

EAR AND BALANCE CLINIC

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OTOSCLEROSIS

The ear is divided into three parts: the external ear, which includes the visible part of the ear (auricle and the ear canal); the middle ear, which is an air-filled chamber behind the eardrum containing three bones of hearing; and the inner ear which contains the sensory organs of hearing (cochlea) and balance (semicircular canals and otolith organs).

WHAT IS OTOSCLEROSIS?

Otosclerosis is a disease of the bone which surrounds the inner ear. In the early stages of the disease, known as otospongiosis, areas of softened bone appear. These soft patches contain enzymes which are capable of dissolving bone. With time, the bone destruction stops and the otospongiotic lesions undergo calcification. This results in an inactive scar which is known as otosclerosis.

Otosclerosis has a notable tendency to run in families and occurs more often in certain ethnic groups than in others. For example, otosclerosis is much more common in northern Europeans than it is in blacks and Asians. Although much is known about the effect of otosclerosis on the ear, the exact cause of the disease remains unknown.

SYMPTOMS OF OTOSCLEROSIS:

1. Hearing Loss: The hearing loss of otosclerosis usually begins in the second, third or fourth decades of life and worsens slowly over many years. Although gradual progression is the rule, periods of accelerated hearing loss may occur during pregnancy or with certain types of hormonal treatment. The disease usually affects both ears, although often unequally. The eventual outcome of untreated otosclerosis is variable, with most people's hearing stabilizing at the moderate to severe loss level. Total deafness may occur, but is quite uncommon. Two kinds of hearing loss occur with otosclerosis: conductive and sensorineural. Conductive loss results from the mechanical effects of otosclerosis on the stapes bone and is potentially correctable. Sensorineural loss results from the effects of otosclerosis on the inner ear and is irreversible.

- A. Conductive Hearing Loss: In the normal ear, sound vibrations entering the ear canal are amplified by the eardrum and pass through a chain of three bones to the inner ear. The third of these bones, the stapes, acts like a piston, pumping vibrations into the fluid-filled inner ear. When the stapes becomes stiff or immobile due to the effects of otosclerosis, it resists the passage of sound vibrations and a hearing loss results. This mechanical type of hearing loss is surgically correctable in the majority of cases.

- B. Sensorineural Hearing Loss ("Nerve Loss"): When active otosclerotic lesions form next to the hearing organ (cochlea), a slowly progressive form of hearing loss occurs. This results from the toxic effect the otosclerosis has on the sound sensing cells (hair cells). Once the damage is done, a permanent hearing loss occurs. Medical treatment is designed to prevent further damage to the inner ear. Surgery will not correct sensorineural hearing loss.
2. Tinnitus: The presence of abnormal ear and head noises is a condition known medically as tinnitus. Sometimes the sounds heard are described as a ringing or buzzing sound, although many other sensations are possible. Otosclerosis may cause tinnitus by damaging the inner ear's sound sending cells or the hearing nerve. This results in nerve impulses being generated, even though no sound of this frequency is coming into the ear. The tinnitus of otosclerosis often improves after surgery, but may persist in some cases. If it does persist after surgery, or if your ears are not suitable for surgical therapy, there are medical treatments which can help to relieve the problem.
3. Dizziness and Vertigo: The lesions of otosclerosis often surround the balance canals of the inner ear. Fortunately, this seldom results in dizziness or vertigo. On rare occasions otosclerosis may cause such symptoms and numerous medical treatments to alleviate them are available.

TESTS FOR OTOSCLEROSIS:

1. Audiologic Studies: Hearing tests are essential in the diagnosis of otosclerosis and in monitoring the results of therapy.
2. Radiographic Studies: Although not often required, X-ray studies may help to confirm the presence of inner ear otosclerosis. In order to visualize the minute structures of the inner ear, special computer enhanced studies are necessary (CT scans).

