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OTOSCLEROSIS: DEAFNESS AMENDABLE TO SURGERY

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TABLE OF CONTENTS

	Page
I. Introduction	1
II. Classification of Deafness	3
III. Pathology	5-11
Phases of Development Histopathologically	5
General Histopathologic Picture	7
Effects of Stapes Involvement	9
IV. Etiology	12-16
V. Diagnosis	17-29
Incidence	17
Diagnostic Tests	20
Signs and Symptoms	22
Sensorineural loss	25
Effects of Pregnancy	26
VI. Surgery	30-39
Early development	30
Fenestration	32
Stapes Surgery	36
Complications	38
VII. Summary and Conclusions	40-42
VIII. Bibliography	43-46

INTRODUCTION

Throughout time many people have been forced into a world of silence by a disease known as otosclerosis. Deafness generally as a handicap is not appreciated by the "normal" population to the same extent that other handicaps are. The main reason for this fact is that in deafness there are no outward signs that show that this individual is "different", while with other handicaps such as loss of a limb or blindness there are outward manifestations which call the attention of the onlooker to the handicap. The world of silence is a very lonely world as two notable men in history have stated. Beethoven and Swift are these men referred to; one known in music and the latter in English literature. These men were creators, and both of their works changed following the onset of their deafness. Swift's work, for example, seemed to show a sense of bitterness toward the world. This type of reaction is not unusual as hearing loss has been known to produce profound psychologic changes in an individual who is faced with the situation of losing hearing. People who are losing hearing have a tendency to withdraw from activities which normally interested them; they make various excuses for their withdrawal but the basic reason is that they are embarrassed to continue to ask "what did you say?". In situations in which there are large numbers of people they have a tendency to miss much of that which is going on around them. Otosclerosis is a major cause of deafness and is manifest by a progressive hearing loss which has its onset in relative early life i.e. in the early twenties. Before the last 25 years the person with otosclerosis was faced with a very dim prospect of progressive hearing loss for which there was no hope. In these last 25 years there have been great surgical advances in the treatment of the disease. In the last decade otologic surgery has become a common thing and the correction of otosclerosis by operation has become a reality. Now the person with otosclerosis no longer faces the inevitable world of silence but now there is hope that his hearing can be partially or entirely restored via operation. It might be pointed out that operations for otosclerosis are not universally successful, however, there is a positive result in approx-

imately 85 per cent of the cases.

This paper will consider the entire subject rather briefly. Otosclerosis will be considered from four major viewpoints. These are pathology, etiology, diagnosis, and surgery or treatment. A classification of deafness will also be provided in order to make the reader more familiar with other causes of deafness, more specifically conduction loss.

CLASSIFICATION OF DEAFNESS

Whenever one considers the problem of deafness the various types of deafness should be kept in mind. This paper dealing with otosclerosis represents only one of many causes of hearing loss. The hearing loss attributable to otosclerosis represents approximately 10-15 per cent of hearing loss due to all causes. It is for this reason that a classification of deafness should be borne in mind by the reader. This classification was taken primarily from the textbook Fundamentals of Otolaryngology by Laurence Boies⁶.

- I. Conduction- impairment of sound passage to the cochlea
 - A. Obstruction of the external canal
 - 1. Impacted cerumen
 - 2. Otomycosis
 - 3. Swelling of the canal walls
 - 4. Foreign bodies
 - 5. Atresia - acquired or congenital
 - B. Abnormalities of the tympanic membrane - usually associated with abnormalities of the of the middle ear also
 - 1. Marked retraction and thickening
 - 2. Excessive scarring
 - 3. Perforation
 - C. Pathologic changes in the middle ear
 - 1. Congenital
 - 2. Secondary to infection or other pathologic process infection being the most common (suppurative or adhesive otitis media)
 - D. Occlusion of the Eustachian tubes
 - E. Stapes fixation - OTOSCLEROSIS
- II. Sensory-neural loss
 - A. Presbycusis or nerve degeneration which accompanies aging
 - B. Trauma
 - 1. Noise exposure

2. Explosive blasts - usually associated with rupture of the tympanic membrane along with nerve damage
 3. Blows to the head and ears
- C. Toxins
1. Infective - mumps, neuritis, syphilis, scarlet fever, and measles,
 2. Drugs - most notably quinine, streptomycin, dihydrostreptomycin, neomycin, kanomycin, and heavy metals
- D. Vascular lesions - cerebral vascular accidents of all types and in locations associated with hearing function
- E. Tumors- for example, neuronoma of the VIII nerve, cerebellar pontine angle tumors, and others
- III. Simulated hearing loss
- A. Psychic
1. Psychogenic overlay on organic disease
 2. Situational - exposure of circumstances which ppt. loss
 3. Malingering
- IV. Congenital - usually sensory-neural in nature and may be associated with congenital malformations of the cochlea

By the above classification one sees that otosclerosis represents only a part of the total picture of deafness; but it is a very important cause because there is a possibility of correction by operative techniques.

PATHOLOGY

Otosclerosis is a primary disease of the labyrinthine capsule rather than the middle ear. It must be kept in mind that these two areas, the capsule and the middle ear, develop embryologically independent of each other. The labyrinthine capsule develops primarily from ectoderm while the middle ear draws its origin from the first and second branchial arches. Histologically the labyrinthine capsule is composed of three distinct layers of bone, an inner layer of endosteal bone, an outer layer of periosteal bone, and a layer of endochondral bone. It is this middle layer of endochondral bone which is involved in otosclerosis. Also there may be found in this layer of bone small remnants of cartilage which might be classified as primitive in nature.

The area of the oval window is the site of predilection for the otosclerotic process which seems to be a very localized condition, that is, the entire bone may not be involved, and the pathologic bone may appear in a very localized area. Also in the area of the oval window there are two anatomic structures which are termed the fissula ante fenestram and the fissula post fenestram. These seem to form a fibrous connection between the periotic connective tissue and the middle ear tissues⁵⁰. Of these two areas it is the fissula ante fenestram which has attracted the most interest for two basic reasons, first, it is in this area where otosclerotic foci are most often seen, and in this area cartilagenous rests are found most frequently. The fissula post fenestram may vary considerably in its form, size, and consistency; while the fissula ante fenestram seems to be a more stable anatomic structure. For the reasons mentioned above, it was felt by many that the area of the fissula ante fenestram was the site of origin of the otosclerotic process. Guild^{22,23} in a large series of temporal bones, that is 1161 selected at random, showed that the area of the fissula ante fenestram was not involved in cases in which the entire area of the temporal bone anterior to the oval window was not involved. Thus, he feels that the fissula ante fenestram is not the primary site of the disease process.

The process of development of otosclerosis has been described by Altmann⁵⁰ as occurring in four developmental stages. First, there is osteoclastic destruction of the old capsular bone, and the formation of new, pathologic bone. This bone is in the form of marrow deposition in the spaces from which old bone has been resorbed. This marrow is very rich in cells and poor in fibrils. In this early phase the process is in the area of the bascular channels. Next is the stage of replacement of the absorbed bone by newly formed pathologic bone which is rich in cementum, poor in fibrils, and stains blue in Hematoxylin-Eosin staining. Also there are irregular shaped and distributed osteocytes which tends to give the area the appearance of being similar to cartilage i. e. chondroid bone. The appearance at this point is sieve-like. The third stage is the replacement of the earlier formed bone by more mature type. There is an increase in the amount of fibrils within the marrow and repeated destruction and replacement of the first formed bone until the entire area is richer in fibrils and poorer in cementum and will stain red in Hematoxylin-Eosin stains. Throughout these last two stages the bone being formed has a web-like appearance. The last stage is the deposition around the blood vessels of areas of true lamellar bone (red staining). Depending upon the amount of blood vessel proliferation it may show a vascular or fibrous character. Thus, the final picture of these events is one of pathologic bone which may have a very irregular appearance.

