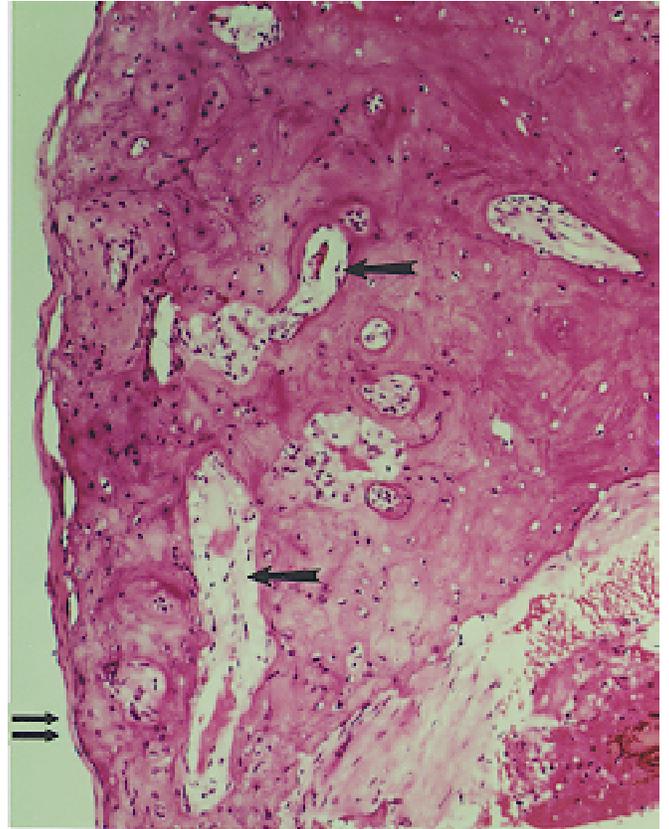


OTOSCLEROSIS

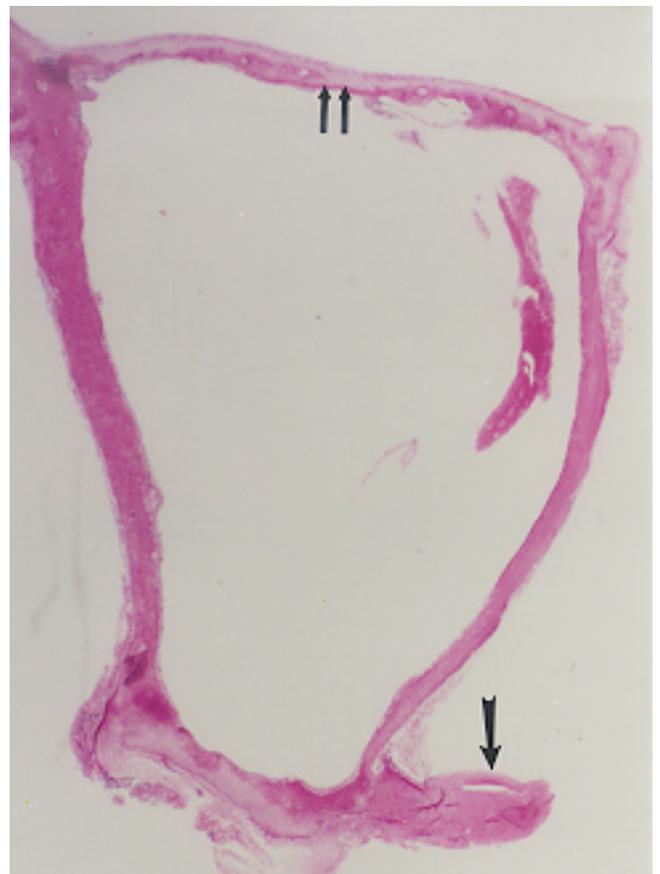
Otosclerosis affects various parts of the temporal bone but causes symptoms only when it fixes the stapedial footplate or invades the cochlea. Using the operating microscope, the distinction between normal bone of the stapedial footplate and abnormal otosclerotic bone is quite clear. There is a thickening and whitening in the abnormal area (early disease). Often only the posterior part of the footplate is involved leaving the anterior half clear. Areas of the middle ear adjacent to the footplate may also be affected and there may be sufficient “overhang” of otosclerotic bone from the cochlea or fallopian canal that the footplate becomes partially obscured and stapedectomy hindered. The disease process may bulge inwardly to encroach on the vestibule and outwardly to partially fill the intercrural space and then the otologist speaks of a “doughnut” footplate.

