

An Overview Of

OTOSCLEROSIS

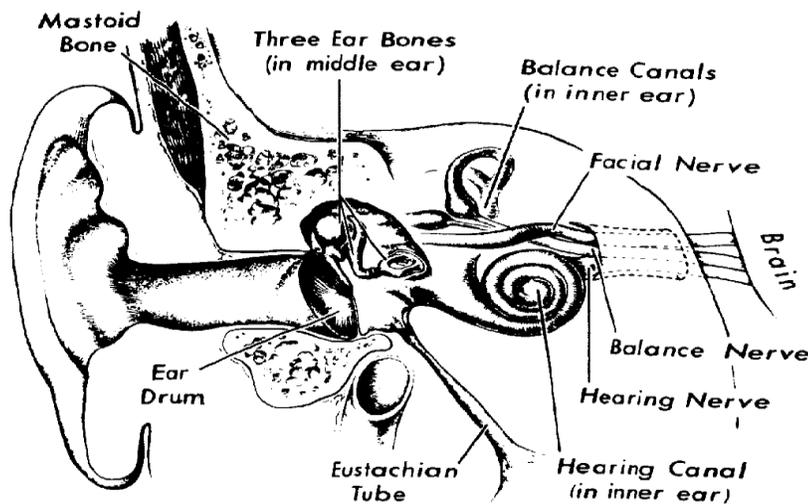
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INTRODUCTION

Otosclerosis is a disease of the inner ear bone (capsule of the inner ear and bottom portion of the stapes or stirrups bone). Otosclerosis is a common cause of hearing impairment and is, at least in part, hereditary. This means that the condition is often passed down from parent to child. Not everyone with otosclerosis has severe enough problems to make it noticeable. Common diseases such as scarlet fever, ear infection, and influenza are thought to be unrelated to the development of otosclerosis. Some evidence suggests that certain viruses may play a role in triggering this disease.

FUNCTION OF THE NORMAL EAR



The ear is divided into three parts: the external ear, the middle ear, and the inner ear. Each part performs an important function in the process of hearing.

Sound waves pass through the external ear canal to vibrate the eardrum, which separates the external from the middle ear. The three small bones in the middle ear (hammer or malleus, anvil or incus, and stirrup or stapes) transmit the sound vibrations from the eardrum to the inner ear. Vibrations in the fluid of the inner ear stimulate delicate cells that convert the vibration energy into a nerve signal. The hearing nerve then transmits impulses to the brain where they are interpreted as understandable sound.

TYPES OF HEARING IMPAIRMENT

If there is some difficulty in the external or middle ear, a **conductive** hearing impairment occurs. If the trouble lies in the inner ear, a **sensorineural** or nerve hearing loss is the result. When there is difficulty in both the middle and inner ear, a combination of conductive and sensorineural impairment exists, it is known as a **mixed** hearing loss. Conductive and mixed impairments are most common in otosclerosis, but purely sensorineural hearing loss also occurs.

HEARING IMPAIRMENT FROM OTOSCLEROSIS

In most healthy bones, old bone is constantly being removed and new bone is constantly being produced. In contrast, healthy hearing bones and inner ear bone do not normally change much over a lifetime. Microscopic examination of ears with otosclerosis shows abnormal areas of bone absorption and new bone formation. This process may involve the stapes, the inner ear, or both of these areas.

Cochlear Otosclerosis: When otosclerosis involves the hearing portion of the inner ear a sensorineural hearing impairment may result due to interference with the nerve function. This nerve impairment is called cochlear otosclerosis and once it develops it may be permanent. In selected cases medications may be prescribed in an attempt to prevent further nerve impairment. Such hearing loss may rarely progress to deafness.

Vestibular Otosclerosis: On occasion the otosclerosis may spread to the balance canals and may cause episodes of unsteadiness or spinning sensations. This may occur in the absence of any measurable hearing loss.

Stapedial Otosclerosis: Otosclerosis commonly involves the bone around the stapes or stirrup bone, the final link in the chain of middle ear bones. This stapes rests in the small groove, the oval window, in intimate contact with the inner ear fluids. Anything that interferes with its motion results in a conductive hearing impairment. This type of impairment is called stapedial otosclerosis and is usually correctable by surgery.

