

Management of Sporadic Vestibular Schwannoma



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KEYWORDS

- Vestibular schwannoma • Acoustic neuroma • Cerebellopontine angle
- Internal auditory canal • Skull base

KEY POINTS

- The majority of patients with small to medium tumors experience high rates of tumor control and excellent facial nerve outcomes, regardless of treatment modality.
- At 10 years, less than a one-quarter of patients who started with serviceable hearing will maintain class A or B hearing regardless of the treatment modality employed.
- There is compelling evidence that, in the long term, patient-related factors are the primary drivers of quality of life, and treatment strategy has less impact.
- Long-term follow-up should be prioritized in vestibular schwannoma outcomes, where the majority of patients are expected to survive many decades beyond their diagnosis and treatment.
- Patients should receive individualized management based on personal priorities, health status, anticipated life expectancy, and symptoms.

INTRODUCTION

Understanding the history of vestibular schwannoma (VS) treatment is to know the development of modern lateral skull base surgery. VS is the model disease that defined a specialty, leading to the refinement of middle and posterior fossa skull base approaches, intraoperative cranial nerve monitoring, and collaboration between the neurotologist and neurosurgeon. The first surgeon to successfully remove a VS remains disputed secondary to uncertainties over tumor pathology, with either Charles Ballance in 1892 or Thomas Annandale in 1895 deserving credit.¹ The first significant breakthrough in the treatment of VS came with Cushing's introduction of

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the mastoid-to-mastoid bilateral suboccipital craniotomy with subtotal resection in place of finger enucleation, decreasing mortality from 80% to less than 20%.² Subsequently, Walter Dandy, Cushing's protégé and later rival, advocated a unilateral approach with complete tumor removal.^{3,4}

In 1951, Swedish neurosurgeon, Lars Leksell introduced the concept of stereotactic radiosurgery (SRS) with the first VS treated in Stockholm with a fixed cobalt 60 source unit in 1969, citing the significant morbidity of surgery and Cushing's own reference to the need for noninvasive treatment methods.^{5,6} Simultaneously, microsurgical advances came in the 1960s with William House and the adoption of the operating microscope and otologic drill, resulting in a further reduction in patient mortality and improved prospects of cranial nerve preservation with surgical removal of VS.^{7,8} It was during this time that the middle fossa craniotomy and the translabyrinthine approaches were revisited after being abandoned for 6 decades because of technical limitations of the time.^{9,10} The last significant advancement within the field of VS microsurgery came with the utilization of cranial nerve monitoring, first pioneered by Delgado and colleagues in 1979.¹¹

Current management options for VS include observation with serial imaging, external beam radiation in the form of SRS (1–5 fractions) or stereotactic radiotherapy (>5 fractions), and microsurgical resection. Practically speaking, the viability of modern treatment modalities differ for small to medium tumors (<3 cm) compared with large VS; therefore, these 2 populations are usually discussed separately. In contrast with large tumors with problematic brainstem compression and symptoms of mass effect where surgery is strongly preferred, many smaller tumors can be managed effectively with observation, radiation, or microsurgical resection. Despite the significant volume of literature analyzing VS outcomes, the management of small to medium VS remains highly controversial. The fact that many VS centers are either dominated by SRS or microsurgery suggests that in most environments, provider bias remains the strongest factor dictating treatment. However, it also should be acknowledged that a number of patients seek out various centers with the intent of receiving a particular therapy. The remaining discussion is dedicated to understanding the advantages, limitations, and controversial aspects of the 3 available treatment modalities.

Microsurgery

Historically, and before the widespread availability of SRS, surgical resection was the preferred treatment for VS. The primary advantages of gross total surgical resection (GTR) include a high rate of long-term tumor control, improvement in symptoms of mass effect, definitive histopathologic confirmation of benign schwannoma, and potentially less intense imaging surveillance going forward. It is also sometimes presented that clinical outcomes of SRS after microsurgery, in cases of recurrence or less than GTR, are more favorable than microsurgery after SRS, in cases of continued tumor growth after radiation. This, in addition to the fact that recurrence after gross total resection is lower than reported SRS failure, is often used as an argument for microsurgery when all else is equal. Conversely, the primary disadvantages of surgery include a greater risk of permanent facial palsy, headache, treatment-related hearing loss, cerebrospinal fluid leak, meningitis, and very rarely stroke or death. Additionally microsurgery requires general anesthesia and a 3- to 5-day hospital stay on average with increased upfront cost, whereas SRS is performed with local anesthesia as an outpatient. Although somewhat controversial, frequently cited indications for primary microsurgery include younger patient age, larger tumor size with symptoms attributable to mass effect, ongoing dizziness, cystic tumors, and small anatomically favorable tumors with good hearing.¹²

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