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Global Incidence of Sporadic Vestibular Schwannoma: A Systematic Review

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Abstract

Objective. Ubiquitous throughout the literature and during patient counseling, vestibular schwannoma is often quoted to affect about 1 per 100,000 people. Yet, reports from distinct international populations suggest that the incidence is likely much higher. The objective of the current work was to systematically characterize the global incidence of sporadic vestibular schwannoma.

Data Sources. Scopus, Embase, and PubMed.

Review Methods. Population-based studies reporting incidence rates of sporadic vestibular schwannoma between January 2010 and August 2020 were searched with language restrictions requiring reports to be published in Chinese, English, German, Italian, or Spanish. The protocol was registered with PROSPERO (CRD42021228208) prior to commencement of data collection. PRISMA guidelines for transparent reporting of systematic reviews were followed.

Results. Among 424 citations, 6 publications covering 4 distinct populations from Denmark, the Netherlands, Taiwan, and the United States met inclusion criteria. Most recent incidence rates of among all ages ranged between 3.0 and 5.2 per 100,000 person-years. Highest incidence rates were reported among patients aged $\geq$70 years, peaking at 20.6 per 100,000 person-years. One study from the United States reported the incidence of asymptomatic, incidentally diagnosed tumors at a rate of 1.3 per 100,000 person-years from 2012 to 2016.

Conclusions. Recent international incidence rates of sporadic vestibular schwannoma exceed the commonly quoted "1 per 100,000" figure by up to 5-fold among all ages and by up to 20-fold among age groups at highest risk. Based on modern incidence rates, the lifetime prevalence of developing sporadic vestibular schwannoma likely exceeds 1 per 500 persons.

Keywords

vestibular schwannoma, acoustic neuroma, sporadic, unilateral, incidence, prevalence, epidemiology, population-based, review

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"How common are vestibular schwannomas (VSs)?" is an often-asked question throughout the literature and within patient counseling. Most commonly, the response is "about 1 per 100,000 in the general population."[1,2] There exist 2 fundamental problems with this response. First, when patients ask this question, they are really asking, "What are the chances that I would have developed this tumor at some point during my lifetime?" This question is not best answered with disease incidence rates, which by definition include only new diagnoses in a population every year; rather, for a disease where new diagnoses accumulate over time in a population, the best answer to the disease "commonness" question is lifetime prevalence. Instead of describing just the number of new diagnoses every year, lifetime prevalence encompasses all people who eventually develop VS at some point throughout the life span. For sporadic VS, the modern lifetime prevalence likely exceeds 1 in 500 persons.[3-7]

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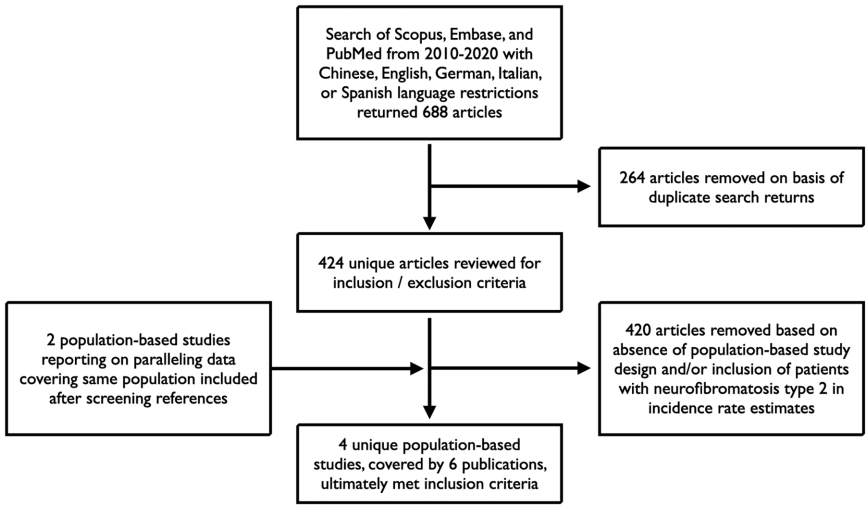

Figure 1. Schematic overview of study selection for systematic review.

