Ministry of Defence

Synopsis of Causation

Otosclerosis

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Disclaimer

This synopsis has been completed by medical practitioners. It is based on a literature search at the standard of a textbook of medicine and generalist review articles. It is not intended to be a meta-analysis of the literature on the condition specified.

Every effort has been taken to ensure that the information contained in the synopsis is accurate and consistent with current knowledge and practice and to do this the synopsis has been subject to an external validation process by consultants in a relevant specialty nominated by the Royal Society of Medicine.

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1. Definition

1.1. Otosclerosis is a localised progressive disease of bone remodelling. The disease involves principally the stapes bone in the middle ear. The stapes bone has a footplate and a head joined by two arches. In otosclerosis, new spongy bone forms around the footplate and may cause the stapes to become fixed. There is resorption of the stable otic capsule followed by a reparative phase with bone deposition.[1] Hearing is impaired, as movements of the ossicles are essential for sound conduction through the middle ear. Otosclerosis may also involve the inner ear sensory organ known as the cochlea.

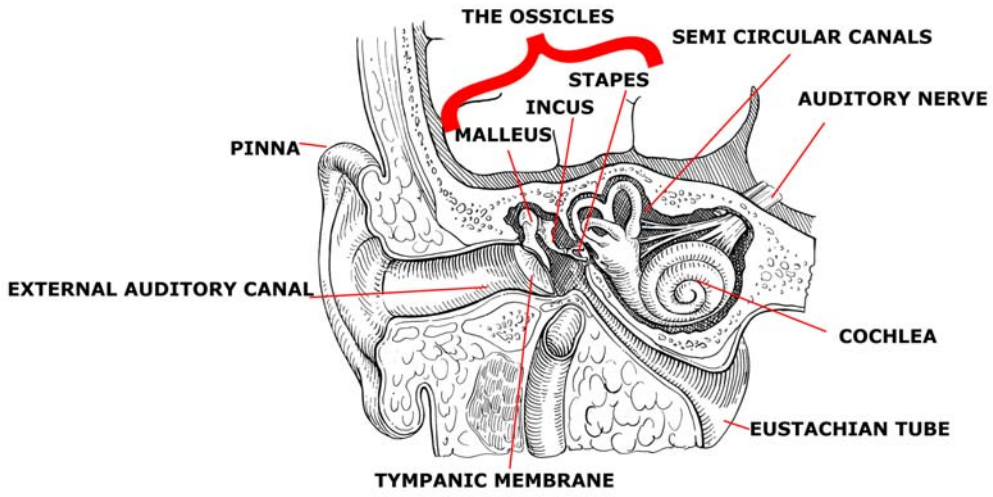

Figure 1: Anatomy of the ear

2. Clinical features

2.1. Otosclerosis is an uncommon, but well recognised, cause of conductive hearing loss in adults. The condition affects both sexes although there may be a female predominance. The apparent male to female ratio is 1:2. It is more common in the white population, with a clinical prevalence of 0.3-0.4%. Histological otosclerosis is present in 1% of cadaver temporal bones,[2] so that about 50% of the disease is sub-clinical. The onset is usually insidious and disease progression is characteristically slow, although it may be rapid at times especially in pregnancy. The age of onset, clinically, is usually between the ages of 20 and 30 years.

2.2. **Hearing loss.** The hearing loss is conductive in stapedial otosclerosis, although an apparent sensorineural element may be noted at 2000 Hertz (Hz) on pure tone audiometry. This is the basis of the so-called Carhart's notch, an important diagnostic clue. The Carhart's notch is a dip on the audiogram at 2000 Hz; the dip is apparently sensorineural although in reality the deafness is conductive, reasons for this being physiological and complex. This does not however indicate a cochlear involvement.[3] When the otosclerotic lesion extends from the footplate of the stapes to involve the cochlea, the deafness becomes of mixed type, the conductive element from stapes fixation and the sensorineural element from the cochlea. It may rarely be purely sensorineural.

2.3. **Paracusis.** This is a phenomenon in which the patient hears better in a noisy environment and is characteristic of otosclerosis.

2.4. **Tinnitus.** Patients with otosclerosis may experience noise in their ears. The tinnitus is usually on one side and is not necessarily associated with cochlear degeneration. The tinnitus may be mild and is usually not troublesome. The cause of the tinnitus is not known. Treatment may or may not affect this, although a successful operation that restores hearing may reduce or eliminate the tinnitus.

