# Otosclerosis

**By Justin Cole, MD and Michael Bergstein, MD**

Otosclerosis is a pathological remodeling of the bones within the otic capsule that occurs at a faster rate than normal, resulting in bony deposits that damage ossicles and other conducting structures. If untreated, it can lead to a combined conductive and sensorineural hearing loss as damage impinges on the cochlea and other inner-ear structures.

## Signs and Symptoms

Otosclerosis presents as progressive loss of hearing that is worse at lower frequencies. Patients report hearing deeper voices and vowel sounds during normal conversation. Some 50% of patients have tinnitus, and 10% complain of vertigo.If otosclerosis is present, there is about an 80% chance that it exists bilaterally. Progression is directly proportional to the degree of hearing loss.

Otoscopic exam is typically normal. Some patients exhibit Schwartz sign, which is a pink-red spot on the promontory of the tympanic membrane. Results of the Hearing Handicap Inventory for the Elderly Screening Version (HHIE-S) questionnaire correlates with hearing loss verified on audiometric findings. Results of the Whisper test and audioscope also correlate with audiometric findings. Use of the tuning fork, including the Rinne and Weber tests, are not recommended in assessing otosclerosis.

NOTE: Hearing screening may be a useful diagnostic aid, although it is not a substitute for formal audiometry, which should be conducted in all cases of suspected otosclerosis.

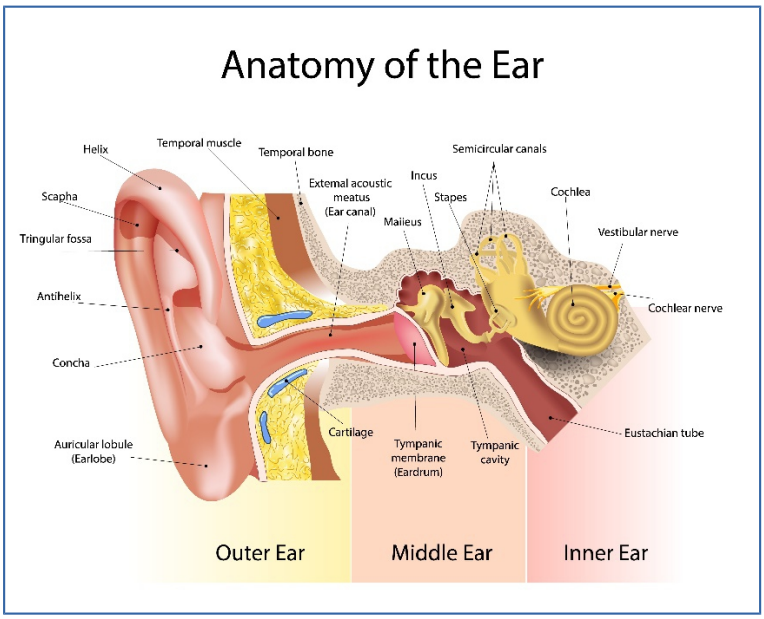
**Causes and Risk Factors**

Otosclerosis may be autosomal-dominant with incomplete penetrance; ~ 60% of patients report a family history of otosclerosis. Measles exposure may predispose for the condition, but the mechanism behind this process has not been elucidated.

**Pearl to Know**

Suspect otosclerosis if there is a family history of hearing loss or if hearing loss becomes worse during times of increased hormone production (eg, puberty, pregnancy, menopause). The disease is histologically present in 12% of Caucasians; only 0.2%-0.3% exhibit signs and symptoms. It is much less common in all other ethnic groups. It affects females more frequently than it does males.

**Treatment Options**




Stapes surgery corrects otosclerosis-associated conductive hearing loss, but it will not restore sensorineural hearing loss from extension of disease into the inner ear. In a stapedectomy, the stapes footplate and crura are removed and replaced with a prosthesis. In a stapedotomy, a small hole is created in the stapes footplate through which a prosthesis is placed, without removal of the stapes. A cochlear implant is an

option for patients with sensorineural hearing loss, although it may be difficult to implant in otosclerotic patients. Furthermore, there is increased risk of cochlear ossification. The bone conduction implant is an option for patients with conductive or combined conductive/sensorineural hearing loss. This device is implanted in the temporal bone behind the ear and transmits sound waves directly to the cochlea. It typically requires only unilateral implantation; it is strong enough to stimulate the contralateral cochlea. A middle-ear implant requires the presence of, and amplified conduction through, the ossicular chain and are only implanted concurrently or shortly following stapes surgery.

There is limited evidence supporting the use of pharmacologic therapy in otosclerosis.

**References:**

Batson L, Rizzolo D. Otosclerosis: An update on diagnosis and treatment. *JAAPA.* 2017;30:17-22.

Michels T, Duffy M, Rogers D. Hearing Loss in Adults: Differential Diagnosis and Treatment. *Am Fam Physician.* 2019 Jul 15;100(2):98-108.

Toscano M, Shermetaro C. Stapedectomy. StatPearls [Internet]. Last Update: January 12, 2021.