Lederer³⁵ reports that there are three basic phases of development of otosclerosis which differ somewhat from Altmann's. First is the stage of absorption, in which there is dilatation of capillaries in Haversian canals with disappearance of the normal walls. Osteoblasts accumulate between vessel walls and where the bony were. Next is the stage of otospongiosis characterized by increased bone absorption. There is a decrease in the bony framework and enlarged marrow spaces and an increase in the capillary size. The bone at this point stains blue with H and E. The last phase is the stage of otosclerosis or rebuilding. The vessels decrease in size and cells collect around them. The stain here is also basically blue but the edge is red and represents the area of new bone. The

Marrow spaces become smaller and now assume the function of Haversian canals.

The overall pathologic picture varies from one which resembles a neoplasm to one which resembles a callus. Generally it is characterized by dark blue staining material which seems to be intracellular and is found most frequently immediately adjacent to the blood vessels. The picture is one of immature, web-like bone; however, there are many phases of development within the single lesion by virtue of what has been described in the foregoing. The entire area may show an element of swelling.

Blue mantels deserve some mention at this point as they have frequently been reported by investigators of otosclerosis. (Manasse seems to be the first to describe them). These are blue staining finger-like projections which are apparently extensions of the process just described. Initially the process begins in areas immediately surrounding blood vessels, but after the process has begun it extends into the bone without regard for blood vessels. Blue mantels seem to represent otosclerotic bone in the resorption spaces which have developed as an extension of the original process. Weber⁵⁶ points out the fact that there are very thin membranes which cover the inner surface of the lacunae and canaliculi of bones as well as the vascular channels. These membranes stain dark blue with H and E stains, the same as blue mantels but merely represent part of the normal structure of bone. Lempert and Wolff⁵⁷ have said, "Blue mantels are the walls of Haversian canals which take an intensely blue dye with hematoxylin. Blue mantles may occur in bone that is not otosclerotic but they are always conspicuous in bone that is from patients with otosclerosis." Many investigators have felt that blue mantels represent the preliminary stages of otosclerosis, however due to the confusion which has developed around the so called "normally appearing blue mantels" the question has yet to be resolved. Due to the frequency and consistency with which they are found in association with otosclerosis there is a definite relationship with the early progression of the lesions. Wolff⁵⁸ feels that they represent the type of calcium being deposited. In the case of oto-

sclerosis she feels that it is due to the formation of new bone and thus, there is blue mantle formations as this is the calcium which is most susceptible to the staining characteristics of the blue mantels.

Lembert and Wolff³⁷ described somewhat different events in the progression of the otosclerotic lesions than have been previously described in this section. They feel that the process is basically vascular in origin, and in the vascular system they have described some changes which they have noted from 100 cases of otosclerosis. In 23 per cent of the cases they found clear globules other than marrow spaces within the vascular channels. In 27 per cent of the cases they found the accumulation and fragmentation of red blood cells. In 21 per cent of the cases they found sludging of the blood and 3 of these cases were ones in which globules were found. In 40 per cent of their cases they found the accumulation of polymorphonuclear leucocytes in the diseased area. The blood vessels seemed to show the pattern of first constriction and then tortuosity, followed by periarteritis and adventitis. They point up these changes basically to emphasize their theory of vascular origin to otosclerosis. The process which results from the above mentioned facts results in seven steps in the pathogenesis of otosclerosis. First there is blue mantle formation followed by hypervascularity in the affected area. Next there is decalcification without osteoclasts, that is, osteoclasts are not demonstrable in the numbers which would be expected with the resorption of bone as is seen in this disease. Osteofibrosis is the next stage followed by osteoporosis and malformations develop within the diseased bone. The final stage in the process is ankylosis of the footplate of the stapes in the oval window.

Now that some views as to the progression of the disease process have been presented, let us consider points of origin of the lesion, that is, sites of predilection. There are two basic types of otosclerosis; the diffuse type in which almost the entire cochlear area is involved, this represents only about 10 per cent of the cases of otosclerosis; and the most common type, focal otosclerosis, representing approximately 90 per cent of the cases of otosclerosis⁵⁰.

There are three large series which best report the areas of predilection of the disease; that of Guild²⁷ who had 49 cases with 81 temporal bones, that of Nylen who studied Nager's collection of 77 cases with 125 temporal bones, and that of Fleischer who analyzed Lange's material of 98 temporal bones from 68 patients. Reports on the results of the latter two investigator's data were taken from Altmann's⁵⁰ translation of their work. The work done by Guild was taken from the routine examination of 1161 temporal bones while the cases in the other two series were highly selective.

Unilateral otosclerosis was present in 25 per cent of Guild's cases, 30 per cent of Nylen's, and 26 per cent of Fleischer's. Thus one may conclude that otosclerosis is unilateral in approximately 25-30 per cent of the time. In about one-third of all of the series there were more than one foci present simultaneously. In 80-90 per cent of the cases in all three series the area anterior to the oval window was involved; between the anterior edge of the footplate of the stapes and the upper border of the promontory and the cochleariform process----the so called "otosclerotic angle". The proposed theory that the process began from the cartilagenous rests in the area of the fissula ante fenestram was not supported as many small foci were found in this area but were not associated with the structures in question. The otosclerotic foci were found to extend back along the upper and lower borders of the oval window but Guild reports that there was a tendency to extend further back along the lower border.

The most important factor in otosclerosis is involvement of the footplate of the stapes, that is, extension of the otosclerotic process through the annular ligament into the stapedia footplate and thereby cause ankylosis. Where this is the case there is clinical manifestation of conduction hearing loss which is so characteristic of otosclerosis. Guild found stapes ankylosis in 15 per cent of the cases, Nylen 50 per cent, and Fleischer 50 per cent. The large discrepancy on this point is most likely due to the highly random sampling of Guild's material. The process of stapes ankylosis seems to be one of invasion, most generally anteriorly in the region of

of the annular ligament. On the tympanic side of the stapes footplate there is often much fibrous proliferation of the otosclerotic focus. The cartilagenous covering or rim of the oval window is destroyed by the ingrowth of otosclerotic bone. Calcification occurs in a large number of cases in the area of this previously described fibrous tissue proliferation and in the annular ligament, thus, forming a calcified bridge from the temporal bone to the stapes footplate and producing ankylosis and an area for further progression of the otosclerotic process through its mode of spread and proliferation already described.

Guild^{22,23} was the first to report on a large number of unselected cases; therefore, his results concerning the general population must seem more valid. He was the first to call attention to the difference between histological otosclerosis and clinical otosclerosis. The most important criterion being involvement of the stapes footplate by the disease process. He found that only 15 per cent of his cases had stapes involvement.

Thus, one sees that there is a large number of people who have histologic otosclerosis without any evidence of hearing loss. He found that the otosclerotic foci might be within 0.1 mm of the footplate without any clinical evidence of the disease. The one fact that must be borne in mind in the investigation of otosclerosis is that many people may harbor the disease and yet show no manifestations clinically.

Foci other than the oval window region bear some mention as the process has been observed in other areas. The most frequent site other than the oval window is the area surrounding the round window. Various authors have reported foci in other areas but these are considered quite rare. In diffuse otosclerosis there may be involvement of the entire middle ear with destruction of the basal turn of the cochlea. Covell¹⁰ has reported otosclerotic foci in the heads of the malleus and incus but this fact has not been confirmed by other investigators.

Covell and Feinmesser¹¹ report on the changes found in the ossicles in otosclerosis. The most consistent finding was the displacement of the ossicle chain. They attribute this to an attempt by the ossicles to overcome the fixation of the stapes to the footplate displacement of the ossicle chain. In their series they found that 50.5 per cent of the ears (otosclerotic) bear ex-

by the ossicles to overcome the fixation of the stapes with resultant displacement of the normal alignment. In their series they also found that 50.3 per cent of the ears (otosclerotic) they examined showed degenerative changes in the malleo-incudal joint. The explanation for this fact is the same as for the distortion of the alignment.

