

CONGRESS OF NEUROLOGICAL SURGEONS SYSTEMATIC REVIEW AND EVIDENCE-BASED GUIDELINES UPDATE FOR THE ROLE OF AUDIOLOGIC SCREENING IN THE DIAGNOSIS AND MANAGEMENT OF PATIENTS WITH VESTIBULAR SCHWANNOMAS

Sponsored by: Congress of Neurological Surgeons (CNS) and the Section on Tumors

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Abbreviations: Vestibular schwannoma (VS); computed tomography (CT); magnetic resonance imaging (MRI); Electronystagmography (ENG); American Association of Neurologic Surgeons (AANS); Congress of Neurologic Surgeons (CNS); Sensorineural hearing loss (SNHL); Cerebellopontine angle (CPA)

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ABSTRACT

Background: Vestibular schwannoma(VS) represents a benign tumor of the vestibulocochlear nerve that presents with otologic dysfunction. Although magnetic resonance imaging (MRI) remains the most common technique for imaging diagnosis of VS, there are no unifying guidelines to suggest when a practitioner should obtain a screening MRI for new otologic complaints to rule out VS.

Objective We aim to assess the diagnostic yield of MRI in the detection of VS in patients presenting with asymmetric SNHL, unilateral tinnitus, and sudden SNHL.

Methods: The questions from the previously published guideline were updated to PICO format. A comprehensive literature search from 1/1/2015 to 5/20/2022 was carried out to answer preconceived research questions drafted by the joint tumor task force. A systematic review of the existing body of evidence was conducted based on predefined inclusion criteria to determine the diagnostic yield of MRI for the diagnosis of VS in patients presenting with (1) asymmetric SNHL (2) unilateral tinnitus and (3) sudden SNHL.

Results: Of the 704 articles initially reviewed, 15 individual publications incorporating 13,733 patients met inclusion criteria. When considering non-redundant data sets, the diagnostic yield of MRI for VS remains low in patients presenting with SNHL (1.68%), unilateral tinnitus (1.56%), and sudden SNHL (3.66%).

Conclusion This document serves as CNS's's most up to date and current recommendations on the audiometric screening of VS expanding upon the previous 2018 version. Patients presenting with otologic complaints of asymmetric SNHL, tinnitus, or sudden SNHL have an estimated 1-3% chance of a VS being the causative lesion. Current screening protocols have an approximate 15% rate of abnormal MRI leading to a diagnosis other than VS meaning 85% of patients presenting with asymmetric SNHL, tinnitus, and sudden SNHL will have no structural cause on imaging studies.

UPDATED RECOMMENDATIONS

Question 1: In adult patients with asymmetric sensorineural hearing loss (SNHL) on audiometric testing, is MRI or other diagnostic tests (such as CT, Electronystagmography [ENG]) more effective in the diagnosis of VS?

Patient Population: These recommendations apply to adults presenting with asymmetric SNHL on audiometric testing.

Recommendation

Level 3: MRI remains the most effective diagnostic measure that can differentiate VS from other labyrinthine conditions (e.g., labyrinthine hemorrhage), and is suggested when patients present with asymmetric hearing loss especially in the setting of abnormal auditory brainstem response test results.

Question 2: *In adult patients with subjective complaints of asymmetric tinnitus, is MRI or other diagnostic tests (such as CT, ENG) more effective in the diagnosis of VS?*

Patient Population: These recommendations apply to adults presenting with asymmetric tinnitus.

Recommendation:

Level 3: MRI screening is suggested for patients presenting with asymmetric tinnitus to minimize the incidence of undiagnosed VS, although the diagnostic yield is low.

Question 3: *In adult patients with verified sudden SNHL on audiometric testing, is MRI or other diagnostic tests (such as CT, ENG) more effective in the diagnosis of VS?*

Patient Population: These recommendations apply to adults presenting with verified sudden SNHL on audiometric testing.

Recommendation:

Level 3: MRI is suggested for the diagnosis of VS in the setting of sudden SNHL.

INTRODUCTION

Rationale:

While our understanding of VS has improved since the initial publication of these guidelines due to investigations of genetic dysregulation, observational studies of natural history, and an ever increasing body of surgical series and radiosurgical series, the optimal screening protocols for tumor diagnosis remain unclear.¹ Contrast-enhanced high resolution MRI remains the gold standard imaging screening

modality for the diagnosis of VS given its high sensitivity.^{2,3} The challenge is to identify which patients presenting with otologic complaints in the absence of neurologic deficits are best suited to undergo screening MRIs to rule out VS. Given the widespread increase of healthcare costs and the ever-limited resources of health care systems, it is neither practical nor cost effective to screen all patients with otologic complaints with MRI. This necessitates the development of evidence based guidelines to assist the practitioner in identifying which patients are most likely to benefit from an MRI in the diagnosis of a suspected VS.^{4,5} The previous evidence based guidelines were constructed from pertinent data published at the time of its creation, however, sufficient time has passed to warrant an update to these guidelines.

Objectives:

The task force aims to analyze the presenting otologic symptomatology as they relate to the diagnosis of VS. As the approach to the VS varies widely by institution and practitioner, there remains a wide variety of screening protocols for the detection of this tumor. Presenting symptoms often associated with VS such as SNHL or tinnitus are vague and nonspecific, complicating the creation of meaningful yet cost-effective MRI screening protocols. The ideal protocol would minimize the probability of either a missed tumor diagnosis (false negative screen) or an unremarkable scan (false positive screen). To achieve these objectives, the following questions were addressed:

1. In adult patients with asymmetric SNHL on audiometric testing, is MRI or other diagnostic tests (such as CT, ENG) more effective in the diagnosis of VS?
2. In adult patients with subjective complaints of asymmetric tinnitus, is MRI or other diagnostic tests (such as CT, ENG) more effective in the diagnosis of VS?
3. In adult patients with verified sudden SNHL on audiometric testing, is MRI or other diagnostic tests (such as CT, ENG) more effective in the diagnosis of VS?

METHODOLOGY

Writing Group and Question Establishment:

The Joint Tumor Section of the AANS and the CNS identified the need for a unifying set of guidelines for the management of VS. The first step of the guidelines task force was reviewing the questions from the 2018 publications and modifying them to PICO (patient/intervention/comparison/outcome) format. Members of the Tumor Section, as well as other neurosurgeons and otolaryngologists, involved in the management of VSs were appointed to form the VS Evidence-Based Practice Guidelines Task Force. Once relevant topic sections were identified, a team of writers were divided and tasked with the

development of pertinent questions for their topics. These questions were then circulated for approval by the task force at large. With the scope of questions clearly identified, the subsequent literature searches were carried out. Further information regarding the literature search and review methodology can be found in the Introduction and Methodology Chapter. The qualifying evidence derived from the searches was then collected and compared to the evidence reported in the 2018 publication which incorporated evidence until 12/31/2014. This guideline was then developed using multiple iterations of written review conducted by the authors, then by members of the task force, and finally approved by the AANS/CNS Joint Guideline Review Committee.

Literature Search Method

Search Method:

The task force, in conjunction with medical librarians, conducted a search for articles published between January 1, 2015 to May 20, 2021. Two electronic databases were searched (Ovid Medline, EMBASE). Strategies for searching electronic databases were constructed by the taskforce members and the medical librarians using previously published search strategies to identify relevant studies (Appendix I). The exact search parameters for each electronic database are identified below.

Study Selection and Eligibility

Seven hundred and four citations were manually reviewed by the task force with specific inclusion and exclusion criteria as outlined below. These criteria were also applied to articles provided by guideline task force members who supplemented the electronic database searches with articles from their own files. To reduce bias, these criteria were specified before conducting the literature searches. Two independent reviewers reviewed and abstracted full-text data for articles meeting initial screening criteria. Inconsistencies regarding inclusion/exclusion were refereed by a third reviewer. Citations that considered the audiologic symptom profile of patients with VSs were considered.

Articles that do not meet the following criteria were, for the purposes of this evidence-based clinical practice guideline, were excluded. To be included as evidence in the guideline, an article had to be a report of a study that:

- Investigated patients with VSs.
- Was a full article report of a clinical study.
- Appeared in a peer-reviewed publication or a registry report.

- Enrolled a minimum of 30 patients.
- Was of humans.
- Was published between January 1, 2015, and May 20, 2021.
- Quantitatively presented results.
- Was published in English.
- Diagnosis was made radiographically or by pathology:
 - Enhanced MRI or heavily weighted T2 sequence (i.e., FIESTA sequences)
 - Histopathologic confirmation of VS
- Verified pure tone thresholds and word recognition with formal audiometry

Articles were excluded if it was determined they:

- were an in vitro study.
- were performed on cadavers.
- were medical records reviews, meeting abstracts, historical articles, editorial letters, or a commentary.
- Were a systematic review, meta-analysis, or guideline developed by others.
- Involved a distinct analysis of VS patients in reviews that included various pathologies of the IAC and Cerebellopontine angle (CPA)

Systematic reviews or meta-analyses conducted by others, or guidelines developed by others were not included as evidence to support this review due to the differences in article inclusion/exclusion criteria specified compared to the criteria specified by the Guidelines Task Force.

