A REVIEW ON COCHLEAR OTOSCLEROSIS

AN INDEPENDENT PROJECT
SUBMITTED IN PART FULFILMENT OF I. M.Sc. (SPEECH & HEARING)
UNIVERSITY OF MYSORE

1984.

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Reg. No.8411
Vijaya Shree

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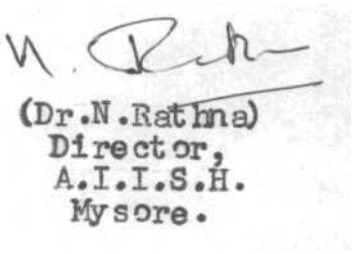
CERTIFICATE

This is to certify that the Independent Project "A REVIEW ON COCHLEAR OTOSCLEROSIS" is the bonafide work in part fulfilment for I. M.Sc. (Speech & Hearing) of the student with Reg. No. 8411



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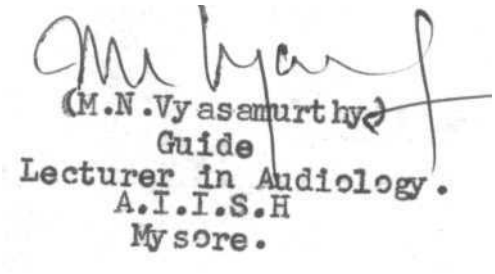
This is to certify that this Independent Project has been prepared under my supervision and guidance.



DECLARATION

This Independent Project is the result of my own work undertaken under the guidance of Dr. M.N. Vyasamurthy, Lecturer in Audiology, All India Institute of Speech & Hearing, Mysore, and has not been submitted earlier at any University for any other diploma or degree.

Reg.No.8411

Place: Mysore

Date:

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CHAPTER-I

INTRODUCTION

Many different diseases of diverse etiology can affect the bone of the otic capsule or labyrinthine capsule. Most of them are generalized diseases which involve the temporal bones as part of the skeletal system; a few of them, otosclerosis in particular, are confi to the otic capsule. Irrespective of the pathogenesis they have one thing in common, their ability to cause deafness and vestibular symptoms (Morrison 1979).

Otosclerosis is a common hereditary localised disease of the ban derived from the otic capsule. Mature lamellar bone is removed by osteoclasis and replaced by unorganised woven bone of greater thickness, cellularity and vascularity.

The misnomer "otosclerosis" was fist applied to cases of stapes ankylosis nearly a century ago in 1881, by Von Troeltsch in the mistaken belief that sclerosing changes in the tympanic mucosa were responsible for the fixation of the stapes. Unfortunately, this name has persisted, despite Politzer's (1894) demonstration that a primary bone disease of the labyrinthine capsule caused the ankylosis of the stapes. Siebenmann (1912) proposed the more accurate pathological term "otospongiosis" still preferred and used by many European otologists.

An otosclerotic focus may cause no symptoms, its presence being detected by postmortem histologic section; it may replace the footplate of the stapes causing progressive osseious ankylosis and conductive deafness; it may involve other parts of the labyrinthine capsule giving rise to sensori-neural changes, both cochlear and vestibular; and it may produce a combination of these effects which are sometimes referred to as "histological", "clinical" and "cochlear" otosclerosis.

Histological otosclerosis means that one or mare foci of otosclerosis are located in the bony labyrinth without causing stapes fixation.

Clinical otosclerosis means that the otosclerotic lesion has caused stapes fixation.

Cochlear otosclerosis implies that otosclerosis can cause sensori-neural hearing loss without an associated conductive loss. I implies that the otosclerotic focus has involved the endosteum sufficiently to cause degenerative changes in the membranous labyrinth while not causing footplate fixation.

That otosclerosis can cause a pure sensori-neural loss without stapes fixation has only recently begun to be appreciated. Shambsugh (1955) was the first to describe a case of tosclerosis presenting at first with pure perceptive deafness and only showing signs of stapedia ankylosis years later. He assumes that in these cases, the focus of otosclerosis first penetrates the endosteum of the otic capsule and forms shunts with the vein of the inner ear. The perceptive bearing loss arises as the first clinical sign of the venous congestion of the inner ear. The conductive hearing loss typical of otosclerosis usually does not arise. In these cases, until later when the otosclerotic proces has extended to ankylose the stapes. In exceptional cases the stapes never become fixed and the patient goes through life with a pure otosclerotic perceptive deafness.

The enzymatic concept of otosclerosis explains the progressive sensori-neural impairment which has been thought of by numerous authors, but the cause of which had remained obscure until now. The spread of hydro3ytic enzymes, and of proteases of cellular destruction by the otosclerotic microfoci of the otic capsule allows us to explain pure cochlear otosclerosis as well as cochlem constant in the

conductive-perceptive audiometric types of surgical otosclerosis.

A double consequence results from this concept: on one hand, the necessity of early detection of cochlear otosclerosis, in its pure type as well as in the cochlear component of the mixed audiometric type of surgical otosclerosis. On the other, the importance of an extended treatment by an enzymogenesis inhibitor for stopping cochlear deterioration. Sodium fluoride acts in more than half of these cases

Sanders (1965) feels that some of the bearing losses of the type the otologist is tempted to label "SN etiology unknown" are actually the result of otosclerotic foci deep to the oval and round windows. That the otosclerotic process can and does invade the cochl space and the internal auditory meatus has been well established by histological and radiographical observations. Otosclerosis is a result of such invasion generally has been referred to as "cochlear otosclerosis" or "labyrinthine otosclerosis", although, Carhart (1963] has suggested "retrofenestral otosclerosis" (as differentiated from fenestral otosclerosis) as a more definitive term.

Cochlear otosclerosis may occur in isolation or it may be associated with conductive loss. Of course, the conductive lesion will not affect the hearing impairment brought about by cochlear otoscleros itself, although its presence may complicate the task of determining how much loss is due to the retrogenestral component.

Cochlear otosclerosis brings about sensori-neural loss by inducing changes in the composition of the inner ear fluids or also by altering the blood supply in the cochlear or both (Gussen 1975).

Progressive sensori-neural hearing loss following successful stapedectomy may be due to cochlear otosclerosis. The course of the

otosclerotic process is uncertain and thus unpredictable. Active and inactive forms may coexist. An inactive focus may become activated. Both forms may be present without producing symptoms. The age of the individual patient with cochlear otosclerosis does not determine acti vity or quiescence of the lesion (Freeman 1979).

Otosclerosis occurs most frequently in the white population. It occurs twice as commonly in women as in men. Otosclerosis starts in early adult life. Rarely it starts before the age of 10 years or after the age of 40 years. It runs in families. The chances of oto-sclerosis being passed on to a child is about 10%, although where the is a strong family history it may be greater. Pregnancy is liable to initiate or accelerate the progress of deafness.

Otoaclerosis is predominantly a disease of Caucasoid man, being a very common cause of deafness throughout Europe, the Balkans, the Middle East and the subcontinent of India, together with the Cauaasiar peoples of North and South America, Australia, New Zealand, South Africa and elsewhere. It also occurs, though less frequently in the negrito peoples of Malaya, New Guinea, and the Philippines and in the Japanese, a mixed race of mongol, ainu and negrito blood (Morrison 197

It is relatively rarely found in mongoloid and negroid man, though it is encountered in the negro population of America and the West Indies, presumably due to hybridization. In this latter group the disease is 10 times less frequent than among Caucasians.

In the Indian subcontinent otosclerosis is more common in South India than in the North where its incidence is 1 to % (Shah 1973 and Sinha 1973).

Elucidation of the incidence is befogged by the differentiation into "histological", "clinical" and "cochlear" otosclerosis. In

Caucasians, Guild (1944) found postmortem "histological" otosclerosls in 38 out of 585 temporal bones (6.5%) and "clinical" otosclerosis in 6 out of 585 (1%). "cochlear" otosclerosls was not recognized.

The prevalence of the "clinical" condition has been estimated at about 0.5% by Shambsugh (1949) and by Cawthome (1965), and at 0.3% by Morrison (1S67). It should be emphasized that this means that between 3 and 5 per 1,000 of the adult Caucasian population have clinical conductive deafness due to stapedial otosclerosis.

Estimation of the frequency of "cochlear" otosclerosis is more difficult. Shambsugh (1969) considers that pure sensori-neural impairment will prove to be as frequent as stapedial otosclerosis. At first acquaintance this figure seems staggering, but increasing awareness of the entity and of its clinical and radiological manifestations makes the idea more acceptable. His personal records indicate that pure cochlear otosclerosis was diagnosed once for every 20 cases of clinical otosclerosis, and the proportion is increasing with experienc Bosatra (I960) in his clinical study of otosclerosis of the inner ear wrote, "The real 'cochlear type' of otosclerosis is believed to represent about 5% of total cases.

Now, pure cochlear otosclerosis is slowly becoming recognized as a serious cause of sensori-neural bearing loss comparatively early in adult life.

Aim of the study:-

Here, an attempt has been made to review the available literature as far as possible, emphasizing the essence of the problem and of modern ideas on its etiology, diagnosis and treatment.

CEAPTER-11

PATHOGENESIS AND ETIOLOGY OF COCHLEAR OTOSCLEROSIS

From the histological point of view, otosclerosIs begins with an osteoclastic destruction of the old capsular bone, the formation of pathological bone marrow and the replacement of the absorbed old bone by a network of new3y formed web-like bone, rich in cement and poor i fibrils. But such an interplay of destruction and formation of bone is typical for all vital bone changes. This is why, there is similar ity between the histological findings in clinically different conditions such as von Rechlinghausen's osteitis fibrosa, Paget's disease and otosclerosis (Ruedi, 1963).

Causes of otosclerotic bone changes:-

Many theories have been advanced concerning the etiology. Constitutional, local and general activating factors are considered.

I. The constitutional factor:-

This factor accounts for the heredity of cochlear otosclerosis. Observations on heredity in clinical otosclerosis have favoured a autosomal dominant type of inheritance, although a recessive autosomal type is not excluded (Schwartz and Becker, 1964).

Cochlear otosclerosis either without stapedial involvement or wit minimal fixation also follows a dominant hereditary pattern. Families are not infrequently encountered having the expected ratio of members with pure sensorineural loss and the rapidity of progress, audiometric pattern and arrest of deafness are often similar in the sibships. Pur cochlear otosclerosis is rarely encountered in families whose other members all have clinical stapedial otosclerosis, yet it does occurr in 2% of families investigated. Combined stapedial and cochlear otosclerosis on the other hand is a common familial phenomenon (Morrison 1971).

II. Local factors:-

In the etiology of cochlear otosclerosis, these factors consist of developmental, vascular and mechanical factors and have been studied by ear histologists for a considerable time.

A. Developmental factors:–

The otic capsule is a box-like structure performed in cartilage which is gradually replaced by enchondral and intrachondral bone, starting from 14 ossification canters. The ossification of the enchondral labyrinthine capsule is complete at the end of the second year of life. Normally no further bone formation takes place, so that the enchondral layer consists throughout life of embryonic bone. The labyrinthine capsule is threaded through with strands of connective tissue from the fissula ante fenestram, and a further strand. These strands are partly surrounded by cartilaginous rests which have remain there since embryonic tines. B ast & Anson (1949) have described other similar remnants of cartilage in the newborn and infants, in the infrs cochlear region, the canalicular region, and in the region of the round window.

If it is assumed that the basic material for otosclerotic bone formation consists of cartilage remnants, or newly formed cartilage within the enchondral labyrinthine capsule, a further (equally unknown general activating factor must also be postulated. This could be a blood-borne osteogenic hormone which reaches the labyrinthine capsule and stimulates the cartilage remnants there into ossification and pathological bone formation. Once established, this process proceeds steadily, possibly due to some failure of the statie functions of the labyrinthine capsule.