The amount of hearing loss due to involvement of the stapes and the degree of nerve impairment present can be determined only by audiometric examination (hearing tests).

TREATMENT OF OTOSCLEROSIS

Medical: There is no local treatment to the ear itself or any medication that has been shown to **improve** the hearing in persons with otosclerosis. In some cases fluoride may be helpful in preventing further loss of hearing. A supplement containing fluoride and calcium (Florical®, fluoride 3.75 mg & calcium 145 mg) is available through health food stores and pharmacies, online (eg, amazon.com), or through the manufacturer (Mericon Industries, Inc. Peoria, IL 61615. mericon-industries.com, 800.242.6464). Your physician may recommend taking one Florical® tablet once, twice, or three times daily with meals, often for a year, to stabilize the bone of the inner ear.

Medicines that have been used to treat osteoporosis (ie, loss of bone strength commonly seen with advancing age) have also been used to treat otosclerosis. The bisphosphonates are the most common class of drugs used for this purpose. Limited information is available to support the use of these drugs in the treatment of otosclerosis. Uncommon, but serious side effects have been reported with bisphosphonate therapy. These may be considered, particularly in patients that are rapidly losing inner ear function as a result of otosclerosis.

Hearing Aids: Most patients with early otosclerosis can be helped with the use of hearing aids. As the use of hearing aids carry little risk, you should consider the use of hearing aids if your hearing sensitivity is poor enough to cause you difficulty in daily situations. Hearing aids are the preferred course of treatment for patients that have enough inner ear hearing loss, such that surgery alone would not be capable of establishing acceptable hearing. Surgery may be necessary to allow hearing aids to work if the hearing loss is severe.

Surgical: The stapes operation (stapedectomy) is an option for many patients with otosclerosis who are candidates for surgery. This operation is performed under local (awake) or general anesthesia (asleep) and requires either no hospitalization or a short period of hospitalization and convalescence. Over 90 percent of these operations are successful in restoring the hearing (to within 10 decibels of optimal inner ear function) on a long-term basis. Surgery is usually helpful for individuals that have little inner ear hearing loss, such that hearing aids would likely not be needed with a successful operation. Surgery may also be needed for individuals that have such severe hearing loss that they cannot use hearing aids. If the inner ear is severely affected by otosclerosis, neither conventional stapes surgery nor hearing aids may be of benefit. In these rare cases, cochlear implant surgery (ie, a “bionic ear”) might be necessary.

THE STAPES OPERATION

Stapedectomy or stapedotomy is performed through the ear canal under local or general anesthesia. At times an incision may be made above or behind the ear to remove tissue for use in the operation. With the use of the operating microscope, the eardrum is turned forward. The laser may be used to vaporize parts of the stapes. The remainder of the stapes is removed with an instrument. A small opening is made in the footplate of the stapes with a laser or a drill. A stainless steel, platinum, or teflon piston (or prosthesis) is then placed into this opening and connected to the second bone of hearing, or the incus. The eardrum is returned to its normal position. Antibiotic ointment or packing is placed in the ear canal to hold the eardrum in the proper position. This will plug up the normal transmission of sound until it is removed.

The stapes prosthesis allows sound vibrations to again pass from the eardrum membrane to the inner ear fluids. The hearing improvement obtained is usually permanent.

Stapes surgery can usually be done on an outpatient basis. Occasionally, the person is admitted overnight and may return to work in seven days, but this is dependent upon occupational requirements.

One should not plan to drive a car home from the hospital. Automobile travel is permissible

immediately. Driving may be resumed as soon as rapid head movements don't cause any dizziness that could interfere compromise safety. Air travel is permissible three to four weeks following surgery.

REVISION STAPEDECTOMY

Hearing improvement achieved with stapedectomy is usually long-lasting. Hearing impairment may develop in ears treated with stapedectomy. Usually, this is a sensorineural hearing loss that results from aging, noise exposure, injuries or other insults to the ear that are not related to the stapedectomy.