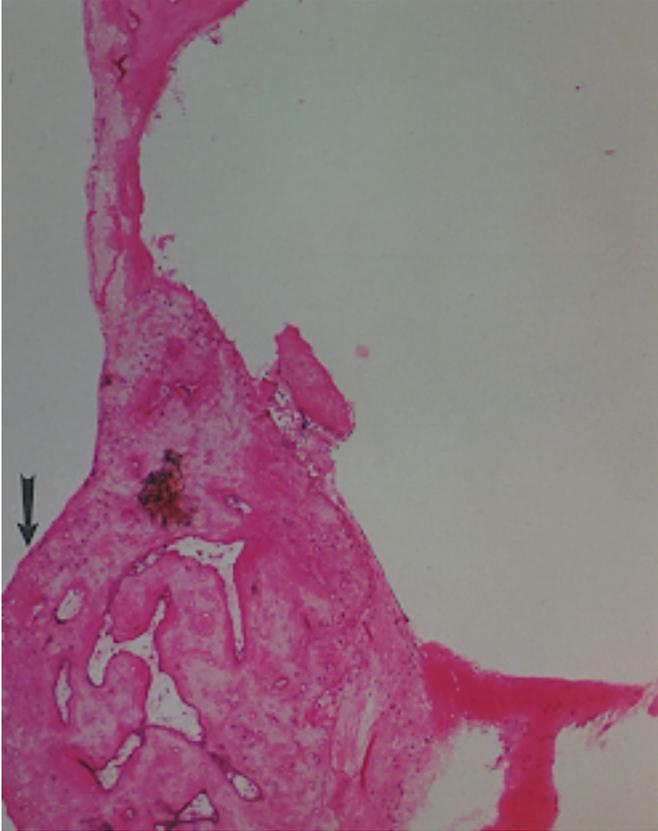
Microscopically, the appearance of otosclerosis differs depending on the stage of the disease. At first (active otosclerosis) there are numerous fibrovascular spaces and increased osteoblasts and osteoclasts. The new otosclerotic bone is soft and bleeds if scraped. At times when this vascularity includes the promontory (basal whorl of cochlea) a faint pink blush (Schwartz sign) is seen through the normally translucent drumhead indicating a poor prognosis for hearing. In the inactive phase the fibrovascular spaces look less active, less vascular, and may resemble the normal adjacent bone of the cochlear capsule.

Shows a large focus of inactive otosclerosis involving the entire footplate and extending deeply into the vestibule (double arrows). The fibrovascular spaces (single arrows) become filled with lamellar bone. This specimen was acquired at the time of stapedectomy. After reconstruction of the ossicular chain, hearing improved dramatically.

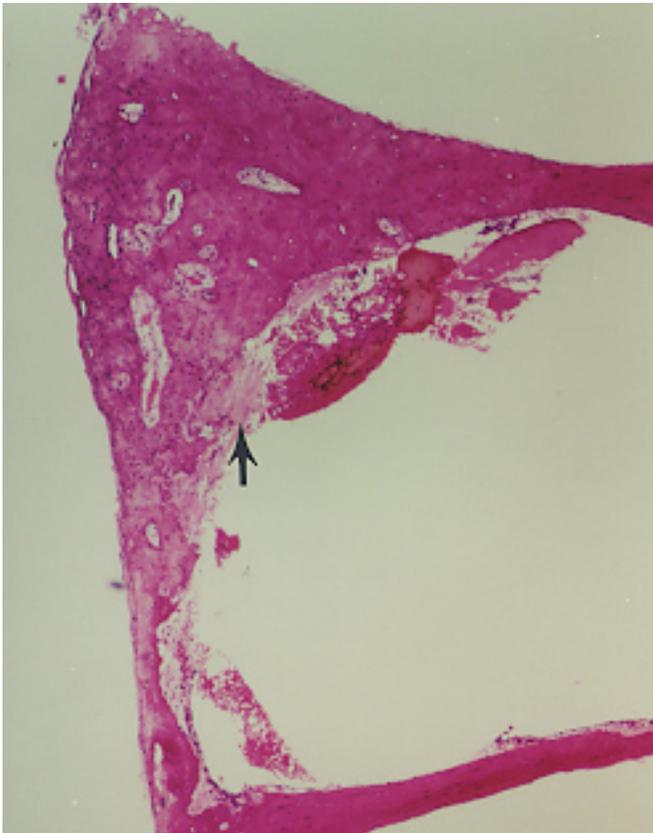


Normal stapes for orientation. Large arrow points to stapedius tendon which attaches to junction of head of stapes and posterior crura. Footplate (double arrows) measures about μ # 3 mm and fits into oval window where it is held in place by an annular ligament.



