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The Rise and Fall of Otosclerosis: A Population-based Study of Disease Incidence Spanning 70 Years

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Abstract

Objective: Although it is commonly held that otosclerosis has become increasingly uncommon over recent decades, no population-based data exist to characterize this trend. Moreover, because most large epidemiologic databases within the United States primarily include cancer data, even the modern incidence of otosclerosis is unknown. The chief objective of the current work was to characterize the trend in the incidence of otosclerosis over 70 years using the unique resources of the Rochester Epidemiology Project.

Study Design: Population-based study.

Patients: Residents of Olmsted County, Minnesota diagnosed with otosclerosis.

Main Outcome Measure: Disease incidence from 1950–2017.

Results: From 1950–2017, 614 incident cases of otosclerosis were identified. The incidence rose from 8.9 per 100,000 person-years in the 1950s to a peak of 18.5 from 1970–1974. From this peak, the incidence significantly declined to 6.2 per 100,000 person-years by the early-1990s and reached a nadir of 3.2 from 2015–2017 ($p < 0.001$). From 1970–2017, age at diagnosis ($p = 0.23$) and the proportion of bilateral cases ($p = 0.16$) did not significantly change; pure-tone average at diagnosis did not clinically appreciably change over the study period (median difference < 5 dB across decades, $p = 0.034$).

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Conclusions: The incidence of otosclerosis drastically declined since the early-1970s. Historically considered one of the most common causes of acquired hearing loss, the low modern incidence of otosclerosis renders it legally a “rare disease” within the United States. These trends require consideration when determining trainee case requirements and developing practice guidelines.

Keywords

otosclerosis; stapes surgery; juvenile; incidence; epidemiology; Rochester Epidemiology Project

INTRODUCTION

In 1956, John J. Shea Jr. performed the first successful stapedectomy and ushered in the modern era of stapes surgery.¹ Throughout the remainder of the 20th century, the treatment of otosclerosis played a fundamental role in characterizing the field of otology. The success of stapedectomy surgery was followed by an immediate influx of new patients seeking surgical treatment.² As a result, disease incidence rapidly rose through the 1960s,³ and otologic surgeons of that era quickly accumulated significant case numbers.^{4,5} However, by the late-1970s, reports surfaced suggesting that the incidence of otosclerosis was declining.⁶ In the following decades, several studies confirmed this decline, showing that stapes surgery was occurring with less frequency over time.^{7–9}

Today, it is commonly believed that the incidence of otosclerosis is less than it was 50 years ago. However, the degree to which the incidence has declined is unknown. To this end, no population-based evidence exists reporting the modern incidence rates of the disease, nor are data reported characterizing the evolving epidemiology of otosclerosis over the past several decades. Instead, most of the reported trends are limited to institutional reports regarding declining surgical volume,^{2,6,7,9} or an amalgamation of surgical volume data.⁸ Importantly, these studies provide no information about the incidence of otosclerosis (i.e., the rate at which new cases are being diagnosed in a population). The primary reason for this lack of information results from the existing limitations of most large epidemiologic databases. Within the United States, large population-based databases are often restricted to the inclusion of cancer or brain tumors. Therefore, data on diseases such as otosclerosis are not centrally collected within a defined geographic region or population. Robust epidemiologic data and knowledge of recent disease trends are necessary when determining trainee case requirements, developing practice guidelines, and informing patient counseling. Furthermore, examination of trend data can elucidate potential sex-specific, race-specific, or environmental factors associated with disease risk.

In 1966, Leonard T. Kurland founded the Rochester Epidemiology Project (REP) after establishing a medical records-linkage system that linked patient data from all healthcare providers in Olmsted County.¹⁰ Virtually every resident of Olmsted County is registered with the REP either from birth or from the date of first healthcare encounter as a resident.¹¹ Therefore, the REP is a unique resource which allows researchers the ability to study disease incidence in a well-defined population over a time period that exceeds five decades. Moreover, the REP contains data that are not limited to a certain subset of diseases.¹²

Fortuitously, for the study of otosclerosis, Dr. Kurland initially developed the REP infrastructure in 1950 within the city of Rochester, Minnesota (the largest city in Olmsted County, comprising approximately 70% of the Olmsted County population). What is more, Dr. Kurland, along with co-authors Richard Pearson and Mayo Clinic otologist Thane Cody, published their data on the epidemiology of otosclerosis from 1950 through 1969.³ Starting where they finished, the purpose of the current work was to describe the evolution in the epidemiology of otosclerosis from 1970 through 2017, therefore allowing examination of disease trends in this population over a period of almost 70 years.

