

Clinical Otosclerosis

Management in Pilots

C. M. Kos, M.D.

OTOSCLEROSIS as a relatively common cause of hearing impairment in young and middle-aged adults has in recent years become more widely recognized. It is a peculiar osteochondrodystrophy which has a predilection for the circumferences of the oval window and footplate of the stapes. It may also invade other areas of the otic capsule and the internal auditory meatus (Fig. 1).

If the involvement is limited to the oval window and stapes, it produces a mechanical type of hearing impairment which is recognized as such by pure tone air and bone conduction audiometry (Fig. 2). Otherwise, examination reveals no obvious evidence of etiology. Hence, the diagnosis is a presumptive one until microsurgical or histologic evidence is found to confirm it. However, the clinical diagnosis is not difficult for one who has reliable audiologic facilities and is familiar with the insidious progressive nature of otosclerosis and its effect on the hearing mechanism.

Tinnitus which is a subjective sensation usually accompanies and is an expression of hearing loss. Tinnitus resulting from mechanical hearing loss is frequently described as the sensation of covering the ear with a sea shell, or in terms that suggest it to be similar to a surging or pulsating white

noise. Tinnitus resulting from cochlear or nerve degeneration is usually described as a whistling or hissing sound either continuous or intermittent, which may interfere with radio signal reception. Since otosclerosis of the stapes may be accompanied by neural degeneration a mixture of tinnitus characteristics may be present. A mechanical defect of hearing, such as otosclerosis causes, may be corrected. With the consequent improvement in hearing the tinnitus usually is reduced or eliminated. Most cases of neural loss are irreversible and irremediable; consequently the associated tinnitus fails to respond favorably to therapy.

The annoyance of tinnitus varies somewhat with the emotional stability, degree of physical and mental fatigue, or the general tolerance threshold of the individual. Otherwise, tinnitus must be considered a natural symptom of hearing loss which does not respond satisfactorily to separate therapy. The only effective and reliable treatment for tinnitus is improvement of the hearing which is made possible, in some cases, by surgical correction or by the wearing of a hearing aid.

The higher incidence of otosclerosis in the middle span of life makes it a health problem of some concern to those who are responsible for the care of flying personnel. It is distributed between the sexes in the ratio of three men to four women.³ Otosclerosis usually begins to impair hearing in the late teens or early twenties and reaches

From the Iowa Clinic of Otology, 309 Iowa Avenue, Iowa City, Iowa.

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its maximum mechanical effect within ten to twenty years after the onset. This usually covers that very important period of pilot or air crew training

It is distance and the consequent decrease of loudness that place a handicap on the subject of otosclerotic hearing loss. As the hearing loss increases



Fig. 1. Arrows indicate point of common sites of predilection for otosclerosis.

and of acquiring career experience, during which a considerable investment is made in the afflicted individual before the hearing loss becomes obvious or possibly before it is revealed by examination.

Generally it may be regarded as a conditional hearing loss, that is, hearing impaired by otosclerosis seems to be better in some situations and worse in others. Hearing is usually adequate in a noisy environment such as in operating aircraft simply because conversation is necessarily forced to a high intensity by the participating individuals. Hearing is usually efficient when using a telephone, and in close-range conversation situations even in a relatively quiet environment because the spoken voice is often uttered at intensities which easily penetrate the elevated threshold barrier of this type of impairment.

to levels exceeding 30 to 35 decibels, the subject becomes less and less capable of concealing his impairment while his handicap becomes more and more obvious to those around him.

This hearing loss is of the chronic progressive type which produces varying degrees of communication interference depending upon environmental and occupational circumstances, hence it may escape the detection of the casual observer or examiner. It is for this reason among several others that accurate hearing testing should be included in the general examination procedures. This proposal has met with some opposition because it requires additional facilities, equipment, time, and personnel to conduct the necessary examinations. These apparent burdens are easily cancelled by the estimated cost of \$300,000 or more to train a person for a pilot's career.⁵

The incidence of hearing loss of all types is not of the minor proportions that some uninformed persons seem to think. In our general population it is

ant periodic care which is a disqualifying physical factor for military pilot duty. This effective procedure was eventually threatened with abandon-

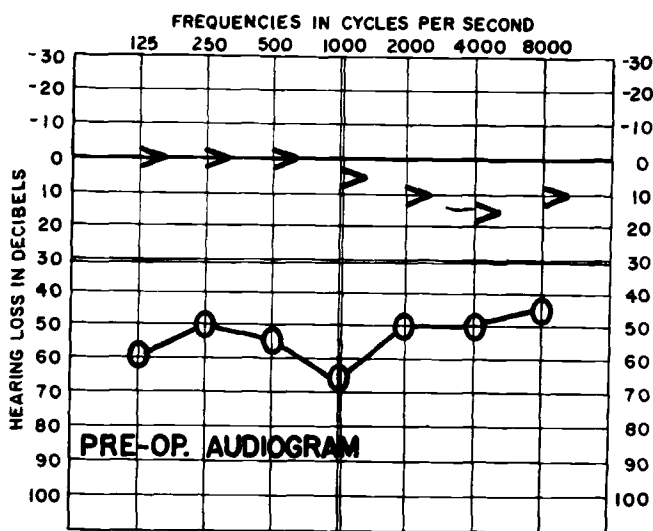


Fig. 2. Air (O-) and Bone (>) conduction thresholds indicate a mechanical type of hearing loss.

estimated to exceed the incidence of blindness, muscular dystrophy, epilepsy, poliomyelitis, multiple sclerosis, tuberculosis, and cerebral palsy combined.¹⁰ Because of its apparently evasive nature, particularly in its early stages, hearing loss due to otosclerosis is probably much more prevalent than currently available statistics indicate.