Assessment for Risk of Bias

Query of largely retrospective reviews of screening paradigms from multiple tertiary care centers is undoubtedly subject to a degree of selection bias given differing institutional protocols. It is likely that some VS patients were not effectively captured by screening protocols, and not all patients that met inclusion criteria were actually enrolled and able to complete an MRI.^{6,7} This would imply that the resulting data set does not represent the true number of VS cases screened by the tertiary centers. As the majority of citations meeting criteria came from institutions with dedicated otology/audiology centers, these centers will encounter a patient population with a higher incidence of hearing loss compared to the general population effectively overestimating

rates of audiology complaints and thus diagnosis of VS.⁸

Rating Quality of Evidence and Linking It to Recommendations

All included citations were retrospective in nature. All evidence incorporated into the current guidelines originate from mostly retrospective review of the screening protocols implemented by tertiary referral centers. Quality of evidence was classified using the Congress of Neurological Surgeons Guidelines Development Methodology (<https://www.cns.org/guidelines/guideline-development-methodology>). In summary, for diagnostic testing as is being addressed in this document, class I evidence is evidence provided by one or more well-designed clinical studies of a diverse population using a “gold standard” reference test in a blinded evaluation appropriate for the diagnostic applications and enabling the assessment of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios. Class II evidence is similar to class I evidence, but with the study being conducted in restricted population. Class III evidence is evidence provided by expert opinion or studies that do not meet the criteria for the delineation of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios. Class I evidence is used to support recommendations of the strongest type, defined as Level I recommendations, indicating a high degree of clinical certainty. Studies with less strength and designated as Class II evidence are used to support recommendations defined as Level II, reflecting a moderate degree of clinical certainty. Other publications including expert opinion, and studies with flaws that do not statistical calculations are considered Class III evidence and support Level III recommendations, reflecting unclear clinical certainty.

Revision Plans

In accordance with the National Academy of Medicine’s standards for developing clinical practice guidelines, the task force will monitor related publications following the release of this document and will revise the entire document and/or specific sections “if new evidence shows that a recommended intervention causes previously unknown substantial harm; that a new intervention is significantly superior to a previously recommended intervention from an efficacy or harms perspective; or that a recommendation can be applied to new populations.”⁹ In addition, the task force will confirm within five years from the date of publication that the

content reflects current clinical practice and the available technologies for the evaluation and treatment for patients with VS.

SUMMARY OF PREVIOUS GUIDELINE

The first version of these guidelines¹ addressed the same three study questions, yielding class III recommendations. With regard to patients presenting with asymmetric hearing loss with >10 dB of interaural difference at 2 or more contiguous frequencies or ≥ 15 dB at one frequency, MRI should be pursued to confirm or exclude the diagnosis of VS. In patients presenting with unilateral tinnitus, MRI again is recommended for the diagnosis of VS, though the yield is low (<1%). Lastly, with patients presenting with sudden onset of SNHL, an MRI again is recommended to minimize the incidence of undiagnosed VS, though again the yield is low (<1%). Since the publication of these results, there have not been any significant difference in institutional protocols for the screening of VS, and therefore the conclusions of the current guidelines remain similar to previous conclusions.

RESULTS

The literature search yielded a total of 704 abstracts from the electronic databases, with 704 remaining after removing duplicates. The authors reviewed all abstracts yielded from the literature search and identified the literature for full text review and extraction, addressing the clinical questions, in accordance with the literature search strategy. Task force members identified the published evidence meeting the stated inclusion criteria to answer the targeted clinical questions.

The task force selected 48 full-text articles for full text review. Of these, 34 were rejected for not meeting inclusion criteria or for being off-topic. Fifteen were selected for evidence table creation, text development and ultimately recommendation creation (Figure PRISMA). Assessment of this literature resulted in an update in the recommendations for each of the questions.

QUESTION 1

In adult patients with asymmetric SNHL on audiometric testing, is MRI or other diagnostic tests (such as CT, ENG) more effective in the diagnosis of VS?

Recommendation(s)

Level 3: MRI remains the most effective diagnostic measure that can differentiate VS from other labyrinthine conditions (e.g., labyrinthine hemorrhage), and is suggested when patients present with asymmetric hearing loss especially in the setting of abnormal auditory brainstem response test results.

STUDY SELECTION

704 abstracts were identified during the literature search process. The task force members first reviewed all abstracts from the targeted literature review search and then placed those articles meeting the inclusion criteria. Studies needed to have included formal audiometry, including verified audiometric thresholds and word recognition. After applying these criteria, the task force selected 48 full-text articles for full-text review. The task force pulled the papers for full-text extraction. Thirty-four were rejected for failing to meet inclusion criteria or for being off topic.

RESULTS OF INDIVIDUAL STUDIES

Seven articles met the inclusion criteria for this question.¹⁰⁻¹⁶ All studies were classified as III based on the single-center retrospective nature of the study design. In addition, one study was a single institutional cross-sectional study over two-time frames¹², and another was a single institution prospective series spanning five years.¹⁰ A summary of the study type, objective, methodology, summary, and the conclusion is included in the evidence table.

Asymmetric SNHL is a common, early symptom of retrocochlear pathologies, and thus audiometry could serve as an early, cost-effective marker of VS compared to MRI.^{17,18} However, there are many labyrinthine conditions that can present with similar symptom complaints including labyrinthitis, vascular conditions, and even labyrinthine hemorrhage to name a few. Often MRI is the only means to differentiate between retrocochlear and cochlear conditions. Kim et al. investigated MRI findings in patients with labyrinthine hemorrhage as a source of sudden asymmetrical SNHL.¹⁵ The retrospective review included 59 patients who met the inclusion screening (1512 initially screened), with 19 presenting with abnormal MRI results. Only six patients had high labyrinthine signals on pre-enhanced T1-weight and 3D FLAIR VISTA MRI with no contrast suggesting labyrinthine hemorrhage, and two patients had VS. The remaining

patients (n=11) were diagnosed with labyrinthitis. The work highlights the importance of MRI in diagnosing the cause of asymmetrical SNHL.

Aguilar and colleagues conducted a cross-sectional study from 2009-2010 and 2016-2018 to determine the best audiometric pattern that predicts VS in patients with asymmetric SNHL.¹² A total of 107 patients with asymmetric SNHL documented with pure tone audiometry were included in the retrospective review, nine of whom had confirmed VS. Asymmetric SNHL was defined as a difference of 15 dB in one or more frequencies between both ears. In addition, patients with abnormal audiology screening underwent MRI with contrast. Clinical decision analysis techniques (sensitivity, specificity, positive and negative predictive values, and accuracy) were conducted based on audiometric patterns. The patients were grouped based on the presence of VS (as determined by MRI). Interestingly, there was a significant difference in auditory brainstem response testing between the two groups ($p < 0.001$); however, no significant differences were observed based on audiometric patterns. No particular audiometric pattern demonstrated a high diagnostic yield for VS. The best clinical performance was indicated for greater than 20dB at 4000Hz with a sensitivity and specificity of 77.78% and 30.61%, respectively. However, when considering the area under the receive operating curve (AUC) of only 0.542, this audiometric variable's diagnostic utility for VS approaches simple chance performance. The authors concluded that the difference of >20dB at 4000Hz is a useful screening metric though is not a sufficient substitute for MRI.

As stated above, MRI is the gold standard diagnostic tool for identifying VS; however, patients presenting with asymmetrical SNHL often first present for audiometric testing to confirm reported symptoms. The use of audiometry to screen for VS presents many cost-saving values over MRI. Abbas et al.¹⁰ conducted a prospective study of 1126 patients first seen in audiology with reports of asymmetric SNHL or unilateral tinnitus to determine if this is a safe and cost-effective first or only mean to screen for VS. MRI was suggested when audiometric thresholds demonstrated a difference in ear thresholds of 15 dB or > at two or more adjacent frequencies. Out of 1126 patients, only 25 (2.22%) were diagnosed with VS. The authors stated that patients with asymmetrical SNHL can be screened by audiologic measures as a first and sometimes only means of evaluation. However, it is essential to note that the tumor size identified in this study

ranged from 3mm to 20mm. Audiometric behavioral and objective screenings can miss identifying tumors of smaller sizes. Therefore, even with a known decrease in sensitivity with small tumors, audiometric screening presents a cost-effective first step to evaluate for VS. Bielinka et al.¹⁴ argued that patients with asymmetrical SNHL should have auditory brainstem response (ABR) testing in addition to comprehensive audiometric testing. The authors reviewed patient charts from 2011-2016 to demonstrate that VS is responsible for inner ear dysfunction. Of 3,456 patients, 252 had abnormal ABRs (7.3%) and were referred for gadolinium-enhanced MRIs. Of these patients, 13 were diagnosed with VS. The ABR criteria warranted MRIs included the following: 1) waves I-II >2.55 ms, 2) waves III –V > 2.35 ms, and 3) waves I-V > 4.6 ms. The authors concluded that prolonged ABR waveform patterns should trigger further referral for neuroimaging (MRI of IAC with contrast). However, the sensitivity of ABR decreases with smaller tumor sizes: ~85% for smaller tumors compared to ~96% for more extensive tumors (Koors et al., 2013).

Kim et al.¹³ also evaluated the patterns of audiometric testing in 171 patients identified with CPA tumors (115 VS tumors) from 2001 to 2013. All patients completed pure tone and speech audiometry and ABRs. Audiometric results varied based on tumor size and location. Audiogram patterns were classified into the following groups: descending high-tone hearing loss type based on if the hearing threshold was lower at a high-tone than at a low-tone frequency, ascending low-tone hearing loss type if a hearing threshold was lower at a low tone than at a high-tone frequency, flat hearing loss type when similar thresholds were observed at all tested frequencies, concave hearing loss type when the primary frequency lowered was mid-tone, convex hearing loss type with preserved mid-frequency ranged hearing. VS tended to follow the descending pattern of hearing loss with increased pure-tone thresholds and symptoms of tinnitus as compared to non-VS-type tumors. Pure tone averages were significantly higher in patients with VS than in non-VS tumors of 11–25mm in size ($p<0.05$); tumors outside of this size range did not reach statistical significance. ABR testing was abnormal in 89.7% of the VS group and 81.8% in the non-VS group but it is unclear if this was statistically significant. Interestingly, there was no biopsy confirmation of tumors after surgery.