B. The Vascular Factor:-

This is no longer accepted by the majority of authors as a local

cause. Otto Mayer (1982) used to defend the view that "the otosclerotic foci were formed in response to localized disturbances in the arterial blood supply of the capsule. This theory has already been given up by Mayer himself because the distribution of the arteries did not always coincide with the distribution of the otosclerotic foc

By means of electrocoagulation of the otic capsule, Bellucci and Wolff (1960) have observed certain vascular and bony changes bearing some resemblance to the changes associated with cochlear otosclerosis. Thus, they suggest some factors altering the blood supply of the otic capsule may play a role in the pathogenesls of cochlear otosclerosis.

Reedi (1969) has observed abnormal vascular connections, called shunts between the veins of the membranous labyrinth and the vascular spaces of active foci of otosclerosis which have penetrated the endosteum of the bony labyrinthine capsule. The resultant venous congestion of the inner ear persists until the bony canals transmitting the vessels through the otic capsule become sufficiently wide to accept the increased blood supply. The venous congestion causes damage to the spiral ganglion cells in the modiolus. The organ of corti and the stria vascularis probably also suffer damage. According to the histological findings the theory of a vascular pathogenesis of all forms of cochlear otosclerosis is established.

C. Mechanical Factors:-

These might possibly give rise to otosclerosis by stress and strain damage to the labyrinthine capsule. The otic capsule is for its greater part deeply embedded in the petrous pyramid and protected by the surrounding softer temporal bone. Otto Mayer (1932) was the first to demonstrate the intra vital organ of microfissures in the enchondral labyrinthine capsule. Mayer ascribed these spontaneous

fractures to the overloading of the petrous pyramid in relation to the base of the skull. He suggested that the labyrinthine capsule could develop spontaneous cracks in the areas of special strisn, with repair by plexiform bone (Looser) and finally, an otosclerotic focus would arrive there.

Sercer (1961) pursued his anthropological theory of the origin of otosclerosis on the basis of a mechanical overloading of the labyrinthine capsule. Instead of extrinsic forces, Sercer attributes the main part in this to intrinsic forces, acting on the base of the skull during growth changes. Due to the transition from quadrupedal to the upright gait of man an angulation of the base of the skull tak place producing a symmetrical rotation and torsion of the two pyramid However, the labyrinthine capsules within the pyramids do not participate in the rotation. The resulting strains in the region of the brittle labyrinthine capsules cause bilaterally symmetrical spontaneos fractures through direct pressure.

Sercer postulates a second local factor for the origin of otosclerosis in the form of venous stasis of the blood draining from the labyrinthine capsule. The venous obstruction is also said to result from the angulation of the base of the skull. This affects the petro temporal bone by narrowing the canals and fissures transmitting the veins. Sercer regards this venous stasis as an indispensable condition in the origin of otosclerosis. As a result of stasis in the vessels draining the region of the fenestra, an osteophilic hormone accumulates also in the area of special mechanical stress. The mechanical forces thus determine the site of the bony metaplasia and the necessary venous congestion, while the osteophilic hormone will contra the modeling of the local bone structure.

III. General Activating Factors:- (Biochemical Concept)

General activating factors such as metabolic disturbances must be considered in the genesis of cochlear otosclerosis. Clinical observations suggest that certain hormones may be involved in the pathogenesis and progress of otosclerosis perhaps as an activating factor. The principal experimental approach toward ascertaining the significance of hormones in the pathogenesis of cochlear otosclerosis has been the measurement of levels of urinary steroids.

A study on the pathogenesis of otosclerosis with special reference to its relationship to level of urinary 17-ketosteroids was made by Sinha and Samant (1967). 62.5% of cases studied showed a decreased or low normal activity of androgenic hormone. They suggest that a subnormal level of androgen may act as a trigger in activating the disease in persons with hereditary predisposition.

Maurer (1968) investigated the calcium phosphorous metabolism in otosclerosis and in other types of deafness but found no difference in the calcium levels in either group. On the other hand, otosclerotics show a decreased alkaline phosphatase activity, the normal level being in the region of 5 king-Angstrong units. This decrease ir the phosphatase activity is said to be associated with diminished production of androgenic hormones in the majority of otosclerotic patient These hormones stimulate osteoblastic activity and a decreased androgen secretion results in decreased activity of the osteoblasts. Thus the alkaline phosphatase activity which is almost wholly dependent on oste blasts, aiio becomes diminished.

Clinically, the onset of otosclerosis is frequently observed to coincide with puberty, the incidence of onset increasing with age. This suggests a role for hormones. Pregnancy, appears to increase the

severity of otosclerotic conductive deafness. This observation further implicates a hormonal activation process.

The question arises as to how subclinical lowering of androgen secretion will result in the genesis of cochlear otosclerosls. This could be explained as follows:

1. The blood supply of the otic capsule is probably in a delicate balance with its physiological needs and therefore even slight variations in the blood level of androgens is sufficient to deviate the osteoblastic activity in an abnormal direction resulting the production of web-like fibrous bone of a poor quality.
2. The hereditary predisposition makes the otic capsule more vulnerable to these changes. A slight decrease in the level of androgen may not affect a normal person but may be the determining factor in projecting the disease in persons with hereditary predisposition.

The distribution of the otosclerotic focus along the blood vessels, at least in the early stages, in the form of figger like processes, further strengthens the possibility of the existing factor, being blood borne.

Soifer et al (1971) however, report that estrogen may ben an influencing factor in the pathogens sis of cochlear otosclerosis. Their findings were based on the study of urinary excretion in 32 patients otosclerosis. Excretion of estrogens was found to be elevated in the otosclerotic male compared to the nonotosclerotic male. Based on urinary excretion, it was concluded that a change in the levels of androgens or corticoids is not involved in otosclerosis.

According to Arslan (1968), general and constitutional factors may cause an alteration of the fundamental substance of connective tissue. The so-called collagen diseases arise as a result and among the well known examples of periarteritis nodosa, scleroderma, midline facial granuloma. Arslan also includes otsclerosis. Ricci (1961),

based on histochemical investigation of otosclerotic bone observed an irregular excess of acid mucopolysaccharides and an increase in the a line phosphatase in the intercellular substance and in the cellular elements (the osteocytes, osteoblasts, and young fibroblasts). The accumulation of acid mucopolysaccharides causes the collagen fibers to dis integrate, and this has also been observed by Chevance (1962) with the electron microscope. The synthesis of these fibers is at the same time delqred by the excess alkaline phosphatase. Changes in the affinity of the calcium salts to the intercellular substance are said to cause decalcification, changing the bony into connective tissue with is finally ossified to produce a focus of otosclerosis.

According to Ricci and Arslan (1961), the disease in the bony capsule of the labyrinth begins with change in the cellular activity of the osteocytes following a constitutionally caused mesenchymopathy.

An enzymatic study of the perilymph from otosclerotic patients was conducted by Chevance et al (1972). A high activity of especially acid phosphatase, collagenase, and chymotrypsin was demonstrated. The also found a striking correlation of hydrolytic activity to the deve-lopment of the perceptive hearing loss.

Chevance (1976) in a study of 250 otosclerotic perilymphatic fluids found statistically that in the presence of developing inner ear deafness, 90% of cases will demonstrate increased trypsin activity. Conversely, elevated anti-trypsin levels are found in the absence of developing inner ear deafness.

Electron microscopic studies of biopsies from the labyrinthine capsule by Bretlau (1971) has confirmed that cochlear otosclerosis is an enzymatic disease whose destructive phase might be explained by the action of the hydrolases found in the lysosomes. The intracellular as

as well as extracellular, localization of the acid phosphatases agree vith the electron microspic findings of the typical morphological features of the otosderotic focus.

Electron microspic study of otospongiotic foci by Causse et al (1977) has led to the notion that contrary to Siebenmann's theory concerning the primary role of osteoclasts, the essential features of the destruction of the normal bone of the otic capsule, which characterizes the initial stage of otospongiosis, are the histiocytes, producing marked hydrolysis and lysis of the collagen fibrils.

They have identified 6 enzymes: phosphatasic acid, collagenase, α-chymotrypisn, lactqte dehydrogenase, ribonuclease and trypsin in the perilymph of otospongiotic patients, operated on by stapedectomy. This enzymatic process starts from one or more of the numerous cartilaginous rests scattered through the enchondral layer of the very unst ble otic capsule.

Hydrolytic enzymes and proteoses of cellular destruction can spread from the focus into different parts of the cochlea situated in more or less remote vicinity. The enzymes may remain inside or in the immediate proximity of the focus and never each the inner ear? When proteases and proteolytic enzymes reach the inner ear, the result is a sensori-neural hearing loss.

Thus the enzymatic concept of otosclerosis explains the progressi SN impairment which has been thought of by numerous authors, but the cause of which had remained obscure until now. The spread of hydrolyti enzymes and of proteases of cellular destruction by the otospongiotic microfoci of the otic capsule allows us to explain pure codhlear otosclerosis as well as cochlear component in the mixed conductive-percept audiometric types of surgical otosclerosis.

CHAPTER-III

PATHOLOGY AND HISTOPATHOLOGY OF COCHLEAR OTOSCLEROSIS

Pathology of Cochlear Otosclerosis:

Pathological studies an the otosclerotlc process were presented by Habermann (1903), Siebenmann (1912), Mayer (1917), Weber (1935), Nylen, Ruedi & Spoendlin and Altmann (1966). These investigators found that severe otosclerosis not only fixed the footplate but also caused degenerative changes in the inner ear. The literature is replete with examples of severe involvement of the cochlear bony labyrinth in association with degenerative changes in the spiral ligament, stria vascularis, organ of corti and cochlear neurons.

The otosclerotic process generally develops in areas of the labyrinthine capsule, in which embryonic cartilage often persists. The first indication of the disease comes as an enlargement of the perivascular spaces.

The otosclerotic lesion may originate in almost any region of the labyrinthine capsule. In the great majority of instances (70 to 90%), however, it islocated in the so-called area of predilection, anterior to the oval window (Beickert, 1965). This region is also characterized by the frequent presence of embryonic cartilage and fibrous tissue. During its evolution, the lesion may well replace parts of the fissular ante fenestram but it does not necessarily originate in that structure. In 30% to 50% of instances, the otosclerotic disease manifests itself in the form of two independent lesions which develop in dif. regions of the labyrinthine capsule.

Otosclerotic foci are much less frequently encountered in other areas of the labyrinthine capsule or in the walls of the internal auditory meatus (15 to 25%). Very rarely are they found beyond the

confinements of the otic capsule.

According to Guild (1944), in the great majority of cases, the otosclerotic focus of new spongy vascular bone remains quite small and asymptomatic. These cases are called histologic otosclerosis. In about 12% of the cases, the focus expands sufficiently to reach th oval window and fix the stapes, causing the familiar and easily diagnosed stapedial otosclerosis. In some of these, the expanding focus reaches the cochlear endosteum as the stapes is becoming fixed or sometime after fixation causing a combination of cochlear otosclerosi and stapedial otosclerosis. In an unknown number of cases, the patie presents with a pure progressive nerve loss that has been caused by a focus of otosclerosis in the cochlear capsule without fixation of the stapes. These are cases of pure cochlear otosclerosis.

