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Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines on Surgical Resection for the Treatment of Patients With Vestibular Schwannomas

Please see the full-text version of this guideline (https://www.cns.org/guidelines/guidelines-management-patients-vestibular-schwannoma/chapter_8) for the target population of each recommendation listed below.

QUESTION 1: What surgical approaches for vestibular schwannomas (VS) are best for complete resection and facial nerve (FN) preservation when serviceable hearing is present?

RECOMMENDATION: There is insufficient evidence to support the superiority of either the middle fossa (MF) or the retrosigmoid (RS) approach for complete VS resection and FN preservation when serviceable hearing is present.

QUESTION 2: Which surgical approach (RS or translabyrinthine [TL]) for VS is best for complete resection and FN preservation when serviceable hearing is not present?

RECOMMENDATION: There is insufficient evidence to support the superiority of either the RS or the TL approach for complete VS resection and FN preservation when serviceable hearing is not present.

QUESTION 3: Does VS size matter for facial and vestibulocochlear nerve preservation with surgical resection?

RECOMMENDATION: Level 3: Patients with larger VS tumor size should be counseled about the greater than average risk of loss of serviceable hearing.

QUESTION 4: Should small intracanalicular tumors (<1.5 cm) be surgically resected?

RECOMMENDATION: There are insufficient data to support a firm recommendation that surgery be the primary treatment for this subclass of VSs.

QUESTION 5: Is hearing preservation routinely possible with VS surgical resection when serviceable hearing is present?

RECOMMENDATION: Level 3: Hearing preservation surgery via the MF or the RS approach may be attempted in patients with small tumor size (<1.5 cm) and good preoperative hearing.

QUESTION 6: When should surgical resection be the initial treatment in patients with neurofibromatosis type 2 (NF2)?

RECOMMENDATION: There is insufficient evidence that surgical resection should be the initial treatment in patients with NF2.

QUESTION 7: Does a multidisciplinary team, consisting of neurosurgery and neurotology, provides the best outcomes of complete resection and facial/vestibulocochlear nerve preservation for patients undergoing resection of VSs?

RECOMMENDATION: There is insufficient evidence to support stating that a multidisciplinary team, usually consisting of a neurosurgeon and a neurotologist, provides superior outcomes compared to either subspecialist working alone.

QUESTION 8: Does a subtotal surgical resection of a VS followed by stereotactic radiosurgery (SRS) to the residual tumor provide comparable hearing and FN preservation to patients who undergo a complete surgical resection?

RECOMMENDATION: There is insufficient evidence to support subtotal resection (STR) followed by SRS provides comparable hearing and FN preservation to patients who undergo a complete surgical resection.

QUESTION 9: Does surgical resection of VS treat preoperative balance problems more effectively than SRS?

RECOMMENDATION: There is insufficient evidence to support either surgical resection or SRS for treatment of preoperative balance problems.

QUESTION 10: Does surgical resection of VS treat preoperative trigeminal neuralgia more effectively than SRS?

RECOMMENDATION: Level 3: Surgical resection of VSs may be used to better relieve symptoms of trigeminal neuralgia than SRS.

QUESTION 11: Is surgical resection of VSs more difficult (associated with higher facial neuropathies and STR rates) after initial treatment with SRS?

RECOMMENDATION: Level 3: If microsurgical resection is necessary after SRS, it is recommended that patients be counseled that there is an increased likelihood of a STR and decreased FN function.

The full guideline can be found at: https://www.cns.org/guidelines/guidelines-management-patients-vestibular-schwannoma/chapter_8.

KEY WORDS: Acoustic neuroma, Neurofibromatosis type 2, Stereotactic radiosurgery, Surgical resection, Vestibular schwannoma

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Vestibular schwannomas (VSs) are slow-growing, benign tumors that typically arise from the vestibular portion of the eighth cranial nerve. Over 95% of VSs are sporadic in nature, while approximately 5% are associated with neurofibromatosis type 2 (NF2), an autosomal dominant syndrome hallmarked by the development of bilateral VSs.[1] Complete tumor removal and cranial nerve preservation are the goals of any VS surgical resection. The success of surgical resection of VSs may be impacted by the surgical approach and serviceable hearing status of the patient, tumor size and location, NF2 status, multidisciplinary team management, combination treatment with stereotactic radiosurgery (SRS), prior SRS treatment, and preoperative symptoms.