SURGICAL TREATMENT OF OTOSCLEROSIS:

Conductive hearing loss caused by mechanical fixation of the stapes can be reversed through surgery in the majority of cases. Surgery on the stapes, which is slightly larger than the head of a pin, requires the use of an operating microscope and sophisticated surgical techniques. Since first developed in the late 1950s, stapes surgery has become widely practiced around the world in the treatment of otosclerosis. Early procedures involved attempts at mobilizing the stapes by fracturing the binding otosclerotic bone. However, the otosclerosis tended to return and the hearing loss recurred within a few years.

Contemporary surgical techniques (see drawings) involve removal of all or part of the stapes. A replacement prosthesis is attached to the second ear bone, the incus. One of two slightly different techniques is employed, depending on the findings encountered during surgery. Total stapedectomy involves removal of the entire stapes and placement of a wire over a tissue graft which seals the inner ear. This tissue graft is usually taken from the cartilage in front of the outer ear canal. In the second technique, called the small fenestra stapedotomy, the upper part of the stapes is removed, but the bottom portion, the footplate, is left in place.

A small hole is created in the footplate using either a diamond drill or laser. A tiny piston (0.6mm) is then placed through this opening into the inner ear.

A person undergoing stapes surgery is admitted to the hospital on the morning of surgery. The surgery is performed under local anesthesia with sedative medications. The anesthetic is injected into the ear skin. The surgical approach is through the ear canal which generally leaves no visible scars and takes from one to two hours. The patient may be discharged later the same day or may stay one night in the hospital and is able to return to work within a few days to one week. Complete healing of the ear takes between one and three months. Precautions advised during this period include keeping the ear dry for one month (earplugs advised for showers or swimming), avoiding vigorous nose blowing, and refraining from lifting heavy objects for six weeks.

MEDICAL TREATMENT OF OTOSCLEROSIS:

Unfortunately, no medicines are yet available which are able to "loosen" a stapes bones frozen by otosclerosis. The treatment of this mechanical problem is either through surgical repair or the use of a hearing aid. No treatment is available that can repair existing inner ear damage or restore sensorineural hearing loss. Some scientific evidence suggests that the mineral, fluoride, may retard or arrest the progression of inner ear otosclerosis. To be effective, fluorides must be taken daily over a prolonged period of time. The presumed action of fluoride is to convert the active bone-destroying otospongiotic lesions to inactive otosclerotic ones. Side effects of fluoride therapy are minimal. Occasional stomach irritation may be avoided by taking the medicine with meals rather than on an empty stomach. Since fluorides are readily incorporated into growing teeth and bones, we are hesitant to prescribe them to pregnant women or children.

RISKS AND COMPLICATIONS OF STAPES SURGERY:

1. Loss of Hearing: Hearing is improved following stapes surgery approximately 90% of the times. Hearing remains unchanged in about 7% and is actually worse than before surgery in from 1% to 3%. These rare cases of hearing loss are usually due to some complication in the healing process. The loss may be severe enough to prevent the use of a hearing aid in the operated ear. In some cases, additional surgery may be necessary.
2. Dizziness: Since the inner ear has a role in both hearing and balance, it is common that dizziness occurs particularly during the first 12 hours after stapes surgery. Some unsteadiness is common during the first few post-operative days and some dizziness with rapid head motion may occasionally persist for several weeks. On rare occasions dizziness may be prolonged and in very rare instances may require additional surgery.
3. Tinnitus: After healing from stapes surgery, tinnitus is usually unchanged or lessened. On occasion it may be more pronounced, particularly during the first few weeks or if the hearing has not been improved.
4. Taste Disturbance: A temporary taste disturbance is not uncommon following stapes surgery. This is due to swelling of the nerve which runs through the middle ear near the stapes and supplies taste sensation to the side of the tongue. A metallic taste on one side of the tongue may last several weeks. Prolonged changes in taste sensation are rare.
5. Eardrum Perforation: A hole in the eardrum develops in less than 1% of cases. If spontaneous healing does not occur, surgical repair may be required.
6. Weakness of the Face: A very rare complication of stapedectomy is temporary weakness of the face due to swelling of the facial nerve.

