

Otosclerosis in New Zealand and the Pacific Islands – a review article

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INTRODUCTION

Hearing loss is the total or partial inability to detect sound waves or to understand sounds that are detected. It can be caused by various environmental, genetic, and etiological factors. Each sound must reach a certain level, known as the hearing threshold, in order to be detected. The hearing threshold is the quietest sound which an average ear with normal hearing can detect in a noiseless environment.¹ Approximately 1 in 10 people have some sort of hearing impairment.² This means, in essence, there are about 400,000 people affected in New Zealand alone. A cross-sectional view of the ear is shown in figure 1.³

Otosclerosis is a disorder in which the abnormal growth of the stapes bone in the middle ear, is confined to the endochondral layer of the otic capsule of the temporal bone.⁴ This condition gradually impedes the free movement of the stapes, leading to the abnormal conduction of sound. Hence, the resultant condition has been termed conductive hearing loss. Otosclerosis is usually a hereditary disorder.³ In 70% to 80% of patients, otosclerosis occurs in both ears, usually with the same distribution and extension.⁵ Figure 2 shows the pathological changes seen in an otosclerotic stapes bone.

Otosclerosis is usually diagnosed through a combination of family history, determination of a conductive pattern of hearing loss, and a computed tomography (CT) scan of the temporal bone.³ Although hearing loss due to otosclerosis can show a sensory pattern in initial hearing tests, it usually shows the classical conductive pattern sometime later. CT scans are specific, but not very sensitive for the diagnosis of otosclerosis.³

METHODS

Types of studies

All studies on the topic were included, regardless of the study's date of publication, language, publication status, or strength. All four studies, and an unpublished essay, 6-10 were included in this review.

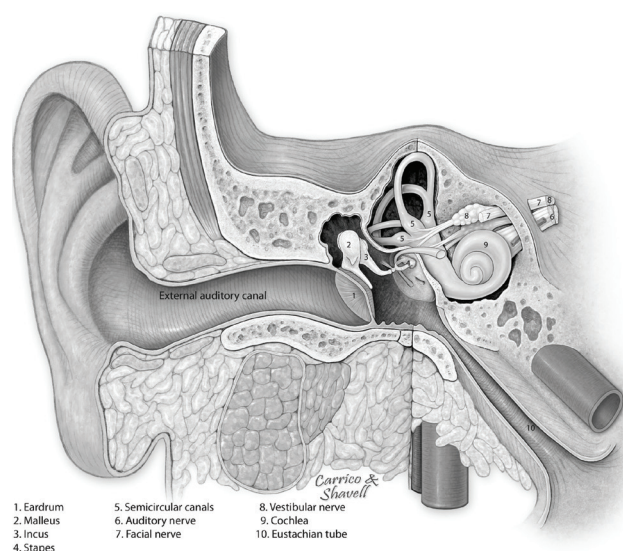


Fig 1. Cross-section of a normal ear.³

Search methods for the identification of relevant studies

Medline through PubMed search was carried out by combining the keyword "otosclerosis" or "otology" with one of the following terms: "New Zealand", "Māori", "Melanesia", "Micronesia", "Polynesia", "Australasia", "Oceania", and "Pacific Island". In addition, Google was used to find unpublished studies on the topic, and the authors were contacted to provide any missing information. Papers were searched for both online and manually, via PubMed/Medline and the Otago University Library catalogue.

RESULTS

Mode of inheritance

Although a variety of modes of inheritance have been proposed, autosomal dominant inheritance with low penetrance (about 40%) seems the most plausible. Many studies on monozygotic twins and familial pedigrees, as well as many epidemiological studies, support this hypothesis.⁴ This disease is reported to be more common in females than males (ratio of 2:1), though histological studies of the temporal bone do not show any sex-specific changes. The average age of onset ranges from 15-45 years old. However, cases in early childhood and after the age of 60 have also been reported.⁵

Prevalence

Otosclerosis can be histological, with only microscopic changes evident. On the other hand, it can progress to such an extent that it causes symptoms.

In these cases, it is referred to as clinical otosclerosis. Different studies and research papers have recorded the prevalence of otosclerosis as either histological or clinical. Despite this inconsistency, otosclerosis has been found to be prevalent to differing extents in different races:

Caucasian population (general):

- Studies have reported different estimates, ranging between 0.2%-2%4. Some studies, however, indicate that histological otosclerosis is more likely prevalent, being present in as much as 10% of the Caucasian population.⁵ Most studies agree that the Caucasian population has a markedly higher prevalence than other races.