METHODS

Clinical Data

After obtaining institutional review board approvals from Olmsted Medical Center and Mayo Clinic (021-OMC-18 and 18-005225, respectively), the medical records of all residents of Olmsted County who were diagnosed with otosclerosis between 1 Jan 1970 and 12 Dec 2017 were reviewed. The diagnosis of otosclerosis was confirmed either surgically or through the combined clinical assessment of classic audiometric and examination findings as well as radiologic imaging when available.¹³ Pure-tone averages (PTA) over 0.5, 1, 2, and 3 kHz were used to report pure tone audiometric data for each diseased ear. The average of 2 and 4 kHz was used when 3 kHz was not tested. Cases of juvenile otosclerosis were defined by diagnosis of disease before age 18 years. The accuracy of the REP medical records-linkage system has been validated previously, and it is estimated that the rate of under-inclusion of one or more medical records for a unique person is approximately 1%.¹⁴ Almost 95% of Olmsted County residents have been seen by a provider in the REP system at least once every 3 years.¹²

Statistical Methods

Continuous features were summarized with medians and interquartile ranges (IQRs) and categorical features were summarized with frequency counts and percentages. Trends in clinical features by decade of diagnosis were evaluated using Spearman rank correlation coefficients, Kruskal-Wallis tests, and Wilcoxon rank sum tests. Incidence rates per 100,000 person-years were calculated using incident cases of otosclerosis between 1970 and 2017 as the numerator and age- and sex-specific counts of the population of Olmsted County, Minnesota as the denominator. The denominators were obtained from a complete enumeration of the Olmsted County population provided by the REP.¹⁴ Incidence rates were directly standardized to the total United States population from the 2010 United States Census. Associations of age, sex, year of diagnosis, and treatment (surgically confirmed vs not surgically confirmed) with incidence rates were evaluated using Poisson rate regression models. Statistical analyses were performed using version 9.4 of the SAS software package (SAS Institute; Cary, NC). P-values <0.05 were considered statistically significant.

RESULTS

Clinical Data

From 1970 through 2017, 504 incident cases of otosclerosis were identified with a 1.6:1 female-to-male ratio and a median age at diagnosis of 42 years (IQR 32–53). The median age at time of diagnosis remained fairly constant throughout the study period ($p=0.23$; Figure 1, Table 1). The proportion of cases diagnosed among non-white races significantly increased over the study period ($p<0.001$). Although statistically significantly different due to the large sample size, the degree and type of hearing loss at diagnosis were clinically indistinguishable across the study period (i.e., median PTA difference <5 dB across decades).

Incidence of Otosclerosis

The incidence of otosclerosis over the study period is shown in Table 2 and Figure 2. Combined with the data from 110 incident cases reported by Pearson, et al. between 1950 and 1969 using the initial REP infrastructure,³ the incidence of otosclerosis rose from 8.9 per 100,000 person-years in the 1950s to a peak of 18.5 between 1970 and 1974. From this peak, the incidence significantly declined to 6.2 per 100,000 person-years by the early-1990s, reaching a study-period low of 3.2 between 2015 and 2017 ($p<0.001$). The incidence of otosclerosis also differed significantly by age (highest in mid-life between ages 36 and 55, $p<0.001$; see Table 1, Supplementary Digital Content) and was higher for women compared to men ($p<0.001$).

The incidence of surgically confirmed cases paralleled the trend observed in all cases of clinical otosclerosis and significantly decreased over the study period from a peak of 10.4 per 100,000 person-years between 1970 and 1974 to 1.9 between 2015 and 2017 ($p<0.001$; see Table 2, Supplementary Digital Content). The rate of decline in incidence rates over time among surgically confirmed cases compared to non-surgically confirmed cases was not statistically significantly different ($p=0.36$).