Second, the ''1 per 100,000'' figure represents a historical estimate of incidence rates of VS from the era prior to the advent of magnetic resonance imaging (MRI) and widespread adoption of screening protocols for asymmetrical sensorineural hearing loss.[8,9] In recent years, several distinct international populations have displayed significantly higher incidence rates of sporadic VS.[8,10,11] For this reason, the current work was undertaken to systematically characterize the global incidence of sporadic VS in the modern era, where adoption of screening protocols for asymmetrical and sudden sensorineural hearing loss with MRI is commonplace. In this way, this review intends to draw attention to the global incidence of sporadic VS in the post-MRI era and its several clinically relevant ramifications.[12]

Methods

This systematic review was performed and reported in accordance with the PRISMA guidelines (Preferred Reporting Items for Systematic Reviews and Meta-analyses).[13] The protocol was registered with PROSPERO (CRD42021228208) prior to commencement of article review and data collection. Data extraction was performed with the help of an experienced medical librarian (C.J.B.). Scopus, Embase, and PubMed were searched for population-based studies reporting the incidence of sporadic (ie, unilateral) VS on populations from January 2010 through August 2020. Article eligibility criteria also required reports to be published in Chinese, English, German, Italian, or Spanish. Exclusion criteria stipulated that the population-based study cannot report combined incidence rates of patients with neurofibromatosis type 2 and those with sporadic tumors. The complete enumerated search strategy by database can be viewed in Supplemental Appendix S1 (available online). Full-text articles were reviewed unless the abstract provided definitive grounds for exclusion. Reference lists of eligible studies were reviewed for potential inclusion. Each retrieved article was reviewed independently by 2 reviewers (J.P.M. and I.D.E.), and disagreement was resolved by consensus among all authors. Data amalgamation and reporting of study heterogeneity were performed by descriptive statistical methods.

Results

Systematic Review

A total of 688 citations were initially retrieved. Review of retrieved articles identified 264 duplicate search returns across the 3 databases, and 424 unique citations were ultimately reviewed (**Figure 1**). Six publications covering 4 population-based studies from Denmark, the Netherlands, Taiwan, and the United States met inclusion criteria.[8-11,14,15] All studies exhibited low risk of bias, as all were strictly descriptive, non–hypothesis testing population-based incidence studies of a complete population in a well-defined geographic region over a recent period. All excluded studies either lacked per-year incidence rates following 2010 or included patients with tumor-predisposing conditions (eg, neurofibromatosis type 2) within the incidence rates of VS.

Clinical Features and Global Incidence Rates

Across all studies, most recent incidence rates of sporadic VS ranged between 3.0 and 5.2 per 100,000 person-years with a median age of 60 years at diagnosis (**Table 1**).[8,10,11,14,15] Intracanalicular tumors composed 48% to 72% of newly diagnosed cases. The incidence of cerebellopontine angle tumors was similar between Denmark and the United States (1.6 vs 1.5 per 100,000 person-years, respectively), whereas the incidence of intracanalicular tumors was higher in the US cohort (1.8 vs 3.7). Age-specific incidence rates were cited in 2 studies, and the highest rates were observed among patients aged $\geq$70 years, peaking at 20.6 per 100,000 person-years.[11,15] All 4 studies noted no significant difference in incidence rates between male and female sex. The population-based study from Taiwan demonstrated a nonstatistically significant female predominance among newly diagnosed cases with a ratio of 1.3 to 1.0; however, the Danish and US data reported higher incidence rates among men in the last decade.[9,11,14] Two studies had multiple years of data during the study period, and both showed modest increases in disease incidence over this period.[8,15] One study from the United States noted the incidence of asymptomatic, incidentally diagnosed tumors

Table 1. Clinical Features at Diagnosis and Global Incidence Rates of Vestibular Schwannoma by Country of Origin of Population-Based Study.