In normal hearing sound waves reach the tympanic membrane and cause it to vibrate. This vibration is transferred in turn to the bones of the middle ear-- to the malleus, the incus, and stapes in that order. The footplate of the stapes is placed in the oval window and it transmits these vibrations to the cochlea and hence to the organ of Corti. The purpose of the middle ear bones is to intensify the sound waves as they are not readily transmitted from one medium to another, as in this case from air to the endolymphatic fluid of the cochlea. The ear ossicles then act as a transformer in the conduction of sound waves. Best and Taylor⁴ report that the ear bones intensify the velocity of the sound waves approximately 17 times. In otosclerosis the footplate of the stapes is fixed in the oval window and thus cannot transmit the sound waves to the cochlea. Therefore an interruption of the normal pathways of transmission is brought about. The result is hearing loss of a conduction type.

ETIOLOGY

The etiology of otosclerosis has remained a mystery in spite of much investigation. Many theories have been proposed and several of these have been rejected. Part of the problem stems from the fact that no investigator has been able to produce otosclerosis experimentally, i.e. anything that would closely resemble the otosclerosis that is seen in humans. Altmann¹ and others have reported on otosclerotic-like lesions seen in chickens and other fowls, but have generally agreed that this condition is different than the one that is seen in humans. Some investigators have stated that they have created an otosclerotic-like state in lower animals by the local injection of a sclerosing solution into the vascular system of the ears of these animals. The big objection to accepting this method is the fact that the circulatory system of the ears of these animals has been shown to be different than that of the human. Much investigation has been with the use of monkeys because their ears, in all respects, seem to resemble that of the human more closely. No success has been reported in this field however. Now a consideration of the proposed theories is in order.

Habermann in the late 1800's proposed the theory that otosclerosis was a manifestation of a latent complication of syphilis. This theory stems from the time when the axiom "to know syphilis was to know medicine" was in vogue. During this period almost every thing that could not be explained was attributed to syphilis. Syphilis will produce changes in bones but nothing quite as localized as otosclerosis. This theory needless to say has lost its appeal at this time.

Some authors have felt that the basic cause is of a congenital nature. They feel that this is a pre-formed but latent process of fetal development. This theory is essentially the basis of the embryonic rest theory which has gained much popularity as the possible cause of neoplasia. In connection with otosclerosis this theory was originated by the finding of otosclerotic foci in a 7 month

old fetus and in a 6 month old infant. Also the presence of the previously described cartilagenous rests seen in the endochondral layer of bone has undoubtedly helped to strengthen this theory. Guild²² showed however that the otosclerotic foci seem to develop independently of these areas of cartilage. Also his series showed that the incidence was considerably lower in subjects less than 5 years old.

Many investigators felt that otosclerosis is primarily an inflammatory disease. The foci which develop were then in effect a result of the inflammatory process. This theory has declined in popularity with the advent of antibiotics and the decreasing frequency of middle ear inflammatory disorders. Also otosclerosis is seen in persons who have no history of an inflammatory disorder.

Another theory which has been proposed has been that of physiologic spongification, which means the process develops during the normal process of ossification. In other words, it is a pathologic variant of a normal process.

Mechanical irritation has been proposed as the possible etiology of the disease. It is felt that there is irritation of the bone producing a hyperostosis and the end product is otosclerosis. Some investigators have shown that with the annular ligament in place, as is the case in otosclerosis, there is a minimum of irritation transmitted to the labyrinthine capsule in the area of the oval window, the most common site of origin of otosclerosis. Irritation certainly does not answer all the questions that are involved in the etiology of the disease process.

Otosclerosis has been compared to many of the diseases of bone, most specifically Paget's disease because of the multi-stages of development found in histopathologic slides of both diseases. The idea that this disease is a variant of one of the bone diseases has been proposed but basically rejected because of their generalized manifestations and the localized pathology encountered in otosclerosis. In this same line of thought otosclerosis has been considered to be possibly a localized bone tumor such as a hamartoma but the fact that this condition occurs only in the temporal bone and not in any areas has pretty well ruled out this proposal.

The possibility of altered metabolism has been in consideration. Calcium metabolism is under prime consideration as abnormalities of calcium are seen in so many other bone disorders. In fragilis ossium associated with otosclerosis there has been found a lowered serum calcium. This decrease in calcium is only found when these two conditions are found simultaneously in the same individual. When these conditions are present alone there is no alteration from normal limits of the calcium concentrations, thus, tending to rule out calcium metabolism as the primary pathologic alteration. In this same line of investigation no variance from normal limits of the blood levels of phosphate could be shown either. The fact that this disease is so localized it seems possible that there could be a disturbance of the calcium and phosphate metabolism at a local level without an alteration in blood concentrations, which could help explain the disease process. Again, this proposal would only represent part of the entire picture.

Endocrine disturbances have been considered as the basic etiologic factor or possibly as playing the role of an activating or "exciting" factor. The exact nature of this disturbance or influence has not been postulated. The popularity of this theory stems from the fact that the onset of otosclerosis in many cases is at times in life when there are tendencies toward endocrine disturbances. It might be a little too strong to refer only to the onset of the disease but just as importantly worsening of the condition when already present. These periods of life referred to above are adolescence, pregnancy, lactation and the climacteric. Although no concrete disturbance can be detected it seems logical that endocrine disturbances cannot necessarily explain the primary etiology but may play a role as an activating substance.

A theory of intoxication has been proposed. This theory contends that there may be substances which act as toxins on the middle ear capsule, and either start the process or tend to activate it. Substances which have been incriminated by this idea have been alcohol, tobacco, toxic products resulting from focal infections, and various other possible toxins which stem from altered body metabolism

of either a general or a local nature.

A vascular basis of etiology has been proposed by many. Mayer⁵⁰ felt that the disease was basically secondary to arterial disease either vasomotor or organic. He felt that otosclerosis developed in certain areas supplied by end arteries, and that the disease was the result of altered blood flow through these arteries. Venous stasis has played a part in the theory of the origin of otosclerosis as proposed by Wittmack. Wolff⁵⁸ among others has attempted to explain the disease on the vascular basis. Certain vascular changes found in her series of 200 temporal bones has already been described in the section of this paper on pathology. She has presented some very interesting changes which she has found in connection with otosclerosis. Another theory which has had some popularity is that of hypervascularity. This idea stems from the fact that one sees an apparent increase in vascularity most of the time histologically but the appearance of the lesion grossly is often one of increased vascularity. Because of the lack of consistency with which the many vascular abnormalities are found and the exact location of these abnormalities, one cannot accept the vascular basis entirely but certain aspects of it definitely have merit.

Perhaps the most irrefutable theory as to origin is the hereditary or constitutional theory. The man perhaps most noted for work in this field is Larsson³⁴. He studied a large number of pedigrees of persons with otosclerosis. He found that quite often there was a family history of similar hearing loss. Even in early literature one of the signs of otosclerosis is the presence of a positive family history for a hearing loss. The mode of inheritance has been discussed to a great extent, but because of the variability with which otosclerosis seems to make its appearance, no simple explanation is possible. Concerning the overall picture of the hereditary theory Larsson has concluded, after studying 257 patients who were operated upon for otosclerosis, that parents, sibling, and offspring of persons who have otosclerosis have a greater incidence of otosclerosis than does the general public--approximately 15 per cent greater incidence. The exact method of transmission is unclear but it certainly does not follow the simple Mendelian inheritance patterns. Larsson feels that it is due to a "monohybrid autosomal

dominant inheritance with a penetrance of the pathological gene of between 25 and 40 per cent." This would indicate that it is a dominant gene which expresses itself only about 25 to 40 per cent of the time that it is present in an individual. This expression it should be kept in mind is in the form of clinical otosclerosis. As Guild has pointed out there is an entity of histological otosclerosis with no manifestations of otosclerosis; and the incidence of this condition is unknown exactly.

