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## DISORDERS

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### ABNORMAL GROWTH

An abnormal growth of bone in the middle ear can cause hearing loss.

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### ARTICLE

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# Otosclerosis

By Habib G. Rizk, MD & Sunny Shah, MD - Medical  
University of South Carolina, Department of Otolaryngology  
- Head & Neck Surgery

## WHAT IS OTOSCLEROSIS?

Otosclerosis is defined by abnormal and unregulated growth of bone within the bony structures that make up the middle ear.<sup>1</sup> Any bone within the middle ear can be affected, however, the footplate of the stapes is most commonly the site of insult. Patients who suffer from otosclerosis are more likely to suffer from mild to severe conductive or mixed hearing loss.

## CONDUCTION OF SOUND AND HEARING IMPAIRMENT BY OTOSCLEROSIS

Sound transduction begins with sound waves passing from the external ear into the ear canal. Within the ear canal lies the tympanic membrane, a structure that vibrates when exposed to sound waves.<sup>1</sup> The middle ear is made up of three small bones referred to as ossicles. The ossicular chain begins at the malleus, a structure attached to the tympanic membrane, then moves to the incus and finally to the stapes, which is attached to the oval window.<sup>1</sup>

Sound waves travel through the ossicular chain and make their way to the cochlea in the inner ear where the sound waves pass through a fluid apparatus causing hair cells within the inner ear to shift.<sup>1</sup> The hair cells then transform the sound into neural impulses which are sent via a branch of the eighth cranial nerve, the cochlear nerve, to the brain where sound is processed and interpreted. Different hair cells within the cochlea are stimulated based on the frequency of the sound waves.

When an insult occurs to any part of the sound transduction system, hearing loss can occur. An insult to the bones of



the inner ear, specifically, leads to a conductive hearing loss.<sup>1</sup> When increased bone deposition occurs at the footplate of the stapes, the vibrations sent through the ossicular chain can be dampened at the point between the stapes and the oval window. This dampening leads to hearing loss.

**HOW DOES OTOSCLEROSIS OCCUR?**

There are a variety of factors that can lead to otosclerosis of bone. External environmental factors such as sodium fluoride, viral antigen exposure, and exogenous hormone intake have been shown to cause dysregulation of bone formation.<sup>2,3</sup> Internal factors such as autoimmune/inflammatory diseases, hormone dysregulation, and genetic anomalies can also lead to otosclerosis.<sup>2,4</sup>

About 50-60% of patients suffering from otosclerosis have a genetic predisposition for the disease and a majority of patients report a family history of otosclerosis.<sup>2,5</sup> Studies are currently underway to narrow down specific genes that are implicated in causing this disease. Although a majority of otosclerosis cases affect hearing and interfere with communication between the middle ear and inner ear, studies have also found that otosclerosis can occur on the vestibular apparatus leading to vertigo.<sup>6,7</sup>

**WHAT ARE THE SYMPTOMS OF OTOSCLEROSIS?**

Progressive conductive or mixed hearing loss is the most common symptom experienced by those with otosclerosis. People may experience improvement in their hearing in noisy environments, a phenomenon known as Paracusis of Willis.<sup>5</sup> Tinnitus, or ringing in the ears, may also be present.

Vertigo, dizziness, or imbalance may

SYMPTOMS
Tinnitus
Hearing Loss
Vertigo
Imbalance
Dizziness

also be present in people suffering from otosclerosis depending on the severity of inner ear involvement.<sup>1</sup> It is crucial to accurately diagnose the cause of vertigo as patients with otosclerosis may also have a concurrent undiagnosed vestibular disease.<sup>5</sup>

The clinical presentation of patients with otosclerosis can be vastly different. While the hearing loss is the typical presentation, dizziness has been associated with the otosclerosis pathology as well. Typically, vestibular symptoms have a lag time following conductive hearing loss.<sup>8</sup> Studies show that almost half of patients with otosclerosis experience vestibular dysfunction during the disease process. Vestibular testing such as caloric testing, oVEMP, and cVEMP have been found to be abnormal in patients with otosclerosis, even if asymptomatic.<sup>9</sup> Thus, vestibular testing should be considered when a patient with otosclerosis presents with dizziness.