Earlier diagnosis of VS is essential to preserve facial nerve and hearing outcomes. Lee et al.¹⁹ explored audiometric test findings (pure-tone and speech audiometry), ABR results, and symptom characteristics in 114 patients prior to MRI. Asymmetric hearing loss with associated tinnitus was most frequently reported in the patients. Hearing loss is commonly reported in tumors ≤ 25 mm in size. In tumors greater than 25mm, tumors often present with dizziness. Like Kim et al. (2016), this study also found that descending hearing loss patterns are most common in VS. Lower speech discrimination scores correlated with severe to profound SNHL. Abnormal ABR patterns (prolonged latencies of waves I, III, and V or absent waves) were observed in 30-52% of the patients, and abnormal interaural latencies differences (waves I-III and III-V) in 68.1 to 40.9% of patients, respectively. However, there was no observed significant correlation between tumor size or site with a degree of hearing loss or speech discrimination score. The authors concluded that MRIs are necessary to confirm the suspicion of VS when patients have asymmetrical SNHL, tinnitus, abnormal ABR patterns, and poor speech discrimination scores. MRI is the only measure to confirm the size and site of the lesion.

Krane et al.¹¹ conducted a retrospective review from 1980-2013 to determine the clinical utility of comprehensive neurotologic testing in patients with primary symptoms of hearing loss, tinnitus, and dizziness. The retrospective review included 1170 patients who underwent various testing (audiometric, ABR, ENG, and MRI). Hearing loss was the most common symptom ($n = 762/1170$ patients) with 91% ($n = 1059$) with documented hearing loss and of those 82% ($n = 870$) had SNHL. ABR was performed in 911 patients, with 26% ($n = 234$) demonstrating abnormal results. ENG testing was completed in 1010 patients, and 34% ($n = 344$) had central findings and 35% ($n = 350$) with peripheral findings, and 31% with both central and peripheral findings. MRI was obtained in 1120 patients and identified 560 cases with abnormalities, yet only 68 (6%) were stemming from VS. The authors omitted to indicate the criterion used to recommend MRI screening, as all but 50 patients received neuroimaging. The study highlighted that a comprehensive neurotological approach to care could yield diagnoses that may go unrecognized otherwise.

Synthesis: MRI remains the gold standard diagnostic measure that can differentiate VS from other labyrinthine conditions (e.g., labyrinthine hemorrhage) and should be ordered when

patients present with asymmetric hearing loss and abnormal auditory brainstem response test results.

QUESTION 2

In adult patients with subjective complaints of asymmetric tinnitus, is MRI or other diagnostic tests (such as CT, ENG) more effective in the diagnosis of VS?

Recommendation(s):

Level 3: MRI screening is suggested for patients presenting with asymmetric tinnitus to minimize the incidence of undiagnosed VS, although the diagnostic yield is low.

STUDY SELECTION

A total of 704 studies were screened and evaluated for eligibility under the previously defined criteria, and 15 publications were included for the final review.^{10-14,19-21} The objective of this recommendation was to evaluate the presenting symptom of asymmetric tinnitus in the general population as a screening tool for the diagnosis of VS on MRI. Further, we sought to identify the frequency at which VS patients reported asymmetric tinnitus at the time of their presentation. To fulfill the objectives of this recommendation, we only considered studies reporting the presence of subjective, asymmetric tinnitus as either a solitary symptom or as part of a symptom profile in a patient screened for or diagnosed with VS. With the application of this exclusion criteria, 8 studies were included.^{10-14,19-21} Data extraction included study design, study objective, methodology, level of evidence, number of patients, number of tumors found in the setting of asymmetric tinnitus, and individual study conclusions (Appendix IV: Evidence Table). There were no included institutions with multiple papers, therefore there was no risk of duplicate reporting of data.

RESULTS OF INDIVIDUAL STUDIES

Of the 8 studies meeting inclusion criteria and addressing this question, only one encompassed a cohort of patients with asymmetric tinnitus as the sole presenting symptom without any form of hearing loss.²⁰ The remaining 7 studies were comprised of patients with asymmetric tinnitus in conjunction with some other form of otologic complaint, most often with asymmetric SNHL.^{10-14,19,21} All studies represent class III data due to lack of blinded assessment and absence of a validation set. Specific data from each citation can be found in Appendix IV, the Evidence Table.

Saxby et al.²⁰ employed MRI in the investigation of patients with unilateral non-pulsatile tinnitus without asymmetrical hearing loss over a 5-year period. In their retrospective analysis, 566 MRIs were conducted which ultimately led to the diagnosis of a VS in 3 patients (0.53%). Two of these three patients had tinnitus ipsilateral to their tumor, measuring between 3-5mm. The third patient was found to have a 4mm VS contralateral to the tinnitus. With such a low diagnostic yield for VS, the authors call into question the utility of MRI for patients presenting with asymmetric tinnitus without associated SNHL.

Lee et al.¹⁹ retrospectively investigated the presenting symptoms of clinical manifestations of known VS patients to determine factors associated with VS before an MRI is performed. The study involved 113 patients from a tertiary referral center over a 12-year period. The main presenting symptoms was asymmetric SNHL, present in 64.6% of patients, with accompanying symptom of tinnitus in 48% of patients. Interestingly, the rates of tinnitus at time of presentation correlated with increasing tumor size up to 30 mm in maximum diameter, then tapered off thereafter. The authors ultimately conclude that an MRI is warranted in patients presenting with asymmetric SNHL, especially if accompanied by tinnitus, though do not comment on the diagnostic yield of MRI for VS in patients with tinnitus alone.

The remaining 6 citations focused on all otologic presenting symptoms of patients referred to tertiary referral centers that eventually led to the diagnosis of VS.^{10-14,21} Presenting symptoms in these publications included SNHL, tinnitus, gait instability, and other signs of mass effect caused by VS, though the objective of each citation differed. Kim et al.¹³ examined two cohorts of patients, one being known VS patients while the other representing non-VS lesions of the CPA, to investigate any potential differences in presenting otologic complaints. Of the 171 patients included, 116 were diagnosed histologically with VS while the remaining 5 had some other form of CPA tumor. The most common presenting symptom in each cohort was expectedly SNHL, however VS tended to present with tinnitus as an additional symptom while non-VS tumors had a higher incidence of gait instability and dizziness. Tinnitus as the chief complaint was equally represented amongst cohorts (7.2% versus 7.9%), though tinnitus was recorded as a presenting symptom in the VS cohort in nearly double the rates of the non-VS cohort (40% versus 22.2%).

Abbas et al.¹⁰ describe the development of a protocol resulting from investigating whether an audiologist is sufficient to screen patients presenting with a variety of otologic complaints without the need for further otolaryngology assessment. The authors conducted a prospective cost-analysis study at a tertiary referral center screening 1126 patients in an audiology clinic with asymmetric SNHL and/or unilateral tinnitus to identify the diagnostic yield of MRI for VS prompting additional otolaryngology referral. All

presenting patients underwent MRI for screening purposes which diagnosed a VS in 2.22% (n=25/1126) of the screened patients. However, this study is limited in that there is no mention of rates of SNHL or tinnitus in the individual patients diagnosed with VS. Therefore, it is not possible to draw meaningful conclusions other than to acknowledge that the majority of patients presenting for complaints typically associated with VS are often not actually caused by VS and will not require referral to otolaryngology.

The remaining four studies investigated the prevalence of VS as a causative lesion for first signs of hearing dysfunction or tinnitus.^{11,14,21} While the primary focus of Yang et al²¹ was to describe the rate of VS being the causative lesion for sudden SNHL, the authors' cohort did also specify the existence of tinnitus as an additional presenting symptom. Amongst the 1249 patients undergoing screening MRI for a chief complaint of sudden SNHL, 1.12% (n=14/1249) were ultimately diagnosed with VS. Of the VS patients, 78.6% (n=11/14) had co-existing tinnitus at time of diagnosis. However, this study is limited in that the rates of tinnitus in the non-VS population is not reported. Krane et al¹¹ aimed to determine the clinical efficacy of comprehensive neurotologic testing in patients presenting with complaints of hearing loss, tinnitus and/or dizziness in the ultimate diagnosis of VS. 1170 patients were assessed over a 33 year span, all undergoing screening MRI. Of the 1170 patients, 56% (n=657/1170) complained of tinnitus at time of presentation, with an abnormal MRI in only 30% of tinnitus patients. VS was diagnosed in 6% (n=48/1170) of cases, although the MRI was diagnostic for some form of causative lesion in 48% (n=536/1170) of cases. The authors conclude that tinnitus is not a reliable screening symptom for VS. Similarly, Bielinskha et al¹⁴ investigated the rates of VS as a causative lesion of initial signs of inner ear dysfunction. 3456 patients presented to the authors' tertiary care center for otologic complaints though only 13 (0.4%) were ultimately diagnosed with VS. The VS cohort of patients reported tinnitus as a presenting symptom in 92% of cases, although this matched the rate of tinnitus in non-VS patients also at 92%. The authors conclude concur with previous findings in that tinnitus is often present with a new diagnosis of VS but is not specific to the pathology. Aguilar et al¹² also investigated the rate of VS diagnosis with new otologic complaints with screening MRI conducted in 107 patients. Of the 107 patients, only 8.4% (n=9/107) were diagnosed with VS. Again, the rates of tinnitus at time of presentation was similar between the VS cohort (100%) and non-VS cohort (94.8%) making tinnitus non-specific to VS.