In contrast to most of the disabilities mentioned above, the mechanical hearing impairment of otosclerosis now can be fully corrected with a high degree of success. This has come about as the result of modern microsurgical techniques which began with the development of the Fenestration operation by Lempert some twenty-odd years ago.⁶ The results were good but missed by some 20 to 30 decibels of restoring normal hearing. It also failed to eliminate the mastoid cavity and its attend-

ment following the re-introduction of Stapes Mobilization by Rosen approximately six years ago.⁷ Like the Fenestration operation, experience with the Stapes Mobilization technique has revealed its limitations and disadvantages. While initial successes with the Mobilization operation seemed to be satisfactory, frequent regressions often within three to twelve months continued to undermine the permanency of hearing improvement.²

The natural sequence to these results was the introduction of stapes substitution procedures such as those recently described by Schuknecht, Shea, and Kos.^{4,8,9} These procedures require the removal of the entire otosclerotic stapes, which is then replaced with an elastic tissue connected to the incus by an artificial strut using polyethylene tube, steel or tantalum wire.

Such a technique has been developed following experience with over 1,000 Stapes Mobilization operations. It consists of replacing the stapes footplate

vein-plug stapedioplasties performed five to ten months previously, ninety produced the maximum hearing improvement permitted by the functional



Fig. 3. Arrow points to vein plug prosthesis in oval window connected to incus with steel wire.

with a section of vein approximately 2 x 4 mm. to which is tied a 4½ mm. length of stainless steel suture wire to substitute for the crura and head of the stapes. The resulting vein plug is inserted into the oval window with the distal end of the wire looped over the long process of the incus and crimped firmly to reconstruct the continuity of the ossicular chain between the vestibule and tympanic membrane (Fig. 3).

Preliminary studies indicate that this technique provides for unprecedented success and should be permanent in most cases because otosclerotic bone involving the stapes footplate is removed and replaced with a tissue which remains suitably elastic. Hearing is consistently restored to the maximum levels permitted by the function of the hearing nerve. In a recent survey of the results of 100 consecutive

level of the hearing nerve. Fifty-two operations achieved normal hearing (0 to 10 db. hearing level) out of fifty-four which had been predicted on the basis of the preoperative bone conduction thresholds. Among the ten which did not produce maximum hearing improvement, six resulted in a hearing level of better than 30 decibels.⁴ Hence, rehabilitation in most instances is complete and secure as long as the hearing nerve continues to function serviceably (Fig. 4).

The operation is performed through a small peritympanic incision which heals without evidence of surgical intervention in three to six weeks. Therefore, it is easily possible for the candidate who has had hearing restored by this method to undergo the most exacting examination without detection unless history and x-ray of the

middle ear reveal the presence of the steel wire, and a small scar on the back of the hand is found to identify the source of the vein plug.

cursions which the ossicular chain might be subjected to in the event of decompression is probably no more than that forced upon the incus dur-

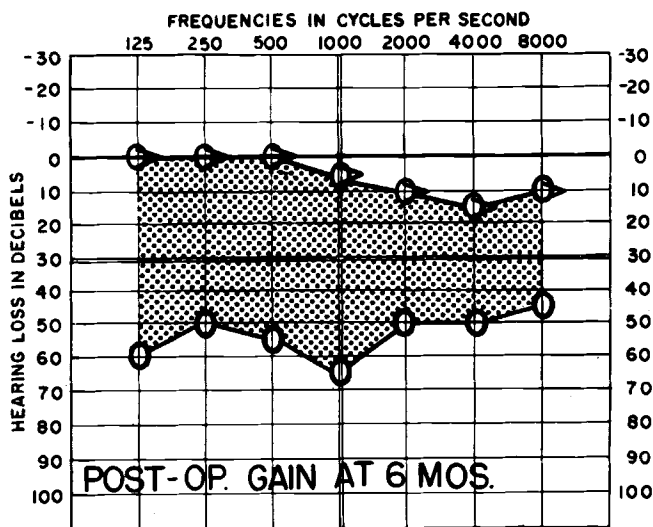


Fig. 4. Same case as noted in Figure 2, six months later. Shaded area represents the pure tone hearing gain resulting from vein-plug stapedioplasty.

