



Acoustic neuroma: What the evidence says about evaluation and treatment

Tumor size and a patient's age, condition, and personal preference are key factors in choosing among watchful waiting, microsurgery, or stereotactic radiosurgery.

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PRACTICE RECOMMENDATIONS

- *The gold standard for diagnosis of acoustic neuroma is contrast magnetic resonance imaging. (A)*
- *Consider watchful waiting for tumors <1 cm in the elderly, medically infirm, or patients with serviceable hearing who opt for a more conservative approach. (C)*
- *The best treatment option for a tumor >1 cm and <3 cm varies. Base your decision on tumor size and the patient's age, comorbidities, and preference. (B)*
- *Recommend microsurgery for tumors >3 cm. (C)*

Strength of recommendation (SOR)

- (A) Good-quality patient-oriented evidence
- (B) Inconsistent or limited-quality patient-oriented evidence
- (C) Consensus, usual practice, opinion, disease-oriented evidence, case series

When a patient presents with unilateral hearing loss—especially with a report of gradual onset—that is accompanied by tinnitus, consider an acoustic neuroma (AN). Also known as vestibular schwannoma, ANs represent just 9% of all intracranial tumors. But its relatively slow growth rate can insidiously lead to impaired quality of life and even complete hearing loss.¹ Vestibular symptoms, such as vertigo, may be present. And in some instances, tinnitus may be absent. Even without hearing loss or other symptoms, ANs may be detected incidentally on magnetic resonance imaging (MRI) performed for other reasons. In this article, I describe the diagnostic work-up for AN and 3 options for treatment.

How acoustic neuroma arises

ANs evolve from the abnormal growth and proliferation of Schwann cells, or neurolemmocytes, at their junction with glial cells surrounding the vestibular nerve. ANs represent 80% of all cerebellopontine angle tumors.¹ The incidence in the general population is about 1 in 100,000 people per year, with equal distribution between men and women.¹ Among adults with sensorineural hearing loss, about 1% have ANs.² Tumor growth within the internal auditory canal and resultant compression of cranial nerves VII and VIII causes associated symptoms. Unabated growth can lead to prolapse into the cerebellopontine angle with compression of the brainstem and cerebellum.¹

There are 2 major types of AN. The sporadic type, which will be the focus of this discussion, occurs in 95% of all cases, is unilateral, and usually affects individuals 40 to 60 years old. AN associated with neurofibromatosis type 2 is typically bilateral, autosomal dominant, and usually affects teens and young adults.¹

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Watchful waiting may be appropriate for elderly or infirm patients with tumors <1 cm.

The only known possible risk factor for the development of AN is the use of cell phones, but data are conflicting. Although 2 meta-analyses did find a correlation between AN and ipsilateral cell phone use >10 years, these studies were limited by possible recall bias and misclassification.^{3,4}

How acoustic neuroma can present

Slowly progressive, high-frequency, sensorineural hearing loss is typical for AN. In most cases, hearing loss is unilateral. Nerve compression and stretching from the growing neuroma cause hearing loss of gradual onset.¹ However, up to 25% of patients may present with sudden onset hearing loss due to total occlusion of the internal auditory canal and the artery supplying the cochlea.¹ Tinnitus is reported in up to 70% of patients with AN.¹ The mechanism of injury is the same as for hearing loss.

A feeling of imbalance or unsteadiness occurs in more than 50% of patients with AN.¹ One research group found that vertigo was the more common symptom in patients with smaller tumors, whereas patients with larger tumors complained more of dysequilibrium.¹

Other physical findings can include trigeminal nerve dysfunction (found in 50% of patients with AN, but patients rarely note the absent corneal reflex), facial nerve motor dysfunction (2%), increased intracranial pressure (rare in tumors <3 cm), and brainstem and cerebellar symptoms (rare).¹

■ **Asymptomatic AN.** Small tumors may be detected by MRI incidentally before the onset of hearing loss. Such a finding accounts for about 12% of all patients diagnosed with AN.⁵ A 2005 study by Lin et al found the prevalence of incidental AN to be about 2 per 10,000 people.⁶ This is higher than the 1 in 100,000 suggested by epidemiologic studies,⁶ but Lin's study was performed at a large tertiary center.

Diagnostic evaluation

Contrast MRI is the gold standard for diagnosing AN.² One study did find that a heavily T2-weighted noncontrast scan in the hands of an experienced radiologist reduced the

procedural cost and was as effective as contrast MRI in evaluating the VII and VIII cranial nerves within the cerebellopontine angle and internal auditory canal.⁷

Although not as sensitive for small tumors, the auditory brainstem response can be used in certain circumstances. During the initial evaluation, other diagnoses to consider are facial neuroma and jugular foramen tumors.¹

Three options for treatment

The goal of treatment is to slow or eliminate both tumor growth and deterioration in hearing and neurological function. Lin's study found that 43% of patients with incidental AN had abnormal audiometry findings.⁶ A hearing evaluation may therefore be helpful in guiding patients through a meaningful discussion about prognosis, further testing and consultations, and the 3 therapeutic options—watchful waiting, microsurgery, and stereotactic radiosurgery.⁸

■ **Watchful waiting** has been recommended for the elderly and for infirm patients with tumors <1 cm who would be poor candidates for surgery or radiation. In up to 57% of AN cases, no further tumor growth occurs after diagnosis; in about 8% of these cases, tumor regression is noted.⁹ A little more than half of patients will experience further hearing loss.⁹

Unfortunately, there are no prediction rules for determining who is most at risk for tumor growth and hearing loss. A recent study found that conservative management was a cost-effective approach for tumors <1.5 cm in any age group,¹⁰ provided there was no increase in complications from continued tumor growth. This, then, is a third group of patients for whom watchful waiting might be an option.¹⁰ Follow-up MRI can be used as a surveillance tool.^{8,9,11}