Race-specific population counts of Olmsted County were available starting in 2000. Between 2000 and 2017, the incidence of otosclerosis was 5.2 per 100,000 person-years in whites, 6.1 in blacks, and 5.1 in Asians. Among those of Hispanic origin, the incidence over the same time period was 6.1 per 100,000 person-years.

Juvenile Otosclerosis

Seventeen (3%) cases of juvenile otosclerosis were identified within the larger cohort of 504 incident cases, including 9 girls and 8 boys with a median age at diagnosis of 14 years (IQR 8–16). During the first half of the study period from 1970 through 1994, the incidence rate was 1.4 per 100,000 person-years ($n=10$), whereas the incidence rate from 1995 through 2017 was 0.8 per 100,000 person-years ($n=7$).

DISCUSSION

Declining Incidence and its Implications

Due to limitations of most large epidemiologic databases, it is difficult to characterize the evolution in the epidemiology of otosclerosis over recent decades. Corroborated by previous reports on declining surgical volume data,^{2,6-9} the current study showed that the incidence of otosclerosis drastically declined from its peak in the 1970s. Neither the age at diagnosis nor the frequency of bilateral involvement significantly changed over the study period, suggesting that the etiopathogenesis of otosclerosis over the 50 years has remained stable. This idea is further supported by the lack of clinically significant differences in severity and type of hearing loss over the study period. Of note, our findings contrast with a single institutional series from the Warren Otologic Group that showed a decreasing proportion of bilateral cases and a small decrease in the degree of conductive hearing loss among surgically treated cases from the 1960s through the 1990s.¹⁵ These differences could be secondary to the varying methodologies between this population-based study and their single institutional series.

Understanding the generalizability of any population-based study is fundamental to accurate interpretation and application. Broadly, two primary categories exist that contribute to the detection and development of disease: Features of a population that either facilitate or prevent the diagnosis of disease (e.g., access to healthcare or social and cultural norms) and features inherent to the population that affect the likelihood of developing the disease (e.g., age, sex, or genetic factors). Regarding the detection of otosclerosis, the Olmsted County population is comprised of persons with a higher median level of education and a higher median household income than the United States at-large.¹¹ It is well-established that the detection and treatment of hearing-related diseases are affected by socioeconomic factors, and it is likely that the population of Olmsted County is more apt to seek healthcare after noticing changes in hearing status. This rationale is substantiated by the high detection rate of vestibular schwannoma within this population.^{16,17} Therefore, as it pertains to socioeconomic factors, it is likely that the detection of otosclerosis in this population is higher than in some other populations and in the United States as a whole. However, it should be emphasized that this does not imply that disease incidence is truly higher in Olmsted County (i.e., socioeconomic factors do not fundamentally alter someone's likelihood of developing the disease). Rather, it simply suggests that the detection of otosclerosis is likely higher in this population and closer to the true incidence. As will be discussed shortly, this observation has several implications.

Pertinent to the likelihood of developing otosclerosis, the age and sex distributions of Olmsted County resemble the United States distributions – an important consideration given both features' significant influence on the incidence of otosclerosis.¹¹ Most critical to contextualizing the current study, the racial profile of Olmsted County is historically less diverse than the United States as a whole.¹¹ Data from the 2010 United States Census show that approximately 86% of Olmsted County is white versus only 72% of the complete United States population. Although robust investigation into the epidemiology of otosclerosis is extremely lacking among non-white racial groups, several prior studies –

most published over 50 years ago – showed that otosclerosis is much more common among whites than Asians, blacks, or those of Hispanic ethnicity.^{18–22} Therefore, because the incidence rates reported in the current work are derived from a largely white population, the incidence of otosclerosis in Olmsted County could exceed national incidence rates on the historical basis of persons' risk of developing the disease. Of note, the magnitude of this effect on the extrapolation of these data to national rates is temporized by the comparable modern incidence of otosclerosis by race reported in the current study.