A large focus of ocesclerosis which is present in the otic capsule in some cames invades the Fallopian canal of the facial nerve and the course of superior division of the vestibular nerve. This focus invades the endosteal layer of the basal, Middle and apica coils of the cochlea. The outer layer of this focus appears to be inactive; whereas that portion involving the endosteum is quite active i.e. blue staining with marked vascularity. The spiral ligament is atrophied in those areas of the otic capsule in which the otosclerosis has invaded its endosteum. The atrophy of the spiral ligament is found to be most prominent in the middle coil. In thse same areas, atrophy of the stria vascularis with large dilated blood vessels may also be observed (Sando et al 1968).

According to Gussen (1975), the pathological changes affecting the spiral ligament in severe cochlear otosclerosis appear to be:
1. loss of capillaries and pericapillary spaces in the spiral ligament

behind the stria vascularis and 2. erosion or resorption of cochlear capsular bone with an increased vidth of soft tissue endosteum separating the spiral ligament from the bone surface.

Benitez and Schuknecht (1962) in a case report showed that otosclerosis can cause atrophy of the spiral ligament of such magnitude as to result in rupture of the cochlear duct and profound deafness.

The otosclerotic focus involving the endosteum of the otic capsule causes an alteration in the vascularity of the inner ear. (Linthicum 1966). There is normally no link between the vessels of the otic capsule and the membranous labyrinth but otosclerotic vascular bone may lead to an anastomosis with an altered blood supply of to stria (Ruedi 1S65).

Kerth (1967) has observed the following sites of cochlear involvement in patients with cochlear otosclerosis in order of decreasing frequency: basilar turn of the cochlea, remainder of the cochlea, semicircular canals, and internal auditory canal.

Valvassori (1969) recognized three degrees of involvement of cochlea shown by tomography in patients with cochlear otosclerosis: changes limited to the capsule of the basilar turn of the cochlea and widespread changes throughout the labyrinthine capsule.

The atlas of Manasse (1917) illustrates the focus reaching the lumen of the cochlea or vestibulum after traversing the capsule. His description of the process follows: on the external wall of the spiral ligament one observes thick, globular connective tissue prominences; they can protrude far into the Chimen of the cochlear duct. Hydropic degeneration of the spiral ligament is never lacking; the coarse

otosclerotic bone transformation produced obvious distortions of the contours of the cochlea with pathologic changes in the striatum fibro sum of the spiral ligament and stria vascularis. There were extensive degenerative changes in the organ of corti and peripheral cochlear neuron. He concluded that these changes were responsible for the ultimate total loss of cochlear function.

Hallemann and Harril (1967) found otosclerotic involvement of the endosteal layer with regressive changes in the stria vascularis and spiral ligament, such as atrophy, fibrosis, hyalinization; however, abnormal vascular shunts were not a significant feature.

In the entire process of otosclerotic penetration of the cochlea, it remains noteworthy that the contours of the intracochlear space are so widely preserved. Frequently, only some flattening of the concave bony contour is seen, and definite penetration by exostotic protru-berances is restricted largely to the basal turn. The penetrating power peters out somehow; seemingly there has to be capsular bone present to be transformed and without this "nourishment", otosclerosis loses its momentum. This might well be what Brunner called atrophy of the advancing edge.

The inner meatus frequently is a veritable showplace of otoscle-rotic proliferation resulting in coarse alterations of the lumen. Concentric narrowing or excrescences into the lumen impinge on the nerve. Here lies possibly the core of the problem of cochlear deafness.

Otosclerotic destruction involving the internal auditory canal can be divided into three distinct steps: 1. Metaplasia reaches the walls of the canal 2. New bone protrudes into the lumen of the canal

3. The eighth nerve is directly attacked by otosderotic excrescences

1. The canal wall can show considerable masses of the new bone, but, besides the breaking up of the originally smooth contour, the lumen is fully conserved. This form is infrequent as the focus generally penetrates deeper and causes more disorder.

2. The new bone surrounds the canal, narrowing it in a very irregular way. Into the center of a large resorption islands, in a focus around the inner meatus, a spiny exostosis has grown. In other cases of otosclerosis, a non otosclerotic exostosis may grow into the inner meatus. Over the focus located in the wall of the inner meatus the thickened dura engorges producing a 'Schwartze' sign in the inner meatus.

3. Direct attack against the eighth nerve takes different forms. Buckling inward of the newly formed mass cuases deviation, and streth-ching of the fibers. Sharper protrusions causes a more conspicuous bend in the nerve. Finally, spine like protrusions penetrates the trunk. (Kelemen & Linthicum, 1969; and Manasse 1917).

Potential danger to nerves by a sharp bony spurs at their divi-sions (facial and chorda; cochlear and vestibular branches of the 8th nerve, in the inner meatus) was pointed out by Kelemen (1934).

Ruedi (1961,1969) described deposits of endosteal bone in the scala tympani of the basal turn of the cochlea which together with the associated labyrinthine degeneration, he regarded as sequelae of otosclerosis.

Wright & Schuknecht (1972) and Kirchner & Schuknecht (1974) observed the following changes when the otosclerotic focus invaded

endosteum of the otic capsule in patients vith cochlear otosclerosis. There was spiral ligament atrophy. This atrophy usually occurs in th lateral arcs of the cochlear turns. When the focus of otosclerosis reaches the cochlear endosteum, there is flattening of the normally rounded contour of the bony cochlear wall and reduction in spiral ligament width. The external laryer of the the spiral ligament adjacent to the involved bone was replaced by a dense acidophilic homogenous tissue.

Altmann, Kornfeld, and Shea (1966) concluded in their paper that specific sensori-neural hearing loss is encountered in otosclerosis in those cases in which the focus extends into the endosteum of the cochlear scalae. They felt that the damage to the sensori-neural elements is produced by the release of substances into the labyrinthi fluids either by the focus or abnormally functioning stria vascularis

HISTOPATHOLOGY OF COCHLEAR OTOSCLEROSIS

The Normal Otic Capsule:-

An understanding of certain unusual features of the otic (labyrinthine) capsule is necessary for the intepretation of histopatlogic characteristics of otosclerosis and other disorders of petrous pyramids (Paparella et al 1973).

The otic capsule differs from other bones of the long and flat variety in that it is a single osseious case or box which encloses the internal ear. The otic (endolymphatic) and periotic (perilymphatic labyrinths, which occupy the internal ear space, have reached their permanent size about midway in fetal life; hence, the inner layers of the osseious capsule are not subject to the changes which occur in long bones to provide for growth. Increase in bulk of the otic capsule is achieved by addition of periosteal bone to the outer

surfaces (Paparella et al 1973).

The osselous capsule is preceded developmentally by a cartilaginous model which persists until the periotic labyrinth has formed around the otic labyrinth and growth of these structures has been completed except for certain appendages - the otic (endolymphatic) duct, the periotic (perilymphatic) aqueduct, the fissula antefenestra and the fossula post fenestram. Ossification centers, 14 in number appear successively in the capsule from the 16th to the 21st week of fetal life (Bast and Anson, 1949). Vascularization and absorption of cartilage between canals and formation of the subarcuate fossa provides for rapid enlargement of the canals during this period. The posterior and lateral semicricular canals are the last to reach full growth, at about 23 weeks. Ossification of the enchondral layer of the capsule begins at the periosteum.

At the time of puberty, the normal otic capsule consists of 3 layers; the narrow endosteum, the wider enchondral layer; and the periosteal layer.

The endosteal layer:

The endosteal layer of bone, which is a thin dense layer surrounding the periotic space, is formed by the internal perichondrium, which titan becomes the endosteum,. It forms a smooth surface and remains throughout life, limiting further expansion of the periotic space (Paparella et al 1973). (Fig.1&2)

The enchondral layer:-

The absorption of the cartilaginous capsule in the process of ossification of the endochondral (enchondral) layer is incomplete. Irregular spicules of calcified cartilaginous matrix and occasional

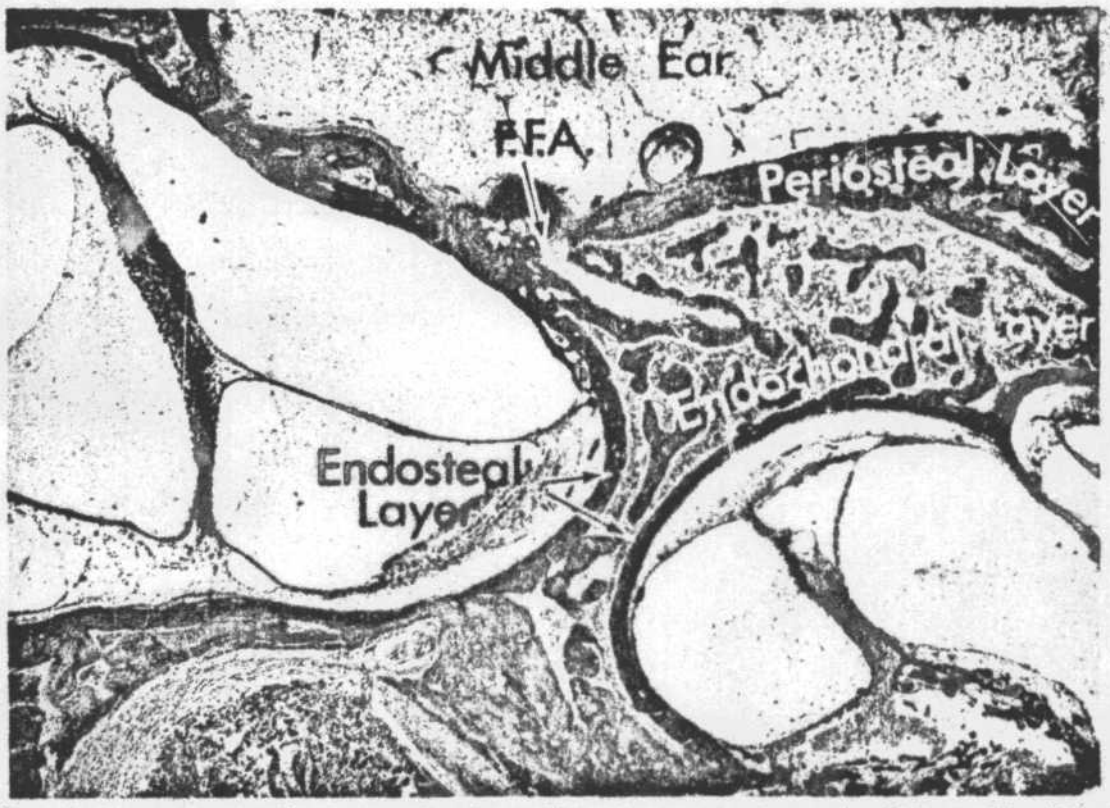

Figure 1. **Photomicrograph illustrating the three layers of the otic capsule in a fetus at about full term. The middle ear contains mesenchymal tissue. The fissula ante fenestram (F.F.A.) at this level leads to the middle ear.**

