroma** • **skull base surgery**

The translabyrinthine approach to the cerebellopontine angle is a versatile cranial base approach for the removal of both small and large acoustic neuromas.[14,16,22] This approach is generally preferred for removal of all acoustic tumors, resulting in nonserviceable hearing, and also for large tumors (> 3 cm) when the likelihood of hearing preservation is low.[4] By using the presigmoid corridor, this approach offers the advantage of virtually no cerebellar retraction as well as early identification of the facial nerve at the distal IAC. First described in 1904 by Panse,[21] the translabyrinthine approach was not fully developed and standardized until 1964 by House and Hitselberger.[13] Translabyrinthine resection of acoustic neuromas requires a presigmoid dural opening that extends over the IAC and results in a skull base dural defect, which has historically been difficult to reconstruct.[23] These surgeries are often complicated by postoperative CSF leaks and fistula formation, which increases the incidence of development of other complications such as wound infection and meningitis.[5,27,30] Postoperative CSF leak rates for all acoustic neuroma surgery have been reported to range between 0% and 30% in the literature, but the mean leak rate remains approximately 10%.[1,8–10,15,17,26,27] The postoperative CSF leak rates after translabyrinthine removal of acoustic neuromas range from 0% to 13% as reported in the most recent literature.[3,6,7,11,18,19,29] Although appropriate medical and surgical management of CSF leaks is generally feasible, the ultimate goal is directed at prevention of postoperative CSF leakage. Achieving a watertight dural closure of

Abbreviation used in this paper: IAC = internal auditory canal.

these presigmoid dural defects after translabyrinthine resection poses a significant challenge to the acoustic neuroma surgeon. A meticulous reconstruction and dural closure technique is required for the prevention of CSF leak formation.

Traditionally, the standard closure technique when using the translabyrinthine approach involves an abdominal fat graft that is packed through the dural defect and mastoidectomy cavity.[10,31] In this technique, the fat graft is in direct contact with the facial nerve, brainstem, and cerebellum. When the fat is overpacked and under pressure, symptomatic compression of the brainstem, cerebellum, and facial nerve can occur.[15] Other complications of autologous fat grafting include fat necrosis, subarachnoid fat embolism, lipoid meningitis, wound discharge, and local fistula formation.[23–25,28] Various other dural reconstruction techniques have been described, including covering the defect with a free temporalis muscle graft or a pedicled temporal fascial flap.[20] Some have also described occlusion of mastoid and temporal bone air cells using bone wax, bone paste, fat, muscle, and hydroxyapatite cement and titanium mesh.[1,2] However, even with the evolution of current dural repair techniques for reconstruction after translabyrinthine surgery, postoperative CSF leak rates are still reported to be as high as 9.5%.[27]

In this report, we describe a novel dural "sling" reconstruction technique to repair presigmoid dural defects created after a translabyrinthine approach for removal of acoustic neuromas. An autologous fascial graft is sutured to the presigmoid dural defect to suspend the fat graft within the mastoidectomy defect. This technique prevents the fat from coming in direct contact with the facial nerve and brainstem. In addition, less fat is required to occlude the mastoid cavity. The operative technique is described and illustrated. We also report the results of our preliminary experience using this technique in a small cohort of patients.

Operative Technique

A standard translabyrinthine approach is performed to remove the acoustic neuroma. The presigmoid dura is incised in a T-shaped fashion that extends directly over the IAC (Figs. 1A and 2A). After removal of the tumor, the cerebellopontine angle structures including the facial nerve, brainstem, and cerebellum are visualized within the dural defect (Figs. 1B and 2B). Due to retraction and shrinkage of the dural edges, primary dural approximation is not possible. A dural sling is created by suturing a piece of autologous fascial graft (either abdominal fascia or fascia lata from the thigh) to the edges of the dural defect using interrupted 4-0 Nurolon (Ethicon) sutures (Figs. 1C and 2C). A redundant portion of the fascial graft is carefully placed over the contents of the IAC as an onlay because this area cannot be directly sutured. A monolayer of Surgicel (Ethicon) is then placed over the suture line. The aditus ad antrum is occluded with a small muscle graft followed by hydroxyapatite cement (Hydro-Set, Stryker) to prevent any fistulous communication into the middle ear. Any visible mastoid air cells are occluded with bone wax or cement. Once the fascial sling is secured in place, an initial autologous fat graft is placed on top of the dural sling within the deeper aspect of the mastoid cavity (deep to the level of the fallopian canal; Fig. 2D). This initial fat layer is used to occlude any areas of CSF egress, and care is taken not to overpack the defect so as to avoid compression of the facial nerve. Continuous facial nerve monitoring is used during the reconstruction to detect any nerve irritation. Fibrin glue is placed over this initial fat graft followed by another layer of Surgicel (Fig. 2E).

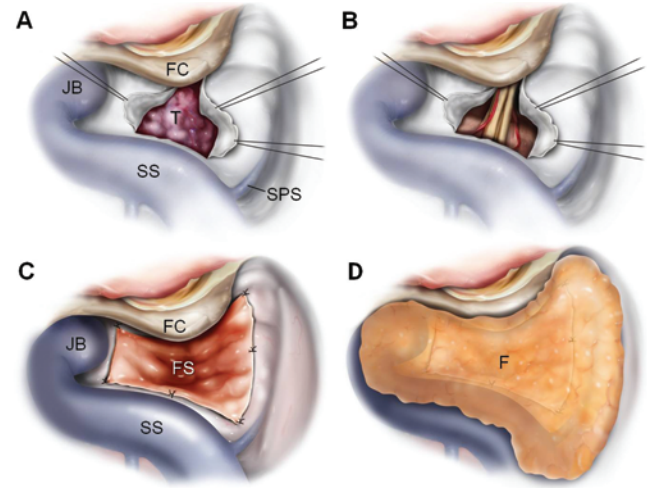

Fig. 1. Illustrations demonstrating the fascial sling technique for translabyrinthine reconstruction using a left-sided approach. **A:** The tumor (T) is exposed after a T-shaped presigmoid dural incision. **B:** The facial nerve is preserved and visualized after tumor removal. **C:** An autologous fascial sling (FS) is sutured into the edges of the presigmoid dural defect using interrupted 4-0 Nurolon sutures. A redundant portion of the fascial graft is carefully placed over the contents of the IAC as an onlay, because this area cannot be directly sutured. **D:** Once the fascial sling is secured in place, an autologous fat graft (F) is placed on top of the dural sling to fill the mastoid cavity. FC = fallopian canal; JB = jugular bulb; SPS = superior petrosal sinus; SS = sigmoid sinus. Reproduced with permission. Copyright Neurological Institute of New Jersey.

The remaining superficial mastoid cavity is filled with another layer of autologous fat graft (Figs. 1D and 2F). Care is taken to avoid compression of the sigmoid sinus, as this can sometimes result in venous outflow obstruction and possible venous hypertension.[15] The fat graft is then bolstered by a titanium mesh plate embedded in porous polyethylene (Medpor Titan implant, Stryker). The plate is secured to the edges of the bony mastoid defect with titanium screws. Meticulous multilayered wound closure is then performed. We do not use routine postoperative lumbar drainage after translabyrinthine approaches to avoid complications of intracranial hypotension.