When attempting to apply these data to suspected national incidence rates, the impact of the distinct socioeconomic and racial features of Olmsted County on the incidence rates of otosclerosis bears considerable implications. When considering these rates in light of determining trainee case requirements or developing practice guidelines, it must be considered that likely *less than 3* new cases of otosclerosis will be diagnosed per 100,000 persons per year nationally, and *less than 2* per 100,000 persons per year will undergo surgery. Diseases with incidence rates of surgically treated cases within this range, such as microsurgical resection of vestibular schwannoma,²³ are essentially reserved for fellowship-trained practitioners in the modern era. The technical skill required to perform stapes surgery with its attendant risks and low volume of stapes surgery in residency training secondary to the declining incidence has resulted in a very limited number of general otolaryngologists performing stapedectomy.^{8,24} The debate surrounding whether or not stapes surgery should remain a key indicator case for general otolaryngology training or be reserved for those with advanced otologic subspecialty training persists. However, the data from the current work provide evidence to support the latter.

Similarly, throughout the otolaryngology literature and during patient counseling, otosclerosis is frequently referred to as “one of the most common causes of acquired hearing loss.” Yet, modern incidence rates of otosclerosis indicate that residents of Olmsted County are more likely to acquire hearing loss secondary to the sporadic development of vestibular schwannoma – ironically often considered a “rare disease” within the literature and during patient counseling – than from otosclerosis. (Figure 3, Table 3).^{16,17} Undoubtedly, the immense evolution observed in the incidence of otosclerosis and sporadic vestibular schwannoma represents perhaps the greatest shift in the epidemiology of diseases managed by otologists in the modern era.

What is more, the incidence rates of otosclerosis observed in recent years render otosclerosis legally a “rare disease” in the United States (Rare Disease Act of 2002, Public Law 107–280). For illustration, the most recent United States Census data showed that there are just over 300 million people living in the United States. At an incidence rate of 3 per 100,000 person-years, there would be 9,000 new diagnoses of otosclerosis per year nationally. Because roughly two-thirds of new diagnoses undergo surgical correction, the prevalence of otosclerosis likely falls significantly under the threshold that the Rare Disease Act of 2002 set, which legally defined a rare disease as one that affects less than 200,000 individuals nationally. Furthermore, the likely lower incidence of otosclerosis across the United States than in Olmsted County underscores the small number of new cases observed nationally each year.

Etiology of the Declining Incidence

The etiology behind the precipitous decline in the incidence of otosclerosis remains unknown. Several hypotheses have been examined throughout the years, though few have stood the test of time.⁸ The development of otosclerosis is likely multifactorial, and more than one molecular pathway may result in the clinical otosclerosis phenotype.^{25,26} Still, one of the prominent remaining hypotheses regarding the declining incidence of otosclerosis stems from the isolation of measles viruses from the perilymph of patients with the disease.^{27–30} If the measles virus drives pathogenesis of otosclerosis, then the widespread vaccination against measles could be responsible for the rapidly declining incidence.³¹ Following the advent of the measles vaccine in 1963, the number of reported cases of measles in the United States dropped by over 15-fold from 1964 to 1968.³² Further decline in incidence was observed after recommendation of a second vaccine at school age, resulting in near elimination in US residents by 2000.^{33,34} Most cases in the US reported to the CDC since 2000 are import associated reflecting travel of susceptible individuals to countries where measles remains endemic.³⁵ Changes in otosclerosis incidence over time would reflect the latency of disease development from initial infection to onset of clinical symptoms.

It is important to first note that the initial fluctuations observed in the incidence of otosclerosis over the 1960s and 1970s likely stem from a backlog of surgically correctable cases that followed the advent of stapes surgery in 1956. For example, the first stapedectomy performed in Olmsted County occurred at Mayo Clinic in 1959. By the early-1960s, the number of stapedectomies had rapidly risen to about 220 per year; however, by the mid- to late-1970s, this number had halved.² This declining surgical volume at Mayo Clinic parallels other institutional series.^{6,7,9} Pearson, et al. report the incidence of otosclerosis in Olmsted county in the 1950s as 8.9 per 100,000 person-years.³ This rises to 17.1 per 100,000 person-years in the 1960s, reflecting increased interest in the disease and its surgical management, rather than altered population demographics. Therefore, if one assumes the true incidence of otosclerosis was in the range of 10–14 per 100,000 person-years before the measles vaccine became available, then the decline in the incidence of otosclerosis that could be attributable to the measles vaccine does not occur until approximately 1988–1990, predicting an average of 20–25-year latency of disease development. All data show otosclerosis is rarely observed before age 20, implying a long latency for disease development. These observations further support the theory that widespread measles vaccination contributed to the decrease in the incidence of otosclerosis observed over the past several decades.