Synthesis: These 8 studies analyzed the examined the association of asymmetric tinnitus with the diagnosis of VS. The diagnosis of VS was made based upon MRI and confirmed pathologically in the majority of studies. A total of 9459 MRIs performed for the presenting symptom of asymmetric tinnitus, the diagnostic yield for VS was approximately 1.56%, though most were in conjunction with other

otologic complaints such as SNHL. In total, only 2066 MRIs were performed for the sole complaint of asymmetric tinnitus, with a diagnostic yield of less than 1% (n=3) or VS. However, of the VS patients included in the cohort, a majority of patients did report asymmetric tinnitus at time of diagnosis. In this regard, it appears asymmetric tinnitus correlates more with asymmetric hearing loss as opposed with VS. Although the incidence of VS in the asymmetric tinnitus population remains low, it is suggested the practitioner still screens for IAC pathologies with MRI.

QUESTION 3

In adult patients with verified sudden SNHL on audiometric testing, is MRI or other diagnostic tests (such as CT, ENG) more effective in the diagnosis of VS?

Recommendation

Level 3: MRI is suggested for the diagnosis of VS in the setting of sudden SNHL.

STUDY SELECTION

Seven hundred and four abstracts were identified during the literature search process. The task force members reviewed all abstracts against inclusion criteria and selected those appropriate to address clinical question 3. Inclusion criteria pertained to 1) studies with at least 30 patients and diagnosis of VS either radiographically or histopathologically, and 2) formal audiometry, including verified audiometric thresholds and word recognition. The diagnostic effectiveness of MRI vs. alternative diagnostic tests in identifying VS for patients with sudden SNHL was the basis for this review. After applying these criteria, the task force selected 48 full-text articles for full-text review. The papers were pulled for full-text extraction (see PRISMA diagram) and summarized to answer the clinical question. Seven retrospective review articles (class III data) met the inclusion criteria for question 3.^{16,21-26}

RESULTS OF INDIVIDUAL STUDIES

VS typically presents with progressive hearing loss, tinnitus, and balance concerns; however, cases of sudden SNHL can occur with VS. However, the prevalence of sudden SNHL arising from VS is relatively low. In a recent retrospective review, Fujita et al.²³ analyzed MRI findings in 499 patients with sudden SNHL. Only 15 patients (prevalence 3.0 %) were identified as having VS, with tumor sizes ranging from eight grade 1 (intracanalicular tumor), six grade II (up to 2 cm) to one grade III tumor (up to 3 cm). No grade IV tumors were found. The authors stated that additional screening beyond audiometric testing for documenting hearing loss is necessary to screen for VS. This work also found that non-

contrast, high-resolution, three-dimensional T2WI or T2*WI MRI is a cost-effective alternative to contrast MRI for VS screening.

Sudden SNHL is defined as sensorineural hearing decrease > 30 dB over at least three consecutive frequencies in pure tone audiometry, occurring within 72 hours.²⁷ While there is a clear definition for sudden SNHL, there is a lack of consensus on the clinical features that can distinguish VS from other causes of sudden SNHL.²⁴ Labyrinthitis, labyrinthine hemorrhage, and vascular causes may all present as sudden SNHL.²⁸ Moreover, the recovery and audiometric patterns are similar between VS and other non-tumorous causes of sudden SNHL. According to the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) guidelines, MRI or ABR should be used for the retrocochlear pathology evaluation of patients with sudden SNHL.²⁸ Jeong et al.²⁴ examined the value of MRI of the internal auditory canal (IAC) within ten days of symptom onset to determine the cause of sudden SNHL. A review of 291 patients revealed that only 4.5% (13 of 291) presenting with sudden SNHL had an abnormal MRI of the IAC, and the most common abnormality detected in these patients was VS (9/13 or 69.2%). However, tumor sizes were small to medium, and tumors were not verified with histopathologic examination. A strength of this work is that MRI-identified VS in patients with sudden SNHL further supports the inclusion of neuroimaging when patients present with symptoms of sudden SNHL. Cho et al.²⁵ also investigated the frequency of IAC lesions in patients with sudden SNHL. The ten-year retrospective review included 200 consecutive patients with sudden SNHL who had an MRI within three days of symptom onset and received 60 mg of prednisone over seven days. Twenty-five patients (12.5% of the sample) had abnormal MRI, with VS identified in 9/25 (36%) patients in this group, which was 4.5% of the total (n=200). Interestingly, patients with IAC tumors had poorer hearing recovery following prednisone therapy than the idiopathic sudden SNHL group. This study also examined the presence of spontaneous nystagmus, headshake nystagmus, and canal paresis via a videonystagmography system. There were no significant differences in canal paresis between tumorous and non-tumorous groups. Nevertheless, statistically significant differences between spontaneous and headshake nystagmus were observed between groups, with more nystagmus findings observed in the non-tumorous groups. Additionally, more patients in the non-tumorous groups presented with secondary vestibular disorders (benign paroxysmal positional vertigo [BPPV]). Non-tumorous causes should be considered when patients present with vascular risk factors, BPPV, canal paresis on VNG, and symptoms of acute vertigo. Larger scale studies are necessary due to the small sample of patients identified with IAC lesions.

Cho et al.²⁵ suggested that patients with retrocochlear pathologies such as VS have poorer hearing recovery outcomes after steroid treatment. It stands to reason that documenting the hearing improvement

with steroids may be a sensitive predictor of the presence of VS. In a large sample (n= 420) retrospective review of patients presenting with sudden SNHL, Ungar et al.²² conducted a comparative analysis for those patients with and without VS responding to 5 days of oral prednisone. Twenty patients were identified as having VS (4.76%), and 80% (16/20) of those with VS had documented improved hearing after the course of steroids. In contrast, only 60% (240) of patients without VS had improvement in audiometric testing after treatment. Based on these findings, two clinical indicators are needed to recommend MRI scanning: 1) documentation of improvement in hearing within one week of steroid treatment and 2) a low speech discrimination score.

Additional work by Lee et al.¹⁶ reviewed 31 patient charts to determine the value of MRI in cases of isolated acute audiovestibular loss. Thirty-one patients presenting with isolated acute audiovestibular loss were retrospectively reviewed. All 31 patients had positive findings on a three-step bedside examination of head impulse, nystagmus type, test of skew (HINT). Fifteen patients demonstrated increased labyrinthine FLAIR signal on MRI: 11 had negative MRIs, two had AICA territory strokes, and three had confirmed VS (9% prevalence). All of the VS were confined to the internal auditory canal.

Additionally, there was no significant difference between pure tone audiometry and recovery rates amongst pathologies. Isolated acute audiovestibular loss is most commonly due to labyrinthitis as opposed to AICA stroke or VS. However, the authors recommended an abbreviated MRI protocol to identify the exact etiology. The work presents some limitations most notably a small sample size limiting generalizability to a larger population.

Ren and colleagues examined clinical predictors in 120 patients with sudden SNHL who completed audiometric testing (pure-tone testing, acoustic immittance, and ABR) in addition to MRI.²⁶ Of the 120 reviewed patients, 42 (35%) had abnormal MRI results, yet only three (2.5%) were identified with VS. All of the patients completed MRI after lack of improvement of hearing loss after a course of steroids. Of the three patients with identified VS, two presented with no response on ABR testing and had severe SNHL, while the third patient (with moderate sudden SNHL) had normal ABR results. The authors recommended MRI when patients with sudden SNHL demonstrating severe SNHL and no response to ABR. The authors further recommend MRI for patients with sudden SNHL with ineffective response after one week of steroid treatment. The authors surmised that when patients with sudden SNHL present with severe SNHL and no response to ABR tests, MRI imaging is clinically beneficial, especially for patients with profound sudden SNHL. One noted concern with this summary statement is that ABR is often absent when hearing loss is in the severe SNHL range, particularly for high-frequency hearing loss, and is not

necessarily a marker for retrocochlear pathology. This summary adds further support for MRI scanning to rule out retrocochlear pathology rather than alternative measures when patients present with clinical characteristics of sudden SNHL, unilateral tinnitus, and vestibular symptoms. However, it also underscores the lack of consensus regarding patient response to high dose steroid administration in the VS population presenting with sudden SNHL. The conclusions of Ren.²⁶ and Cho²⁵ suggested both suggest that sudden SNHL in the VS population does not improve after steroid administration. However, Ungar²² concludes that symptoms of sudden SNHL do improve following steroid administration. Given the evidence presented, it is not possible to discern if VS patients presenting with sudden SNHL are more likely to respond to steroids. It is also important to realize several pathologies more common than VS also present with sudden SNHL that are improved by rapid steroid treatment. It is often the case that patients will have received steroids prior to being able to undergo MRI to diagnose a potential VS.

Auditory brainstem response testing may have a high diagnostic yield as an initial screener, but sensitivity is dependent on tumor size, and as aforementioned, the ability to complete the test is contingent on hearing loss status. Yang et al.²¹ conducted a review from 2009-2019 in southern China to determine the prevalence and clinical features of VS in patients with sudden SNHL. A large sample (n = 1249) of patients who underwent ABRs and MRI were included in the study. One thousand two hundred forty-nine patients were included in the review, with only 14 (1.12%) diagnosed with VS. ABRs were abnormal in 12/14 of the patients identified with VS, yielding a sensitivity of 85.7%. However, the majority of the patients had small tumors (grade 1, or < 10mm), which significantly reduced the sensitivity (71.4%) for ABR to identify VS.