ABBREVIATIONS: AAO-HNS, American Academy of Otolaryngology-Head and Neck Surgery; **AN,** acoustic neuroma; **CPA,** cerebellopontine angle; **DHI,** dizziness handicap index; **FN,** facial nerve; **FSRT,** fractionated stereotactic radiotherapy; **GBI,** Glasgow Benefit Inventory; **GKRS,** gamma knife radiosurgery; **GTR,** gross total resection; **HB,** House-Brackmann; **IC,** intracanalicular; **IOM,** intraoperative monitoring; **MF,** middle fossa; **MPNST,** malignant peripheral nerve sheath tumor; **NF2,** neurofibromatosis type 2; **NTR,** near total resection; **QOI,** quality of life; **RS,** retrosigmoid; **SRS,** stereotactic radiosurgery; **STR,** subtotal resection; **TL,** translabyrinthine; **VS,** vestibular schwannoma

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_8) and within the methodology article (https://www.cns.org/guidelines/guidelines-management-patients-vestibular-schwannoma/chapter_1) of this guideline series. A total of 2949 citations were manually reviewed by the team with specific inclusion and exclusion criteria as outlined below. Two independent reviewers evaluated and abstracted full-text data for each article that met criteria, and the 2 sets of data were compared for agreement by a third party. Inconsistencies were re-reviewed, and disagreements were resolved by consensus. Citations that considered adult patients focusing on surgical treatment of VSs were considered. The selected studies were classified according to criteria for evidence on therapeutic effectiveness as detailed in the Joint Guidelines Committee guideline development methodology.

RESULTS

Successful hearing preservation and facial nerve (FN) function were found in patients undergoing a middle fossa (MF) microsurgical approach for resection of their VS.[2-9] The MF approach is selected mainly for patients with intrameatal VS tumors.

When comparing FN function preservation rates in patients either undergoing a retrosigmoid (RS) or translateral (TL) approach for complete VS resection at the same center, some studies stated that a TL approach provided better FN function

preservation.[10,11] Other studies did not show a difference in FN function preservation.[12-16]

The primary preoperative predictors of hearing preservation were tumor size/grade and preoperative hearing levels. Collectively, these data demonstrate that tumor size is among the most reliable prognostic factors for hearing preservation and FN function following microsurgery of VSs. While little controversy accompanies the management of large VSs that abut or compress the brainstem, the appropriateness of surgery for intracanalicular VSs continues to inspire debate. In patients with NF2, early surgical therapy is an option aimed at preserving the patient's long-term quality of life (QOI) as the goal of therapy. This can be meaningfully achieved with hearing preservation when possible, or prevention of side effects secondary to tumor progression and mass effect. However, it should be noted that in expert hands hearing loss or complications can occur with surgery. Observation or SRS appear to be viable alternatives when considering QOI.

Neurosurgeons, neurotologists, and radiation oncologists routinely manage patients with VSs, and treatment may be done by a single surgeon or as a team. With surgery, however, subjective balance seemed to improve consistently in patients who presented with impaired balance.[17-19] Notably, vestibular dysfunction was not associated with decreased QOI.[20] Surgery can become necessary after SRS treatment of VS when tumor regrowth or recurrence is detected.

DISCUSSION AND CONCLUSION

Both the MF and RS surgical approaches can permit preservation of hearing and FN function. Small, lateral-based VS tumors in the internal auditory canal may permit greater hearing preservation by an MF approach. FN preservation rates are reported higher with an RS approach in patients with serviceable hearing undergoing surgical resection of their VS. The evidence for this guideline was drawn from studies with class III evidence; currently, no class I or II evidence exists to guide recommendations for this subject. These data should be utilized when counseling patients regarding the probability of long-term maintenance of serviceable hearing and FN preservation following microsurgery for sporadic VSs.

Both the TL and RS approaches permit FN function preservation in patients with no serviceable hearing undergoing complete removal of VSs. These data should be utilized when counseling patients regarding the probability of FN preservation following microsurgery for sporadic VSs when nonserviceable hearing is present. Excellent rates of resection, FN preservation function results, and hearing preservation have been reported after surgery for internal auditory canal VSs. However, there are insufficient data to support a firm recommendation that surgery be the primary treatment for this subclass of VS.

Class III evidence suggests hearing preservation surgery using both the MF or the RS approach for removal of small to medium VSs in patients with good preoperative hearing function. The definition of hearing success after VS resection remains controversial. Many audiologic classification schemes have been developed to determine "hearing preservation," and the fact that there are multiple schemes indicates that none is universally accepted. Limited literature is available to guide superiority of one surgical choice over another in relation to preservation or recovery of vestibular or trigeminal function.

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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