Of note, the rate of decline in the incidence of otosclerosis in the early-1990s does not match the decline in measles in the 1960s (approximately 2-fold decrease in otosclerosis vs. 15-fold decrease in measles). By 2017, the incidence of otosclerosis had declined about 5.8-fold from its peak (5.5-fold decrease for surgically confirmed cases). Though a more distinct inflection point in the incidence of otosclerosis might be anticipated during this time interval, the differing rate and magnitude of decline in incidence between otosclerosis and measles does not necessarily suggest that the declining incidence is unrelated to a decline in measles infections. The most likely reason for a slower change in clinical incidence of otosclerosis is that the latency period varies among susceptible individuals. Similarly, it is unlikely that the magnitude of decline in these rates would match because virtually all

unvaccinated persons are susceptible to infection with measles, whereas only a subset of the population likely harbors genetic susceptibility to the development of otosclerosis. Other factors include variable duration of conductive loss prior to presentation and variance in age at initial infection. In addition, any measles-independent pathway for otosclerosis development would not be altered by vaccination programs. Interestingly, the incidence of otosclerosis continued to decline even since the early 2000s. This observation may be related, at least to a limited extent, to the recognition of superior semicircular canal dehiscence syndrome as a unique entity. While there are many clinical and audiometric features that distinguish otosclerosis from superior canal dehiscence, both exhibit normal otoscopy and often present with a low-frequency air-bone gap. Therefore it is plausible that a proportion of clinically diagnosed cases of otosclerosis were, in fact, superior canal dehiscence, prior to its official recognition and characterization at the turn of the century. However, since the epidemiological trend of surgically confirmed cases mirrors the overall trend of clinically suspected cases, this explanation alone cannot account for the continued global decline in incidence.

Incidence Rates Among Non-White Races

The proportion of cases of non-white races significantly increased over the study period. However, this is likely attributable to the evolving racial demographics of Olmsted County rather than a true increase in disease incidence among these races. Specifically, a significant trend towards an increasing proportion of non-white races exists in the Olmsted County population over recent decades.¹¹ Nevertheless, with the majority of epidemiologic studies on non-white races dating back almost 50 years ago, modern epidemiologic data on the incidence of otosclerosis in non-white races are extremely limited.^{18–22} The current study shows that the incidence of otosclerosis in Olmsted County between 2000 and 2017 is 5.2 per 100,000 person-years in whites, 6.1 in blacks, and 5.1 in Asians. Among those of Hispanic origin, the incidence over the same time period was 6.1. These findings do not support a substantial race-specific difference the incidence of otosclerosis since 2000. Given the racial homogeneity of Olmsted County 50 years ago and the lack of race-specific denominators in the REP prior to 2000, it is unknown whether the incidence among non-white races has significantly changed or remained stable over the past half-century.

Juvenile Otosclerosis

Similar to otosclerosis at-large, very limited epidemiologic data exist regarding juvenile otosclerosis. The identification of only 17 cases of juvenile otosclerosis over the study period speaks to its rarity. Roughly equal sex distribution was observed, and the median age at diagnosis was 14 years. The incidence of juvenile otosclerosis also decreased over the study period, similar to the adult form. Overall, this suggests that adult and juvenile forms of otosclerosis may share a similar etiologic pathway. Finally, the current work shows that the modern incidence of juvenile otosclerosis is approximately 0.8 per 100,000 person-years, which has not been described in the literature recently.