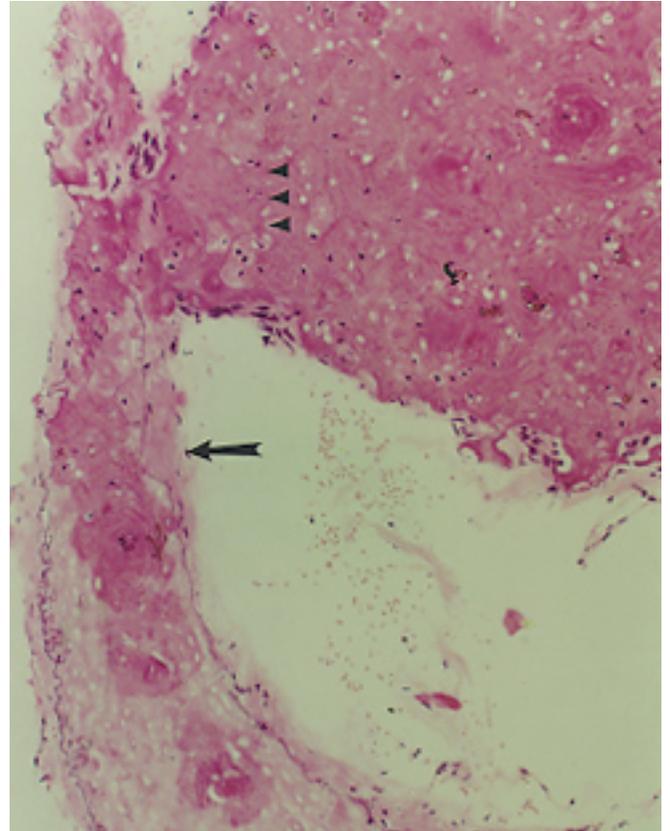


Active otosclerosis. Base of anterior crura is most markedly affected (arrow).



Active otosclerosis involves entire footplate and base of one crura with bulging into vestibule (arrow).

The large focus of inactive otosclerosis represents a greatly thickened footplate (triangles) while the curving piece of bone (arrow) is the base of one of the crura. It too, is invaded by otosclerosis.



CLINICAL ASPECTS

Ankylosis of the stapes causing deafness has been recognized since 1737 (Valsalva). The term otosclerosis was provided by Politzer over a hundred years later. In 1912, the term otospongiosis was proposed as a more descriptive term for the disease which, at least initially, presents a more spongelike or porous appearance than does the hard bone of the otic capsule that it replaces.

Otosclerosis occurs in some 10% of the population but since the focus is minute and often not even in the footplate, there is no hearing loss. Clinically significant otosclerosis may affect as much as 1% of the white population but is less common in other races. The disease begins in the late teens and gradually advances producing progressive conductive hearing loss until a section of the stapes footplate becomes fixed to the margin of the oval window when a slowly progressive conductive hearing loss begins.

Otosclerosis must be distinguished intraoperatively from tympanosclerosis which can cause the same type of hearing loss and which at the footplate level looks much like inactive otosclerosis. The distinction between the two, however, is clear since otosclerosis infiltrates the bone of the footplate whereas tympanosclerosis is

deposited in the mucous membrane covering the footplate and is removable by scraping or peeling. Also in early otosclerosis there is bleeding when the footplate is disturbed surgically but not in tympanosclerosis. Finally, patients who have tympanosclerosis fixing the footplate invariably have additional heavy deposits of tympanosclerosis in the tympanic membrane and often about the heads of the malleus and incus and in mucous membranes of other areas of the middle ear.

In addition to footplate disease, otosclerosis also invades the cochlea in about a third of patients with clinical otosclerosis. When it does the patient develops sensorineural hearing loss in addition to the conductive hearing loss that occurs when only the stapes is fixed. With cochlear invasion there may be sufficient increase in vascularity to produce a so-called Schwartze sign a faint pink blush readily seen through the normal translucent tympanic membrane.

Treatment to restore hearing in a patient with otosclerosis began in the latter part of the 19th century and consisted of stapedectomy, but the operation was largely unsuccessful and was dropped. In the early 20th century the fenestration operation was devised in Europe but was little used until 1938 when Julius Lempert revised and perfected the technique in America. In the fenestration operation, now no longer in use, a bypass around the fixed stapes is created by a new window ("fenestra nov ovalis") drilled in the horizontal semicircular canal. In 1952, Rosen accidentally "rediscovered" the stapes mobilization technique, but this operation, since it did not remove the stapes footplate but temporarily freed it from its otosclerotic attachment, often failed, as the bony fixation recurred. Finally, in 1967, Shea developed the modern stapedectomy operation and now this procedure, or a variation called stapedotomy, is widely and successfully used to restore hearing to patients with conductive hearing loss due to otosclerosis. (Some otologists believe that long-term use of fluoride compounds will prevent or delay the development of cochlear otosclerosis.