Black population (general):

- Blacks have been reported to have a much lower prevalence of otosclerosis than Caucasians, with a prevalence of approximately 1%.⁵

Māori/Pacific Island populations within New Zealand:

- Māori population in New Zealand: There is no evidence that the Māori, as a race, have a higher genetic prevalence of otosclerosis than other races. Some have postulated that it is the increased susceptibility resulting from the environment in which indigenous Māori once lived in, or their general attitude towards disease, that makes it seem as if they are more prone to otosclerosis and ear diseases than other races.⁶ During the early 1960s, a survey was conducted on pre-school Māori children in the Waikato Hospital Board's district. Out of 645 children, 14 (~2.2%) were reported to have "other conditions", including congenital and perceptive deafness, although otosclerosis was not specifically diagnosed.⁶
- Pacific Island population in New Zealand: Insufficient numbers of Pacific Islanders, as well as inconsistency in data from different sources, hinder any true estimate of the prevalence of otosclerosis in Pacific Islanders. The estimates of the prevalence of hearing loss, including hearing loss due to otosclerosis, which were based on a 2001 update of a survey done in 1991 and 1992, are shown in table 1.⁷ The different prevalence percentages may be a reflection of differing genetic makeup among these groups.⁷

Pacific Island populations outside New Zealand:

	Māori (%)	Pacific Island (%)	Other (%)
0-14 years	4.6	2.6	1.9
15-64 years	5.0	3.9	5.5
65+ years	14.3	2.6	20.7
Total	3.4	2.5	6.5

Table 1: Estimates of the prevalence of hearing loss for different age ranges and ethnic backgrounds in New Zealand.⁷

- Natives of Easter Island: Easter Island is a Polynesian island in the southeastern Pacific Ocean.⁸ The population of this small, isolated island consists of three ethnic groups: the natives, the mixed race, and the continentals. The natives are of a Polynesian ethnic background.⁸ A study looking at the effect of an industrialized lifestyle on hearing was done in the mid-1980s. A pool of 347 participants (~19% of the population) underwent full otolaryngological and audiological assessments. From this pool, 90 (~26%) were of the native population of Easter Island. Of this native population, the study reported neither a single case of otosclerosis nor a family history of otosclerosis.⁸ Even though the population size may have been too small, not finding a single case raises the possibility of some "hidden" factors that are yet to be determined.
- Fijian population: Medical problems in Fiji can be represented in very two distinctive populations, due to very little mixing between the two major racial groups, which are the Melano Fijians and Indian Fijians. Hence, there is a marked difference in the incidence of otosclerosis between the two racial groups.⁹ According to Stewart, otosclerosis is virtually nonexistent in Melano Fijians, whereas it has a greater prevalence amongst Indian Fijians.⁹ Although specific numbers were not given, Indian Fijians also reported family histories of otosclerosis and hearing loss related to otosclerosis at a more significant frequency. However, this problem is usually left untreated due to a lack of expertise in stapes surgery.⁹
- Tongan and Samoan populations: Discussions of otosclerosis in these populations are very limited in the medical literature. One study reported a prevalence of otosclerosis in these populations that was "similar" to that in Melano Fijians, but provided no further details.⁹ Otolological conditions, such as chronic otitis media and mastoiditis, were much more common, which is most likely due to a lack of hygienic practices.⁹