It has been attempted here to show that the etiology of otosclerosis is not a simple matter, but there may be a multiplicity of factors involved. One of the most sound theories is that of a hereditary factor or predisposition; however, some of the other ideas presented may be involved in the development of the disease. For example, endocrine or vascular factors may play an important role in activation of the disease of in its progression. The exact etiologic factor and/or factors is at this time unknown as is the possible relationship and interaction of the various proposals herein presented.

DIAGNOSIS

In otosclerosis, as in any disease, a diagnosis must be established before any definitive therapy can be instituted. In the text book the diagnosis does not appear too difficult however clinically this is not always the case. This section will discuss the incidence of otosclerosis including the age of onset, the signs and symptoms, some of the diagnostic tests, and finally a few brief comments on sensori-neural loss associated with otosclerosis and the effect of pregnancy on otosclerosis.

When considering the incidence of otosclerosis one must again go to the work of Guild²² as he had a large series of completely unselected cases and demonstrated the presence of histological otosclerosis. This condition can be detected only by microscopic examination of presumably normal temporal bones. He studied 1161 cases with approximately one-half Negro subjects. He found that the disease is much more prevalent in white subjects than in the Negro. One in 12 white subjects over 5 years old harbored otosclerotic foci while the ratio was only one in 96 among the Negroes. He found that the overall incidence of otosclerotic foci was 5-10 per cent among the general population. In the group under 5 years of age the incidence was 1 in 161. His data seems to support the general belief that the disease is more common in women than in men. The incidence among women was 1 in 8 while in men it was 1 in 15. This is 12.3 per cent in females and 6.5 per cent in males (both refer to white subjects). In only 49 cases was there evidence of clinical otosclerosis, or 4.2 per cent of the entire series. Of these cases both temporal bones were examined in 46 cases, 32 had bilateral otosclerosis while 14 showed unilateral disease. This figure is somewhat higher than other investigators (re: incidence). In these cases there was stapes involvement but actual hearing tests and the like are not known exactly. The incidence of clinical otosclerosis has been reported by various authors. Davenport states that the incidence among the general population is 0.2 per cent which is one of the lowest figures reported. Cawthorne⁹ states "the incidence of otosclerosis cannot be less than 0.5 per cent." Shambaugh has reported the incidence of otosclerosis to be 0.5 per cent which represents the figure which is acceptable by most investigators

of today. Most feel that the disease is more frequent among women, however, Larssen³⁴ reports that in his series of 257 cases and their families the incidence was not significantly greater among women. As stated earlier most investigators do not feel that this is the case, and otosclerosis is indeed more common among women. Shambaugh Jr.⁵¹ reports the ratio of 3:2 with women greater than men. Petersen reports the incidence is 3 times greater in women than in men.

Otosclerosis is a disease of relatively early life. Usually it has its onset sometime before the age of 40 years and may develop very early but generally not before 5 years of age. In table 1 the data from 2000 cases of Cawthorne⁹ is shown. These figures show that in his series 70 per cent of the cases of otosclerosis developed between the ages of 11 and 30. If the upper limit was extended to 40 years of age it would include 431 more cases. Geothals¹⁹ in a series from the Mayo Clinic in table 2 shows that in the age groups from 10 to 30 76.3 per cent of the cases developed. The mean age at which otosclerosis developed was 25 years. Shambaugh⁵¹ reports that the mean age of onset in his series was 22.4 years. This would agree with most of the other figures reported in the literature. One problem concerning itself with the exact age of onset is that the onset usually tends to be insidious in nature and the hearing loss may not be apparent to the individual until a considerable period of time has elapsed.

In this same line of consideration otosclerosis is a disease which is characterized by progressive hearing loss. The rate of progression of this loss is directly proportional to the amount of activity in the otosclerotic foci. Many investigators have reported that in microscopic sections of the affected temporal bones there may be foci that are not actively growing and are in a resting stage. When this is the case there will be no progression of the disease. The rate of progression is a variable thing and no accurate estimation can be made once the disease has started. Progressive hearing loss is characteristic of the disease process, and is the point which is found in the family history of many patients afflicted with this disease.

TABLE 1

AGE AT ONSET	NUMBER OF CASES
0 to 10	37
11 to 20	545
21 to 30	806
31 to 40	431
41 to 50	58
51 to 60	19
60 plus	3
No record	101

Summary: 70 per cent of the cases developed between the ages of 11 and 30
(from Cawthorne⁹)

TABLE 2

AGE	NUMBER OF PATIENTS	PER CENT OF TOTAL
0 to 9	10	2.7
10 to 14	18	4.8
15 to 19	45	12.0
20 to 24	106	28.3
25 to 29	117	31.2
30 to 34	57	15.2
35 to 39	18	4.8
40 to 50	<u>4</u>	<u>1.0</u>
Total	375	100.0

Mean age at onset - 25 years
(from Goethals et al¹⁹).

The audiogram is probably the most important single diagnostic tool in otology. In otosclerosis there is hearing loss of the conductive type. This indicates that the loss of the audiogram (air) will be more marked for the low frequencies. On the audiogram 250, is generally the lowest frequency tested. 500, 1000, and 2000 are the frequencies which are most important in the conversation ranges. This points up the fact that much of the loss in otosclerosis is in the range of normal conversation and represents a serious problem. The presbycusis or the hearing loss which is normally seen as part of the aging process is of the sensorineural type and its loss on the audiometer is usually in the high frequency ranges initially. Thus, high tone loss is more characteristic of nerve loss. Otosclerosis will generally make its appearance as a low tone loss on the audiogram.

There are other tests which will confirm the diagnosis of conduction loss. The Weber test is performed by placing a vibrating tuning fork (256 double vibrations is normally used) on the patients forehead or the top of his head in the midline and normally the sound will be heard equally in both ears. If there is a conduction loss then there is greater bone conduction of sound in the affected ear and the test will be localized to the ear with the greater bone conduction. It should be noted that this test is certainly not diagnostic, and its value in otosclerosis is questionable. The Rinne test is of much more value in otosclerosis. This test is ideally performed with a tuning fork of 512 double vibrations. The vibrating fork is placed on the mastoid bone of the ear being tested and the patient signals when he can no longer hear the tuning fork. Thus the length of time of bone conduction is found. The fork is then held approximately 1 inch from the subjects ear, and he signals when he no longer hears it in air. Normally the air conduction is approximately twice as long as bone conduction. When the air conduction is greater than the bone conduction, it is termed a positive Rinne. If the reverse is true, it is spoken of as a negative Rinne. With a nerve hearing loss, the Rinne will remain positive, but the length of conduction will be decreased. In otosclerosis there is a conduction loss

so the Rinne will be negative, i.e. the bone conduction is greater than air conduction. Also this bone conduction can be measured on the audiometer and compared with the air conduction for a more accurate evaluation of bone conduction versus air conduction. The ranges can be measured at the various frequencies routinely tested during the normal audiogram. When the bone conduction is greater than air conduction as measured by the Rinne, and then more accurately measured by the audiometer as to the exact amount of difference, this difference is spoken of as the air bone gap and is measured in decibels. When the bone conduction is greater than air there is significant hearing loss, and the person may be classified at best as hard of hearing. It is the goal of treatment to attempt to bring the air conduction up to at least the level of bone conduction, and thereby produce serviceable hearing.