CONCLUSION

Although the incidence of otosclerosis initially rose following the advent of stapedectomy surgery in 1956, these incidence rates drastically declined starting in the early-1970s. Historically considered one of the most common causes of acquired hearing loss, the low modern incidence of otosclerosis renders it legally a “rare disease” within the United States. These trends require consideration when determining trainee case requirements and developing practice guidelines.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Figure 1.

Patient age at diagnosis by decade from 1970–2017.*

*The borders of the box represent the 25th and 75th percentiles, grey dots represent individual patients, the median is drawn in the middle of the box, and the bars at the bottom and top of each decade extend to the minimum and maximum observed ages.

Figure 2.

Age-adjusted incidence of otosclerosis by sex and year of diagnosis from 1950–2017.*

*The dotted lines represent incidence rates prior to 1970 from the study by Pearson, et al.³

A

B **Figure 3.**

Comparison of disease incidence among otosclerosis and sporadic vestibular schwannoma in Olmsted County, Minnesota. (A) Incidence rates by decade from 1970 through 2016. (B) Incidence rates by age at diagnosis from 2012 through 2016.*

*The restriction of years through 2016 was used due to the absence of 2017 data on vestibular schwannoma in this population.

Table 1.
Clinical data by decade for the cohort of 504 patients identified with otosclerosis from 1970 through 2017.

	1970–1979 N=161	1980–1989 N=124	1990–1999 N=83	2000–2009 N=85	2010–2017 N=51	
Demographic and Clinical Features			<i>Median (IQR)</i>			P
Age at diagnosis in years	42 (32–55)	36 (29–51)	41 (32–54)	46 (34–53)	43 (39–52)	0.23
Years from symptoms to diagnosis (N=363)	4.0 (1.0–10.0)	1.8 (0.6–7.0)	3.0 (1.0–10.0)	3.8 (1.0–7.2)	2.2 (0.9–6.0)	0.45
Sex			<i>n (%)</i>			
Female	94 (58)	81 (65)	53 (64)	46 (54)	34 (67)	0.76
Male	67 (42)	43 (35)	30 (36)	39 (46)	17 (33)	
Race (N=488)						
Asian	2 (1)	2 (2)	3 (4)	4 (5)	3 (6)	<0.001
Black	0	0	3 (4)	3 (4)	6 (12)	
Other/mixed	0	0	0	4 (5)	2 (4)	
White	151 (99)	121 (98)	72 (92)	72 (87)	40 (78)	
Hispanic ethnicity (N=488)	1 (1)	1 (1)	3 (4)	3 (4)	6 (12)	<0.001
Symptoms						
Gradual hearing loss (N=489)	141 (92)	101 (85)	65 (79)	73 (88)	49 (96)	0.69
Tinnitus (N=476)	53 (34)	47 (41)	49 (63)	34 (44)	25 (49)	0.004
Dizziness (N=477)	20 (13)	10 (9)	5 (6)	7 (9)	6 (12)	0.38
Family history						
Otosclerosis or stapes surgery (N=279)	18 (22)	13 (23)	13 (27)	17 (27)	5 (17)	0.89
Unspecified hearing loss (N=309)	39 (42)	23 (38)	22 (43)	33 (49)	15 (39)	0.66
Pregnancy (N=264)*	17 (22)	17 (23)	8 (17)	5 (17)	7 (21)	0.56
Sidedness (N=501)						
Unilateral	83 (52)	70 (57)	48 (58)	54 (64)	28 (55)	0.16
Bilateral	77 (48)	53 (43)	35 (42)	30 (36)	23 (45)	
Treatment						
Medical therapy						
Fluoride (N=462)	20 (14)	18 (16)	9 (12)	3 (4)	2 (4)	0.008
Hearing aids (N=451)	50 (35)	42 (40)	30 (39)	32 (41)	19 (39)	0.43

Surgically confirmed (N=496)	1970–1979 N=237	1980–1989 N=176	1990–1999 N=118	2000–2009 N=114	2010–2017 N=74	
	85 (54)	57 (46)	29 (35)	42 (50)	26 (51)	0.24
Audiometric Features at Diagnosis[†]			Median (IQR)			
AC PTA in dB HL (N=647)	45 (34–59)	43 (33–53)	40 (33–50)	44 (35–55)	43 (32–51)	0.034
BC PTA in dB HL (N=645)	19 (13–31)	21 (14–28)	21 (15–29)	21 (14–34)	20 (15–27)	0.22
SRT in dB HL (N=640)	45 (32–56)	40 (30–50)	40 (30–50)	43 (35–50)	45 (35–50)	0.042
WRS in % (N=620)	96 (92–100)	100 (92–100)	100 (96–100)	100 (90–100)	100 (100–100)	<0.001

* Feature examined among women only.