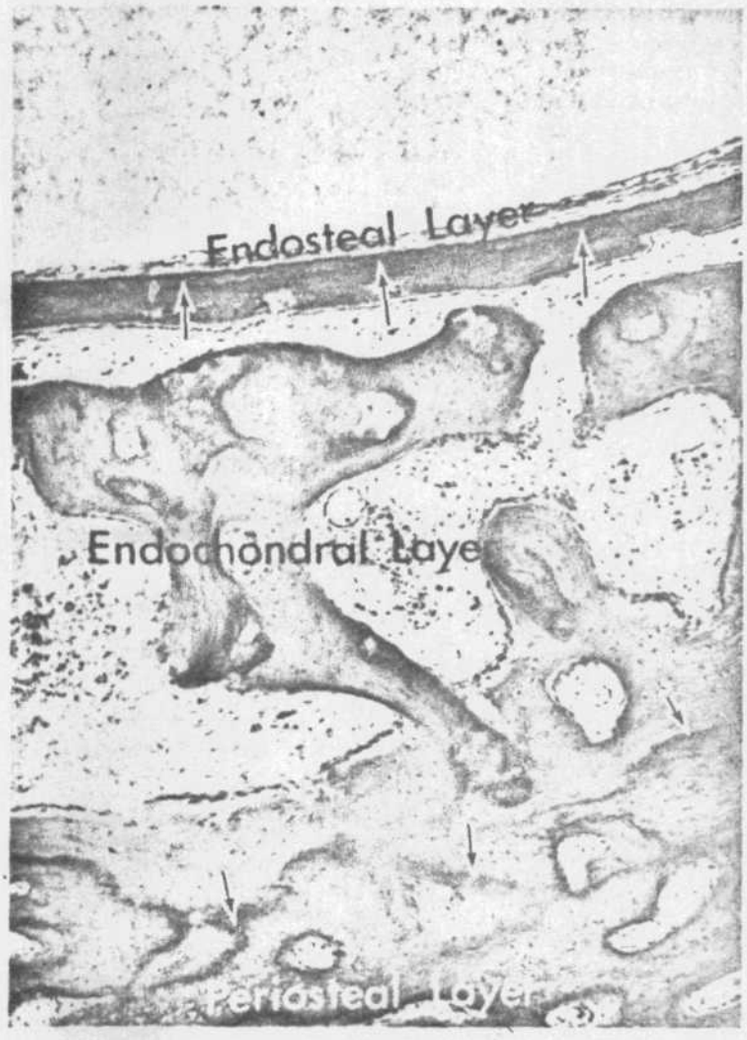

Figure 3 Photomicrograph showing under higher magnification the three layers of the otic capsule of a full term fetus. The junction of endochondral with periosteal and endosteal bone is indicated by arrows. Globuli ossei and interglobular cartilaginous remnants have been included in the endochondral layer.

cartilage cells remain. These spicules have lacunae along their margins where enlarged cartilage cells have bean entered by the capillary buds. Osteobalsts appear and deposit bone in the lacunae, thereby forming small bony globules, termed "globuli ossei" by Manasse (1897). The intervening remaants of calcified cartilaginous matrix and an occasional cartilage cell have been termed "interglobular spaces" by Manasse and "intrachondral bone" by Bast and Anson (1949). These remnants are found throughout life in the human otic capsule and are characteristic of the endochondral layer. A large remnant of cartilage sometimes persists in the cochlear capsule, adjacent to the endosteal layer and may exhibit cartilage cells and many globuli ossei. (Fig. 3)

The ossification centers fuse to form one continuous endochondral layer which is intimately fused with the endosteal and periosteal layers and transmits vascular channels. The layers remain microscopically distinct throughout life.

The primary bone in the lacunae and on the immediate surfaces of the cartilaginous matrix has an interwoven, weblike structure (Meyer 1927) and persists throughout life. On its surface the margins of which are usually delineated by cement lines, new bone is gradually deposited by osteoblasts to fill in the marrow spaces (Paparella et al 1973).

This new endochondral bone is distinctive in that the fibrils are arranged in bundles to give it a skeinlike appearance as contrasted with veblike bone of the globuliossei and the primary periosteal layer or with lamellar haversian bone.

Although the enchondral layer mainly persists throughout life, evidence of resorption and replacement by lamellar bone is seen in so:

areas along vascular channels. The inner wall of the new bone around these vessels stains a deep blue with hematoxylin because of its high mineral content. (Paperalla et al 1973).

The Periosteal layer:-

The third layer of the osseious capsule has alteady begun to appear at about the 21st week of fetal life. It is added to outer surfaces of the cartilaginous capsule by the outer layer of the perichondrlum. (Bast & Anson 1949) which then becomes the periosteum. It adds bulk and strength to the capsule as growth proceeds to the adult stage. Physiological resopption and replacement of membrane bone by lamellar haversian systems occurs to a varying extent in this layer. (Paparella et al 1973).

Histopathological findings:-

The otosclerotic process generally develops in areas of the labyrinthine capsule, in which the embryonic cartilage often perissts.(Fig. The first indication of the disease comes as an enlargement of the perivascular spaces. Bone is resorbed around a vessel and replaced by a cellular fibrous connective tissue. Areas undergoing active resorpt are conspicuous by the presence of numerous osteoclastic giant cells and the unusual amount of vascular channels. (Nager,1969; and Ruedi (1969).

Within the perivascular connective tissue, reticular cells and fibroblasts assume the form of osteoblasts. At the same time calcification begins in the matrix and a new immature plexiform bone is laid down. This bone which consists of much ground substance and few collagen fibers and takes a bluish stain with hematoxylin and eosin, is soon resorbed and replaced by an osseous tissue, with reversed proportion of ground substance to collagen fibers which takes

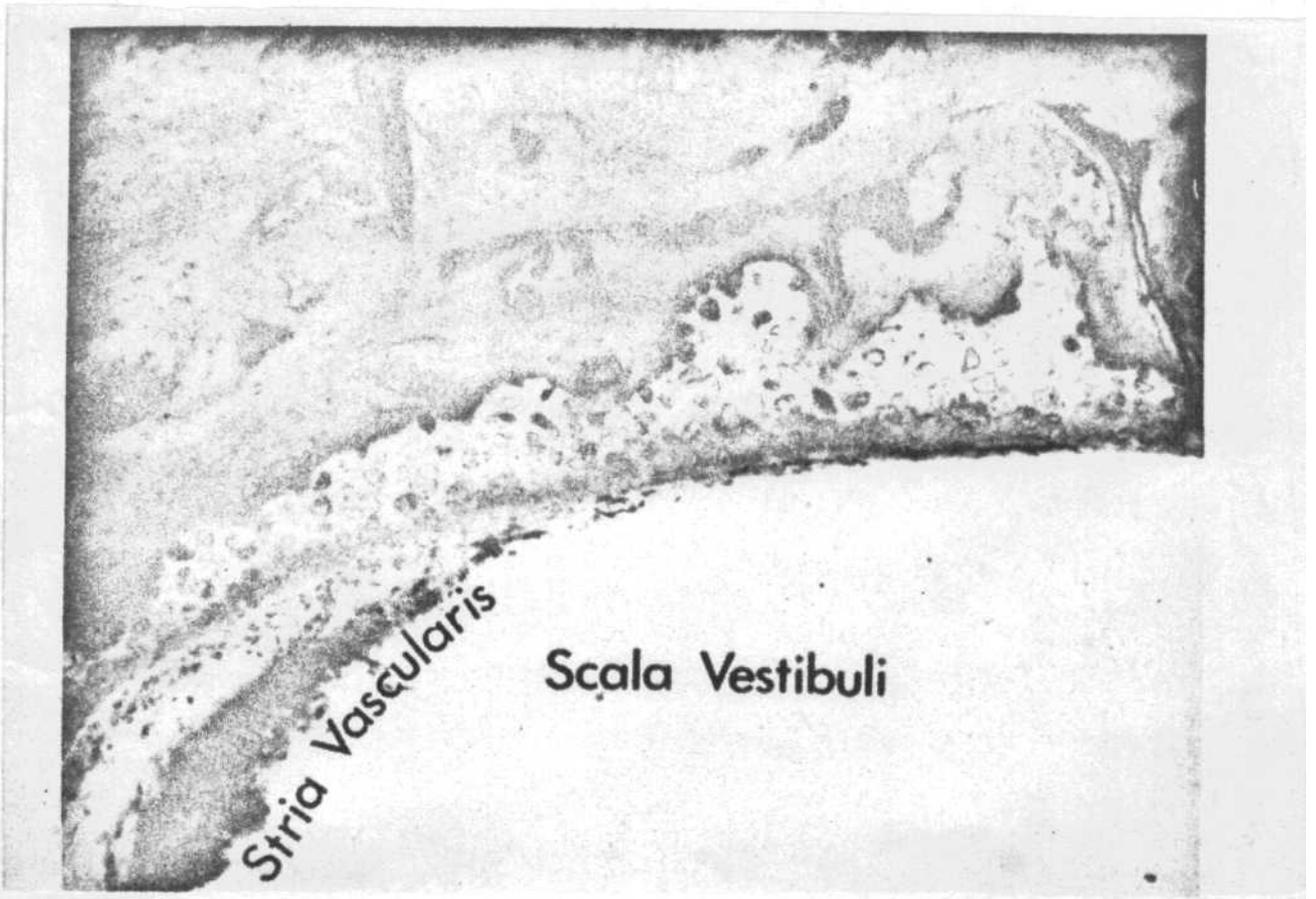

Fig.3: Photomicrograph of a large remnant of cartilage containing "globuli ossei" and cartilage cells bordering on the endosteal layer of the cochlear capsule. (Paparella & Shumrick, 1973)

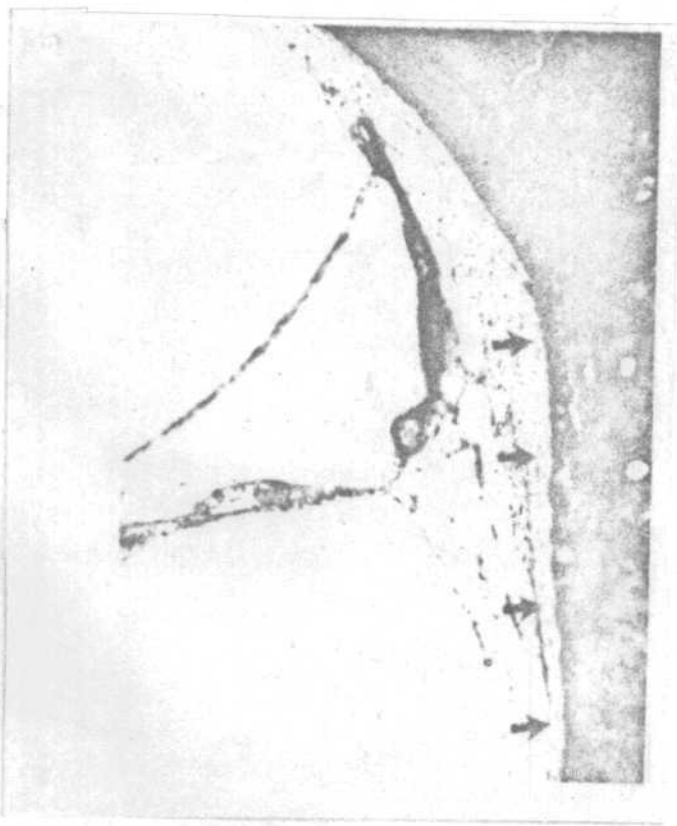

Fig.4: Photomicrograph showing replacement of the endosteal bony layer by otosclerosis bordering the spiral ligament. A seam of osteoid (collagen) containing osteocyt has formed beneath the endoste (Paparella & Shumrick, 1973).

a reddish stain with hematoxylin and eosin (Nager, 1969; & Ruedi 1969

Finally, lamellar bone may be laid down. The process of resorption and replacement goes on irregularly within a focus, with occasion extension to the original bone of the labyrinthine capsule(Fig 5). Succeeding generations of bone therefore be laid down within the lesion in an irregular manner. As some regions of the lesion become more sclerosed, the resulting osseious tissue gets a mosaic-like appearance Deposition of new bone occurs along the margins of the original ossei tissue in the form of a gradually increasing, irregular network of trabeculae. As the otosclerotic focus appproaches the middle ear surface, the overlying mucoperiosteum increases in thickness and becomes more vascular. The vascular hyperemia, in the superficial layer of the focus and in the overlying mucoperiosteum ends a pinkish blue to a large and actively growing otosclerotic process, as seen through a translucent eardrum (Nager 1969; Ruedi 1969).