Results

Eight patients (5 male and 3 female, mean age 55.6 years) underwent translabyrinthine removal of acoustic neuroma followed by subsequent autologous fascial dural sling reconstruction. Fifty percent of the tumors were left-sided and the remaining 50% were right-sided. One patient also had concomitant neurofibromatosis Type II.

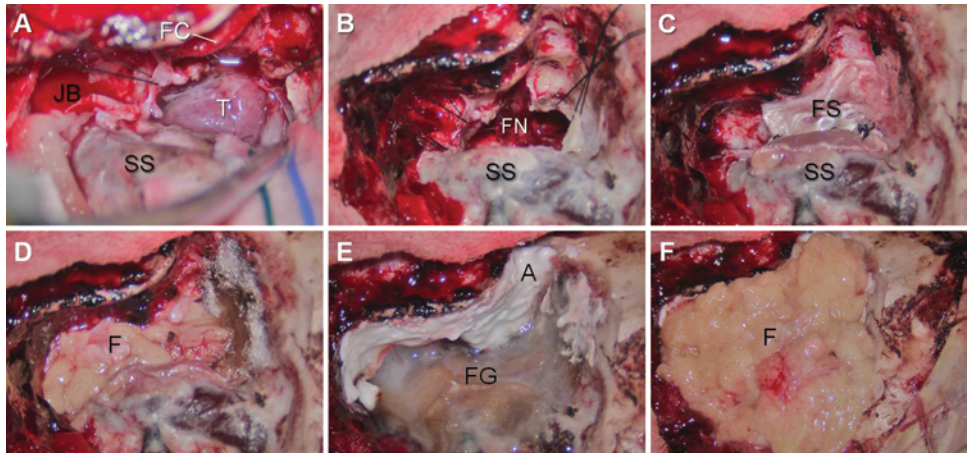

Fig. 2. Intraoperative photographs demonstrating fascial sling reconstruction after translabyrinthine resection of a left-sided acoustic neuroma. **A:** Tumor (T) is exposed through a left-sided presigmoid dural incision. **B:** After tumor removal, facial nerve (FN) is identified and preserved in the presigmoid dural defect. **C:** An autologous fascial sling (FS) is sewn into the edges of the presigmoid dural defect using interrupted 4-0 Nurolon sutures. The redundant portion of the fascial graft is carefully placed over the facial nerve of the exposed IAC as an onlay graft because this area cannot be directly sutured. **D:** An initial deep fat graft (F) is placed over the fascial sling to occlude any areas of CSF egress. **E:** The aditus ad antrum (A) and remaining mastoid air cells are occluded with hydroxyapatite cement. A monolayer of Surgicel and fibrin glue (FG) is placed over the initial deep fat layer. **F:** The remaining superficial mastoid cavity is filled with another layer of autologous fat graft (F).

In 50% of the patients, abdominal fascia was used as the dural sling, and in the remaining cases, fascia lata was used. The mean follow-up period was 13.8 months (range 6–22 months).

Patients were evaluated for the presence of postoperative CSF leakage (incisional leak, rhinorrhea, and otorrhea). There were no postoperative CSF leaks (0%) after fascial sling reconstruction. We compared these results to our own historical control group (n = 5, 1 male and 4 females, mean age 49.6 years) in which a dural substitute (DuraGen [Integra LifeSciences] or Dura Matrix [Stryker]) was used as the sling material, instead of autologous tissue. Mean follow-up in these patients was 35.8 months (range 30–45 months). In this control group, 1 (20%) of the 5 patients presented with a postoperative CSF leak (incisional leak), which was subsequently repaired and required lumbar drain placement.

Discussion

Cerebrospinal fluid leakage remains a common complication following removal of acoustic neuromas via the translabyrinthine approach. The presigmoid dural defect created during the surgery often cannot be primarily reapproximated in a watertight fashion. Exposure of the temporal bone air cells and aditus ad antrum provides multiple potential pathways for CSF fistula formation, thus providing a route for resultant infection and meningitis. Because of the morbidity and mortality associated with these postoperative complications, an effort must be made to prevent CSF leakage. The presentation of CSF leaks may range from incisional, otorrhea (CSF leakage through the tympanic membrane), and rhinorrhea (CSF leakage through the eustachian tube and nasopharynx).[12,30] Following the transmastoid transtemporal petrosectomy required for the translabyrinthine approach, the mastoid antrum and mastoid air cells become exposed, enabling CSF to leak through if the air cells are not properly sealed.[30] Dural opening into the posterior cranial fossa provides direct communication between the subarachnoid space and the mastoid cells. The CSF can then enter the middle ear via the aditus ad antrum, sinus tympani cells, facial recess cells, or retrofacial air cells.[12] Cerebrospinal fluid can also reach the temporal bone through apical air cells, which are located above and below the IAC. Therefore, it is critical to obliterate the mastoid antrum and remaining mastoid air cells, in addition to achieving a watertight dural closure to prevent fistulous pathways for CSF leakage. Meticulous multilayer soft-tissue closure of the wound followed by placement of a compressive mastoid pressure dressing is also important for preventing pseudomeningocele formation and incisional CSF leakage.

One common technique that has been used to reduce the occurrence of CSF fistula formation and leakage is packing or plugging the dural defect and mastoid cavity with an autologous fat graft. However, fat packing may pose a potential danger if overpacking results in mass effect on the brainstem, cerebellum, and/or cranial nerves. In addition, although rare, autologous fat grafting may result in complications including lipoid meningitis, subarachnoid fat embolism, fat liquefaction, and fat necrosis.[23–25,28] In this report, we describe a novel reconstructive technique in which a dural sling is created at the base of the mastoid defect, in which the fat graft is suspended (Fig. 3). The sling serves several purposes. First, the fascial graft provides coverage of the presigmoid dural defect, thereby converting a large dural defect with a "high-flow" CSF leak state to a "low-flow" or minimal CSF leak state. Second, the fascial sling allows suspension of the fat graft to avoid overpacking of fat, which can run the risk of direct compression on the facial nerve or brainstem. The sling also minimizes the volume of fat packing needed to prevent CSF egress through the dural repair.