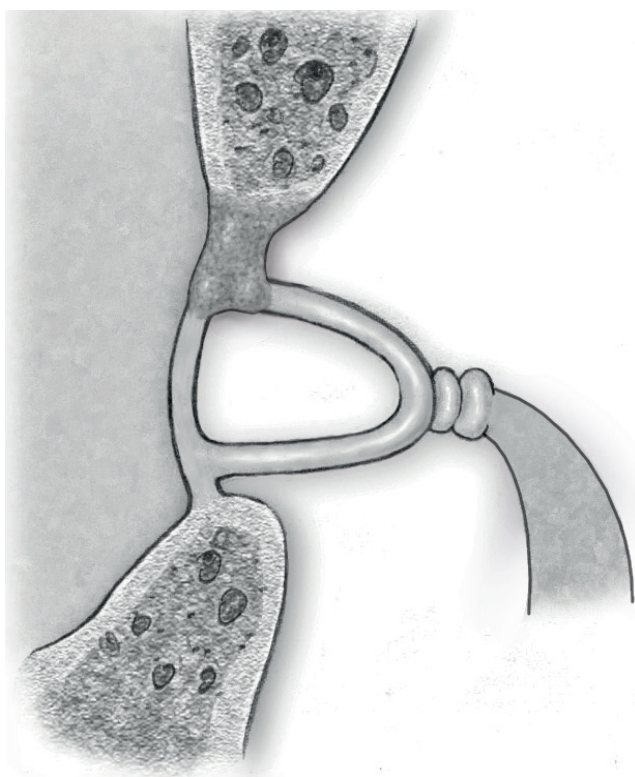


Fig 2. Pathological changes to the stapes bone that are seen in a patient with otosclerosis.³

DISCUSSION

Limitations

Although an extensive review of the relevant literature has been undertaken, it is important to acknowledge the possible limitations of such studies. This is especially important when comparing different studies, as these limitations can distort the true prevalence of otosclerosis. These limitations can be grouped into three categories which will be discussed further:

- Limitations associated with detecting and diagnosing otosclerosis in the wider community.
- Limitations in recruiting patients for such studies.
- Practical limitations.

First, there is usually some difficulty in detecting and diagnosing all cases of otosclerosis in the community. Otosclerosis does not usually present with any pain to the patient, making visits to otologists dependant on social, cultural and economic norms, rather than a sense of urgency and discomfort. This becomes much more evident as we see the distinctive ethno-geographical differences in the prevalence of otosclerosis. Similarly, many cases of otosclerosis are not detected simply because the patient's hearing level has not yet fallen below socially adequate levels.¹⁰ Likewise,

some patients assume that their hearing loss is, for example, noise-induced and, therefore, not treatable.¹⁰

Inclusion criteria for studies of patients with otosclerosis are not ideal. Many studies recruit otosclerosis subjects through their history of stapes surgery. Many patients who are eligible for surgery, however, do not undergo surgery because their air-bone gap is not sufficiently great to justify it.¹⁰ Furthermore, elderly patients are usually averse to having an operation on their ears and would rather continue wearing hearing aids.¹⁰

Finally, conducting such studies poses some practical problems. Factors such as the genetic causality of otosclerosis, as well as the condition's relative mildness, have put otosclerosis on the "back burner" of clinical research. We should also keep in mind the health disparities and discrepancies that occur with patients of Māori and Pacific Island origin, when considering the prevalence of any disease. Even though this problem is not as bad as it used to be 40 or 50 years ago, most of the studies found on this topic have been published during those times, ranging from the 1970s to the 2000s.

Fluoride deficiency hypothesis

Some environmental trace elements are now widely known to be associated with human disease such as iodine deficiency and goiter, and zinc and gastrointestinal problems.¹⁰ Several correlation studies have proposed an association between fluoride deficiency and an increased incidence of otosclerosis. For example, a lower incidence of otosclerosis has been reported in Texas, where the concentration of fluoride in the drinking water is relatively high (1.9 parts per million).¹¹ On the other hand, a lower concentration of fluoride in the drinking water in Missouri (0.6 parts per million) was associated with a higher incidence of otosclerosis.¹¹ Another study showed that fluoride therapy arrested the progression of sensorineural hearing loss in patients with otosclerosis.¹² Unfortunately, both studies included small, unrepresentative populations, so results cannot yet be generalized. It would be interesting to do a randomized, controlled trial on a sufficiently large population to study whether the incidence of otosclerosis truly decreases once a drinking-water fluoridation program has been implemented. To date, only a few observational studies have been carried out, all with small populations, to assess the possibility of an association between fluoride levels in drinking water and otosclerosis status. These studies concluded that the fluoridation of drinking water had a beneficial effect on otosclerotic ears that had not been operated on but no significant effect on the hearing levels of ears after an operation.^{13,14}

CONCLUSION

Otosclerosis is a genetic disease that affects bone homeostasis in the middle ear. Prevalence differs vastly according to genetic makeup, with the White/Caucasian population having the highest prevalence rate all among racial groups. Even though current treatment methods are effective and successful, further research is required, especially in countries and regions with no previous studies on otosclerosis among their populations, such as New Zealand and the Pacific Islands.

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