Otosclerotic lesions may be characterized as active or inactive, depending on their developmental stage and growth tendency. Active lesions can be recognized by their spongy structure and immaturity of osseious tissue, by the extent and size of the marrow spaces that contain a very cellular reactive tissue together with numerous osteoclastic giant cells, and above all by the large number of dilated vascular channels. Inactive lesions which reppresent the end stage of the otosclerotic bone transofrmation process are identified by solid lamellar mosaic-like osseious tissue, which contains few and small blood vessels.(Fig.5) Active lesions consequently are crumbly, bleeding easily and profusely, whereas inactive lesions are hard and poorly vascularized. (Nager 1969).

Lesions located elsewhere in the otic capsulederive their blood principally from the vascular network within the capsule, and secondary

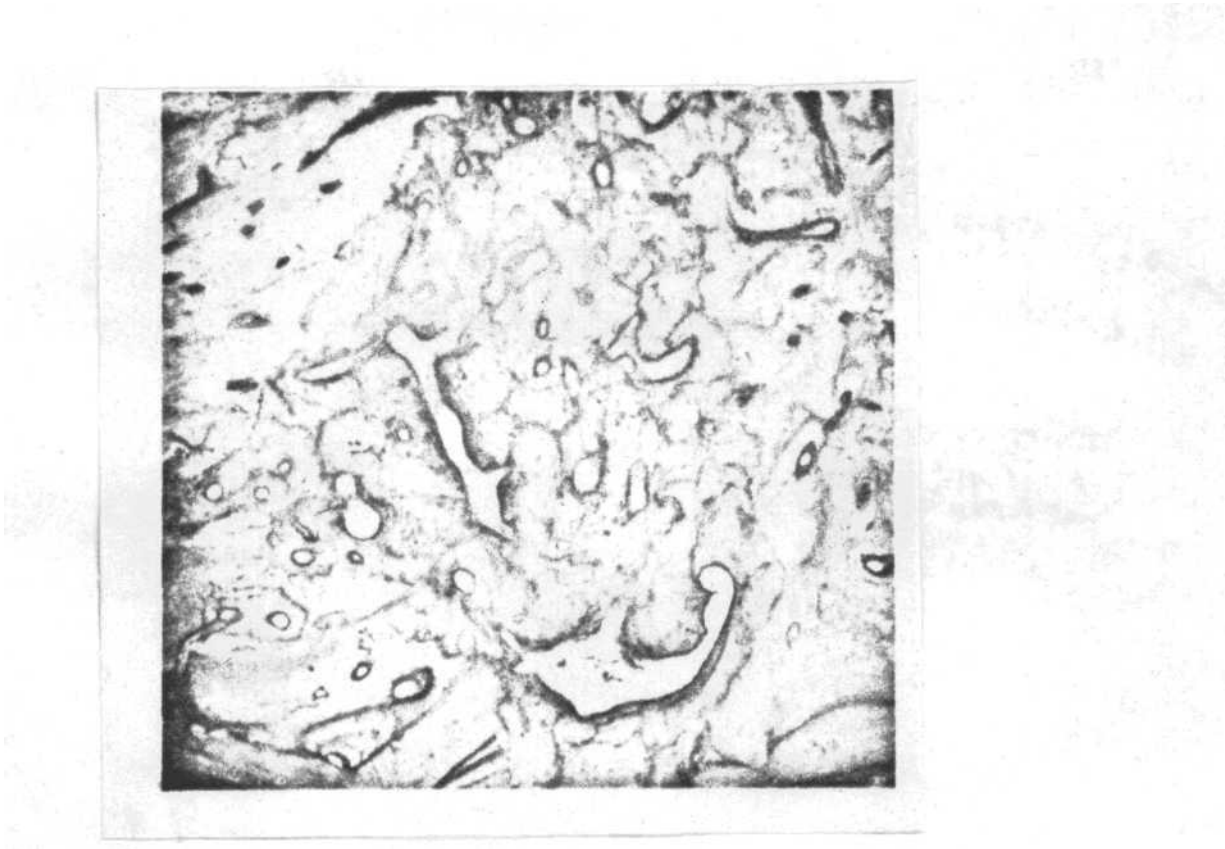

Fig. 5: Photomicrograph of an isolated sclerotic area of nonclinical otosclerosis in the cochlear capsule inferiorly. Dark-staining cement lines outlining areas of resorption and replacement have created a mosaic pattern (Paparella & Shumrick 1973).

ly to a varying extent, from vessels in the adjacent mucoperiosteum and dura. The venous drainage generally follows the arterial supply whereby the superficial petrosal and superior tympanic veins drain the major portions of venous blood from the lseion at the oval window (Nager 1969).

Kelemen and Linthicum (1969) observed the following changes in the cochlear capsule based on the histopathologic study of the tempor bones of patients with cochlear otosclerosis. Conversion of the capsilar bone to otosclerosis has been studied in detail; particularly as it relates to functional changes that might occur when the suspension mechanism of the basilar ligament is distorted.

The otosclerotic focus advances by sheer physical force. At the inner surface of the bony capsule, this shows in the form of cracks. The endosteum is burst open by the mass coming from the periphery. The endosteum may remain intact but a crack appears in the endosteal bony layer indicating the approaching focus. These fissures are not to be confused with the crack around the otosclerotic focus which deli its it from the original capsular bone. This is caused by different contraction coefficients in the fixation fluid, due to the dif. chemical composition of the two kinds of bone.

The pressure will manifest itself as originating with finger like processes protruding from the main mass of the focus, singly or in groups. The contour of the still intact capsular bone may be seen bulging before the pressure of the approaching focus. A protruding otosclerotic finger may proceed bulkier parts of the focus, albeit the latter has already partially replaced the endosteum and has initiated destruction (Kelemen & Linthicum 1969).

Considering the localization of the always sharp boundary between

the original bone and the focus progressing toward the cochlear lumen, one has to distinguish between comparatively comparatively mute areas, lying outside the area of attachment of spiral ligament and functionally significant ones covering partially or totally the area behind the latter.

Contour changes within the cochlea varies markedly from mild flattening of the concave walls to gross alteration with narrowing of the lumen and the occurrence of protruberances in the foam of exostoses. The concave contour may assume an angulated shape. Angulations may become critical when developed behind the anchorage of the basilar membrane. A high degree of deformity transformed the scala tympani into a funnel-shaped space. In some instances, the entire wall seems to advance, while nearby the protrusion assumes the character of an exostosis. (Kelemen and Linthicum 1969).

Assault agains t the cochlea comes from the center and the periphery: the former leads directly into the modiolus and proceeds from the direction of the cribiform plate or from the interscalar septa (Newby 1958). Fowler (1949) illustrated extension into the septum between the basal and middle turn. Altmann (1962) thought that ingrow of otosclerosis into the septa particularly between the basal and midd turn interferes with the vascular supply of the cochlea.

The newly established otosclerotic wall of the scalae may respect the original contour, but rather than smooth, is wavy in a high or moderate degree. Resorption cavities inside the focus are by a fibrous netwrok frequently reach the inner surface, either in a "silent", "mut region or over the spiral ligament. An entire system of similar spaces may form in series (Kelemen & Linthicum 1969).

Replacement of the endosteum is the last step in the progress of the focus against the cochlear Limen. This process follows an essenti lly regular course. But it must be emphasized that pressure exerted

by the advancing new bone established within the external layers of the capsule before the endosteum is reached may already manifest itself within the latter by the appearance of cracks (Kelemen & Linthicum 1969).

The most conspicuous phenomenon of replacement is a sequence which may be called "fragmentation". The endosteal layer is broken up into small particles and the latter form a rosary (row of beads) before disappearing definitely under the advancing otosclerotic mass (Ruedi 1969; Siebenmann 1899; Firedmann 1974). The particles may assume the form of vacuoles. The same may be observed where the otosclerotic mass surrounds a semicircular canal. Sometimes it is hard to decide whether this margin belongs to the transformed endosteal layer or to the invading bone. In the latter case, the much wider meshes of the otosclerotic bone must have changed into smaller and remarkably uniform vacuoles. So interpreted the rosary represents the initial form of resprption of the endosteal layer before definite replacement (Chevance 1962).

The particles of the mosaic layer remain until their disappearance at the area of contact with the terminal layers of the fibers of the spiral ligament. When this phase ends attachment of the spiral ligament to the concavity of the capsular wall will never be reestablished in its original firmness.

This transformation of the attachment of the spiral ligament and with it of the basilar membrane into otosclerotic bone is of prime importance.

Neubert (1960) summarized the mechanism of the peripheral anchoring in the following way: The attachment of the basilar membrane is secured by the connective tissue wedge of the spiral ligament,

which is divided according to the structure of its cellular-Fibrous material into 3 zones not sharply delimited against each other. The inner portion jutting out wedge like, contains as filling mass the fanlike deploying basilar fibers. In the median layer, inter-woven and anastomosing fibers create a honeycomb network with meshes which become richer in fluid the more they approximate the top of the cochlea. The thiid border-layer follows outward, as a compara-tively narrow, dense plate rich in cells; its network forming a vertical lattice. The adjacent periosteum is distinctly thicker, in the extent of the spiral ligament, its fiber bundles meet in acute angles, crossing and penetrating each other and run generally, in longitudinal direction along the cochlear canal. Differences in the firmness of the single constitutents at the borders of the divi-sions guarantee deformability of the spiral ligament.

This deformability, a shifting of the planes along each other offers ample explanation for changes incurred by the approach of the otosclerotic mass (Neubert 1960).

Contact of the otosclerotic foucs with the external wall of the cochlear turns is a candinal point for potential damage through influence exerted on the anchoring system of the basilar membrane. Goodhill (1961) emphasized that massive intracochlear lesions inter-fere with proper basilar membrane dynamics.

Held (1926) emphasized the importance of conditions in the regi of the spiral ligament and explained how tension of the basilar membrane depends on its anchoring in the spiral ligament.