Turn now to a consideration of the possible effects flying may exert on such a rehabilitated ear. The operation does not alter eustachian tube function and there is no reason for expecting that this segment of the middle ear should not perform just as efficiently after surgery as prior to it. Tolerance to barometric pressure changes by the tympanic membrane and ossicular chain should remain as effective as before surgery. The tympanic membrane may be expected to rupture in rapid decompression no more or less frequently than in the normal ear. The oval window being protected by a thick membrane is not likely to be injured by excessive middle ear negative pressure more frequently than an oval window containing a normal stapes.

Actually the magnitude of the ex-

ing the process of attaching the steel wire. Also, the subsequent palpations that are exercised to determine the firmness of the connection and mobility of the prosthesis in the oval window, certainly are far in excess of the flexibility demanded of the ossicular chain during the usual exposures to barometric pressure alterations.

Barotitic trauma causing middle ear effusion should not permanently impair the hearing any more than effusion in the normal ear. In fact, three patients in a series of over 100 whose ossicular chains were reconstructed with the vein-plug prosthesis sustained purulent otitis media within three weeks of surgery without evidence of complication or subsequent hearing impairment. This indicates that the vein plug is adequately protective to the

inner ear and apparently resists inflammation as effectively as the normal stapes.

Because the function of the stapedius muscle is eliminated by this procedure the question of resistance or susceptibility to excessive noise trauma arises. On this subject no direct information is available. However, experiments in cats designed to study the effect of noise in ears in which the stapedius muscle has been severed reveals a significant hearing impairment in the affected ears after exposure to loud noise as compared to the contralateral normal ear.¹ Suitable experiments should be set up to determine whether the substitution of a prosthesis (vein plug) for the stapes subjects the ear to any greater damage than for normal ears.

Whether such information is applicable to the effects of substitution prosthesis for the otosclerotic stapes in human ears is entirely speculative because of the wide difference in the ear conditions being compared. This may be an area of research with which aviation-minded investigators may wish to be concerned. Until the advent of stapes replacement techniques the effect of traumatizing noises was not considered seriously because the Fenestration and Mobilization operations did not require severance of the stapes tendon. Aside from this speculative consideration, the currently superior results of vein plug stapedioplasty threaten to supersede both the Fenestration and the Mobilization techniques. However, because of the relatively short period of experience (one year) with this new approach to the problem, it is recommended that it be employed cautiously until such time as full and complete comparisons can be

made. For the present, comparing all the credits and liabilities of the vein-plug procedure with those of the Fenestration and Mobilization operations at equal periods of experience with them, the vein-plug technique exceeds the successful results of the others in every respect. It should prove to be the most effective surgical treatment for everyone whose hearing loss is due to otosclerosis of the stapes.

The alternatives to surgery are: (1) The use of a hearing aid and possible occupational readjustment. (2) Discharge from occupation for disability. Neither permits the complete rehabilitation desired nor does either carry the risk, however remote, of inflicting surgical damage to the ear.

The first alternative (hearing aid) provides for only limited and conditional rehabilitation which is no less expensive. The second alternative (discharge for disability) may be expedient from the administrative viewpoint, but it most certainly deprives the industry of the proceeds of a costly investment in training and experience, possibly of special talents difficult to replace at any cost.

These are some of the medical and surgical aspects of hearing loss due to otosclerosis which invite further consideration by the aviation medical services. Additional experience and pertinent research should help to establish the appropriate policies for the management of otosclerotic hearing impairment, as distinguished from other types of hearing loss, in flyers.

SUMMARY

Otosclerosis is a common cause of hearing impairment in young adults. Its insidiously progressive nature eludes detection during its early stages

unless hearing is periodically monitored by audiometry.

The mechanical portion of the hearing impairment may be corrected in most cases by microsurgical techniques.

Whether rehabilitation by such methods alters or hinders the individual's ability to meet all requirements for flying duty is unknown.

In view of the recent success of vein-plug stapedioplasty, it is anticipated that it will supersede previously used techniques for the correction of hearing impairment due to otosclerosis.

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ORNL Scientists Fail to Find Radiation Threshold

W. L. Russell and Elizabeth M. Kelly, Oak Ridge National Laboratory, have reported to the National Academy of Sciences that their attempts to find a threshold for radiation-induced mutations have been unsuccessful. The two lowered their dose rates to mice from 90 roentgens per week to 10 r/wk, then compared the mutations at the lower rate with nonirradiated animals. Mutations at 10 r/kw, they said, were "significantly higher" than with control mice and comparable with the number obtained at 90 r/wk. "Thus," they concluded, "there is no indication of a threshold dose rate (a dose rate below which no mutation occurs) even when the dose rate is reduced to this low level of 10 r/wk. This is the lowest dose rate ever tried in laboratory mutation-rate experiments with animals."

Meanwhile, the National Academy of Sciences-National Research Council issued an updating of its 1956 report on radiation. Its major conclusion: "In general, the potential hazards associated with radiation sources are being recognized by an increasing number of those responsible for their operation. . . . There are still many unknowns, and research on a wide front is urgently needed. As new information is gained, man can expect to derive increasing benefits from the release of nuclear energy with a minimum hazard to himself and his descendants."—From *"Nucleonics,"* June, 1960.