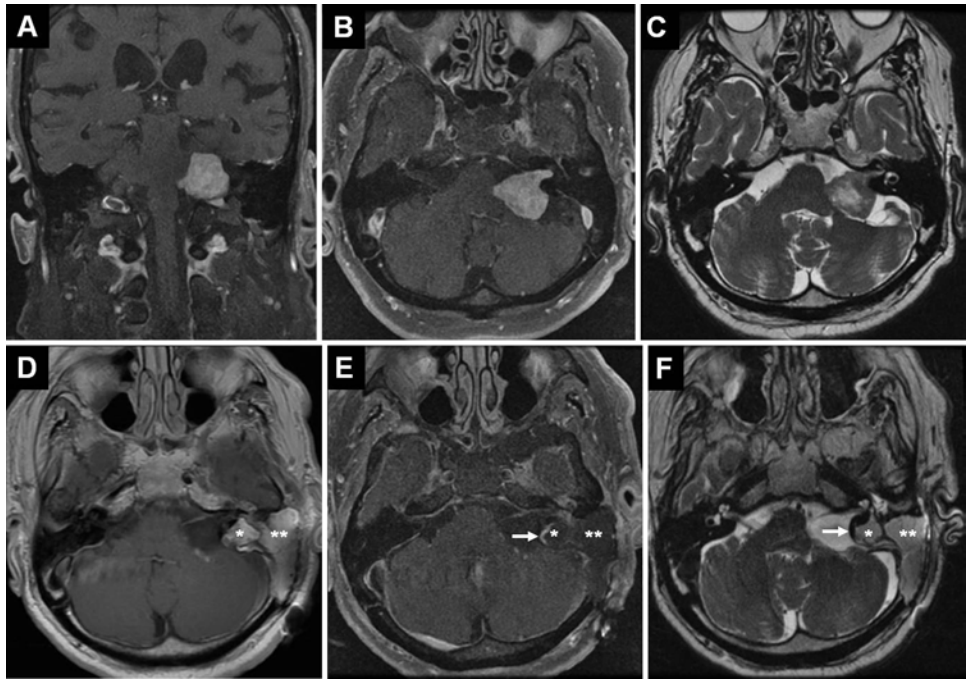

Fig. 3. Preoperative **(A–C)** and postoperative **(D–F)** MR images. **A–C:** Coronal (A) and axial (B) T1-weighted images after Gd administration and an axial T2-weighted image (C) demonstrate a left-sided acoustic neuroma within the cerebellopontine angle with tumor extension into the fundus of the IAC. A left-sided translabyrinthine approach was performed followed by fascial sling reconstruction. **D–F:** Axial T1-weighted images with Gd administration (D) and fat suppression (E), and a T2-weighted image (F) demonstrate complete tumor removal with excellent decompression of brainstem. The fascial sling *(arrow)* allows suspension of the fat graft within the mastoid cavity. Note that the fat graft is not in any direct contact with or compression against the facial nerve, brainstem, and cerebellum. This technique also minimizes the volume of fat required for reconstruction. * deep fat layer; ** superficial fat layer.

Although synthetic graft materials (dural substitute) can be used for the sling, we believe that autologous fascial tissue is a better source for dural reconstruction and postoperative healing than dural allografts. In our report, the autologous fascia group had a lower rate of CSF leak than the dural substitute group (0% vs 20%, respectively). The senior author has had experience with 2 prior cases (unpublished) of midline suboccipital craniectomies in which the dural substitute disintegrated or dissolved upon reexploration for pseudomeningocele repair. Although our experience with the fascial sling technique is limited, our preliminary results in 8 patients suggest that it is a safe and effective method for dural reconstruction of presigmoid dural defects following translabyrinthine resection of acoustic neuromas. Furthermore, additional postoperative lumbar drainage to divert CSF pressure away from the repair site does not appear to be necessary. Therefore, patients can avoid potential risks related to lumbar drainage including intracranial hypotension, CSF infection from an indwelling catheter, and thromboembolic complications from lack of mobilization. Patients who do not require lumbar drainage tend to ambulate sooner with a faster recovery and shorter hospital stays.

Successful prevention of CSF leak after translabyrinthine removal of acoustic neuromas depends significantly on careful and meticulous attention to surgical technique at every step of the closure. These steps include suturing the dural sling, sealing off the aditus ad antrum and mastoid air cells, applying tissue sealant for reinforcement, fat packing of the mastoid cavity, performing a cranioplasty of the mastoid defect to apply pressure on the fat graft, and precise multilayered wound closure.[11]

Conclusions

The dural sling reconstruction technique using autologous fascia can be safely and successfully used to reconstruct presigmoid dural defects after translabyrinthine resection of acoustic neuromas. Additional studies in a larger cohort of patients are warranted to ascertain whether this technique results in significantly lower rates of postoperative CSF leakage than other traditional dural reconstruction techniques.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Liu. Acquisition of data: all authors. Analysis and interpretation of data: Liu, Patel, Podolski. Drafting the article: Liu, Podolski, Patel. Critically revising the article: Liu, Patel. Reviewed submitted version of manuscript: Liu, Patel, Jyung. Approved the final version of the manuscript on behalf of all authors: Liu, Patel. Administrative/technical/material support: Patel. Study supervision: Liu.

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Postoperative imaging of vestibular schwannomas

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Symptomatic vestibular schwannomas can be treated with resection (translabyrinthine, retrosigmoid [suboccipital], or middle cranial fossa approaches) or stereotactic radiosurgery. When appropriate, auditory brainstem stimulators can also be implanted in patients with current or impending hearing loss due to bilateral vestibular schwannomas. Imaging plays a prominent role in determining management following these procedures. In this article, the expected postoperative imaging appearances are depicted. The radiological features of complications are also reviewed, including recurrent tumor, fat graft necrosis, CSF leakage, infection, hydrocephalus, cerebral infarction, venous sinus thrombosis, hemorrhage, and temporal lobe and cerebellar contusions.
(http://thejns.org/doi/abs/10.3171/2012.6.FOCUS12150)

Key Words • vestibular schwannoma • magnetic resonance imaging • computed tomography • postoperative imaging

Vestibular schwannomas are typically benign, slow-growing tumors that most commonly arise from the region of Scarpa's ganglion and comprise approximately 85% of cerebellopontine angle masses.[23] Patients with vestibular schwannomas can present with tinnitus, dizziness, unsteadiness, and vertigo, as well as symptoms due to compression effects, including hearing loss, facial and trigeminal nerve dysfunction, and hydrocephalus.[15] Management options include observation with serial imaging, stereotactic radiosurgery, fractionated radiotherapy, microsurgical resection, or a combination of these. Auditory brainstem implants are a useful adjunct in select patients. Radiological imaging plays an important role in guiding management. In this article, we review the imaging features of surgical approaches, auditory brainstem implantation, and stereotactic radiosurgery along with relevant complications in cases performed by several surgeons from various institutions. Indeed, many of the cases presented in this article were referred to our institution for specialized management of complications.

Abbreviation used in this paper: MRV = MR venography.

Surgical Approaches

There are 3 main types of surgical routes that can be implemented for resection of vestibular cranial nerve tumors, including translabyrinthine, retrosigmoid (suboccipital), and middle cranial fossa approaches.[1,21] These routes are detailed as follows: 1) Translabyrinthine resection involves performing mastoidectomy and labyrinthectomy, including resection of the semicircular canals and at least portions of the vestibule. The ipsilateral cerebellum is retracted, and the internal auditory canal is skeletonized to facilitate tumor resection. Once the tumor is removed, the resection bed is packed with a characteristically triangular-shaped fat graft (Fig. 1). 2) The retrosigmoid (suboccipital) approach consists of performing craniotomy or craniectomy with cranioplasty via a retromastoid incision inferior to the transverse sinus and medial to the sigmoid sinus. Portions of the mastoid air cells are often traversed, but are sealed intraoperatively using bone wax. In addition, drilling through the posterior wall of the internal auditory canal is performed to access the internal auditory canal. This is often repaired with an adipose graft. Cerebrospinal fluid is drained to promote cer-