■ **Microsurgery.** This option is the oldest and best-studied treatment for AN. Microsurgery appears to provide the best tumor control, although morbidity and mortality remain risks. A systematic review by Yamakami in 2003 showed that microsurgery completely removed 96% of ANs, with tumor recurrence, mortality, and major disability rates of 1.8%,

0.63%, and 2.9%, respectively.¹² More recent reviews have shown mortality rates of approximately 0.1%.¹³ Surgery usually involves removal of cranial nerve VIII, with the risk of damage to cranial nerve VII. Nerve-sparing procedures are available. Cerebral spinal fluid leaks and meningitis are occasional adverse events. The experience of the surgical team can affect outcome, including complications and cost.¹⁴

■ Stereotactic radiosurgery. Through the use of sophisticated imaging devices and 3-dimensional treatment-planning computers, stereotactic irradiation allows much more specific targeting of the AN, with significantly less radiation delivered to surrounding healthy tissues.¹⁵ Dynamic beam shaping and intensity modulation provide flexibility and enable delivery of much higher radiation doses to the tumor, resulting in greater control rates and decreased complications. There are 3 delivery technologies: Gamma Knife, proton beam, and specially modified linear accelerators.

Whereas older studies did not provide sufficient evidence to support the use of low-dose over high-dose radiation for long-term control of ANs,¹⁶ a more recent systematic analysis by Yang et al seems to indicate that patients treated with the lower dose (12.5 Gy) did equally as well with better preservation of hearing.¹⁷

Applying the evidence in practice

A large randomized controlled trial comparing these treatment options has yet to be done and, indeed, would be difficult to conduct due to the small number of AN cases, varying surgical expertise among centers, the different treatment goals inherent in the 3 therapies, and the risk involved in each. Evidence to date generally indicates that observation is appropriate for small intracanalicular tumors (<1 cm) in the elderly, medically infirm, or asymptomatic patients who understand and opt for this management approach. For tumors ≥3 cm, evidence supports microsurgery as optimal management.^{13,18} Tumors falling between these extremes pose the real challenge.

Over the last 10 years, numerous studies have demonstrated good tumor control with either microsurgery or radiosurgery,

but with varying degrees of hearing preservation and permanent nerve injury to the facial and trigeminal nerve. There is also a concern for malignant transformation of AN after radiosurgery, with 8 cases reported in the literature.^{8,13,19}

Three evidence-based studies in the last 6 years have compared the 2 interventions. In 2002, Nikolopoulos et al reviewed 111 studies and concluded there was insufficient evidence to support one approach over the other.¹⁸ Pollock conducted a prospective cohort study in 2008 that showed superior outcomes in facial movement, hearing preservation, and Health Status Questionnaire subscales for patients undergoing stereotactic radiosurgery.¹³ This study was limited to nonfractionated radiosurgery, and follow-up varied from 12 to 62 months.

In 2009, a Norwegian prospective study of 91 patients reported better facial nerve and hearing outcomes from radiosurgery for medium and small tumors. This study was well performed, but it looked at only a small, non-randomized population.²⁰ The same author in 2005 had found that, from the patient perspective, cranial nerve function and overall outcomes were better in the radiosurgery group.²¹

Stereotactic radiosurgery does confer lower risks for acute treatment complications than microsurgery, and therefore can be advantageous for patients who are older, infirm, require anticoagulant therapy, are otherwise poor candidates for surgery or anesthesia, or have serviceable hearing and opt for a more conservative approach. Other advantages of stereotactic radiosurgery over microsurgery are its lower cost and its preferred use in patients with permanent hearing loss in the unaffected ear. These advantages of stereotactic radiosurgery may, however, be offset in the long term by cranial neuropathy and eventual hearing loss, which can be comparable to the experiences of patients after microsurgery.⁸

Based on these limited studies, patient-oriented outcomes can be comparable between stereotactic radiosurgery and microsurgery in medium-sized tumors (1-2.9 cm), depending on the patient's clinical presentation. Hearing and preservation of nerve VII



For tumors 1 to 2.9 cm, microsurgery and stereotactic radiosurgery generally provide equivalent outcomes.

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are more likely with radiosurgery than with microsurgery. But several real and potentially large confounding factors limit this interpretation. How does one define tumor control in light of AN's inherently slow growth? What about reports of less than optimal microsurgery outcomes if previous radiosurgery has failed? And, although it is small, a definite risk of malignant transformation exists after irradiation.^{8,13}

For patients to make good decisions, family physicians need to be aware of these issues when discussing treatment options. To aid in patient education and decision making, there is a helpful algorithm from the International RadioSurgery Association in *Stereotactic radiosurgery for patients with vestibular schwannomas* (available online at <http://www.irs.org/AN20%Guideline.pdf>).¹⁵

Follow-up

Follow-up depends on the treatment modal-

ity, but usually relies on MRI to document tumor behavior. MRI studies are typically performed at predetermined intervals such as 6 months and 1, 2, and 4 years. For patients with preserved serviceable hearing, audiograms are recommended at intervals coinciding with clinical and neuroimaging re-evaluations. Tumors proven to be stable over 4 to 5 years can subsequently be reassessed at 2- to 4-year intervals.¹⁵

■ **Enabling recovery.** One of the most important educational opportunities involves early vestibular rehabilitation to facilitate recovery of postural control after treatment. Research has shown benefit from customized vestibular rehabilitation in addition to instructions that stress the need to engage in some type of activity, such as walking or other modes of exercise.²²

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