When one is confronted with a case of conduction type hearing loss, in order to make the diagnosis of otosclerosis, it is necessary to rule out the other causes of a conduction loss as outlined in the classification of deafness. When this has been done the very definite possibility of otosclerosis exists, however there are other diagnostic tests and signs and symptoms which will help confirm the diagnosis.

Bezold has described three tests which will establish the diagnosis of otosclerosis. The first of these is a marked loss of low tones by air with prolonged bone conduction for these same frequencies. This is a positive Schwabach's test. It should be pointed out that with diffuse otosclerosis and involvement of the cochlea there may be high tone loss later in the disease and the Schwabach's test will become negative. This can happen also with very extensive otosclerosis however in both cases this usually does not happen until late in the course of the disease. The second of Bezold's criteria is a strongly negative Rinne to the point where bone conduction is at least five times greater than air conduction. The third is the Gelle test. This is performed by placing a Politzer bag in the external canal. This must be with a tight fitting tip and the air is forced into the external meatus while simultaneously a vibrating

tuning fork is placed on the mastoid. In a normal ear there is a decrease in hearing when this procedure is carried out. In an ear in which there is fixation of the stapes there is no decrease in hearing with this procedure. The mechanism behind this test is that the air moves the tympanic membrane and thereby sets the ossicles in motion thus when the vibrations from the tuning fork reach the ossicles they are not able to react normally. In the otosclerotic ear the footplate of the stapes is not in motion when the stimulus of bone conduction is applied therefore there is no decrease in hearing. These tests have been termed Bezold's triad. When all of these tests are as described above it is considered diagnostic of otosclerosis.

Vuorinen and Newman⁵⁴ have attempted to aid in the diagnosis of otosclerosis by radiographic measurements of the skull. Their work was stimulated by Sercer who felt that the reason that otosclerosis did not develop in quadrupeds was that the basal angle of the skull is in the verticle position and hence there is no otosclerosis. This theory is based on a stress mechanism which in turn produces the disease. They measured the sphenoidal angle in 108 controls and in 25 persons with otosclerosis. The sphenoidal angle is defined by them as the angle formed between the nasion (median point of the nasofrontal suture) to the middle of the sella tursica, and from that point to the basion (median anterior edge of the foramen magnum). This angle was 130.6 degrees for the otosclerotic group while the control group showed an angle of 132.1 degrees. They felt that this difference was not significant and have discarded the notion for either etiology or diagnosis of otosclerosis.

Another sign which might give the physician a clue to the diagnosis of otosclerosis is the appearance of a pinkish cast to the tympanic membrane. This finding is called the Schwartz sign. It is not the tympanic membrane which is discolored but rather it is a reflection from the promontory--the so called promontory flush. Extensive investigation has shown that this pink cast is due to blood vessels on the promontory and there is no abnormality of the tympanic membrane. These blood vessels are thought to be due to the increased vascularity of actively growing otosclerotic foci on the temporal bone in the area of the promontory thus we see the so-

called promontory flush, or Schwartz sign in many cases of otosclerosis. It should be mentioned that not all cases of otosclerosis will have this finding.

There are a few other findings which should be mentioned here. These are not frequently mentioned when one reviews the classic signs of otosclerosis in the modern literature. The first of these is that the person with otosclerosis appears to be quite healthy in all other medical aspects besides hearing. This might be explained on the relative young age of the patient with otosclerosis. Lederer³⁵ reports that the external auditory canal of the otosclerotic patient is usually quite wide and there is very little cerumen present. No explanation is offered for the basis of this finding. Next he mentions that sometimes there is hypesthesia of the auditory canal. These above mentioned findings are quite variable and deserve mentioning only as sometimes present and do not warrant their inclusion in the list of findings characteristic of otosclerosis.

Now a consideration of the patient is in order. The patient with otosclerosis is usually a young person (average age of onset in twenties) and more often a female. The patient first notices a unilateral hearing loss which is progressively getting worse and may become bilateral. In Guild's series of 46 cases of stapedial ankylosis there were 32 cases of bilateral disease and 14 of unilateral findings. Bezold reports that the condition is bilateral 88 per cent of the time. Let it suffice to say that the majority of the time the disease is bilateral; however, the degree of involvement may be different. As mentioned before the hearing loss gets worse with time. The patient will have a positive family history for progressive hearing loss 60-70 per cent of the time, according to Larsson³⁴ and Cawthorne⁹.

The most common symptom other than hearing loss that the otosclerotic patient complains of is tinnitus. Cawthorne reports that 84 per cent of the patients in his series had this complaint. The tinnitus which is associated with otosclerosis is usually quite severe, and it may be this complaint that bothers the patient the most. Another very frequent symptom is paracusis or paracusis

Willisii. This is the phenomenon whereby the patient is able to ^{hear} hear better in noisy surroundings. The reason for this is that the ossicles are already set in motion by the background noise and when someone is confronted with the spoken word it is comprehended more effectively because the ear ossicles are better able to transmit the sound vibrations ~~at~~ the cochlea. By this it is meant that with stapes fixation it will take a greater amount of motion on the part of the ossicles to transmit vibration through thisankylosis, and this is achieved when they~~are~~ are already in motion by background. The most important reason for this phenomenon is that by the noise~~the~~ the auditory nerve is already being stimulated and hence there is more response on the part of the nerve to the spoken word or at least to the sounds that the patient is attempting to hear.

The next symptom that commonly bothers the otosclerotic patient is vertigo. This symptom is somewhat more variable in both its appearance and its intensity than the others mentioned in this section. Cawthorne⁹ reports that 24% of the patients in his series did indeed have vertigo.

When considering the differential diagnosis of otosclerosis there are several bone diseases which may cause deafness in the same basic manner as otosclerosis. These diseases have been mentioned previously and it should be emphasized that they are generalized conditions rather than diseases confined to the labyrinthine capsule. Because of this fact some of the diseases will only be mentioned here. These diseases are fragilitas ossium, osteitis deformans, osteoarthritis, osteogenesis imperfecta, osteomalacia, osteopathyreos, rickets, Paget's disease and chondrodystrophy. There are reports in the literature by House, et al,²⁸ of congenital fixation of the stapes which is not due to otosclerosis but rather is a developmental anomaly. This condition is considered quite rare when considering the causes of hearing loss. They mention that the audiogram seen in this condition tends to be more flat, i.e. loss at all frequencies rather than low frequency loss characteristically seen in otosclerosis.

There are two subjects concerning otosclerosis which deserve mention in any general discussion of the disease. The first of

these is sensorineural loss in association with otosclerosis and the second is the effect of pregnancy on otosclerosis.

Loss other than the conduction type seen in otosclerosis is occasionally seen clinically. This sensorineural loss presumeably associated with or attributable to otosclerosis has been investigated by many students of otosclerosis. Clinically there occasionally is seen a profound hearing loss of bone conduction which cannot be adequately explained by ankylosis of the footplate of the stapes. It seems logical that there might be atrophy of the auditory nerve of a disuse type phenomenon as there is less stimulation in otosclerosis than normally. Generally it is agreed however that there is only slight degeneration of the VIII nerve histologically. Schuknecht⁵⁰ has shown that only 25% of the fibers of the auditory nerve are necessary to maintain normal thresholds in animals. He feels that the basis for the sensorineural loss in otosclerosis is due to atrophy of the hair cells of the organ of Corti, rather than a nerve degeneration per se. It has been postulated by some authors that the severe hearing losses which are sometimes seen in otosclerosis are due to otosclerotic invasion of the cochlea with resultant damage. In this line of thought Ruedi⁵⁰ states that perhaps the otosclerotic process produces metabolites and other breakdown products which contaminate the perilymph and eventually diffuse into the endolymph to act directly on the organ of Corti. Glorig and Gallo⁵⁰ have also studied this problem. They point out that with progression of otosclerosis there is a progressive loss of high tones in the bone conduction audiogram of patients with otosclerosis. This same phenomenon is also seen in the so-called "normal" general population. When they correlated these findings as a function of age, they found there was only slightly more bone conduction loss in the otosclerotic than in the air conduction of quite large samples of the population. For example, they reviewed the sixteen thousand cases of the public health service in 1935 and three thousand and four hundred persons at the Wisconsin state fair. They concluded that there was no more sensorineural hearing loss than that which would be expected on the basis of aging.