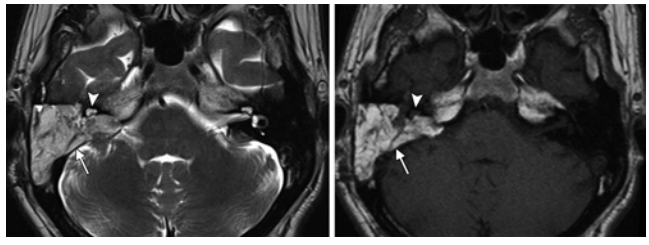

Fig. 1. Translabyrinthine resection. Axial T1-weighted **(left)** and T2-weighted **(right)** MRI studies showing the hyperintense triangular fat graft *(arrows)* within the right mastoid bowl, labyrinthectomy site, and skeletonized internal auditory canal. The right cochlea remains intact *(arrowheads)*.

ebellar relaxation, and the ipsilateral cerebellum may be retracted. This can result in a characteristically flattened lateral edge of the cerebellar hemisphere in association with prominence of the overlying extraaxial CSF (Fig. 2), which should not be mistaken for an arachnoid cyst or hemorrhage. 3) The middle cranial fossa approach involves performing a temporal craniotomy, retracting the temporal lobe superiorly, and decompressing the internal auditory canal by skeletonizing the roof (Fig. 3). A layer of fat or fascial graft is usually applied to the dural defect overlying the internal auditory canal to seal the internal auditory canal.

The translabyrinthine and the retrosigmoid approaches can be used for all tumor sizes, while the middle cranial fossa approach is useful only for removal of intracanalicular tumors. Hearing preservation can be achieved only via the retrosigmoid and middle cranial fossa approaches.[15] Resection can be classified as gross total if there is no visible tumor remaining, near total if the remaining tumor dimensions are less than 5 × 5 × 2 mm, and subtotal if the remaining tumor is larger than that achieved after near-total resection.[4] Alternatively, near-total resection can be designated as incomplete removal with less than 5% residual tumor volume and subtotal resection as greater than 5% of the original tumor volume.[7] Subtotal resection is associated with a 9 times greater likelihood of tumor recurrence compared with gross-total resection.[4]

Magnetic resonance imaging is routinely performed after resection, mainly to assess for residual or recurrent tumor, as well as suspected complications. A typical

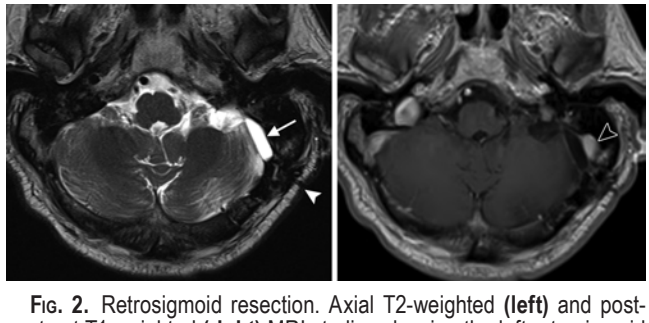

Fig. 2. Retrosigmoid resection. Axial T2-weighted **(left)** and postcontrast T1-weighted **(right)** MRI studies showing the left retrosigmoid approach with a simple lenticular-shaped extraaxial fluid collection *(arrow)* overlying the left cerebellar hemisphere. The left postauricular incision *(white arrowhead)* is posterior to the left sigmoid sinus *(black arrowhead)*.

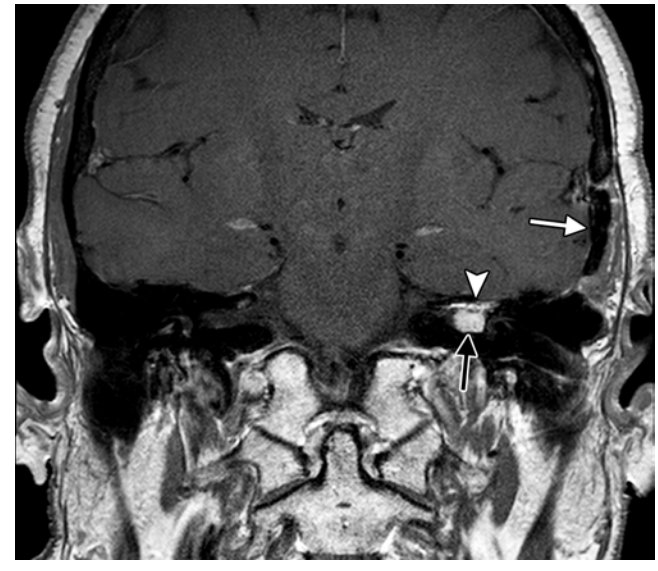

Fig. 3. Middle cranial fossa approach. Coronal postcontrast T1-weighted MRI study showing the left temporal craniotomy site *(white arrow)* used to remove the roof of the internal auditory canal where a small amount of fat graft *(arrowhead)* has been applied. Tumor is present within the internal auditory canal *(black arrow)*.

postoperative MRI examination consists of an internal auditory canal protocol that includes thin-section axial precontrast T1-weighted imaging, axial and coronal thin-section postcontrast fat-suppressed T1-weighted imaging, T2-weighted fast spin echo, T2-weighted FLAIR, diffusion-weighted imaging, and cisternography sequences, such as CISS (constructive interference in steady state) or FIESTA (fast imaging employing steady-state acquisition), at the level of the cerebellopontine angle. The use of fat suppression is helpful for distinguishing enhancing tumor from the intrinsic high signal intensity of the fat grafts, which are commonly implanted in the surgical beds during tumor resection to minimize CSF leakage. Sequences can be added or modified based on the particular indication, such as whole-brain field of view for widespread complications or MRV for venous sinus thrombosis. Computed tomography also plays a role in postoperative imaging for vestibular schwannoma resection, particularly for defining the altered bony anatomy and hemorrhage.

Linear enhancement (Fig. 4) corresponding to scar and/or granulation tissue in the surgical bed is apparent on MRI in the majority of cases during the first 6 postoperative months and can persist for 1 year or longer.[2,4] On the other hand, the presence of nodular enhancement (Fig. 5) on the initial postoperative MRI studies is associated with a 16-fold higher risk for eventual recurrence compared with a linear pattern of enhancement.[4] It is also important to assess the entire extent of the surgical approach for nodular enhancement that may represent intraoperative tumor seeding (Fig. 6). It is sometimes difficult to distinguish between the different patterns of enhancement on baseline MRI, and follow-up imaging is useful for elucidating the nature of the enhancement.