	Denmark[8,9]	Netherlands[10]	Taiwan[11]	United States[14,15]
Clinical features				
Median age, y	60	57[a]		62
Tumor location: intracanalicular, %	48			72
Median CPA tumor size, cm	1.3[a]			1.2
Incidence rate[b]				
Most recent: all ages	3.4	3.3[c]	3.0	5.2
Tumor				
CPA	1.6			1.5
IAC	1.8			3.7
Age, y				
40-49			2.6	3.3
50-59			4.3	11.3
60-69			4.9	13.6
≥70			4.1	20.6
Asymptomatic, incidentally diagnosed tumors				1.3

Abbreviations: CPA, cerebellopontine angle; IAC, internal auditory canal.
[a]This figure represents a mean as reported in the original study (vs median).
[b]Incidence rates per 100,000 person-years. Most recent year varies by study: 2015 for Denmark, 2012 for Netherlands, 2012 for Taiwan, and 2016 for United States.
[c]Several incidence rates were reported in this study; however, for reasons exhaustively described in the original study, the rate of 3.3 per 100,000 person-years likely represents the most accurate estimate reported based on completeness of data.[10]

during neuroimaging workup for unrelated indications at a rate of 1.3 per 100,000 person-years from 2012 to 2016.[15]

Discussion

Although the difference between a disease incidence of 1 per 100,000 person-years and 3 or 5 per 100,000 person-years can appear inconsequential, it actually suggests that substantially more people develop sporadic VS than historically considered. First, the difference between the commonly quoted ''1 per 100,000'' and the incidence rates in this systematic review becomes magnified on a global population–based scale. Second, among older groups within the population that are at highest risk, there may be 20-times more people being diagnosed with sporadic VS per year. Since the majority of new diagnoses among older subgroups of the population comprise small tumors with minimal symptoms, roughly 75% of these patients undergo at least an initial period of observation with serial imaging following diagnosis.[14] As a result, incident cases accumulate over time, and disease prevalence rises. Few population-based studies have reported the prevalence of sporadic VS in recent years.[3,7,16] To estimate the lifetime prevalence of sporadic VS in a clinical setting—that is, during routine clinical practice with modern neuroimaging capabilities, not through review of temporal bones at autopsy—the chance that a person sporadically develops a VS exceeds 1 in 500.[3] As the resolution of MRI continues to improve, the clinical prevalence will likely gradually approach historical temporal bone studies that have cited lifetime prevalence as high as 1 in 100 persons.[4,17] Thus, when a patient asks, ''How common are VSs?'' the most accurate answer based on current knowledge is ''About 1 in 500 develop this tumor during the lifetime,'' at least for most persons residing in the United States and Europe.

Over the last several decades, an ostensibly paradoxical epidemiologic evolution has transpired surrounding sporadic VS. Namely, despite the average age at diagnosis steadily increasing over the last 50 years, the average tumor size at diagnosis has actually decreased and residual hearing increased.[8,9,14] This observation suggests that beyond earlier detection of VS, tumors that historically went undiagnosed due to their previously subclinical disease are now being detected (ie, with current MRI techniques), and the detection of these previously subclinical tumors actually represent the majority of new diagnoses in the general patient population with VS.[18] This rationale is substantiated by the observation from this systematic review that roughly 50% to 75% of new diagnoses are composed of intracanalicular tumors. Moreover, the rising incidence of incidentally diagnosed tumors plays a contributory role.[15] Overarchingly, these findings support that improved detection serves as the chief etiology behind the rising incidence over the last several decades, as opposed to a biological shift in susceptibility to disease development (**Figure 2**). To this end, a substantiated biological explanation behind the rising incidence rates remains absent. Although several survey-based studies have reported an association between noise exposure and the development of sporadic VS[19,20] or between cell phone use and sporadic VS,[21] several robust population-based case-control studies have refuted these claims.[22-24] The apparent discrepancy is most likely accounted for by the susceptibility of the aforementioned survey-based studies to recall bias, where patients who have been diagnosed with a tumor on their hearing nerve are