Synthesis: Sudden SNHL may be secondary to labyrinthine and retrocochlear causes. The overall prevalence of VS causing sudden SNHL is relatively low. MRI scanning remains the gold standard for identification although the expected diagnostic yield is low. Clinical features such as poor word recognition scores, profound SNHL, and abnormal or absent auditory brainstem response testing may also trigger the necessity for performing MRI. The patient response to high dose steroid administration for sudden SNHL should not guide the need for an MRI. Audiometric testing, both behavioral and objective (e.g., ABR), is inefficient in identifying VS in cases of sudden SNHL; only MRI can identify the site and size of the tumor and help to rule out other etiologies presenting with sudden SNHL.

DISCUSSION

The first rendition of guidelines on audiologic and otologic screening for patients with VS ultimately favored the use of MRI for patients with (1) SNHL >10 dB of interaural difference at 2 or more contiguous frequencies or ≥ 15 dB at one frequency, (2) sudden SNHL regardless of how profound, and (3) unilateral tinnitus. The recommendations were based on level 3 evidence. With respect to tinnitus and sudden SNHL, the recommendations identified that the diagnostic yield of MRI for VS in these cohorts approached 1%, though ultimately advocated for image screening in an effort to minimize undiagnosed tumors. Since the initial publication of these guidelines, no major shifts in the field or development of screening protocols have been made that dramatically affect the conclusions in the current work. While the recent data sets support a slight increase in the incidence of VS in patients presenting with otologic complaints up to 1-3%, this is likely reflective of more liberal screening protocols.

It is clear that an MRI offers a high degree of sensitivity such that an existing VS will be detected in virtually all scenarios.²⁹⁻³¹ If the goal of screening protocols were to simply detect all existing VS at time of symptom onset, then the role of the physician would be quite simple: any patient complaining of SNHL >10 dB at 2 or more contiguous frequencies, sudden SNHL, or any degree of asymmetric tinnitus would require an MRI which would in most certainty detect the causative lesion. However, we exist in a world of limited resources, and as such must operate under somewhat restrained conditions. Rather, the more accurate goal is to maximize the number of VS detected based on objective audiologic and otologic data in order to minimize patient morbidity from such tumors going undetected, all the while under the confinement of limited resources of the healthcare system. As the overall cost of an undetected tumor outweighs the risk of a negative MRI in a patient with otologic complaints, we allow for generous screening protocols leading to MRIs with low diagnostic yields given the presenting symptoms. As imaging technologies advance, the question is no longer which imaging platform has the highest sensitivity for the detection of a VS, but which patient is most appropriate to undergo an MRI?

With respect to unilateral tinnitus or sudden SNHL, our cumulative body of evidence is clear: in isolation these presenting symptoms have an exceedingly low chance of being caused by a VS (1.56% and 3.66%, respectively). Similarly, the chance of VS as a causative lesion in a patient presenting with SNHL >10 dB of interaural difference at 2 or more contiguous frequencies or ≥ 15 dB at one frequency is merely 1.66%, though in conjunction with tinnitus the risk profile does mildly increase. While it can be debated that certain factors can increase the likelihood that these symptoms are VS-derived (i.e. improvement of sudden SNHL with steroids)^{32,33}, the end result of obtaining an MRI is not always evidenced-based as

seen by the wide variety of adopted screening protocols at high volume tertiary centers included in these update guidelines. Practitioners continue to screen patients with these symptoms due to high risk aversion for an undetected tumor. For instance, current evidence suggests that the overwhelming majority of sudden SNHL presentations are due to infectious or vascular causes which is logical given the acuity of symptom onset.^{34,35} As VS are slow growing, it should reasonably follow that such an acute onset of symptoms is unlikely to be caused by the progressive slow growth of an otherwise benign tumor. Regardless, we continue to observe screening protocols advocating for MRI on the 3.66% chance we detect VS. Similarly, in the patients presenting purely with tinnitus not coinciding with SNHL the diagnostic yield of MRI was 0.3%.²⁰ Thus, it is unlikely that foregoing an MRI in the setting of sudden SNHL will miss a VS. Furthermore, of the pure tinnitus cohort, the VS was detected at less than 1cm making the clinical urgency to treat non-existent.

With the diagnostic yield of MRI for diagnosis of VS hovers between 1.56-3.66% amongst the current dataset, it is important to realize this diagnosis rate is not too different from other more widespread imaging screening protocols. For example, the diagnostic yield of a mammogram to diagnose breast cancer in a high risk population for breast cancer is estimated to be 1.2%.³⁶ While we do observe low diagnostic yields of specifically for VS on MRI for patients presenting with SNHL, tinnitus, or sudden SNHL, we cannot dismiss the fact that an abnormal MRI leading to a different clinical diagnosis is made in approximately 15% of cases. The MRI with contrast would serve as a useful screen to determine if an MRV is necessary. Incorporating the vascular imaging component upfront would shift the narrative away from tinnitus and onto a dissertation on causes of sinus stenosis. If we believe the goal of screening protocols is to effectively utilize resources to make a diagnosis while minimizing rates of negative MRI scans, then the current screening protocols do seem to approach competency for that criterion.

KEY ISSUES FOR FUTURE RESEARCH AND CONCLUSIONS

VS remain the most common tumor of the CPA, often presenting with some form of SNHL and/or tinnitus. The screening protocols employed to detect VS vary considerably across institutions given economic restraints of their given healthcare system. While an MRI is able to detect existing tumors with exceedingly high sensitivity and sensitivity, the true challenge is deciphering which patient's symptoms are most reasonably caused by a VS warranting an MRI in the first place. Current screening protocols continue to rely upon objective data gathered by less resource intensive modalities such as audiology screening, with a threshold of SNHL required before an MRI is considered judicious. This reliance on

audiology screening harbors the inherent risk of missing the diagnosis of some VS as evidenced by some tumors present without hitting the required threshold to prompt follow up MRI. Similarly, despite the low diagnostic yield of unilateral tinnitus or sudden SNHL, we currently lack more eloquent methods of objectively assessing these presenting symptoms as they relate to VS and therefore opt to screen all such patients.

Future screening protocols will likely need to extend past the current metrics to warrant MRI. As the natural history of these tumors are often slow growing, perhaps multiple audiologic screenings over time are a better use of resources compared to the current threshold necessary to trigger an MRI. Similarly, the incorporation of other factors unrelated to SNHL or tinnitus such as readily available genetic data or social history (noise damage) might offer additional insights to improve the diagnostic yield of MRI for VS. Nonetheless, in current practice the conclusions of these guidelines closely mirror those of the initial publication due to the lack of significant changes to screening protocols during the time interval and continued risk aversion of practitioners that accommodate negative MRIs over a delayed diagnosis of VS.

Conflicts of Interest

All Guideline Task Force members were required to disclose all potential COIs prior to beginning work on the guideline, using the COI disclosure form of the AANS/CNS Joint Guidelines Review Committee. The CNS Guidelines Committee and Guideline Task Force Chair reviewed the disclosures and either approved or disapproved the nomination and participation on the task force. The CNS Guidelines Committee and Guideline Task Force Chair may approve nominations of task force members with possible conflicts and restrict the writing, reviewing, and/or voting privileges of that person to topics that are unrelated to the possible COIs.

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Disclaimer of Liability

This clinical systematic review and evidence-based guideline was developed by a physician volunteer task force as an educational tool that reflects the current state of knowledge at the time of completion. Each chapter is designed to provide an accurate review of the subject matter covered. This guideline is 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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Appendix I: Literature Searches

Ovid MEDLINE(R)

- | | | |
|---|---|---------|
| 1 | exp Hearing Loss/ or HEARING.mp. | 154256 |
| 2 | (hypoacuses or hypoacusis or deafness*).ti,ab,kw. | 24135 |
| 3 | AUDITORY DEFECT*.ti,ab,kw. | 66 |
| 4 | Tinnitus/ | 8968 |
| 5 | tinnitus*.ti,ab,kw. | 13248 |
| 6 | or/1-5 | 165821 |
| 7 | DIAGNOS*.mp. | 5610758 |
| 8 | exp Magnetic Resonance Imaging/ | 511451 |