[†] Audiometric features are reported *per-ear*.

Abbreviations: AC=air conduction; dB=decibels; HL=hearing loss; IQR=interquartile range; PTA=pure tone average; SRT=speech reception threshold; WRS=word recognition score.

Table 2.
Incidence of otosclerosis in Olmsted County, Minnesota between 1970 and 2017 by year of diagnosis.

Year	Women			Men			Total		
	N	Population	Incidence [*]	N	Population	Incidence [*]	N	Population	Incidence [*]
1970–1974	54	248,455	21.7 (27.4) [‡]	31	211,749	14.6 (21.1) [‡]	85	460,204	18.5 (24.2) [‡]
1975–1979	40	242,082	16.5 (18.5) [‡]	36	208,825	17.2 (23.4) [‡]	76	450,907	16.9 (20.8) [‡]
1980–1984	37	262,983	14.1 (14.7) [‡]	28	230,880	12.1 (13.8) [‡]	65	493,863	13.2 (14.3) [‡]
1985–1989	44	274,905	16.0 (17.0) [‡]	15	244,947	6.1 (6.7) [‡]	59	519,852	11.3 (12.0) [‡]
1990–1994	20	293,454	6.8 (6.9) [‡]	15	267,036	5.6 (5.6) [‡]	35	560,490	6.2 (6.2) [‡]
1995–1999	33	319,276	10.3 (11.3) [‡]	15	294,711	5.1 (6.0) [‡]	48	613,987	7.8 (8.5) [‡]
2000–2004	27	350,955	7.7 (8.3) [‡]	23	325,586	7.1 (7.7) [‡]	50	676,541	7.4 (8.0) [‡]
2005–2009	19	380,140	5.0 (5.0) [‡]	16	348,747	4.6 (4.6) [‡]	35	728,887	4.8 (4.8) [‡]
2010–2014	26	399,332	6.5 (6.9) [‡]	10	364,014	2.7 (2.9) [‡]	36	763,346	4.7 (4.9) [‡]
2015–2017	8	247,106	3.2 (3.8) [‡]	7	225,210	3.1 (3.2) [‡]	15	472,316	3.2 (3.5) [‡]

^{*} Incidence per 100,000 person years.

[‡] Incidence per 100,000 person years directly standardized by age to the 2010 US Census total population.

[‡] Incidence per 100,000 person years directly standardized by age and sex to the 2010 US Census total population.

Table 3.

Incidence of otosclerosis and sporadic vestibular schwannoma in Olmsted County, Minnesota between 2012 and 2016 by age at diagnosis.

Age	Population	Otosclerosis		Vestibular Schwannoma [‡]	
		N	Incidence [*]	N	Incidence [*]
0–17	191,925	1	0.5	0	0.0
18–30	142,769	1	0.7	0	0.0
31–40	106,518	12	11.3	0	0.0
41–50	91,695	7	7.6	4	4.4
51–60	105,426	6	5.7	13	12.3
61–70	69,648	2	2.9	10	14.4
71	67,482	1	1.5	13	19.3
All ages	775,463	30	3.9 (3.9 [‡])	40	5.2 (5.1 [‡])

^{*} Incidence per 100,000 person-years.

[‡] Incidence per 100,000 person-years directly standardized by age and sex to the 2010 United States Census total population.

[‡] Of methodological note, the incidence rates of vestibular schwannoma are slightly higher than those reported in the study by Marinelli, et al.¹⁷ due to standardization to the 2010 United States Census total population instead of the 2000 United States Census total population and restriction to the years 2012 through 2016. The restriction of years through 2016 was used due to the absence of 2017 data on vestibular schwannoma.