Other complications related to surgical treatment of vestibular schwannomas that can be readily assessed on

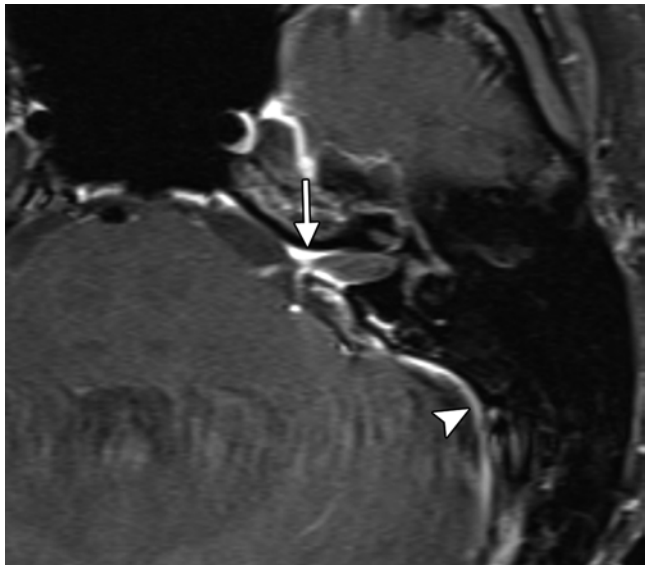

Fig. 4. Linear enhancement. Axial postcontrast fat-suppressed T1-weighted MRI study showing linear dural enhancement within the left internal auditory canal *(arrow)* and in the posterior fossa *(arrowhead)*.

imaging include fat graft necrosis, CSF leakage, infection, cerebral infarction, venous sinus thrombosis, hemorrhage, cerebellar atrophy, and endolymphatic fluid loss. Risk factors for complications include underlying severe general and/or neurological morbidity, cystic tumor, and major cranial nerve deficits.[17]

Fat graft necrosis occurs in approximately 1% of cases of vestibular schwannoma surgery in which a fat graft is used to eliminate dead space and reinforce dural closure.[24] Sequelae of fat necrosis can occur within days to years after surgery and include sterile liquefied fat fistula,

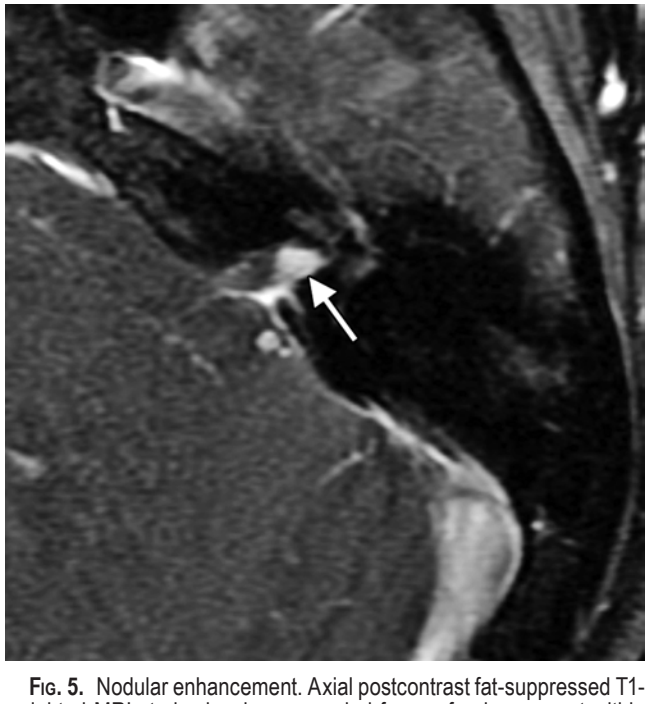

Fig. 5. Nodular enhancement. Axial postcontrast fat-suppressed T1-weighted MRI study showing a rounded focus of enhancement within the left internal auditory canal *(arrow)*.

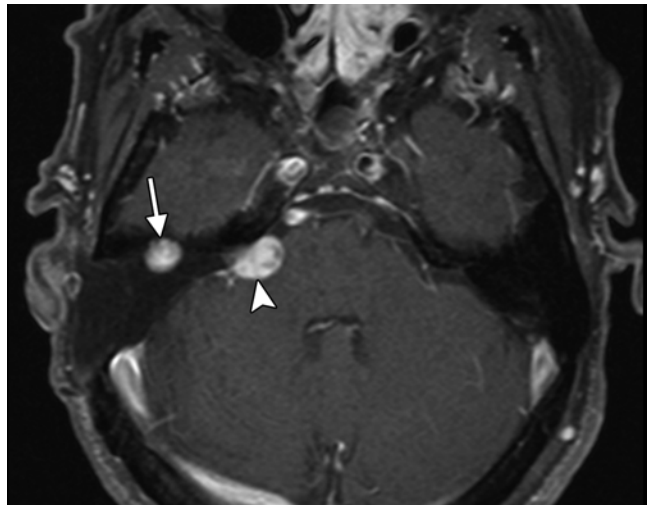

Fig. 6. Intraoperative tumor seeding. Axial postcontrast fat-suppressed T1-weighted MRI study showing a progressively enlarging enhancing nodule anterior to the fat graft *(arrow)*, which was not present preoperatively. Recurrent tumor is present in the right cerebellopontine angle *(arrowhead)*.

CSF leakage, lipoid meningitis, and graft shrinkage.[9,24] Patients with these complications generally experience favorable outcomes with appropriate management.[24] On imaging, fat necrosis can appear as fragmentation of the fat graft, allowing infiltration by fluid (Fig. 7). The fluid can extend to the scalp incision. Aseptic lipoid meningitis can present with meningismus and appears as punctate ovoid droplets of fat density or signal on imaging (Fig. 8).[9]

Cerebrospinal fluid leakage occurs in approximately 1%–8% of cases following schwannoma resection.[5,12,14] Cerebrospinal fluid otorrhea can sometimes occur due to mastoid air cell entry despite the application of bone wax.[21] In the appropriate clinical setting, opacification of the ipsilateral mastoid air cells and middle ear with craniotomy defect that traverses the mastoid air cells suggests mastoid entry as the source of CSF leak (Fig. 9). Cerebral spinal fluid wound leakage usually occurs over the craniotomy or cranioplasty site and can result in pseudomeningocele. These lesions display fluid attenuation on CT and CSF signal intensity on MRI (Fig. 10). Since pseudomeningoceles can become large, potentially

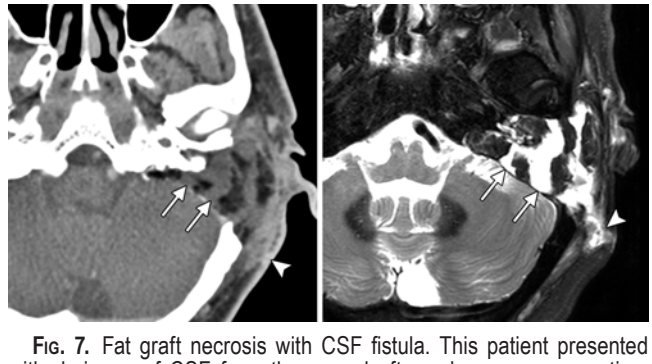

Fig. 7. Fat graft necrosis with CSF fistula. This patient presented with drainage of CSF from the wound after schwannoma resection. Axial CT image **(left)** and axial T2-weighted MRI study **(right)** showing striated bands of fluid *(arrows)* traversing the fat graft, extending to the incision site *(arrowheads)*.