2.5. **Vertigo.** Dizziness may occur. This is usually due to a sudden change in the position of the patient's head and is self-limiting. The duration of the vertigo is not quantifiable.

3. Aetiology

3.1. Otosclerosis is an inherited localized disease of the bone of the ear with typically islands of new bone laid down at certain sites. These have a characteristic pattern. The mechanism is unknown.

3.2. **Heredity.** There may a strong family history of the disease. The inheritance is thought to be autosomal dominant with perhaps variable penetrance. To date, 5 otosclerosis loci (chromosomes) have been reported.[4]

3.3. **Infection**. The presence of measles virus in the otic capsule has been shown in otosclerotic patients.[5] The number of stapedectomy cases has declined over the past 30 years in the United States. The reason for this is uncertain, although widespread immunisation for measles is a plausible hypothesis.[6] It remains to be proven that infection may cause otosclerosis.

3.4. **Pregnancy.** Although pregnancy does not cause otosclerosis there is good evidence that the condition may worsen during pregnancy. Shambaugh, in 1967, looked at 475 mothers who had treatment for otosclerosis and found the risk of increased hearing loss from any one pregnancy in a woman with stapedial otosclerosis to be 1 in 24.[7] Gristwood and Venables found that pregnancy aggravated deafness in women with bilateral otosclerosis from 33% after one pregnancy to 63% after 6 pregnancies.[8] Hormone replacement therapy may have a similar effect.

3.5. **Trauma.** Otosclerosis is not caused by trauma. However, trauma to the head has been reported to accelerate the condition in some patients. The conductive loss due to otosclerosis may also protect the ear from noise damage in patients exposed to loud noise.[9]

4. Prognosis

4.1. Clinical onset of otosclerosis at an early age and rapid progression of hearing loss carries a worse prognosis. Bone density measurement around the footplate of the stapes on CT has recently been shown to be a good indicator of disease progression. The bone density is lower in otosclerotic patients compared to controls and, the lower the bone density, the greater the hearing loss.[10]

4.2. Surgery and aiding are 2 options for the treatment of stapedial otosclerosis. The results of treatment for stapedial otosclerosis by surgery, or aiding, are excellent.

4.2.1. Successful stapes surgery may restore hearing in up to 90% of cases dealt with in specialised centres,[11,12] with less successful surgery improving hearing in 10% of cases. Some of these studies are based, however, on short-term follow up. There is a known deterioration in hearing following a successful stapedectomy which is usually up to 10 dB every 10 years, and which may result in the patient requiring a hearing aid later in life.[13,14]

4.2.2. Total un-aidable deafness is met in 1% of cases. With no treatment, the hearing loss drops to 60 dB and with time may become sensorineural. Aiding with a modern digital aid works very well if the patient is happy to wear one.

4.3. In cases of cochlear otosclerosis, oral fluoride may help to stabilise the sensorineural hearing loss.

5. Summary

5.1. Otosclerosis is a known cause of conductive hearing loss and presents most commonly between the ages of 20 and 30 years. It may involve one or both ears and is usually a progressive disease.

6. Related Synopses

Conductive Hearing Loss

Sensorineural Hearing Loss

7. Glossary

autosomal dominant inheritance	Requiring that only one parent need have the trait (characteristic) in order to pass it to the offspring.
cochlea	The sensory hearing organ of the inner ear.
CT	Computerised tomography.
Hertz (Hz)	A measure of frequency of sound.
ossicles	The 3 middle ear bones. These are the malleus, incus and the stapes.
otic capsule	The bony covering of the membranous inner ear. During development of the embryonic ear, the cartilaginous otic capsule derived from mesoderm surrounds the differentiating inner ear and latterly becomes ossified.
sensorineural	A type of deafness that involves the cochlea and/or the auditory nerve.
stapedectomy	Surgical removal of the stapes or part of the stapes.
stapes	The smallest bone in the human body, connecting the membranous inner ear with the middle ear. It has a head, neck, 2 crurae and a footplate. The footplate lies in the oval window on the lateral surface of the middle ear.
tinnitus	A sensation of noises in the ear and sometimes the head. The term is usually used for subjective tinnitus.

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