



Dr. Storper is a current fellow with The EAR Foundation's Fellowship Program.

INTRODUCTION

In order to hear, sound pressure waves in the air are converted to nerve impulses on the auditory nerve by the ear. These impulses are then relayed to the brain. The ear does this in an ingenious way: first sound waves arrive at the outer ear and are funnelled into the ear canal. When they reach the end of the canal, they hit the eardrum and set it into vibratory motion. The eardrum is connected to a series of three tiny bones, the malleus, incus, and stapes, which are therefore also set into motion. The stapes normally rests on the inner ear, a fluid-filled chamber enclosed in bone, which has the ability to convert pressure waves which the moving stapes generates into nerve impulses on the auditory nerve, allowing us to hear sound. The area where the stapes meets the inner ear is called the oval window.

Otosclerosis is a condition where normal bone of the ear is replaced by abnormal, spongy bone. This abnormal bone can get in the way of normally functioning ear components. There is no known cause of this condition, although a number of factors have

OTOSCLEROSIS

Ian S. Storper, MD

been implicated in the past. In approximately 12% of patients with otosclerosis, the oval window becomes involved with abnormal bone. This bone interferes with the normal motion of the stapes, resulting in hearing loss. In the vast majority of these patients, the problem is treatable.

HISTORICAL ASPECTS

The history of otosclerosis dates back to 1735 in Venice, when the pathologic process of this disease was described by Valsalva, from the autopsy of a deaf patient. The actual involvement of the stapes by this process was first noted by von Troltsch in 1881. The earliest surgical attempt to improve the hearing loss associated with otosclerosis was performed by Kessel in 1876. He attempted to break it free of its abnormal bony attachments to restore the hearing mechanism. If the stapes could not be freed from its attachments, it would be removed. The first American attempts to improve this type of hearing loss were performed by Blake and Jack of the Massachusetts Eye and Ear Infirmary, in 1892 and 1893 respectively. Due to complications associated with breaking the stapes free, these operations were largely abandoned at the turn of the century.

Barany and Holmgren attempted a different, safer operation in Germany in 1923 to improve the hearing loss associated with otosclerosis. They did this by opening the inner ear and allowing sound to be transmitted directly to this area. Partial hearing improvement was frequently noted. In 1938, Julius Lempert of New York

devised a simplified method of performing this operation, which he called the fenestration; it remained the standard procedure until 1952.

The next major advancement occurred in 1952, when Samuel Rosen inadvertently jarred a stapes loose of its surrounding attachments during a fenestration procedure. There was an immediate improvement in hearing, repopularizing this mobilization operation from the nineteenth century. Unfortunately the abnormal bone of otosclerosis often regrows, producing recurrent hearing loss, after one of these procedures. After Shambaugh applied the operating microscope to ear surgery in 1954, the stapedectomy operation was redeveloped in 1958 and remains the gold standard to this day. In this operation, the fixed stapes is removed and bypassed by a prosthesis connecting the incus to the inner ear. By using the operating microscope, the complication rate decreased dramatically.

SIGNS AND SYMPTOMS OF OTOSCLEROSIS

Otosclerosis most commonly presents as progressive hearing loss over many years. The hearing loss is occasionally accompanied by ringing in the ears. Unsteadiness is present only in very rare instances. Over 60% of the time, it is present to some degree in both ears, and almost 20% of the time, it is found to occur in other family members. After many years, the inner ear may also become involved with abnormal bone. If this does occur, unserviceable hearing loss may develop.

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DIAGNOSIS

Otosclerosis is usually diagnosed by the symptoms which a patient experiences. Any patient who has a slowly-progressive hearing loss in both ears in a family with similar symptoms is likely to have this condition. The diagnosis is confirmed by the hearing test, results of which are highly suggestive of a process fixing the bones of the middle ear. The diagnosis cannot be completely established unless the ear is explored and the disease process visualized.

TREATMENT

There are three treatment options for otosclerosis. If the hearing loss is mild, no treatment may be initially necessary; hearing tests should be performed yearly to follow this condition, with further treatment imposed if the hearing worsens.

A hearing aid is an acceptable treatment for this disease, even with severe hearing loss. The advantage of this option is that it is nonsurgical. The disadvantages include the facts that the disease process can continue to grow to involve the inner ear and that the hearing aid must be continually worn for the patient to gain benefit.

The third method of treatment is surgical. The standard procedure is the stapedectomy. This procedure is done directly through the ear canal. In this operation, the eardrum is lifted up and the stapes is maneuvered to assure that it is fixed. If it is, it is removed and replaced with a small prosthesis which connects the incus to the inner ear, *reconnecting the chain.*

Overall, 91% of patients can have their hearing improved to an excellent level by operation. If hearing improvement is *not* initially noted and revision surgery is performed, an total of 96%

of patients can have their hearing restored to an excellent level. As a result of this procedure, there is no need for a hearing aid and the disease process is essentially removed from the inner ear. The risks of this procedure are significant, but few. 1% of patients develop an eardrum perforation which must be repaired. 1% lose the hearing in that ear completely. 0.3% develop partial hearing loss, dizziness, or ringing in the ear.

CONCLUSIONS

When otosclerosis develops in the middle ear, it can result in hearing loss. There is an outstanding chance for hearing improvement when surgery is performed for this disease. A viable alternative, especially in patients who will not or cannot tolerate surgery, is the use of a hearing aid.