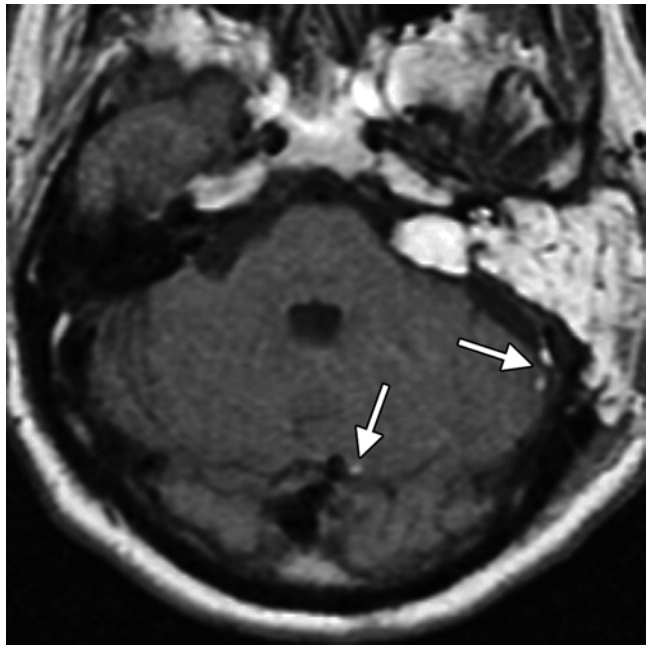

Fig. 8. Lipoid meningitis. Axial T1-weighted MRI study showing scattered punctate foci of high signal *(arrows)* in the left posterior fossa remote from the left translabyrinthine fat graft. The foci showed loss of signal with fat suppression (not shown).

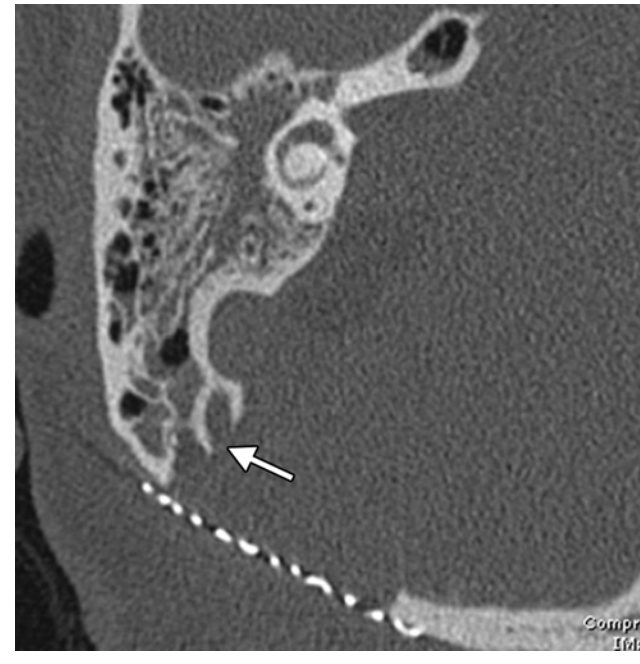

Fig. 9. Mastoid entry with CSF leakage. The patient presented with right CSF otorrhea after surgery. Axial CT image showing a right retrosigmoid cranioplasty and entry into the posterior mastoid air cells *(arrow)*. The mastoid air cells and antrum are partially opacified.

extending toward the apex superiorly and into the posterior neck soft tissues inferiorly, it is important to adjust the imaging field of view accordingly to include the entire lesion, perhaps with the aid of external markers.

Wound infection occurs in up to approximately 4% of cases.[11] Although the diagnosis can be made clinically, imaging is useful for delineating the extent of disease and establishing prompt diagnosis and treatment, which is crucial for preventing severe morbidity. On MRI, postoperative wound infections can appear as rim-enhancing fluid collections (Fig. 11) with variable presence of restricted diffusion. There may also be intracranial involvement, which can manifest as extra- or intraaxial abscess, labyrinthitis (Fig. 12), and meningitis with communicating hydrocephalus (Fig. 13). On MRI, infectious labyrinthitis can manifest as diffuse enhancement of the cochlea, vestibule, and/or the semicircular canals, as well as loss of endolymphatic fluid on T2-weighted sequences. Labyrinthitis ossificans can eventually develop, which appears as hyperdensity within the inner ear structures. Diminished labyrinthine fluid signal on MRI is associated with postoperative hearing loss.[23,25] The imaging differential diagnosis for loss of endolymphatic fluid signal includes labyrinthine fenestration, vascular injury, and blood products.[21] It is important to compare these findings with the aid of preoperative imaging since schwannomas can produce these signal abnormalities.[3,22]

Arterial injury during vestibular schwannoma resection is rare and tends to involve the anterior inferior cerebellar artery and less often the posterior inferior cerebellar artery with retrosigmoid and translabyrinthine approaches and the middle cerebral artery branches with the middle cranial fossa approach. This can sometimes result in cerebral infarction, which produces restricted diffusion and swelling in the acute and subacute stages (Fig. 14). Venous parenchymal injury sometimes occurs in the lateral pons and middle cerebral peduncle during removal of adhesive schwannomas and can manifest as an infarct on imaging.[21]

Venous sinus thrombosis has been reported in up to 5% of suboccipital craniotomies and translabyrinthine craniectomies for tumor resection.[10] The transverse and sigmoid sinuses on the treated side are most often affected, while the contralateral sinuses are typically spared. Magnetic resonance imaging with MRV is a suitable, noninvasive modality for obtaining the diagnosis (Fig. 15). On T1-weighted MRI studies, the thrombus can display a high signal if it is in the subacute stage.[6] Consequently, contrast-enhanced MRV can lead to false-negative results. However, on 3D time-of-flight MRV, the loss of flow-related enhancement confirms the diagnosis.[6] Computed tomography venography is also sensitive for delineating venous sinus thromboses, which appear as filling defects. Patients can present with headache, visual obscuration, and papilledema due to elevated intracranial pressure, even if the affected venous sinus is nondominant.[10] Symptomatic treatment consists of acetazolamide and steroids; however, endovascular thrombolysis or anticoagulation may be warranted in the acute postoperative period.[10]

Postoperative hemorrhage can result from venous injury and occurs in approximately 0.6% of cases.[18] Subdural and brainstem hematomas have been reported in approximately 0.4% and 0.1% of cases, respectively.[18] Noncontrast CT scanning is adequate for assessing acute hemorrhage, which appears hyperdense (Fig. 16). If the hemorrhage is sufficiently large, there can be mass effect

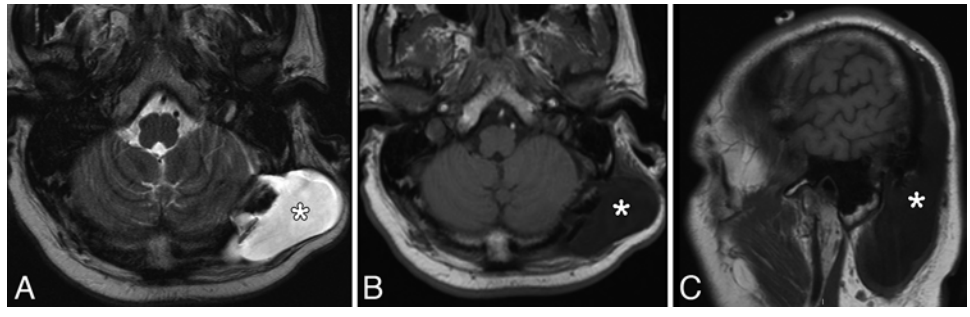

Fig. 10. Pseudomeningocele. Axial T2-weighted **(A)**, axial T1-weighted **(B)**, and sagittal T1-weighted **(C)** MRI studies showing a large subgaleal fluid collection *(asterisk)* that communicates with the intracranial CSF across the left retrosigmoid craniotomy.

