Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines on the Role of Imaging in the Diagnosis and Management of Patients With Vestibular Schwannomas

QUESTION 1: What sequences should be obtained on magnetic resonance imaging (MRI) to evaluate vestibular schwannomas before and after surgery?

TARGET POPULATION: Adults with vestibular schwannomas.

RECOMMENDATIONS:

Initial Preoperative Evaluation

Level 3: Imaging used to detect vestibular schwannomas should use high-resolution T2-weighted and contrast-enhanced T1-weighted MRI.

Level 3: Standard T1, T2, fluid attenuated inversion recovery, and diffusion weighted imaging MR sequences obtained in axial, coronal, and sagittal plane may be used for detection of vestibular schwannomas.

Preoperative Surveillance

Level 3: Preoperative surveillance for growth of a vestibular schwannoma should be followed with either contrast-enhanced 3-dimensional (3-D) T1 magnetization prepared rapid acquisition gradient echo (MPRAGE) or high-resolution T2 (including constructive interference in steady state [CISS] or fast imaging employing steady-state acquisition [FIESTA] sequences) MRI.

Postoperative Evaluation

Level 2: Postoperative evaluation should be performed with postcontrast 3-D T1 MPRAGE, with nodular enhancement considered suspicious for recurrence.

QUESTION 2: Is there a role for advanced imaging for facial nerve detection preoperatively (eg, CISS/FIESTA or diffusion tensor imaging)?

TARGET POPULATION: Adults with proven or suspected vestibular schwannomas by imaging.

RECOMMENDATION: Level 3: T2-weighted MRI may be used to augment visualization of the facial nerve course as part of preoperative evaluation.

QUESTION 3: What is the expected growth rate of vestibular schwannomas on MRI, and how often should they be imaged if a “watch and wait” philosophy is pursued?

TARGET POPULATION: Adults with suspected vestibular schwannomas by imaging.

RECOMMENDATION: Level 3: MRIs should be obtained annually for 5 yr, with interval lengthening thereafter with tumor stability.

QUESTION 4: Do cystic vestibular schwannomas behave differently than their solid counterparts?

TARGET POPULATION: Adults with vestibular schwannomas with cystic components.

RECOMMENDATION: Level 3: Adults with cystic vestibular schwannomas should be counseled that their tumors may more often be associated with rapid growth, lower rates of complete resection, and facial nerve outcomes that may be inferior in the immediate postoperative period but similar to noncystic schwannomas over time.

QUESTION 5: Should the extent of lateral internal auditory canal involvement be considered by treating physicians?
**TARGET POPULATION:** Adult patients with vestibular schwannomas.

**RECOMMENDATION:** Level 3: The degree of lateral internal auditory canal involvement by tumor adversely affects facial nerve and hearing outcomes and should be emphasized when interpreting imaging for preoperative planning.

**QUESTION 6:** How should patients with neurofibromatosis type 2 (NF2) and vestibular schwannoma be imaged and over what follow-up period?

**TARGET POPULATION:** Adult patients with NF2 and vestibular schwannomas.

**RECOMMENDATION:** Level 3: In general, vestibular schwannomas associated with NF2 should be imaged (similar to sporadic schwannomas) with the following caveats:

1. More frequent imaging may be adopted in NF2 patients because of a more variable growth rate for vestibular schwannomas, and annual imaging may ensue once the growth rate is established.

2. In NF2 patients with bilateral vestibular schwannomas, growth rate of a vestibular schwannoma may increase after resection of the contralateral tumor, and therefore, more frequent imaging may be indicated, based on the nonoperated tumor's historical rate of growth.

3. Careful consideration should be given to whether contrast is necessary in follow-up studies or if high-resolution T2 (including CISS or FIESTA-type sequences) MRI may adequately characterize changes in lesion size instead.

**QUESTION 7:** How long should vestibular schwannomas be imaged after surgery, including after gross-total, near-total, and subtotal resection?

**TARGET POPULATION:** Adult patients with vestibular schwannomas followed after surgery.

**RECOMMENDATION:** Level 3: For patients receiving gross total resection, a postoperative MRI may be considered to document the surgical impression and may occur as late as 1 yr after surgery. For patients not receiving gross total resection, more frequent surveillance scans are suggested; annual MRI scans may be reasonable for 5 yr. Imaging follow-up should be adjusted accordingly for continued surveillance if any change in nodular enhancement is demonstrated.

The full guideline can be found at [https://www.cns.org/guidelines/guidelines-management-patients-vestibular-schwannoma/chapter_5](https://www.cns.org/guidelines/guidelines-management-patients-vestibular-schwannoma/chapter_5).