Meurman⁵⁰ has said, "Cochlear involvement is not infrequent in clinical otosclerosis. It may be mentioned, as an example, that a definite cochlear involvement was found in 24.5% of one hundred and sixty three patients under forty years of age examined at the Hearing Center in Helsinki. In most of these cases the loss increased towards the high-frequency end of the range."

Guild²² whose outstanding work has been referred to previously has stated, "Atrophy of the cochlear nerve fibers or of the organ of Corti does not occur more often in ears with otosclerotic areas than in ears free from otosclerosis. When atrophy does occur in otosclerotic ears, it is usually limited to the basal turn and differs in no way from the nerve and end organ atrophy that is often seen in the sections of ears without otosclerosis. The etiology of cochlear atrophy in ears without otosclerosis cannot, in most cases, be established and it therefore does not seem logical to attribute to otosclerosis all the cochlear atrophy found in otosclerotic ears." Feldman¹⁵ studied 122 cases of otosclerosis and concluded that the statistical study did not bear out abnormal nerve loss secondary to otosclerosis when one considers the loss seen physiologically with aging--presbycusis.

Thus the opinions of many investigators are divergent on the point of nerve degeneration but most who have made extensive investigation feel that what is seen histologically cannot be explained as secondary to otosclerosis. Many on the other hand such as Cawthorne feel that there is certainly clinical evidence that there is such degeneration, as manifested by a more profound hearing loss than can be explained on the basis of otosclerosis alone.

The next problem is the effect of pregnancy on otosclerosis. It is pretty well agreed by most investigators that otosclerosis is indeed aggravated by pregnancy and may even be precipitated by it. This is not to say that pregnancy will cause it but merely stimulate it to become manifest clinically. The problem which revolves around this subject is basically one of medical management. There have been suggestions that therapeutic abortions be performed

on otosclerotic women who became pregnant. In the light of modern day advances in the treatment of otosclerosis this is no longer practiced. Goethals, et.al¹⁹ have studied the effect of pregnancy on cases at the Mayo Clinic. Table 3 shows the effect of pregnancy by reporting the loss of hearing in relation to pregnancy as recorded by Goethals on the work of several investigators. It shows a varying results on the effects of pregnancy. Smith reports that 37% had loss with pregnancy while on the other extreme Cawthorne reports that 63% of the women in his series of 419 cases had loss of hearing with pregnancy. The study at the Mayo Clinic included 375 women who had a definite diagnosis of otosclerosis after complete otologic exam, were less than 51 years of age, and experienced pregnancy prior to or during the period of loss of hearing. Their figures showed that pregnancy initiated or increased hearing loss in 211 or 56.2 % of the otosclerotic patients they had selected. 158 of these patients had their hearing loss triggered by pregnancy-- 42.1%. In this group 106 (28.3%) related their hearing loss to the onset of the first pregnancy, 35 to the second pregnancy, and 17 to the third. Of the 106 primagravidas 81 had a second pregnancy. In 25(30.9%) of this group the hearing loss was further increased by the second pregnancy. 24 of these had subsequent gestations and 15 of the 24 had further aggravation of their deafness. Of the 35 patients that noted the onset of otosclerosis with the second pregnancy, 12 had further gestations and only two noted further loss of hearing associated with these pregnancies. 53 of their patients had a history of loss of hearing prior to their first pregnancy. 35 of this group noted progression of their hearing loss with their first pregnancy. 26 of these 35 had a second gestation and 20 of the 26 noted further loss. One sees by this data that otosclerosis is made worse or in some cases initiated by pregnancy, and the statistical data to support this fact is more than casual. Goethals has concluded, "the possibilities in otosurgery and aural rehabilitation, we find that much reassurance can be offered to otosclerotic women who are anticipating further

TABLE 3

LOSS WITH PREGNANCY

AUTHOR	CASES	PER CENT
Smith	73	37.0
Walsh	243	43.0
House	194	45.0
Larsson	104	46.0
Allen	72	48.6
Shambaugh	475	50.0
Schmidt	49	51.0
Barton	133	55.0
Cawthorne	419	63.0

Average loss of all authors above 48.7 per cent
(from Goethals et al¹⁹)

gestations. Certainly one would find it most difficult to justify a therapeutic abortion or sterilization in cases of otosclerosis complicated."

SURGERY

The treatment of otosclerosis is certainly the most important aspect of the disease to the patient. In otosclerosis medical treatment has been abandoned in favor of recent surgical developments. There has been no medical treatment which has been of any benefit in restoration of hearing. In the last 25 years there have been many advances in otologic surgery for otosclerosis. It is also interesting to note that much of the research concerning this disease has now turned toward new techniques for dealing with the disease on a surgical basis. Former unresolved areas such as the etiology of the disease have been neglected in most of the recent work which has been published on otosclerosis. Now with modern surgical techniques and development there may be hope offered to the otosclerotic patient in a large number of the cases. When considering the various types of operations that will be presented here, it is interesting to note that there seems to be as many techniques in performing the operations as there are otologists treating otosclerosis by operations. Because of this fact what will be presented here will be the historical development including the major differences in the approach to the solution of the common problem---restoration of hearing which has been lost due to otosclerosis.

Valsalva was the first to describe the macroscopic picture of otosclerosis. In 1891 Habermann described the staining difference histologically in the disease. Politzer in 1889 is credited with naming the disease otosclerosis; he was also the first to describe the disease as a primary pathologic alteration of the labyrinthine capsule. About the beginning of the twentieth century the first attempts at surgical correction were recorded. Jack reported the effect of 70 stapedectomies but his results were not favorable and he abandoned his operation. Sieberman about this same time is reported to have attempted stapes mobilization, but he also met with little success. The delicate microtechnique which is necessary for these procedures was not developed adequately at this time contributing to their failure along with the absence of antibiotics.

In 1913 the first feasible operations at that time were reported by Jenkins and Barany separately⁵⁰. These operations were of the

fenestration type, that is, they attempted to make a new opening into the semicircular canals by which the sound wave vibrations could reach the cochlea. Barany was led to his work by his inadvertently opening the horizontal semicircular canal in a radical mastoidectomy which was followed by immediate and marked improvement of the patient's hearing. Fenestrae were made at various sites in the temporal bone but the results were far from encouraging. In fact this type of operation for otosclerosis was discouraged by the leading otologists at that time and consequently the work was carried on by only a few. Holmgren²⁵ contributed much to the development of surgical approach although his individual work was not too successful. He stimulated much interest in the possibility of surgical treatment in otosclerosis. He felt that the improvement of hearing following fenestration was due to decompression of labyrinthine fluids. In 1923 he reported a partial success in one case nine months after operation. Sourdille had visited Holmgren and was stimulated by his work. He followed up on the idea of opening the semicircular canals. He developed an operation which he called a "tympanolabyrinthopexy". This operation took place in three or more stages. The first was confined to the external canal; the integument was removed of the posterior portion almost down to the tympanic membrane. This area would form a scar in 6-8 weeks and he termed this the internal plastic. The second stage was removing part of the mastoid cells through a postauricular approach exposing the middle ear. The malleus and incus were separated and the head of the malleus was removed. The internal plastic was then placed over the incus and lateral semicircular canal. The third operation consisted in elevating a portion of this membrane from the horizontal semicircular canal area, and then a fenestra was made in the canal. This fenestra was then covered with the internal plastic. He reported his results on 109 operations with positive results in 81 cases or 74 per cent. He had excellent results in 40 per cent of the cases. The major drawback to this procedure was the length of time involved and the long periods of hospitalization.