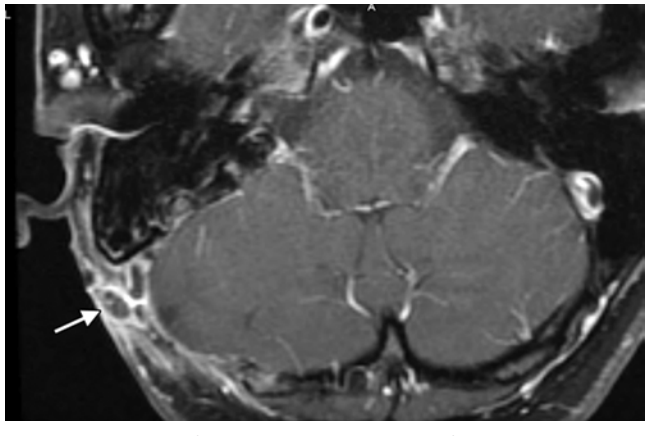

Fig. 11. Wound infection. Axial postcontrast T1-weighted MRI study showing rim-enhancing fluid collections in the subcutaneous tissues overlying the right retrosigmoid craniotomy and in the epidural space *(arrow)*.

upon the brainstem and obstructive hydrocephalus. Organizing hematomas can last several years after surgery and occasionally mimic recurrent tumor.[8]

Auditory Brainstem Implant

Auditory brainstem implants are typically implanted via craniotomy at the time of tumor removal and are intended to facilitate lip reading and, to some extent, enable direct speech comprehension.[20] The devices are used when the contralateral ear provides no hearing or if there is concern of contralateral hearing loss, such as in patients with neurofibromatosis Type 2. The main components of a typical auditory brainstem implant include the receiver-stimulator, grounding ball electrode, and stimulator electrode, which is positioned over the affected cochlear nucleus (Fig. 17), usually via the lateral recess of the fourth ventricle. Computed tomography is well suited to assess whether the device components are intact. However, considerable streak artifact produced by

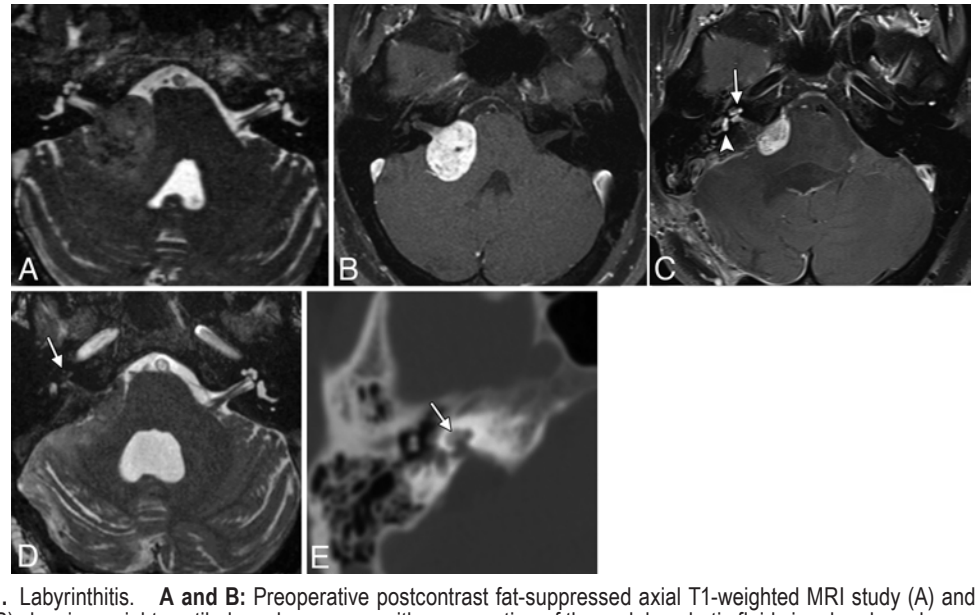

Fig. 12. Labyrinthitis. **A and B:** Preoperative postcontrast fat-suppressed axial T1-weighted MRI study (A) and CISS sequence (B) showing a right vestibular schwannoma with preservation of the endolymphatic fluid signal and no abnormal labyrinthine enhancement. The patient developed postoperative *Klebsiella* meningitis. **C:** Postoperative postcontrast fat-suppressed axial T1-weighted MRI study showing subtotal resection of the tumor and new avid enhancement in the cochlea *(arrow)* and vestibule *(arrowhead)*. **D:** Corresponding CISS sequence showing loss of the fluid signal in the right cochlea *(arrow)*. **E:** Axial CT image obtained 1 year later showing hyperdensity within the cochlea *(arrow)*, consistent with labyrinthitis ossificans.

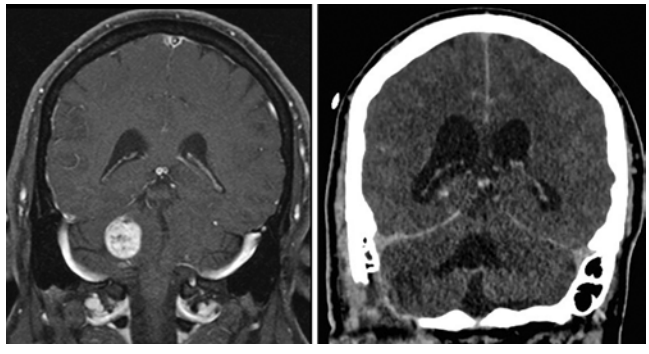

Fig. 13. Hydrocephalus. **Left:** Preoperative coronal postcontrast T1-weighted MRI study showing a right cerebellopontine angle schwannoma. **Right:** Postoperative coronal CT scan showing interval enlargement of the ventricular system following right retrosigmoid resection, requiring ventriculoperitoneal shunt insertion. The patient had postoperative meningitis.

the hardware can limit precise determination of implant positioning. On the other hand, thin-section T2-weighted images are useful for delineating the intracranial course of the stimulator electrode. Complications related to auditory brainstem implant malposition include suboptimal production of auditory stimuli, CSF leak along the course of the wire, and nonauditory stimuli, such as trigeminal neuralgia.[19]

Stereotactic Radiosurgery

Stereotactic radiosurgery (for example, proton beam and Gamma Knife surgery) for vestibular schwannomas consists of delivering a focused radiation dose to halt tumor growth.[13,16] The imaging response to radiosurgery is widely variable. With regard to tumor volume, transient enlargement is observed in 30%–41% of patients, no change or sustained regression in 34%–82%, alternating enlargement and regression in 13%, and continuous enlargement in 12%–16%.[13,16] Transient enlargement generally occurs within 2 years after radiosurgery and is often followed by regression.[13,16] Changes in tumor enhancement characteristics include transient loss of enhancement in 84% of patients (Fig. 18), continuous increase in enhancement in 5%, and no change in enhancement in 11%.[13] An increase in the proportion of tumor cystic components does not appear to correlate with the effectiveness of the treatment.[13] New areas of T2 hyperintensity in the adjacent brain parenchyma can appear in about 30% of cases on average 12 months after treatment

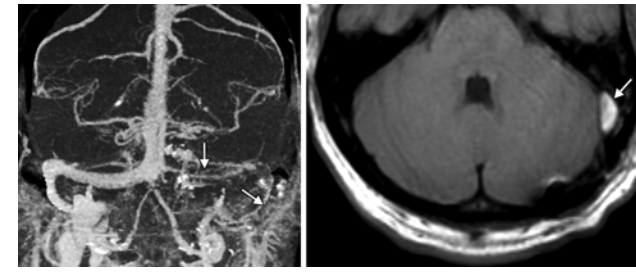

Fig. 15. Venous sinus thrombosis. **Left:** Axial T1-weighted MRI study obtained after left retrosigmoid resection, showing intrinsic high signal within the left sigmoid sinus. **Right:** Corresponding 3D maximum intensity projection MRV showing a paucity of flow-related enhancement in the left transverse and sigmoid sinuses *(arrows)*.