**KEY WORDS:** Acoustic neuroma, Advanced imaging, Cystic, Growth rate, Facial nerve, MRI, Vestibular schwannoma

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**METHODS**

Details of the systematic literature review are provided in the full text of this guideline ([https://www.cns.org/guidelines/guidelines-management-patients-vestibular-schwannoma/chapter_5](https://www.cns.org/guidelines/guidelines-management-patients-vestibular-schwannoma/chapter_5)) and in the methodology article ([https://www.cns.org/guidelines/guidelines-management-patients-vestibular-schwannoma/chapter_1](https://www.cns.org/guidelines/guidelines-management-patients-vestibular-schwannoma/chapter_1)) of this guideline series. A total of 2070 citations, which were identified through a search of PubMed and Cochrane Central, were manually reviewed. Two independent reviewers evaluated and abstracted full-text data for each article. Citations focused on the imaging of VSSs in adult patients largely in the magnetic resonance imaging (MRI) era (January 1, 1990 to December 31, 2014), published in English, were considered; citations meeting the inclusion criteria were reviewed in full and used for evidence tables and to create recommendations. The selected studies

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**ABBREVIATIONS:** 3-D, 3-dimensional; CISS, constructive interference in steady state; FIESTA, fast imaging employing steady-state acquisition; MPRAGE, magnetization prepared rapid acquisition gradient echo; MRI, magnetic resonance imaging; NF2, neurofibromatosis type 2; VS, vestibular schwannoma

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RESULTS

Excellent preoperative identification and surveillance of VSs can be achieved using 3-D T2 constructive interference in steady state or postcontrast 3-D T1 magnetization prepared rapid acquisition gradient echo MRI. Class III evidence supports that the course of the facial nerve may be determined on preoperative MRI, especially with T2-weighted sequences and with tractography reconstruction. Class III evidence supports the conclusion that about two-thirds of patients with VSs may not exhibit measurable growth, while one-third demonstrate growth, defined variably as either any increase in size or a change in diameter >2 mm. Intrameatal tumors are less likely to grow. Cystic vestibular may demonstrate rapid growth, symptomatic deterioration, and be associated with lower rates of complete resection, worse short-term facial nerve outcomes, and unpredictable response to radiation. Class III evidence supports the conclusion that lateral involvement correlated with decreased facial nerve and hearing function; Class III evidence also supports the conclusion that neurofibromatosis type 2-associated VSs demonstrate unique growth patterns and growth rates. Continued surveillance is required in patients without a complete resection.

DISCUSSION

VSs are usually imaged with MRI, with contrast-enhanced scans considered to be the gold standard for the initial evaluation and postoperative assessment of recurrence or residual tumors. The use of high-resolution T2 sequences to follow VSs, however, is supported by class II evidence. Specific imaging features with clinical application—though supported by class III evidence—include, in addition to size and the presence of hydrocephalus, the extent of lateral extension in the internal auditory canal and the presence of cystic intratumoral contents.

Imaging is a crucial tool in the evaluation and management of patients with VSs, with MRI supplanting computed tomography nearly entirely. Bias as to which patients are treated conservatively and interobserver variation in the definition of growth rate may underlie differential reporting; automated size calculations may remedy this. Higher-resolution T2 sequences and refinements in diffusion tensor imaging may render facial nerve identifi-cation even more reliable, providing valuable information to the surgeon, and in turn the patient, preoperatively.

Disclosure

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Conflict of Interest

The Vestibular Schwannoma Guidelines Task Force members were required to report all possible COIs prior to beginning work on the guideline, using the COI disclosure form of the AANS/CNS Joint Guidelines Committee, including potential COIs that are unrelated to the topic of the guideline. The CNS Guidelines Committee and Guideline Task Force Chair reviewed the disclosures and either approved or disapproved the nomination. The CNS Guidelines Committee and Guideline Task Force Chair are given latitude to approve nominations of Task Force members with possible conflicts and address this by restricting the writing and reviewing privileges of that person to topics unrelated to the possible COIs. The conflict of interest findings are provided in detail in the full-text introduction and methods manuscript (https://www.cns.org/guidelines/guidelines-management-patients-vestibular-schwannoma/chapter_1).

Disclaimer of Liability

This clinical systematic review and evidence-based guideline was developed by a multidisciplinary physician volunteer task force and serves as an educational tool designed to provide an accurate review of the subject matter covered. These guidelines are disseminated with the understanding that the recommendations by the authors and consultants who have collaborated in their development are not meant to replace the individualized care and treatment advice from a patient’s physician(s). If medical advice or assistance is required, the services of a competent physician should be sought. The proposals contained in these guidelines may not be suitable for use in all circumstances. The choice to implement any particular recommendation contained in these guidelines must be made by a managing physician in light of the situation in each particular patient and on the basis of existing resources.

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