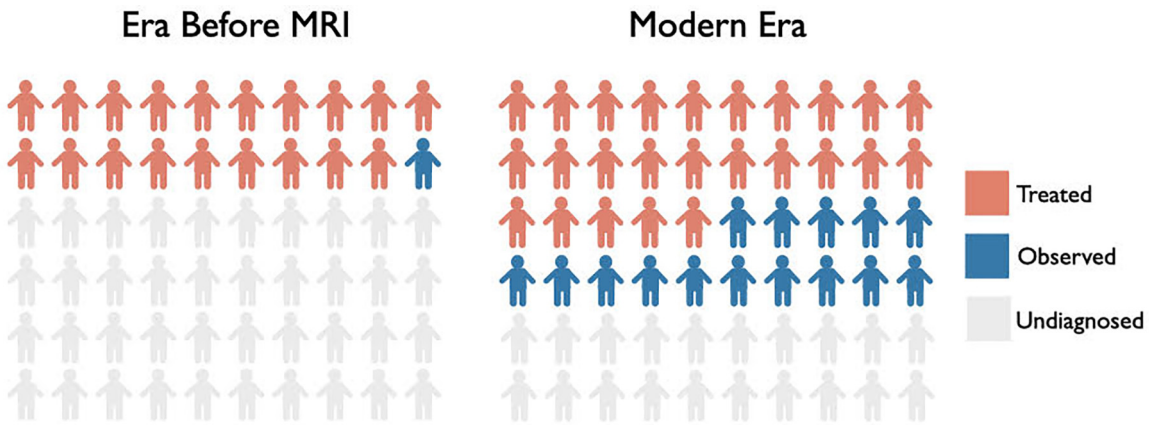

Figure 2. Heightened detection of vestibular schwannoma as an explanation for rising global incidence. MRI, magnetic resonance imaging.

reasonably more likely to note exposures related to hearing. Similarly, patients with significant cell phone use may be more sensitive to asymmetrical hearing loss and consequently undergo workup.

Understanding the influence of detection capabilities on the observed incidence rates helps to reconcile the difference in incidence rates in this systematic review. For instance, the incidence of cerebellopontine angle tumors in the Danish and US studies was essentially indistinguishable. However, the incidence of intracanalicular tumors in the US population–based study exceeded the Danish rate by >2-fold. This finding is furthered by the notably higher proportion of intracanalicular tumors at diagnosis among the US cohort (72% vs 48%). Interestingly, there are 25-times more MRI scanners per person in Olmsted County (ie, the county from which the US cohort resided) than Denmark.[9] By age 70 years, nearly one-third of all adults residing in this US county have undergone a head MRI scan for various indications.[3] Moreover, the uniquely medically oriented demographic of this region likely contributes to an elevated overall health literacy.[14] In combination, these factors likely lead to a greater detection of VS in the US county. Supporting this rationale, higher rates of VS detection have been found in regions of Denmark with elevated access to health care,[25] and the highest published incidence rate of VS to date was reported in Beverly Hills, California, at 5.4 per 100,000 person-years.[26] Of note, given the historical immigration patterns from Nordic countries into Minnesota, it is unlikely that substantial genetic differences in susceptibility to the development of sporadic VS account for the observed differences. Nevertheless, notwithstanding potential differences in detection, the Danish data uniquely represent the only longitudinal national database that covers a complete country since the 1970s.[8]

One of the other important ramifications of the incidence rates described in the current systematic review surrounds the apparent underreporting of disease incidence within national brain tumor and cancer registries (**Table 2**).[27-29] In 2004, the Benign Brain Tumor Cancer Registries Amendment Act (US public law 107-260) nationally mandated the additional registration of benign brain tumors to the already mandated registration of new cancer diagnoses. Yet, with the innumerable types of systemic malignancies and brain tumors, comprehensive manual searches of clinic visits are logistically impractical for most institutions. Instead, most institutions employ software infrastructure that queries pathology reports (eg, biopsied tissue, ablative surgical tissue) and cancer-related treatment codes, such as those related to radiation and chemotherapy, to register most new diagnoses.[30] Unfortunately, the reliance on pathology specimens and cancer-related treatment data introduces a selection bias regarding benign brain tumors that are often diagnosed clinically (ie, without biopsy) and frequently involve observation as a primary treatment modality.[30] This methodologic limitation behind cases in national brain tumor and cancer registries also helps to explain the discrepant clinical data, specifically surrounding the propensity for included patients to have larger tumors at diagnosis, which more frequently require definitive treatment as opposed to observation.[30] Of note, this method of registering new cases in the United States is characteristically different than the national registration of new VS diagnoses in

Table 2. Underreporting of Incidence Rates of Vestibular Schwannoma in National Brain Tumor and Cancer Registries Within the United States.