- 9 (magnetic resonance imag* or chemical shift imaging* or mr tomograph* or magnetization transfer contrast imaging or nmr imaging or nmr tomograph* or proton spin tomograph* or spin echo imaging or spin echo imagings or zeugmatograph* or fmri or magnetic resonance tomograph* or mr imaging* or MRI or MRIs).ti,ab,kw. 366096
- 10 tomography, x-ray computed/ or exp four-dimensional computed tomography/ or exp positron emission tomography computed tomography/ or exp single photon emission computed tomography computed tomography/ 426211
- 11 (ct x ray* or cine ct or cine-ct or computed x ray tomograph* or computed x-ray tomograph* or electron beam computed tomograph* or electron beam tomograph* or tomodensitometr* or transmission computed tomograph* or x ray computer assisted tomograph* or x ray computerized axial tomograph* or x ray computerized tomograph* or x-ray computed tomograph* or x-ray computer assisted tomograph* or x-ray computerized axial tomograph* or x-ray computerized tomograph* or xray computed tomograph*).ti,ab,kw. 8082
- 12 Electronystagmography/ 3093
- 13 Electronystagmograph*.ti,ab,kw. 1684
- 14 (electric nystagmogram* or electric nystagmograph* or electrical nystagmogram* or electrical nystagmograph* or electro nystagmogram* or electro nystagmograph* or electronystagmogram* or photoelectronystagmograph*).ti,ab,kw. 211
- 15 (cat scanning or cat scan or cat scans or CT scan or CT scans or computed tomograph* or computer tomograph* or computerised axial tomograph* or computerised tomograph* or computerized axial tomograph* or computerized tomograph*).ti,ab,kw. 407502
- 16 or/7-15 5951743
- 17 6 and 16 52885
- 18 exp Neuroma, Acoustic/8763
- 19 ((vestib* or acoustic) adj3 (neuroma* or neurilemmoma* or neurilemoma* or neurinoma* or tumor* or tumour* or schwannoma*)).mp. [mp=title, abstract, original title, name of substance word, subject heading word, floating sub-heading word, keyword heading word, organism supplementary concept word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier, synonyms] 11046
- 20 (acoustic nerve cancer* or acoustic neurofibroma* or acusticus neurinoma* or auditory nerve neurinoma* or ear schwannoma* or angle tumor* or angle tumour* or cerebellopontine angle tumor* or neurinoma of the acoustic nerve or neurosensory deafness* or sensoryneural deafness* or sensory neural deafness*).ti,ab,kw. 1211
- 21 18 or 19 or 20 12489
- 22 limit 21 to english language 10469
- 23 Animals/ not Humans/ 4974929
- 24 22 not 23 10374
- 25 comment/ or editorial/ or letter/ or review/ or systematic review/ 5102112

26	24 not 25	8685	
27	exp adolescent/ or exp child/ or exp infant/	3849849	
28	exp Adult/	7797507	
29	27 not 28	2052582	
30	26 not 29	8366	
31	limit 30 to dt=20150101-20220520	2297	
32	in vitro techniques/	387712	
33	Culture Techniques/	47809	
34	Drug Evaluation, Preclinical/	54481	
35	Disease Models, Animal/	383220	
36	Xenograft Model Antitumor Assays/	44247	
37	31 not (32 or 33 or 34 or 35 or 36)	2275	
38	17 and 37	488	

Embase

('hearing impairment'/exp OR 'hearing impairment':ti,ab,kw OR 'hearing loss':ti,ab,kw OR 'hearing losses':ti,ab,kw OR 'auditory defect':ti,ab,kw OR deaf:ti,ab,kw OR deafness:ti,ab,kw OR 'hearing damage':ti,ab,kw OR 'hearing defect':ti,ab,kw OR 'hearing difficulty':ti,ab,kw OR hypacusia:ti,ab,kw OR hypacusis:ti,ab,kw OR hypoacusia:ti,ab,kw OR hypoacusis:ti,ab,kw OR 'impaired hearing':ti,ab,kw OR 'hearing'/exp OR 'hearing':ti,ab,kw,de OR audition:ti,ab,kw OR 'auditory function':ti,ab,kw OR 'auditory perception':ti,ab,kw OR 'noise perception':ti,ab,kw OR 'sound perception':ti,ab,kw OR 'tinnitus'/exp OR 'tinnitus':ti,ab,kw) AND ('diagnosis'/de OR diagnos*:ti,ab,kw,de OR 'nuclear magnetic resonance imaging'/exp OR 'magnetic resonance imaging':ti,ab,kw,de OR 'magnetic resonance tomography':ti,ab,kw OR 'magnetization transfer imaging':ti,ab,kw OR 'mr imaging':ti,ab,kw OR mri:ti,ab,kw OR 'nmr imaging':ti,ab,kw OR mris:ti,ab,kw OR 'chemical shift imaging':ti,ab,kw OR 'mr tomography':ti,ab,kw OR 'magnetic resonance image':ti,ab,kw OR 'magnetic resonance images':ti,ab,kw OR 'magnetization transfer contrast imaging':ti,ab,kw OR 'nmr tomography':ti,ab,kw OR 'proton spin tomography':ti,ab,kw OR 'spin echo imaging':ti,ab,kw OR 'spin echo imagings':ti,ab,kw OR zeugmatograph*:ti,ab,kw OR fmri:ti,ab,kw OR 'x-ray computed tomography'/exp OR 'x-ray computed tomography':ti,ab,kw OR 'x-ray tomography'/exp OR 'ct scan':ti,ab,kw OR 'ct scanning':ti,ab,kw OR 'ct scans':ti,ab,kw OR 'four dimensional computed tomography'/exp OR 'four dimensional computed tomography':ti,ab,kw OR '4-dimensional computed tomography':ti,ab,kw OR '4-dimensional ct':ti,ab,kw OR '4d computed

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Appendix II: Rating Evidence Quality

Classification of Evidence on Therapeutic Effectiveness and Levels of Recommendation

Class I Evidence Level I (or A) Recommendation	Evidence from one or more well-designed, randomized controlled clinical trial, including overviews of such trials.
Class II Evidence Level II (or B) Recommendation	Evidence from one or more well-designed comparative clinical studies, such as non-randomized cohort studies, case-control studies, and other comparable studies, including less well-designed randomized controlled trials.
Class III Evidence Level III (or C) Recommendation	Evidence from case series, comparative studies with historical controls, case reports, and expert opinion, as well as significantly flawed randomized controlled trials.

Classification of Evidence on Prognosis and Levels of Recommendation

Class I Evidence Level I (or A) Recommendation	All 5 technical criteria above are satisfied.
Class II Evidence Level II (or B) Recommendation	Four of five technical criteria are satisfied.
Class III Evidence Level III (or C) Recommendation	Everything else.

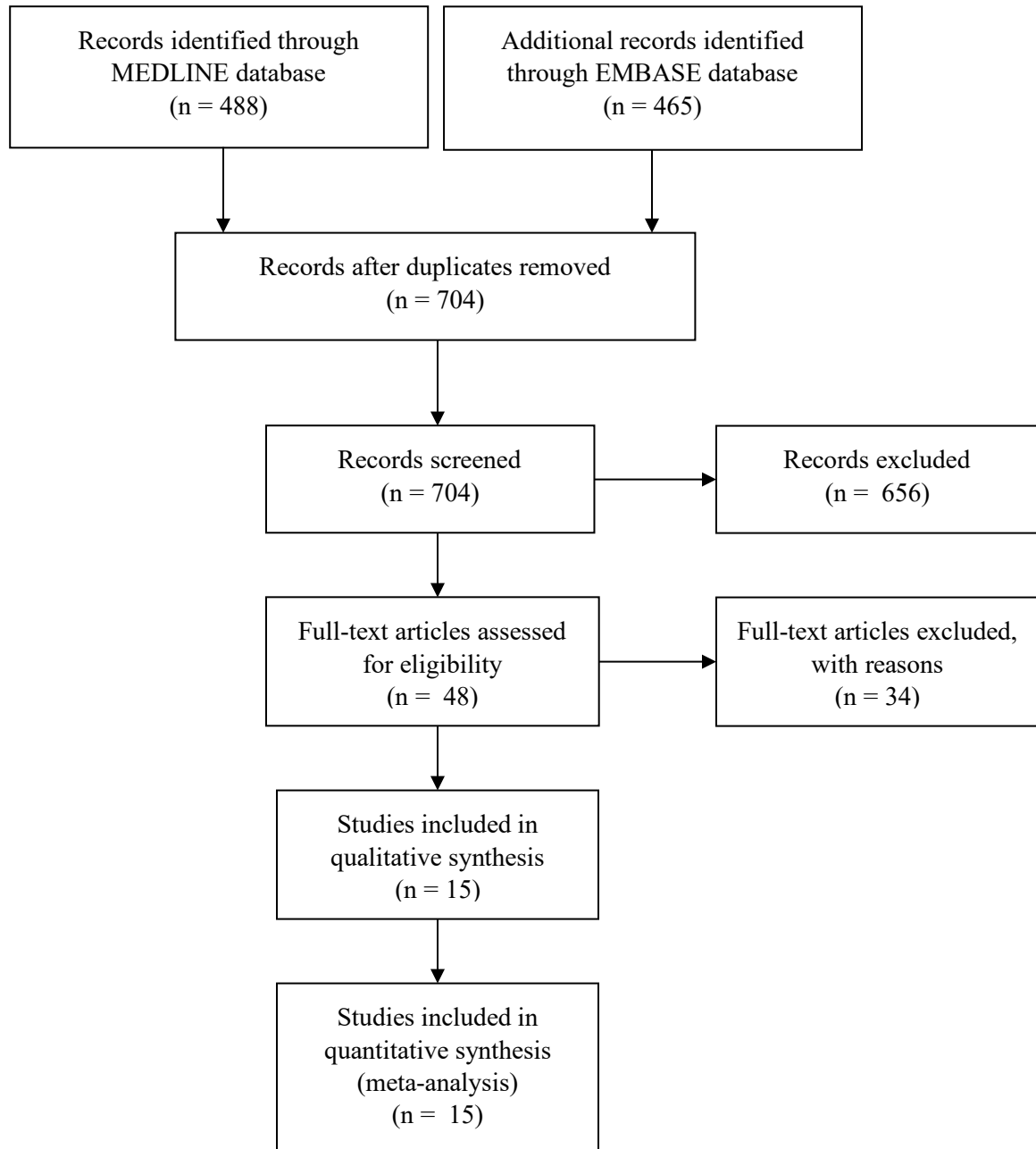
Classification of Evidence on Diagnosis and Levels of Recommendation

Class I Evidence Level I (or A) Recommendation	Evidence provided by one or more well-designed clinical studies of a <i>diverse</i> population using a “gold standard” reference test in a blinded evaluation appropriate for the diagnostic applications and enabling the assessment of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios.
Class II Evidence Level II (or B) Recommendation	Evidence provided by one or more well-designed clinical studies of a <i>restricted</i> population using a “gold standard” reference test in a blinded evaluation appropriate for the diagnostic applications and enabling the assessment of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios.
Class III Evidence Level III (or C) Recommendation	Evidence provided by expert opinion or studies that do not meet the criteria for the delineation of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios.