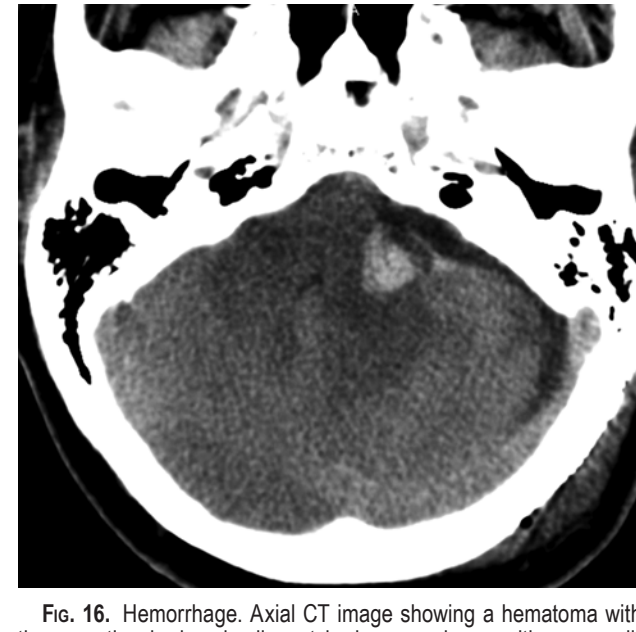

Fig. 16. Hemorrhage. Axial CT image showing a hematoma within the resection bed and adjacent brain parenchyma with surrounding edema and mass effect upon the brainstem and fourth ventricle.

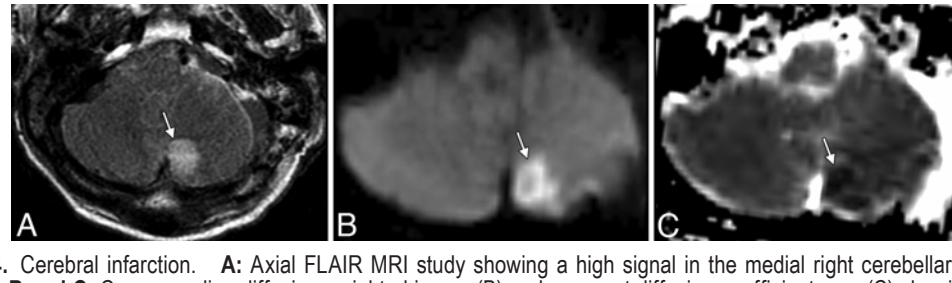

Fig. 14. Cerebral infarction. **A:** Axial FLAIR MRI study showing a high signal in the medial right cerebellar hemisphere *(arrow)*. **B and C:** Corresponding diffusion-weighted image (B) and apparent diffusion coefficient map (C) showing restricted diffusion *(arrows)*.

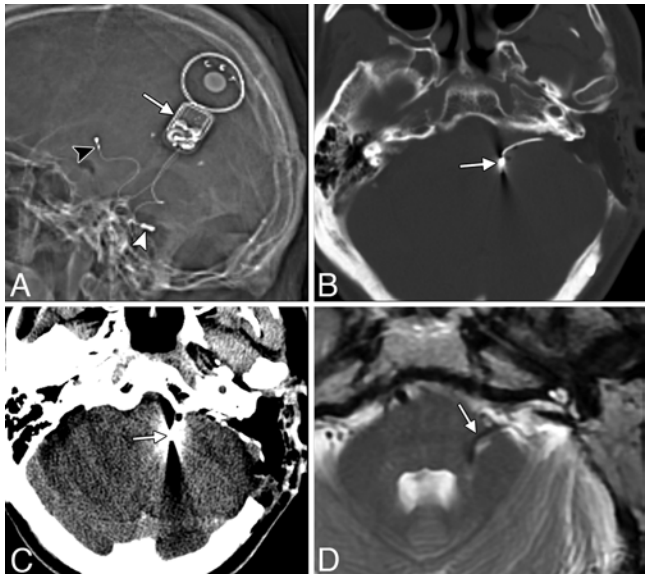

Fig. 17. Auditory brainstem implant. **A:** Scout image showing the components of the device, including the receiver stimulator *(arrow)*, stimulator electrode *(white arrowhead)*, and grounding electrode *(black arrowhead)*. **B and C:** Axial CT images in the bone (B) and soft-tissue (C) windows showing that the electrode *(arrows)* produces considerable streak artifact, which makes precise localization difficult. **D:** The position of the electrode *(arrow)* is better delineated on the axial T2-weighted MRI.

and eventually resolve in most cases.[13] Rarely, secondary neoplasms can arise as a result of the radiation exposure, including sarcomas and meningiomas.[10,15]

Conclusions

Radiological imaging plays an important role in evaluating patients with vestibular schwannomas after microsurgery, auditory brainstem implantation, and stereotactic radiosurgery. Familiarity with expected and complicated imaging findings is necessary for optimal posttreatment management.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Ginat. Analysis and interpretation of data: both authors. Drafting the article: Ginat. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors. Approved the final version of the manuscript on behalf of both authors: Ginat.

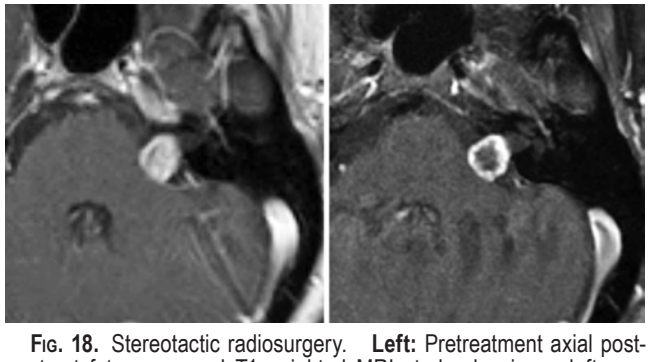

Fig. 18. Stereotactic radiosurgery. **Left:** Pretreatment axial postcontrast fat-suppressed T1-weighted MRI study showing a left vestibular schwannoma that contains minimal nonenhancing components. **Right:** Axial postcontrast fat-suppressed T1-weighted MRI study obtained 1 year after stereotactic radiosurgery, showing interval development of an extensive area of central nonenhancement.

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