	Incidence rate[a]
National brain tumor and cancer databases	
SEER[29]	1.3
CBTRUS[27,28]	1.9
Denmark[8]	3.3
Netherlands[10]	3.3
Taiwan[11]	3.0
US REP data[14,15]	5.2

Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States; REP, Rochester Epidemiology Project; SEER, Surveillance, Epidemiology, and End Results.
[a]Incidence per 100,000 person-years. Most recent incidence rate varies by publication, ranging from 2012 for the Netherlands and Taiwan, 2015 for SEER and Denmark, and 2016 for the REP and CBTRUS.

Denmark, for example, where every newly diagnosed patient is at least initially seen at the University Hospital in Copenhagen. Perhaps most significant, there exists no reason to believe that this underreporting of VS incidence does not equally affect other common benign brain tumors that frequently undergo observation with serial imaging, such as meningioma and pituitary adenoma, as these tumors are also diagnosed clinically and tissue specimens are never obtained.[30,31] Since these studies tend to shape understanding of benign brain tumors in the United States, the potential for widespread selection bias warrants careful further investigation, especially given the possibility that potentially more VSs are treated today than ever before as a result of rising incidence rates.[12] Of note, the incidence rates from these national registries did not meet the inclusion criteria of the current study, as cases of neurofibromatosis type 2 were included in their estimations—a feature that accentuates the apparent underreporting of sporadic VS incidence.[30,31]

An important limitation of the current work surrounds the limited epidemiologic data from multiple other regions of the world. Non–population based work has suggested that other populations may exhibit significantly different incidence rates of sporadic VS. For example, at a large tertiary referral center in sub-Saharan Africa, only 2 sporadic VSs were identified during a 13-year period, and review of nearly 7500 neuroimaging scans revealed no incidentally diagnosed tumors.[32] A report covering the Icelandic population from 1979 to 2009 showed an increasing incidence of VS, along with incidentally diagnosed tumors, and incidence trends that paralleled European data during the years covered.[33] A study from Israel describing the incidence of all brain tumors noted a modestly increasing rate of benign brain tumors from 1990 to 2015, although significant VS-specific incidence data were lacking.[34] Data from the United States suggest the potential for varying incidence rates among different races nationally, though access to health care may contribute to the observed findings.[35] Additionally, it is unclear whether the population-based data from Taiwan in the present study include exclusively unilateral tumors, but the very low incidence of neurofibromatosis type 2 likely renders this potentiality inconsequential.

Conclusion

Recent international incidence rates of sporadic VS exceed the commonly quoted ''1 per 100,000'' figure by up to 5-fold among all ages and by up to 20-fold among age groups at highest risk. In this way, it is likely that at least 3 times as many people are living with sporadic VS in the United States than what national cancer and brain tumor databases suggest. The rise in incidence rates over the last several decades is best explained by improved detection secondary to widespread adoption of screening protocols for asymmetrical sensorineural hearing loss with MRI. As a result of rising incidence rates, the chance of developing a sporadic VS throughout a person's lifetime likely exceeds 1 in 500.

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Author Contributions

John P. Marinelli, substantial contributions to the conception or design of the work; acquisition, analysis, or interpretation of data for the work; drafting the work and revising it critically for important intellectual content; final approval of the version to be published; agreement to be accountable for all aspects of the work; **Cynthia J. Beeler**, acquisition, analysis, or interpretation of data for the work; drafting the work and revising it critically for important intellectual content; final approval of the version to be published; agreement to be accountable for all aspects of the work; **Matthew L. Carlson**, analysis and interpretation of data for the work; revising it critically for important intellectual content; final approval of the version to be published; agreement to be accountable for all aspects of the work; **Per Caye-Thomasen**, analysis and interpretation of data for the work; revising it critically for important intellectual content; final approval of the version to be published; agreement to be accountable for all aspects of the work; **Samuel A. Spear**, analysis and interpretation of data for the work; revising it critically for important intellectual content; final approval of the version to be published; agreement to be accountable for all aspects of the work; **Isaac D. Erbele**, substantial contributions to the conception or design of the work; acquisition, analysis, or interpretation of data for the work; drafting the work and revising it critically for important intellectual content; final approval of the version to be published; agreement to be accountable for all aspects of the work.

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Supplemental Material

Additional supporting information is available in the online version of the article.

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