Gussen (1975) observed the spiral ligament changes occuring predominantly adjacent to the stria vascularis and consisting of a decrease or loss of the capillary-perilymph system.(Fig 6). It is suggested

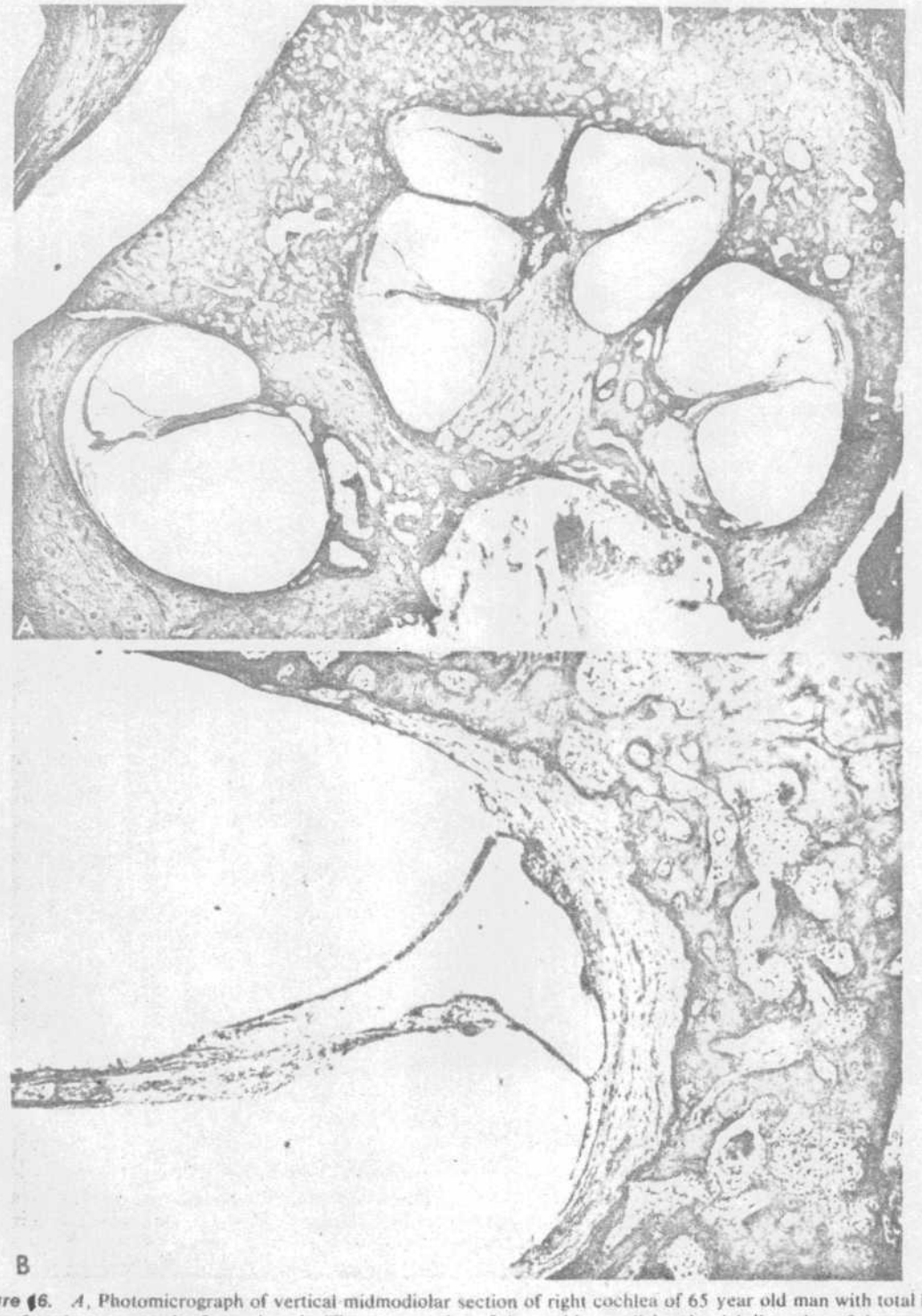

Figure 16. *A*, Photomicrograph of vertical midmodiolar section of right cochlea of 65 year old man with total loss of cochlear function as a result of otosclerosis. The compression of the cochlea, radial and axial distortion of the modiolus, and changes in the spiral ligament were induced by the otosclerotic process. *B*, A higher magnification of the upper middle turn shows distortion of the basilar membrane, partial degeneration of the stria vascularis, and reactive changes in the deeper layer of the spiral ligament and endosteum overlying the otosclerotic process. (Courtesy of Dr. George Nager, Johns Hopkins University School of Medicine.)

that this results in increased concentrations of metabolic products in the cochlear fluids that can no longer be absorbed effectively.

The function of the caplllary-pericapilary space system of th spiral ligament is not clear; however most authors consider that it is characteristic of tissue activity involved infiltration or resor tion of fluid. Electron microscopy studies have demonstrated minutes openings in the lining of the spiral ligament facing the scala vesting buli and scala tympani; so that it is assumed that the spiral ligament spaces contain perilymph.

Shambaugh (1959) noted formation of a thin layer of amorphous collagen material beneath the adjacent connective tissue after otosclerosis had absorbed the endosteal layer of the bony capsule.

Manasse (1922) explained how the walls of the cochlear duct undergo hydropic degeneration at the spiral ligament, the coarse connective band lining the concavity of the capsule becomes permeate by many partly rhombic spaces, which are either empty or show finely pulverized or mucous content.

The otosclerotic tissue itself may go thorough a retrograde step when approaching the cochlear space. According to Brunner (1952) the otosclerotic bone approaching the endosteum frequently becomes atrophic.

Hildyard et al (1972) were able to study the histopathological findings in the temporal bone of a case with far-advanced otoscle-rosis. In the inner ear some interesting observations were made. Acellularity of the spiral ligament was seen in the area where the focus invaded to the endosteal layer of the otic capsule. In addi-tion, in the same area, an eosinophilic collagenous tissue layer

was present in the spiral ligament next to the focus.(Fig.7). Atrophy of the stria vascularis was also observed in these particular areas. Sporadic outer hair cell loss was found in portions of all 3 turns of the cochlea and appears to correspond with the areas of oto-sclerotic foci invading the endosteal layer. Partial ganglion cell loss in all cochlear turns was also present.

Ruedi (1969) described the changes in the circulatory pattern when the otosclerotic foci invaded the endosteum of the otic capsule New vessels, so called shunts develop between the vascular spaces of these active otosclerotic foci, penetrating through the endosteum and the capillaries of the membranous labyrinth. The se shunts are present in the region of the spiral ligament in different turns of the cochlea. Similar shunts have been found in the region of the basal scala tympani, near the round window. Thus the normally separate vascular systems of the membranous labyrinth and the bony otic capsule communicate with each other at these points. It would appear, that the vascular, actively growing focus of oto-sclerosis needs further outlets for venous drainage. These newly formed shunts mainly serve to carry venous blood from the congested vascular spaces of the otosclerotic focus into the venous capillaries of the spiral ligament. The otosclerotic focus is breaking across ti endosteum and the blood from it is being carried into the normally narrow vessels of the membranous labyrinth giving rise to congestion resulting in a wide dilatition of the labyrinthine vessels. The effect of the congestive pressure is most marked in the bony spaces of the modiolus and within the small vascular canals runnint through the bony otic capsule into the sinuses of the durameter. As a result of thispressure, the bony canals are widened by osteoclasts and the vessels become dilated to provide adequate venous drainage

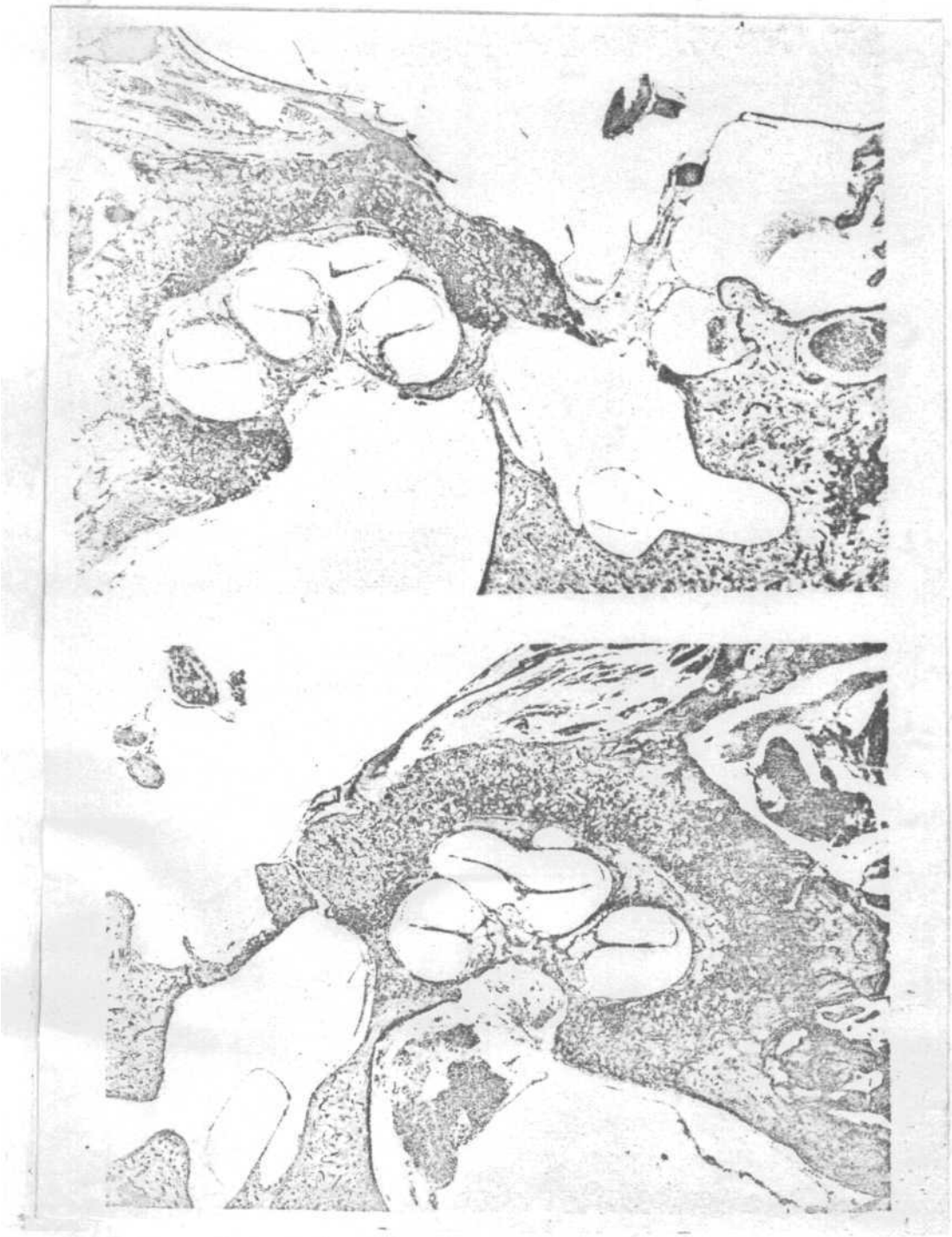

Fig.&7: Bilateral diffusle otosclerosis of the otic capsule (By courtesy of Dr.H.F.Schuknecht)

from the membranous labyrinth.

Consequences of venous congestion within the inner ear:- (Ruedi 1969) In the region of the spiral ligament and in the basa scala tympani, the congestion might produce a deposit of the lamell bone. The newly produced osteoid tissue forms a red band inside of the endosteum. Such a narrow layer of lammellar bone at the attachment of the spiral ligament is hardly of functional importance. On the other hand, a massive deposit of lamellar bone within the scala tympani is very probably also caused by venous congestion The obstruction of the cochlear lumen leads to a significant interference with sound conduction within the inner ear. But this is an uncommon occurrence.

Much more important from the functional point of view is the effect of congestion of the membranous labyrinthine veins on the neuroepithelial elements of the cochlea.

Penetration of the cochlear endosteum can be achieved not only by the two components of the otosclerotic focus, the spongiotic and sclerotic, but also by a third, the fibrous-grannulomatous mass which is surrounded by the two bony portions (Kelemen & Alonso 1980) This mass is not merely filler material; its destruction and penetrating power equals that of the two bony components.

The space occupied by this mass can be extensive comprising a considerable part of the focus. This space has been identified by various names, such as fissula antefenestram and post fenestram; resorption foci; vascular space and acellular space. When the third component surrounds the corner of the stapedial footplate, it gives rise to what is called a "loose stapes" or a "floating footplate" (kelemen and Alonso 1980).

Attention has been frequently directed to the role of the vascular stria changes in the stria vascularis caused by the approach ing focus as a possible source of lesions in the organ of corti, vere mentioned by Rutten, Mayer (1917), Lange (1926), Wolff (1950) and Ruedi (1964).