In 1938 Lempert³⁸ reported a one stage fenestration procedure

which was feasible. He previously had developed the endaural technique of mastoidectomy, and it was an enlargement of this approach that allowed him to develop this new operation. His approach was through an endaural incision and elevation of the posterior portion of the external canal being careful to retain its attachment to the tympanic membrane. The next step is the removal of some of the mastoid cells thus exposing the horizontal external canal. Then with careful technique there is the creation of the cutaneous tympanomeatal membrane, and it is then liberated from the attachment to the ossicular chain which involves removal of the head of the malleus. Then a troughlike fenestra is created in the horizontal semicircular canal down to the perilymphatic space. The final step is the placement of the tympanometal membrane in the fenestra with Shrapnell's membrane or the membrana flaccida portion of the tympanic membrane in the fenestra. This technique was termed the "fenestra non-ovals." This one stage operation for the correction of otosclerosis was a great advance in otologic surgery. It is interesting that this operation made its appearance about the time when the sulfonamides were introduced and later other antibiotics; thus, infections following otologic surgery of this nature were reduced until they became a negligible factor in post operative complications. In his original article he reported the results of 23 cases on which he had operated upon. He found "a good practical improvement in hearing was obtained and maintained." In 4 cases there was no improvement but in each of these cases there was a low level of bone conduction prior to operation. He stated that if the fenestra would remain open for six months then it would remain patent permanently. Prevention of closure of the fenestra was one of the problems which faced the early pioneers of the fenestration type of operation. The physiologic basis behind the success of the fenestration operation is that shearing forces are transmitted from the tympanomeatal membrane to the fluid of the inner ear by these forces. The fenestra created must be out of phase with the movement of the round window or they will act as canceling forces upon each other. This mechanism is at work in the normal ear between the oval and round windows. There were refinements of technique on the fenestration procedure by Lempert and others but the basic fundamental of a tympanomeatal flap inserted into a fenestra in the area of the semicircular canals

has persisted. The fenestration operation of this type was the first real promise of success in the surgical treatment of otosclerosis. There were otologists who began to participate in the fenestration operation and their results were equally encouraging. Hall⁵⁰ has compiled the data presented in table 4 which shows the results of six series of fenestration procedures for otosclerosis. As the table shows the results of the six series vary but the success is between 75 and 91 per cent but nothing is mentioned as to the type of patients which were operated upon. By type it is meant the advisability of the operation in the first place regarding the status of their VIII nerve. Shambaugh⁵¹ has divided the prospective patients into various groups which are on the basis of amount of involvement as shown by clinical tests. Therefore the group of patients with the least otosclerotic involvement and the ones with the best auditory nerves are the ones which have the best post-operative results. Success of the operation is generally considered to be restoration of hearing to 30 decibels which is the minimum which is necessary for socially acceptable hearing. Some feel that the closure of the air-bone gap to 10 decibels is considered to be the criterion for success but if the patient has very poor bone conduction then he is not a candidate for the operation.

Shambaugh in reviewing 3091 fenestration operations with a follow up of 8 months to 13 years found that 13.5 per cent of these cases failed to show a rise of 10 db. for air conduction post operatively. Half of these were early failures. Of these initial failures 70 per cent were due to serous labyrinthitis, 27 per cent to poor selection of cases, and 3 per cent to technical error. Of the late failures, 44 per cent were due to osseous closure of the fenestra, 44 per cent to cochlear deterioration and 12 per cent to endolymphatic hydrops. Still these results give a positive result in 86.3 per cent of the cases.

Campbell⁸ reported on the long term results of the fenestration procedure that 65 per cent of his series obtained practical hearing for the three speech frequencies. Of the remaining there were some in which there was a gain in hearing to a serviceable level but

TABLE 4
RESULTS OF THE FENESTRATION OPERATION

AUTHOR	LENGTH OF FOLLOW UP (YEARS)	SUCCESS IN PERCENTAGE OF CASES
Ruedi et al	2	70.0
Cawthorn	4	74.0
Farrior	2-5	83.0
Venker	4-8	76.4
Hilleman and Shambaugh	1	91.0
Hall	2-18	82.9

(from Hall⁵⁰)

this gain was not maintained. There was also a number of these in which the operation was redone which brought the overall success rate up to 75 per cent.

House²⁷ in reporting on 500 patients after fenestration found that 79 per cent of 266 cases with ideal conditions had serviceable hearing. 66 per cent of 211 cases considered borderline had serviceable hearing following fenestration; while only 1 (4%) of 23 had a good result in the group considered unsuitable for operation. Williams at the Mayo Clinic⁵⁰ reported on 494 ^{follow}fp;pw-up cases of fenestration and reported results as excellent if the hearing was above 30 db. in the three speech frequencies and good if there was 30 db in two of the three frequencies in question. Fair was serviceable hearing in one of the above frequencies (500, 1000, and 2000). Their results showed 40 per cent excellent, 35 per cent good, and 7 per cent fair while only 18 per cent showed poor results. Thus, the overall gain was in the neighborhood of 88 per cent.

When one considers all the reports of results on the fenestration operation one finds that most investigators report that there is a favorable increase in hearing in about 70-80 per cent of the cases which are followed up to an average of about 5 years. When considering positive results with the fenestration procedures one should keep in mind the report of Davis and Walsh who showed an average acuity in fenestrated ears of 27 decibels less than the normal. Sometimes after fenestration there may be much more hearing than this but they explain this fact on the variance of the normal and the normal for these ears in question would certainly be above the normal of zero. They felt that the ears that after fenestration show a particularly fortunate result are ones that showed considerably better than average hearing before the onset of otosclerosis.

The fenestration operation was indeed a boon to otosclerotic treatment but its fame was not to be long lived. In 1953 Rosen⁴⁷ while preparing to do a fenestration operation decided that he would see how much stapes involvement there was and he moved the stapes thus freeing the ~~an~~kylosis and there was a sudden and dramatic

improvement in hearing on the part of the patient. He tried several of these stapes mobilizations but on follow up examination it was found that this simple procedure was not entirely suitable as the patients had a tendency to lose the hearing that they had gained due to reankylosis of the stapes footplate. This procedure tended to focus the attention on the stapes and the oval window rather than the fenestration type operation. The stapes has been fractured, replaced by several different materials, sawed, loosened, and removed in attempts to improve hearing via operation in otosclerosis.

Stapes surgery has become a reality in the last decade and there are several different methods of approaching the problem of otosclerosis. Again each surgeon has his individual technique but there are some pioneers in the field with a few basic principles of technique and this work is what will be presented herein.

One of the prominent names in otosclerotic surgery is Shea^{50,52} who was one of the first to advocate complete removal of the stapes and replacing the footplate area in the oval window with a graft to keep it open. He chose a vein for this purpose after watching veins being used in place of arteries as grafts. During his procedure he completely removed the stapes and after the vein graft was in position in the oval window he advocated replacement of the stapes by a prosthetic device and he felt that polyethylene tubing should be placed in the oval window on top of the graft with a connection to the incus, thus completing the ossicular chain. The basic principle of this procedure is removal of the entire stapes, vein graft in the oval window, to prevent otosclerotic reclosure, and replacement of the stapes with a polyethylene strut.

Hough²⁶ has proposed partial stapedectomy as the ideal surgical principles in that he removes the diseased portion but has preserved all normal tissue. For example, both crura of the stapes are removed or separated near the base. The footplate is removed and gelfoam is inserted into the oval window; next the longest crus remaining is placed in the gelfoam and thus the ossicular chain is reestablished. With his procedure Hough found an average gain of 31.6 decibels regarding the air-bone gap, however his series was without long

follow-up results.