Occasionally, a problem with the ear drum, incus bone, or stapes prosthesis will develop, resulting a conductive hearing loss. These problems can often be corrected with a procedure similar to that described above. In revision surgery, the stapes prosthesis may need to be tightened, replaced, or connected to the malleus bone. The ear drum may need reinforcement.

In some cases where hearing fails to improve with the stapes operation, a small hole in one of the balance canals may be responsible. Treatment of this problem may require a different surgery, such as a procedure in the mastoid bone or in the middle cranial fossa, above the ear.

Complete resolution of the conductive hearing loss is achieved in approximately 75% of patients with revision stapedectomy. Approximately 15 - 20% will have less than complete resolution of the conductive hearing loss. Two to five percent of patients may have the hearing impairment worsen. In rare cases all hearing in the operated ear may be lost.

STAPES OPERATION AFTER FENESTRATION SURGERY

The stapes operation can be performed on previously fenestrated ears, providing the hearing nerve function is essentially the same as necessary for an initial stapes operation. Seventy percent of these operations are successful in improving the hearing.

HEARING IMPROVEMENT FOLLOWING STAPES SURGERY

Hearing improvement may or may not be noticeable at surgery. If the hearing improves at the time of surgery, it usually regresses in a few hours due to swelling and packing in the ear. Improvement in hearing may be apparent within three weeks of surgery. Maximum hearing, however, is obtained in approximately 6 – 8 weeks.

The degree of hearing improvement depends on how the ear heals. In the majority of patients the ear heals perfectly and hearing improvement is as anticipated. In some the hearing improvement is only partial or temporary. In these cases the ear usually may be reoperated upon with a good chance of success. In two percent of the cases the hearing may be further impaired due to the development of scar tissue, infection, blood vessel spasm, irritation of the inner ear or a leak of inner ear fluid (fistula). In less than one percent, complications in the hearing process may be so great that there is severe loss of hearing in the operated ear, to the extent that one may not be

able to benefit from an aid in the ear. For this reason the poorer hearing ear is usually selected for surgery.

TINNITUS & STAPES SURGERY

Most patients with otosclerosis notice tinnitus (head noise) to some degree. The amount of tinnitus is not necessarily related to the degree or type of hearing impairment, as many conditions may contribute to its development. Tinnitus may develop due to impaired transmission of sound through the ear and irritation of the delicate nerve endings in the inner ear. Since the nerve carries sound, this irritation is manifested as ringing, roaring or buzzing. It is usually worse when the patient is fatigued, nervous or in a quiet environment.

Once the healing is complete following successful stapedectomy, tinnitus is often decreased in proportion to the hearing improvement, but this is far from universal.

RISKS AND COMPLICATIONS OF STAPEDECTOMY

Dizziness: This is normal for a few hours following stapedectomy and may result in nausea and vomiting. Some unsteadiness is common during the first few postoperative days; dizziness on sudden head motion may persist for several weeks. On rare occasions dizziness is prolonged.

Taste Disturbances and Mouth Dryness: Taste disturbances and mouth dryness are not uncommon for a few weeks following surgery. In five percent of the patients this disturbance may be prolonged.

Loss of Hearing: Further hearing loss develops in two percent of the patients due to some complications in the hearing process. In one percent this hearing loss is severe and may prevent the use of an aid in the operated ear.

Tinnitus: Should the hearing be worse following stapedectomy, tinnitus (head noise) likewise may be more pronounced.

Eardrum Perforation: A perforation (hole) in the eardrum membrane is an unusual complication. It develops in less than one percent and usually is due to an infection. Fortunately, should this complication occur, the membrane may heal spontaneously. If healing does not occur, surgical repair (myringoplasty) may be required.

Weakness of the Face: A very rare complication of stapedectomy is temporary weakness of the face. This may occur as the result of an abnormality or swelling of the facial nerve.

Infection: This is very uncommon following stapes surgery. This could affect hearing and balance and could require additional treatment.

Spinal Fluid Leak: The inner ear communicates with the spinal fluid in very young children. If this communication fails to seal normally or if there is an abnormality in the inner ear, spinal

fluid leakage may be encountered during the stapes operation. This is very rare, but may require additional treatment if encountered.