Manasse (1912) saw at the lower end of the stria thick, round, fibrous prominences of the dubious vascular origin, occasionally jutting far into the Inmen of the cochlear duct.

The inner meatus frequently is a veritable showplace of otosclerotic proliferation resulting in corrse alterations of the lumen.(Fig 8) Concentric narrowing or excrescences into the lumen impinge on the nerve. Here lies possibly the core of the problem of cochlea deafness. Manasse (1917) considered the constriction of the lumen of the inner meatus by otosclerotic bone a serious influence on the function of the eighth nerve. The normally smooth wall of the inner meatus becomes frayed, torn up; over the otosclerotic product the dura is engorged, producing a "schwartze sign" in the inner meatus.

The fundus of the inner acoustic meatus is surrounded by several layers of periosteal lamellae; the enchondral layer follows the contour of the periosteal sheath while an endosteum-continuation here of the dural lining of the meatus is hardly discernible. It is possble that the lack of it is responsible for the exuberant proliferation of otosclerotic excrescences in this location.

In the entire process of otosclerotic penetration of the cochlea it remains noteworthy that the contours of the intracochlear space are so widely preserved. Frequently only some flattening of the concave bony contour is seen, and definite penetration by exostotic protruberances is restricted largely to the basal turn.(Lawrence 196

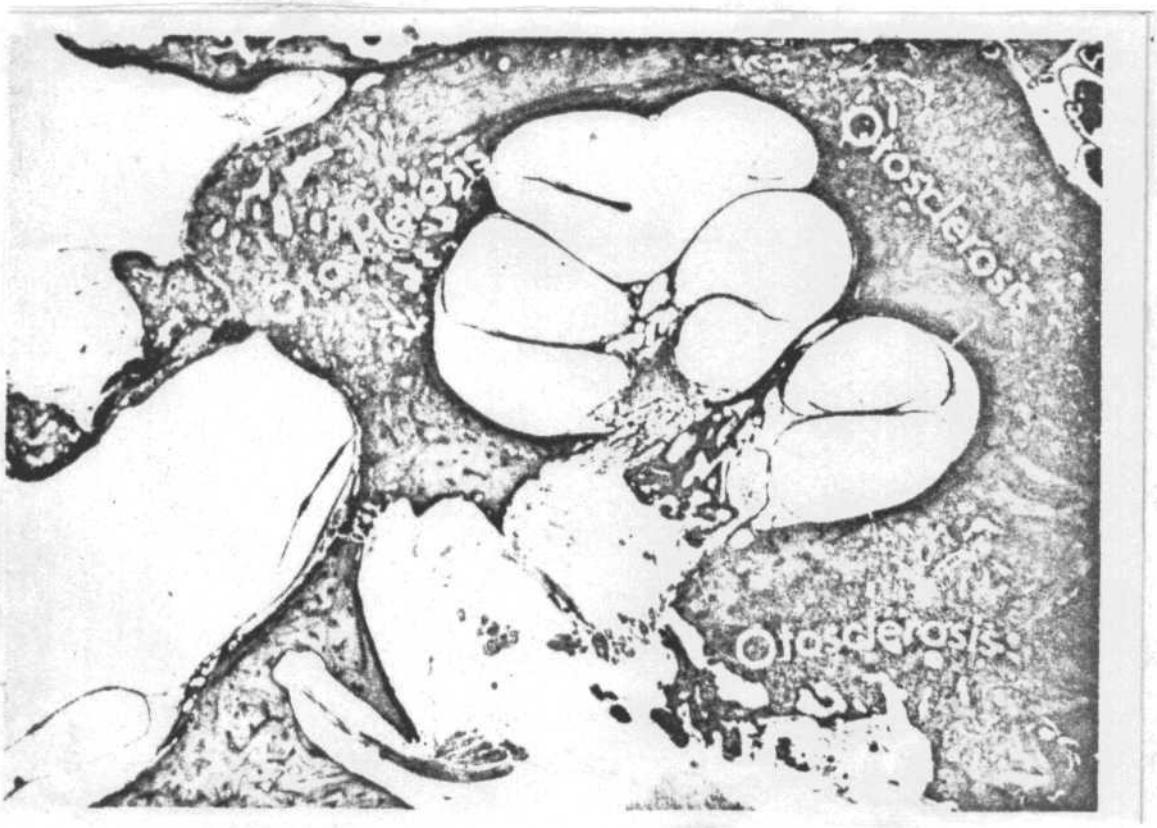

Fig. 8: Photomicrograph showing three foci of otosclerosis located at the oval window, including the stapes, the anterior inner end of the internal meatus, and the anterolateral wall of the cochlear capsule (Paparella & Shumrick, 1973).

CHAPTER-IV

CLINICAL FEATURES

The patient with cochlear otosclerosis complains of a progressed hearing loss, vhich begins in early adult life and which is usually bilateral and symmetrical. Progression is normally slow and may remai stationary for quite long periods of time.

- There is usually a dominant family history (70%) and relatives, may have proven stapedial disease (12%).

- The deafness may have started or increased during preganncy or following the use of oral contraceptives (29%) or there may be a Schwartze sign (8%).

- Tinnitus is encountered especially in the older age group with combined otosclerosis and in those with an early age of onset and cochlear involvement. In pure cochlear otosclerosis arising in pregnancy, it can be the presenting symptom (Morrison 1979).

- Transient episodes of vertigo, often positional occur in 20% of patients with pure cochlear otosclerosis, while the symptoms of hydrops are found in 6%.

Garcia (1972) found greater percentage of caloric abnormalities in those ears with cochlear involvement of the otosclerotic process than the ears with a purely conductive loss. He also found vestibular symptoms to be associated with cochlear otosclerosis. Freeman (1979) also observed vestibular symptoms in 29 (54.7%) out of 53 cases of cochlear otosclerosis.

The occasional finding of labyrinthine hydrops and of degenerative changes in the supporting structure and neuroepithelium of the vestibul labyrinth (Altmann, and Komfeld, 1965); Bretlau and Jorgensen 1968; Johnson and Hawkins 1973) helps to explain the relative frequency and

diversity of vestibular symptoms in patients with otosclerosis, whether stapedial, combined or pure cochlear.

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CHAPTER-V

DIAGNOSIS & DIFFERENTIAL DIAGNOSIS OF COCHLEAR OTOSCLEROSIS

A. Diagnosis:-

Pure cochlear otosclerosis is slowly becoming recognized as a serious cause of sensori-neural hearing loss comparatively early in adult life. Many cases of hearing loss previously ascribed to hereditary progressive snesori-neural loss or presbycusis are in fact due to cochlear otosclerosis without stapes fixation. The importance of correct identification of cochlear otosclerosis is stressed by the possibility that fluoride treatment may effectively halt the advance (Linthicum 1972).

The diagnosis of stapedial otosclerosis and of stapefial combined with cochlear otosclerosis is easily made from the history and hearing tests. The diagnosis of pure cochlear otosclerosis producing a sensorineural hearing loss without a conductive component is a more difficult problem.

At least part of the difficulty in the diagnosis of pure cochlear otosclerosis is due to the fact that the pure tone audiogram for the resulting hearing loss which is sensori-neural does not fit into a consistent definitive pattern. The problem is further complicated by the fact that the pure tone audiometric configuration in cochlear otosclerosis frequently mimic those produced by other sensorineural involvements (Carhart 1963).

The diagnosis of cochlear otosclerosis has been made possible by Case History, Auditory tests, Radiologic Investigations and Histopathological studies.

1. Case History:-

A detailed history is the first step in examination, and this

should include enquiry into the age of onset of deafness, its rate of progression, the degree of social and occupational handicap, if relevant the influence of pregnancy, the presence or absence of tinnitus or vertigo and the presence of paracusis. Paracusis is the phenomenon of bearing better in thepresence of background noise.

Past history of ear disease, of head or blast injury, of exposure to noise, of ototoxic drags administered, of bone or joint disease, and of previous illnesses may be relevant to the differential diagnosis and for management

A detailed family history is desirable and invaluable in assessing the prognosis when relatives are affected. It is found that 70% of the cases with cochlear otosclerosis have a dominant family history and 12% of their relatives have proven stapedial disease (Morrison 1969).

2. Auditory Tests:-

The tests comprise a battery designed to provide qualitative as well as quantitative information regarding the functioning of the peripheral auditory system.

In addition to conventional pure tone audiometry and speech audiometry, the battery includes specialized hearing tests like tone decay test, the SISI, Bekesy Audiometry, the ABLB and Impedance audiometry.

a. Pure tone Audiometry:-

It is a measure of the hearing at different frequencies (250-8000 Hz) of air conducted (AC) and bone conducted (BC) sounds. AC sounds are delivered to an earphone placed on the patient's test ear; and BC sounds are delivered by a bone vibrator placed

usually over the mastoid bone of the test ear. The patient indicates the level at which the sound becomes just audible. This is plotted for all frequencies on a graph called the audiogram. The difference between the AC and BC responses indicates the conductive loss - BC response is a measure of the cochlear (sensory) and neural functions and therefore reflects the sensori-neural level of hearing.

The pure tone audiogram in cochlear otoscelerosis varies considerably from one patient to another. Indeed Carhart (1963) has emphasized the fact that "otosclerosis induces highly varied lesions which can, in turn produce diverse auditory disturbances".

Based on the configuration of the sensorineural component, Carhart (1963) categorised the different patterns of audiograms of patients exhibiting cochlear otosclerosis:

Flat: 500-4000 cps variation of 10 db or less

Gradual falling: Drop between 500 and 4000 cps of greater than 10 db but less than 30 db.

Sharp drop off: Greater than 20 db drop in an octave without return to higher level

Marked falling: Drop between 500 & 4000 cps of 30 db or higher

Notch: Dip greater than 15 db deep and 4 octaves or less in width

Trough: Dip greater than 10 db deep and greater than 4 octaves wide

Low frequency drop: 30 db or more between levels with low frequencies poorer.

Carhart (1966) found 12% with flat or low tone impairments, 31% with notched losses, 15% with trough shaped audiogram and 37% with high frequency deafness.

In Sanders' (1965) patients, the category labeled "marked

falling" accounted for the largest number of ears, with the "gradual falling" and "notch" patterns tied for second in frequency of occurrence. The "low frequency drop" accounted for the fewest number of ears.

Shambaugh (1969) has given a lucid and frequently quoted analysis of the subcategories which he feels characterize sensori-neura loss due to cochlear otosclerosis. He states, three principal varieties of cochlear involvement are encountered in patients with cochlear otosclerosis, a high tone loss resembling the perceptive loss usually seen in presbycusis ...., a diffuse cochlear loss producing a flat bone loss at all frequencies ...., a loealized cochlea loss in the middle frequencies producing a deep notch or valley in both the air and b/c audiograms.

It is also found that pure cochlear otosclerosis is characterized by progressive sensorineural loss usually bilateral and symmetrical.

Kelemen and Linthicum (1969) tried to correlate the area of the cochlea involved by otosclerosis and the amount of bearing loss aid the configuration of the audiogram. No pattern could be ascertained that produced a characteristic audiometric curve. They found, that if the basal coil of the cochlea, plus one other coil or the internal auditory coil was involved by the otosclerosis, there was apt to be a sensori-neural loss.

Thus, we find that varied configurations occur in substantial proportions in consequence of cochlear otosclerosis. This fact leads to the generalization that cochlear otosclerosis cannot be identified on the basis of one or two typical audiometric contours. Instead the clinician is faced with the prospect that almost any

audiometric pattern of the sensorineural variety may bespeak of cochlear otosclerosis.