Classification of Evidence on Clinical Assessment and Levels of Recommendation

Class I Evidence Level I (or A) Recommendation	Evidence provided by one or more well-designed clinical studies in which interobserver and/or intraobserver reliability is represented by a Kappa statistic > 0.60.
Class II Evidence Level II (or B) Recommendation	Evidence provided by one or more well-designed clinical studies in which interobserver and/or intraobserver reliability is represented by a Kappa statistic > 0.40.
Class III Evidence Level III (or C) Recommendation	Evidence provided by one or more well-designed clinical studies in which interobserver and/or intraobserver reliability is represented by a Kappa statistic < 0.40.

Appendix III: PRISMA Flowchart



Appendix IV. Evidence Table

Author/year	Study Description	Data Class	Conclusion
Aguilar et al. 2022	Study Type: Single institutional cross-sectional study from 2009-2010 and then 2016-2018. Objective: To identify the audiometric pattern that best serves as a predictor for VS in patients with asymmetric SNHL Methodology: Retrospective analysis of 107 patients presenting with asymmetric SNHL on pure tone audiometry. Hearing loss was defined by a difference of 15 dB in one or more frequencies between both ears. Patients with abnormal audiology screening underwent MRI with contrast. The sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV) and accuracy of different audiometric patterns were analyzed.	III	Summary: Of the 107 patients with asymmetric SNHL analyzed, only 8.4% (n=9) were determined to have VS while the remaining 91.6% (n=98) had no discernable CPA lesion. No significant difference in audiometric patterns were found in patients with and without VS. A difference of >20dB at 4000Hz was the best audiometric screening test with sensitivity 77.78%, specificity 30.61%, PPV 8.33%, and NPV 93.75%. Conclusions: While a difference of >20dB at 4000Hz is a reasonable screening test for VS in patients with asymmetric SNHL, however, this is not sufficient to forego an MRI. Commentary: This manuscript is retrospective in nature and therefore yields class III data.
Saxby et al. 2021	Study Type: single institution retrospective review from 2014 to 2019 Objective: To evaluate MRI in the investigation of patients with unilateral tinnitus without associated SNHL Methodology: Retrospective review of 2066 patients undergoing RI of the internal auditory canal for symptoms of unilateral tinnitus without associated SNHL.	III	Summary: Of the 2066 patient MRIs reviewed, only 0.3% (n=3) VS were diagnosed, while 134 other incidental findings were discovered. Conclusions: The authors question the utility of MRI in the diagnosis of VS in patients presenting with unilateral tinnitus without associated SNHL. Commentary: No pure tone audiology or speech testing were given in this manuscript as no patients complained of, or were demonstrated to have, SNHL at time of MRI.
Yang et al. 2020	Study Type: Single-institution retrospective review from 2009-2019 Objective: To review the prevalence and clinical characteristics of vestibular schwannoma (VS) in patients with sudden SNHL in southern China. Methodology: Patients presenting with sudden SNHL undergoing audiology screening and subsequent MRI were eligible for inclusion. Sudden SNHL was defined as sensorineural hearing decrease of ≥ 30 dB over at least 3 consecutive frequencies in pure tone audiometry, which developed within a period of 72 hours.	III	Summary: 1249 patients met inclusion criteria, of which 14 (1.12%) were diagnosed with VS. Of the VS, the majority were Koos grade I (n=7), followed by grade II (n=4), and fewer grade III (n=3). Abnormal ABRs were noted in 12/14 patients, with hearing recovery observed in 3/14. Conclusions: VS were diagnosed in 1.12% of patients presenting with sudden SNHL. Predicting the presence of a VS with audiology screening and ABRs is insufficient, and this patient population should undergo MRI to rule out VS. Commentary: This manuscript is retrospective in nature and therefore yields class III data.
Fujita et al. 2019	Study Type: Single-institution retrospective review from 2008-2017 Objective: To assess the prevalence of vestibular schwannoma (VS) in patients with sudden SNHL. Methodology: Retrospective review of 499 patients presenting with sudden SNHL as defined by loss of 30 dB over at least 3 continuous frequencies in pure tone audiometry that develops within a period of 72 hours. MRIs of screened patients were reviewed for presence of VS.	III	Summary: Of the 499 patients meeting inclusion criteria, 15 were found to have VS. Amongst the VS, the majority were Koos grade I (n=8), fewer grade II (n=6), and one grade III. There were no grade IV tumors. Conclusions: The prevalence of VS in patients with sudden SNHL was 3.0%; considering this high prevalence, clinicians should consider detailed examinations in addition to audiometry for patients with sudden SNHL Commentary: This manuscript is retrospective in nature and therefore yields class III data.

Krane et al. 2019	Study Type: Single institution retrospective review from 1980-2013 Objective: Determine the clinical efficacy of comprehensive neurotologic testing in patients presenting with complaints of hearing loss, tinnitus and/or dizziness Methodology: Retrospective review of 1170 patients presenting with neurotologic complaints. A variety of patient variables were evaluated, including audiometric screening, ABR, and MRI data. Hearing loss was not defined.	III	Summary: Of the 1170 evaluated patients, 762 (65%) presented with complaints of subjective hearing loss, 575 (49%) with vertigo, and 657 (56%) with tinnitus. Patients were determined to have an abnormal MRI in 48% (536/1120) of cases, with 6% (n=68) resulting from VS. Conclusions: Comprehensive neurotological workup results in diagnoses that would go unrecognized otherwise, allowing patients to receive prompt treatment for medically important conditions, some of which may be causally related to their neurotologic complaints. Commentary: This manuscript is retrospective in nature and therefore yields class III data. The authors make no recommendations with regard to which patient should undergo MRI, rather they simply report the prevalence of VS in the patients screened in their study.
Ungar et al. 2019	Study Type: Single institution retrospective review from 2013-2017 Objective: Investigate whether there is a different pattern of steroidal treatment response after sudden SNHL in patients with and without a VS in order to determine whether rapid hearing improvement can serve as a predictor of the presence of VS Methodology: Retrospective analysis of 420 patients presenting with sudden SNHL were reviewed. Sudden SNHL was defined by sensorineural hearing decrease of ≥ 30 dB over at least 3 consecutive frequencies in pure tone audiometry, which developed within a period of 72 hours. Patients were administered 5 days of oral prednisone at 1mg/kg/day. A comparative analysis was performed between patients responding to treatment and the presence or absence of a VS.	III	Summary: Of the 420 patients enrolled, 20 (4.76%) were found to have a VS. Audiometric testing 7 days after steroidal treatment initiation revealed that the pure-tone average of 240 patients (60%) without VS improved, and that of the 16 (80%) patients with VS improved by the same audiometric criteria. Conclusions: Improvement of hearing within 1 week after steroidal treatment initiation in patients with sudden SNHLs may suggest the presence of a VS Commentary: This manuscript is retrospective in nature and therefore yields class III data.
Abbas et al. 2018	Study Type: Single institution prospective series from 2013-2017 Objective: To determine whether patients within an ENT department presenting with asymmetrical SNHL and/or unilateral tinnitus can be safely and cost-efficiently screened for VS by audiologists as a first or only point of contact. Methodology: Prospective study of 1126 patients presenting to an audiologist initially for complaints of asymmetric SNHL or unilateral tinnitus undergoing workup inclusive of audiometric screening and MRI. An MRI was indicated if the audiogram shows a difference in the left and right thresholds of 15 dB or greater at two or more neighbouring frequencies. All patients with unilateral tinnitus underwent an MRI.	III	Summary: Of the 1126 patients meeting inclusion criteria, only 2.22% (n=25) were diagnosed with a VS. These patients were referred to ENT for further treatment guidance. Tumor sized ranged from 3mm to 20mm. Conclusions: Patients with asymmetrical SNHL and/or unilateral tinnitus can be safely screened for VS and independently managed by audiologists as a first or only point of contact Commentary: This manuscript is retrospective in nature and therefore yields class III data.