Accurate diagnosis is crucial when considering surgical therapy versus medical therapy, especially if there is dizziness associated with the hearing loss. Meniere’s disease (MD) should be

ruled out given the surgical treatment for otosclerosis, a stapedectomy, is contraindicated in patients with Meniere's disease. MD typically presents with fluctuating hearing loss, tinnitus and vertigo. Furthermore, superior semicircular canal dehiscence (SSCD) may present as a similar type of hearing loss (on audiometry), however, other tests such as the stapedial reflex are still normal and this should prompt imaging before proceeding with a stapedectomy. Patients with SSCD usually have other symptoms such as autophony (hearing their voice, their pulse, or their breathing), a Tullio phenomenon (vertigo induced by sound), vertigo induced by pressure, and/or sensitivity to noise and to internal body bruits (such as hearing their eyes moving). Sometimes, patients with SSCD are asymptomatic.

## DIAGNOSIS OF OTOSCLEROSIS

Otosclerosis is typically best diagnosed by an otologist/neurotologist.<sup>1</sup> Investigation usually involves a comprehensive hearing test, CT imaging of the middle and inner ear to inspect bone density, and vestibular testing depending on whether patients are experiencing associated dizziness or imbalance.<sup>1</sup> Careful consideration should be given to other diagnoses that may present with similar symptoms to properly guide management.



## TREATMENT OF OTOSCLEROSIS

Depending on the severity of symptoms, a neurotologist/otologist may recommend different treatment types.

For milder hearing loss, a trial of bisphosphonates may be attempted to better regulate bone remodeling of the middle and inner ear structures. A recent study highlighted the effectiveness of sodium fluoride therapy and bisphosphonates in treating vestibular and audiologic symptoms. Sodium fluoride and bisphosphonate treatment for 6 months both resulted in stabilization of hearing loss, improvement of vestibular symptoms and delay in progressive tinnitus.<sup>10</sup> Conservative treatment can, at times, also be considered for rapidly progressive otosclerosis with cochlear and vestibular involvement to slow down disease progression. Although, historically, sodium fluoride was the preferred medication, a recent shift towards bisphosphonate use has occurred, especially in patients who are intolerant to sodium fluoride.<sup>10</sup>

Any patient with otosclerosis and clinically significant hearing loss may qualify for hearing aids. Surgical therapy may also be considered unless a contraindication is present (i.e. active Meniere's disease or only hearing ear). The current standard treatment is a stapedectomy or stapedotomy which allows a prosthesis to replace damaged bone. The goal of a stapedectomy is to remove the diseased bone while replacing it with a prosthesis that can re-establish vibration of the fluid within the inner ear for improved sound conduction.<sup>11</sup> While there are multiple prostheses or surgical techniques that can be employed, in the hands of an experienced surgeon very few differences in outcomes exist.<sup>12,13</sup> Patients with far advanced otosclerosis, and severe hearing loss, can still benefit from stapedectomy

followed by hearing aid amplification, however there are cases of significant hearing loss where cochlear implantation is indicated. This surgery can significantly improve hearing. In fact, studies have shown that patients with otosclerosis show similar improvement in hearing as patients without otosclerosis after cochlear implantation.<sup>14</sup> Discussing all treatment options with an otologist is important.

Some risks associated with surgical treatment of otosclerosis with stapedectomy/stapedotomy include severe hearing loss (1%) or post-operative vertigo or dizziness which can happen for a variety of reasons including an inappropriately long prosthesis or misplaced prosthesis that protrudes too far into the inner ear.<sup>15</sup>

## INNOVATIONS IN OTOSCLEROSIS

New techniques for minimally invasive surgical intervention are being developed. Endoscopic stapedotomy has shown to provide a few benefits when compared to the traditional microscopic approach: shorter operation times, similar audiologic outcomes and complication rates, and less invasive.<sup>16</sup>

Multiple genes have been identified and implicated in the incidence of otosclerosis emphasizing the importance of family history. Genome wide studies have identified 23 loci which are associated with an increased risk of otosclerosis.<sup>17</sup>

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