Portmann⁴⁶ has described his interposition operation. This operation consists of disarticulation of the incudo-stapedial joint, followed by removal of the mucosa around the oval window and ablation of stapes from the window. A graft is then inserted into the window and the stapes is then re-inserted and articulated approximately. He found that this procedure produced 94.8 per cent good-excellent results.

Scheknecht^{49,50} was in favor of this same basic type of procedure but he felt that following stapedectomy the graft substance should be fat removed from the patient's sub cutaneous tissue. He advocates replacement of the stapes by a wire prosthesis which is re-attached to the incus. House³⁰ has proposed a very similar procedure, but instead of fat he has found that gelfoam will provide adequate covering of the oval window. He also felt that if the footplate was too much involved to remove completely then fragmentation of the footplate would suffice.

Kos⁵⁰ also followed this type of technique with the wire prosthesis, and his results are reported to be quite good. Goodhill⁵⁰ reports the use of a polyethylene stapedial prosthesis and fat graft technique in his approach to otosclerotic surgery. On occasion he does not completely remove the stapes footplate but merely fragments it and inserts his prosthetic device into the fragmented area. He felt that an articulated device would assume normal function of the stapes more readily than would an unarticulated one. He also describes a combined polyethylene foot prosthesis and stainless-steel malleus clasp.

Thus one sees that the basic principles of stapes surgery today are removal of the stapes entirely, placement of a graft or some material into the oval window to prevent reclosure of the oval window, and substitution of the stapes by a prosthetic device. The results of these procedures are quite good in the neighborhood of 85 per cent success. One thing that should be kept in mind is that any procedure is only as good as the person who is doing it. With careful technique one can expect a high rate of success in surgery

for otosclerosis provided there is meticulous selection of cases. For example, if there is very poor bone conduction, then there is no sense in performing any type of operation as the ear can function only as well as the auditory nerve will allow it to function. Bone conduction may be used as a standard of nerve status as the operation cannot improve hearing except in unusual cases more than the bone conduction is pre-operatively. The newer techniques of stapes surgery are more suitable than the fenestration procedure because they are less extensive and may be done under a local anesthetic. At the present time the fenestration operation is almost a thing of the past but because of the newness of the stapes procedures only time will tell as to which will be the final answer although stapes surgery seems to be the answer at this time. At any rate these wonderful advances in otologic surgery have certainly made the future more bright for the otosclerotic patient.

Otologic surgery is not without its risk and complications. One big risk is certainly poor technique where a mechanical error is made and the entire ear is destroyed by a careless surgeon. This type of error does not occur often however.

Lewis³⁹ has presented some of the complications of stapes surgery. He feels that there are four major complications to this type of surgery. First is surgical trauma manifest by immediate vertigo and nausea and vomiting while still on the operating table. They may lose hearing for a while with either regaining the lost hearing later or progression to total deafness. The mechanism is thought to be direct injury to the utricle and/or saccule. The second complication he describes is labyrinthitis which is not suppurative but aseptic or serous. The onset of symptoms comes any time from the day of the operation until two weeks post operative. The symptoms are a low pitched tinnitus, postural vertigo, ataxia, nausea and vomiting. Many of these patients sustain permanent hearing loss. The third of the complications he describes is that of a fistula in the footplate area or small rupture of the membrane in the oval window. The symptoms of this condition are recurrent symptoms of the inner ear with hearing loss and equilibrium disturbances.

These things may not develop for several months after the operation. The final complication Lewis described is that of delayed inner ear storm which is manifest by sudden loss in hearing, tinnitus, vertigo, nausea and vomiting. This may develop any time post-operatively and constitutes a surgical emergency. This condition is caused by rupture of the membrane covering the oval window. These complications are only some of the ones which have been reported but whatever the complication may be, the frequency of complications is directly related to the technique which is used at the time of operation.

SUMMARY AND CONCLUSIONS

As has been presented in the preceeding, otosclerosis is a disease of many facets. It represents approximately 10-15% of hearing loss due to all causes. Otosclerosis is a primary disease of the labyrinthine capsule, more specifically of its middle layer of endochondral bone. The disease histologically, shows multiple stages of development. The process apparently begins in the area of the vascular channels. The over all picture is one of immature, web-like bone, which is characterized by dark blue staining intra-cellular material. Blue mantels have been described in association with otosclerosis however, they may occur in bone that is not otosclerotic. They have been thought to represent (by many) early stages of otosclerosis.

There are two basic types of otosclerosis; the diffuse type involving the entire cochlear area represents about 10% of the cases, and the most common type, focal otosclerosis, which represents approximately 90% of the cases. The focal type may be active or quiescent. Otosclerosis is unilateral 25-30 % of the time. The foci are seen most often in the area anterior to the oval window. The most important factor of the disease process is involvement of the stapes footplate. When this is the case, there is interruption of the normal ossicular chain with a resultant conduction hearing loss. Guild has described a condition of histologic otosclerosis i.e. the stapes footplate was not involved.

The etiology of otosclerosis is unknown at the present time. Some theories which have been presented are the manifestation of syphilis, an embryonic rest theory, a result of inflammation, a result of mechanical irritation, and that it is a variant of one of the bone diseases i.e. Paget's. A vascular basis has also been processed. This may be arterial in nature, either vasomotor or organic. Venous Stasis has also been proposed as the possible vascular anomaly. Hyper-vascularity has also been considered. Endocrine disturbances have also been incriminated, primarily because otosclerosis makes its appearance at times in life when endocrine disturbances are common e.g. pregnancy. It seems logical that endocrine levels may influence the course of the disease however a direct causal relationship has not been established. The most irrefutable theory as to origin is the

hereditary or constitutional theory as 60-70% of all patients with otosclerosis have a family history of a similar hearing loss. Certainly there is no one answer to the etiology of otosclerosis. Most likely there are constitutional endocrine and perhaps metabolic factors which may affect the course of the disease. The exact possible relationships and interactions of these different theories is unknown.

Otosclerosis is found in approximately 0.5% of the general population. It is most often seen in women. It is a disease of relatively early life--average onset before age 30. The disease is characterized by a progressive hearing loss. On the audiogram in otosclerosis there is a typical conduction loss picture--most marked losses in the low frequencies. Other tests which will help confirm the diagnosis are a negative Rinne, a positive Schwabach's test and a positive Gelle test. The Schwartz sign or promontory flush is present quite often. The patient complains of hearing loss, severe tinnitus, paracusis, and occasionally vertigo.

Most investigators have not been able to show a significant degree of sensorineural loss histologically in association with otosclerosis. However many have described clinical evidence that there is nerve loss in otosclerosis. Otosclerosis may be precipitated or aggravated by pregnancy. With the modern techniques of otosurgery it is felt that therapeutic abortion in the otosclerotic should be a thing of the past.

Many attempts at surgical correction of otosclerosis were made in the early 1900's, but until Lemperts fenestration procedure in 1938 there was little success. His operation utilized a fenestration of the semicircular canal and a tympano-meatal flap. In 1953 Rosen introduced stapes mobilization procedures but the stapes ankylosis recurred. Since that time the concept of stapedectomy has evolved. At the present time the surgical correction of otosclerosis involves removal of the stapes, a graft in the oval window to keep it patent, and replacement by a prosthetic device. Several substances have been used as graft substances---vein, fat, and gelfoam have gained the most

popularity. Many substances have been used to substitute for the stapes---wire and polyethylene struts (some articulated). Portions of the stapes have also been used to complete the ossicular chain. With the modern techniques there is approximately an 85 per cent chance of succes^s of correction of otosclerosis if there is no nerve loss. The otosclerotic patient is no longer faced with the progressive hearing loss and eventual total deafness but chance of correction is offered in the majority of the cases.

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