Lee S et al. 2018	Study Type: Single institutional retrospective review from 2007-2017 Objective: to identify the value of MRI for the evaluation of isolated acute audiovestibular loss Methodology: 31 patients' charts were retrospectively reviewed presenting with isolated acute audiovestibular loss. Acute hearing loss was defined as above 30 dB at three consecutive frequencies on the affected side occurring less than 1 week prior to first evaluation.	III	Summary: Of the 31 enrolled patients, 15 patients demonstrated increased labyrinthine FLAIR signal on MRI while 11 MRIs were negative, 2 patients had AICA territory strokes, and 3 had VS. There was no significant difference between pure tone audiometry and recovery rates amongst pathologies. Conclusions: Isolated acute audiovestibular loss is most commonly due to labyrinthitis as opposed to AICA stroke or VS, however, an abbreviated MRI is recommended to identify the exact etiology. Commentary: This manuscript is retrospective in nature and therefore yields class III data.
Cho et al. 2017	Study Type: Single institution retrospective study from 2006 to 2016 Objective: To investigate the frequency of IAC lesion in patients presenting with sudden SNHL Methodology: Retrospective review of the MRIs of 200 consecutive patients presenting with sudden SNHL. The audiometric criteria for sudden SNHL were a rapid decrease in hearing of more than 30 dB affecting at least three consecutive frequencies within 3 days. All MRI were obtained within 3 days of symptom onset. Patients received 60mg prednisolone for 7 days.	III	Summary: Of the 200 enrolled patients presenting with sudden SNHL, 12.5% (n=25) had an abnormal finding on the MRI believed to be causative for symptom onset. VS were diagnosed in 36% of the abnormal MRI group (n=9/25), or 4.5% of all patients enrolled presenting with sudden SNHL. Patients with a lesion of the IAC had significantly poorer hearing recovery following steroids compared to the iatrogenic sudden SNHL cohort. Conclusions: Sudden SNHL is often caused by non-tumorous causes, the majority of which are not apparent on an MRI. Commentary: This manuscript is retrospective in nature and therefore yields class III data.
Kim D. et al. 2017	Study Type: Single-center retrospective study from 2010-2015 Objective: To evaluate the magnetic resonance imaging (MRI) findings of labyrinthine hemorrhage as a cause of sudden SNHL Methodology: 1512 patients presenting with acute SNHL underwent audiometric screening and MRI. Hearing loss was classified as mild (26–50 dB), moderate (50–70 dB), severe (70–90 dB), profound (90–119 dB), or anacusis (120 dB).	III	Summary: Of the 1512 screened patients, 59 met inclusion criteria. Amongst the included cohort, 40/59 (67.8%) patients had no abnormalities on MRI while 19/59 (32.3%) had an abnormal MRI. High labyrinthine signals on a pre-enhanced T1-weighted and 3D FLAIR VISTA MRI with no contrast on 6/19 (31.6%) of patients, consistent with hemorrhage. A diagnosis of VS was made in 2/19 (10.5%), and the remaining 11/19 (57.9%) had labyrinthitis. Conclusions: MRI using pre-enhanced T1-weighted, 3D FLAIR VISTA, and post-enhanced T1-weighted, 4-h delayed enhanced FLAIR VISTA images is able to identify labyrinthine hemorrhage as the cause of sudden SNHL. Commentary: This manuscript is retrospective in nature and therefore yields class III data.
Ren et al. 2017	Study Type: Single institution retrospective review from January 2015 to October 2015 Objective: To investigate the clinical predictors of MRI in the diagnosis and treatment of sudden SNHL Methodology: 120 patients with sudden SNHL underwent auditory evaluations and MRI examinations. Audiometric and clinical variables were compared to MRI results. Sudden SNHL was defined as sensorineural hearing decrease of ≥ 30 dB over at least 3 consecutive frequencies in pure tone audiometry, which developed within a period of 72 hours.	III	Summary: Of the 120 evaluated patients, 42 (35%) had an abnormal MRI result. Three (2.5%) of screened patients were ultimately found to have a VS; all 3 patients harboring a VS underwent MRI after failing to improve following a week of steroid administration. Conclusions: When patients with sudden SNHL have severe or worse SNHL and no response for ABR tests, MRI imaging seems to be a useful examination, especially for patients with profound sudden SNHL. Commentary: This manuscript is retrospective in nature and therefore yields class III data.

Bielinska et al. 2016	Study Type: Single institutional retrospective review from 2011-2016 Objective: To determine the incidence of vestibular schwannoma being responsible for the onset of inner ear dysfunction. Methodology: Patients being screened for SNHL (range not defined) or tinnitus underwent audiology screening and ABR. Prolonged ABR (I-III above 2.55 ms, III-V above 2.35 ms, I-V above 4.6 ms) underwent MRI with contrast	III	Summary: Of 3456 patient being screened, abnormal ABRs were identified in 252 (7.3%) cases of which 13 (5.16%) had MRI diagnosis of VS. Conclusions: Presentation of asymmetric SNHL warrants audiology screening with ABRs. Presence of prolonged ABRs warrants further investigation with dedicated MRI of the IAC with contrast to screen for neoplasms. Commentary: This manuscript is retrospective in nature and therefore yields class III data.
Jeong et al. 2016	Study Type: Single institution observational study from 2007-2012 Objective: To assess abnormal MRI findings in patients with sudden SNHL and evaluate the value of MRI in identifying the cause of sudden SNHL Methodology: Retrospective review of 291 patients presenting with sudden SNHL undergoing audiology screening and eventual MRI. Hearing loss was defined by loss of 30 dB or more over at least 3 contiguous frequencies in pure tone audiometry that develops within 3 days. All patients underwent a dedicated MRI of the IAC within 10 days of symptom onset.	III	Summary: Of the 291 patients enrolled, the MRI identified the cause of the sudden SNHL in 4.5% (n=13). VS was diagnosed in 3.1% (n=9) of enrolled patients, or 69.2% of patients with abnormal MRI findings. Of the VS, 3 were purely intrameatal, while 6 demonstrated extrameatal extension. Conclusions: The most commonly observed MRI abnormality in patients with sudden SNHL was vestibular schwannoma, and all of the lesions were small or medium-sized tumors involving the IAC. Commentary: This manuscript is retrospective in nature and therefore yields class III data.
Kim S. et al. 2016	Study Type: Single-center retrospective review from 2001-2013 Objective: To evaluate otologic symptoms and audiometric data of VS compared to other neoplasms of the CPA. Methodology: Comparison of otologic symptoms of 171 patients diagnosed with CPA tumors (VS-116, Other-55). Factors analyzed included tumor type, size, and location and the results of audiometric examinations. Audiogram patterns were classified as high-tone hearing loss (descending) type if the hearing threshold was lower at a high-tone than at a low-tone frequency; as flat type when the thresholds of all assessed frequencies were comparable; low-tone hearing loss (ascending) type when a threshold was lower at a low-tone than at a high-tone frequency; concave type when the primary frequency lowered was mid-tone; and convex type when the mid-frequency was relatively well preserved. Tinnitograms were used to measure tinnitus pitch and loudness matching.	III	Summary: The most frequent patterns of hearing loss were the descending type in patients with VS and the flat type in patients with non-VS tumors. Pure tone thresholds tended to increase more in patients with VS than non-VS tumors according to tumor size, and pure tone averages were significantly higher in patients with VS than non-VS tumors of 11–25mm in size. Conclusions: Hearing loss with tinnitus was the most common combination of symptoms in patients with VS, whereas hearing loss with dizziness was more common in patients with other types of CPA tumor. Commentary: This manuscript is retrospective in nature and therefore yields class III data.

Lee SH et al. 2015	Study Type: Single institution retrospective review from 2001-2013 Objective: To determine the neurotologic factors associated with VS by analyzing the clinical manifestations and diagnostic test results of patients with VS before MRI scanning Methodology: 114 patients presenting with neurotologic complaints were enrolled. Multiple patient factors were retrospectively analyzed including: demographics, symptomatology, pure-tone audiometry, speech discrimination score, ABR, and MRI results.	III	Summary: Of the 114 VS patients analyzed, the most common symptom at time of presentation was asymmetric hearing loss associated with tinnitus. More severe deafness correlated with lower speech discrimination scores. Of patients with latencies of waves I, III, and V on ABR tests, 56.1%, 92.4%, and 92.4%, had interaural latency differences ± 0.2 ms. Conclusions: MRIs should be obtained in patients presenting with asymmetric hearing loss, tinnitus, low speech discrimination score, and abnormal ABRs. Commentary: This manuscript is retrospective in nature and therefore yields class III data.
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Appendix V. Conflicts of Interest

Task Force Member	Disclosure
<i>Julie Honaker PhD, AuD</i>	Nothing to Disclose
<i>Ben Allen Strickland, MD</i>	Nothing to Disclose
<i>Eric J. Lehrer, MD</i>	Servier Pharmaceuticals, Novocure Inc.
<i>Sheryl Green, MBBCh</i>	Nothing to Disclose
<i>John P. Marinelli MD</i>	Medtronic
<i>Christopher S. Graffeo MD, MS</i>	Nothing to Disclose
<i>Isabelle M. Germano, MD, MBA</i>	Brianlab
<i>Mateo Ziu, MD</i>	Omniscient Neurotechnology America Ltd; GT Medical Technologies, Inc
<i>Walavan Sivakumar, MD</i>	Stryker Corporation
<i>Sherwin Tavakol, MD</i>	IRRAS USA, Inc.; Globus Medical, Inc.
<i>Lucas Paul Carlstrom, MD, PhD</i>	Kuros Biosciences USA, Inc
<i>Jamie J. Van Gompel, MD</i>	Medtronic, Cadence
<i>Ian Dunn, MD</i>	Nothing to Disclose
<i>Jeffrey J. Olson, MD</i>	Verastem, Inc., Research Grant American Cancer Society, Editorial Consultant; Azurity Pharmaceuticals, Inc.
<i>Ghazal S. Daher MD</i>	Nothing to Disclose
<i>Matthew L. Carlson, MD</i>	Cochlear Americas, Advanced Bionics, Stryker Corporation; iotaMotion, Inc.; Stryker Corporation

<i>Neil S. Patel, MD</i>	Cochlear Americas, Zeiss, Viridian Therapeutics, IotaMotion, Inc.
<i>Michael Sughrue, MD</i>	Omniscient Neurotechnology America Ltd
<i>Constantinos G. Hadjipanayis, MD, PhD</i>	Stryker Corporation; Integra LifeSciences Corporation; Omniscient Neurotechnology America Ltd
<i>Jeffrey Jacob, MD</i>	Stryker Corporation; KLS; Synthes