WHAT TO EXPECT FOLLOWING SURGERY

There are some symptoms that may follow any stapes operation. Please call the ENT clinic if you experience:

- clear, watery drainage from your incision or nose;
- redness, swelling or infected drainage from your ear;
- pain, cramping or swelling in the legs;
- fevers (>101⁰F);
- *severe headache or neck stiffness.*

PAIN: Some degree of pain and tenderness is expected after surgery. This usually originates in spasm of the jaw and neck muscles that are moved during the surgery. Pain should be controlled with oral acetaminophen or an anti-inflammatory medication (eg, ibuprofen), alone or combined with a prescribed narcotic medication. Significant pain should decrease over the first few days following surgery. If the pain initially improves and worsens on subsequent days, you should contact your surgeon's office. Worsening pain may be a sign of an ear infection.

DRAINAGE FROM THE EAR CANAL: This is expected for a few weeks after surgery. Cotton balls are placed in the bowl of the outer ear to catch the drainage. If the drainage saturates more than a dozen cotton balls per day or becomes foul or pus-like, please contact your surgeon's office.

TASTE DISTURBANCES AND MOUTH DRYNESS: Taste disturbances and mouth dryness are not uncommon for a few weeks following surgery. In some patients this disturbance is prolonged.

TINNITUS: Tinnitus (head noise), frequently present before surgery, is almost always present temporarily after surgery. It may persist for one to two months and then decrease in proportion to the hearing improvement. Should the hearing be unimproved or worse, the tinnitus may persist or be worse.

NUMBNESS OF EAR: Temporary loss of skin sensation in and about the ear is common following surgery. This numbness may last for six months or more.

CARE INSTRUCTIONS FOLLOWING SURGERY

MEDICATIONS: Your surgeon will commonly prescribe a medicine to relieve pain for the first few days after surgery. Antibiotics to be taken by mouth are not routinely used, but may be indicated in certain circumstances.

EAR DRESSING: A cotton ball is kept in the bowl of the outer ear to catch drainage from the ear canal. This should be changed with fresh cotton as it becomes saturated. Under the cotton

ball, foam packing, a blood clot, or ointment may be seen. This should not be disturbed. It is normal for some of this packing to fall out with cotton ball changes. This packing will be removed by your surgeon during your postoperative visits.

BATHING & SWIMMING: Washing the hair may be resumed 2 or 3 days following surgery. The incision behind the ear, if any, may get wet at this time. Avoid scrubbing the incision. If a small area of the incision behind the ear opens or bleeds after bathing, simply clean it daily with hydrogen peroxide on a cotton ball. It is important to avoid contamination of the ear canal with water until the canal is healed (usually < 4 weeks). As earplugs are far from perfect, submersing the head, such as with swimming, is not recommended until the ear canal is healed.

NOSE BLOWING & SNEEZING: Raising pressure in the back of the nose by nose blowing or sneezing with the nostrils obstructed and mouth closed can lead to build-up of pressure in the middle ear and displacement of the eardrum and prosthesis. Avoid nose blowing and sneeze with your mouth open until cleared by your surgeon.

ACTIVITIES: Most daily activities will not affect your outcome with surgery. Activities that lead to jarring of the head (eg, boxing or jumping on a trampoline) should be avoided for 4 – 6 weeks after surgery.

TRAVEL: You should have someone drive you from the hospital. Driving is permissible the day after surgery if there is no difficulty with head movements (eg, dizziness). Air travel is permissible 48 hours after surgery but should be avoided, if possible, for four weeks after surgery.

POSTOPERATIVE CLINIC VISITS: The first visit to your surgeon will generally be between 1 and 2 weeks after surgery. Keeping these scheduled visits is important to ensure optimal healing and prompt treatment of any problems. Subsequent visits are commonly scheduled at 4 – 8 week intervals until healing is complete.

GENERAL COMMENTS

Whether you chose treatment with surgery or hearing aids, it is advisable to have careful hearing tests repeated at least once a year. Other, more serious causes of hearing loss may co-exist with otosclerosis. Only with ongoing monitoring can such problems be identified.

Should any questions arise regarding your hearing impairment, feel free to call or write us at any time.