The audiometric effect which results when stapes fixation and cochlear otosclerosis combine:-

When stapedial fixation produces a definite hearing loss, the familiar air-bone gap appears, with the a/c curve representing the sum of hearing losses produced independently by the stapedial fixation and by cochlear lesion. The bone curve now no longer depicts accurately the bearing deficit due to cochlear otosclerosis. Instead the bone curve suffers a mechanical distortion, epitomized by so-called Carhart notch, due to the fenestral obstruction.

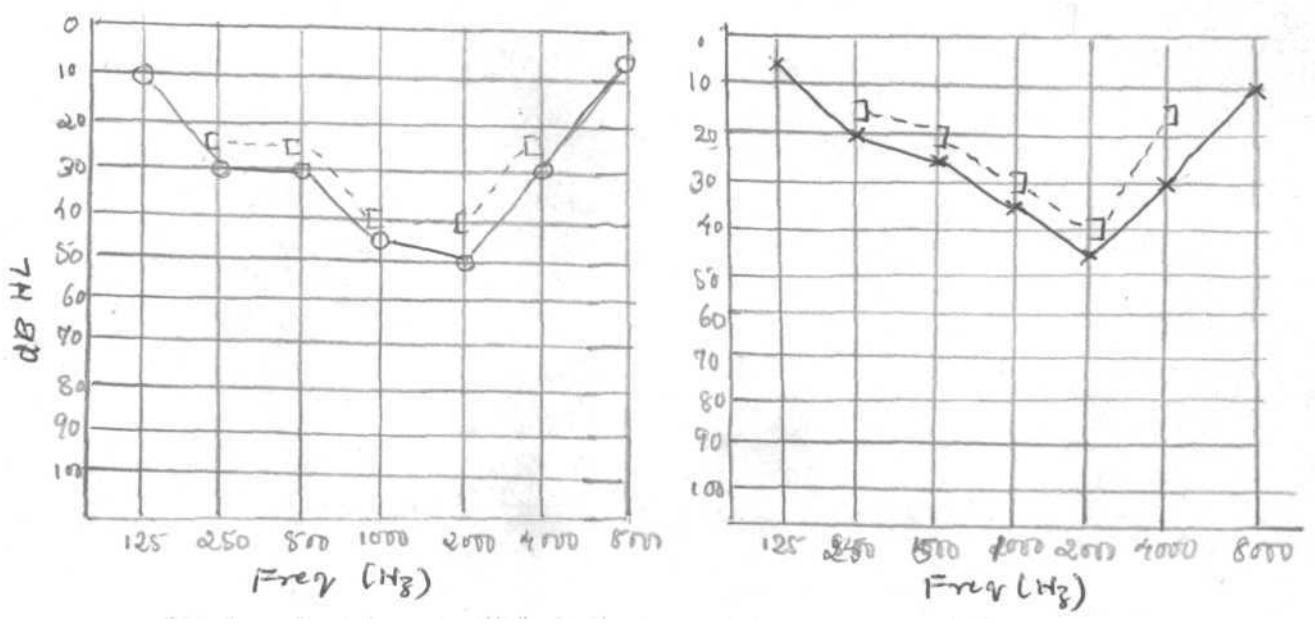
Pure tone audiograms of patients with cochlear otosclerosis and combined otosclerosis (stapedial plus cochlear) are illustrated in the next page.

b. Speech Audiometry:-

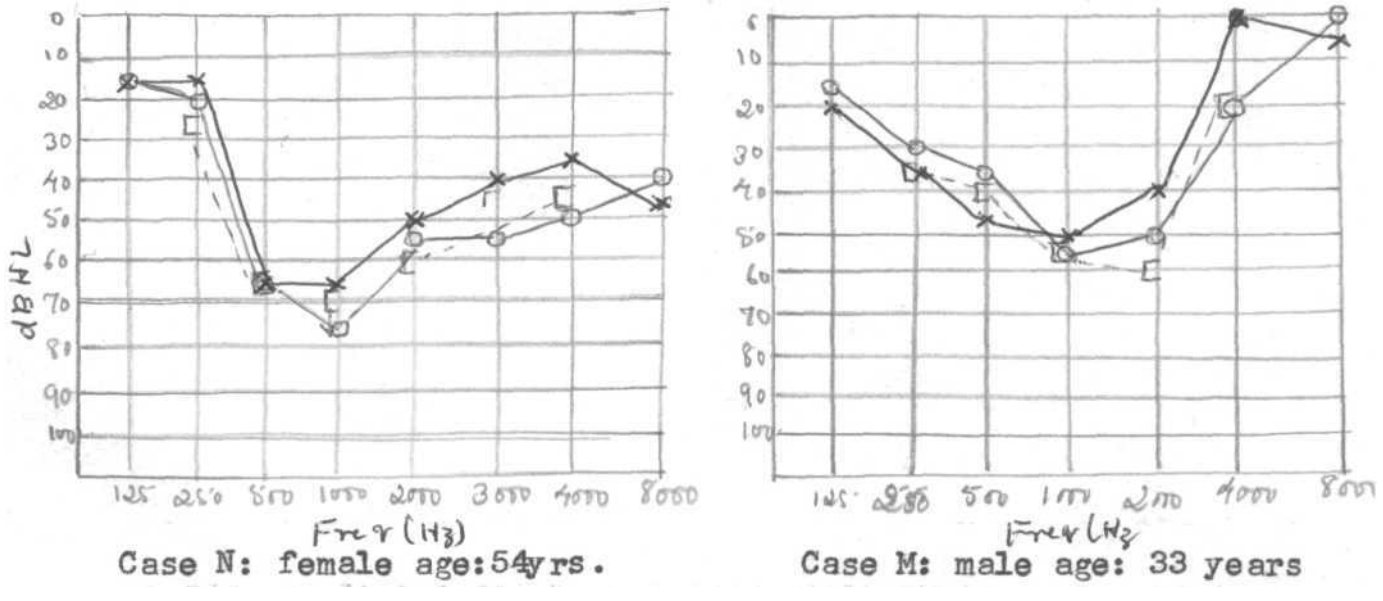
The routinely used speech tests are SRT and speech discriminati score. Speech reception threshold (SRT) is a level at which 50% of all double syllable or spondee, words (eg. baseball, doormat) are heard and repeated by the patient. The SRT level can also be obtain by averaging the pure tone hearing loss at 500, 1000 and 3000 Hz or by averaging the two best responses at these frequencies.

In the speech discrimination score test, phonetically balanced (PB) monosyllable words are delivered to the test ear at 40 db above the patient's SRT. The percentage of words the patient can repeat correctly is expressed as PB-max. The normal range is 90 to percent. This test can also be done with intensities lower than 40 db above the patient's SRT.,and then it is referred to as, for eg. PB score 30, if the sound intensity used was 30 db above the

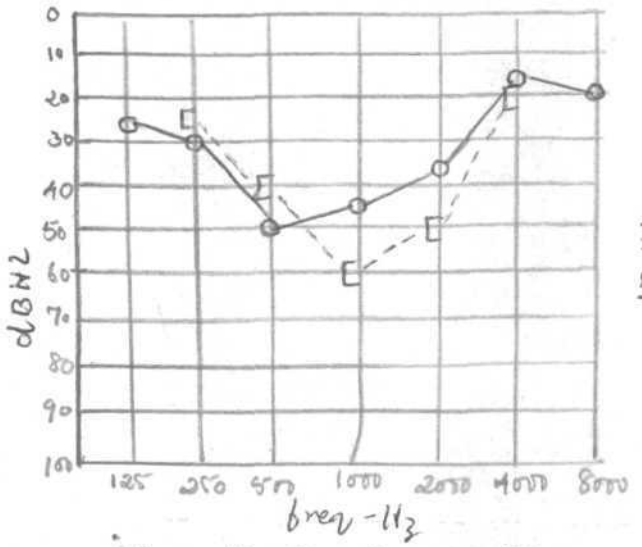
Pure tone audiograms lillustrating? cochlear otosclerosis:



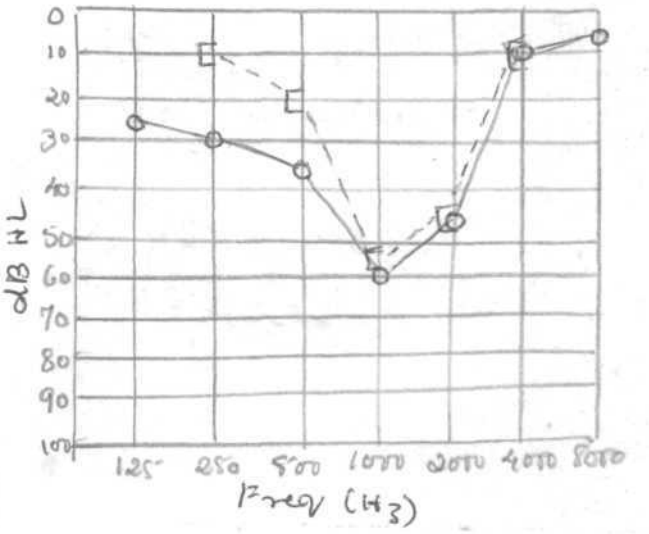
Bilateral pure sensori-neural loss with 'cookie-bite' aspect in pure tone audiogram (Causse & Beziers 1974)



Audiograms of two cochlear otosclerosis illustrating notched (Case N) and trough-shaped (case M) configurations (Carhart & Evanston, 1966).

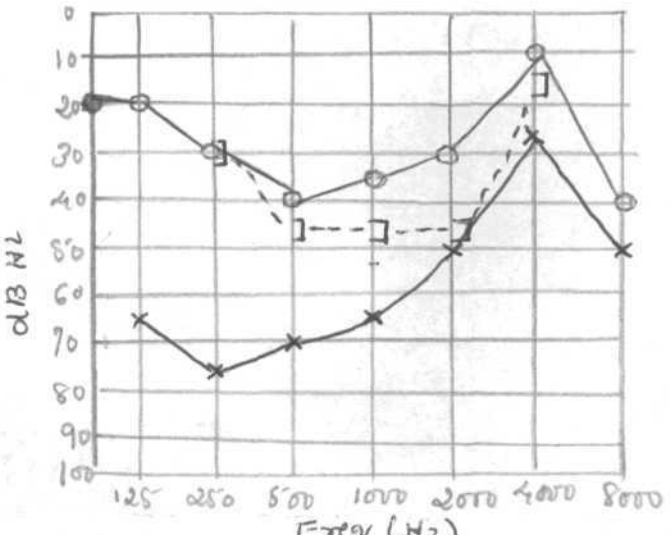


Case M: female age:51years



Case N: female age:35years

Examples of cochlear otosclerosis combined with miner conductive involvements which in themselves would be considered preclinical (Carhart & Evanston 1966).



Case D: female age: 27years

Audiogram of the left ear of a patient combining cochlear & stapes fixations. Curve A - composite a/c loss; Curve B- b/c loss; Curve C- configuration of cochlear otosclerosis estimated by corrective curve B for Carhart notch. The A-C gap is the corrected estimate of the conductive impairment. (Carhart & Evanston 1966).

patient's SRT. These simple speech tests are of diagnostic and therapeutic value.

In pure cochlear otosclerosis, speech discrimination scores of 80-90% are encountered with moderate deafness, of 50-80% with more severe changes and 0-30% with subtotal loss (Shambaugh 1966; Saunders 1965; Carhart 1966; Sheehy 1962; Causse et al 1977; Linthicum 1972).

Poor speech discrimination score is reported in cases of combined otosclerosis depending upon the degree of conductive and sensori-neural invoivement.

SPECIALISED HEARING TESTS

Tone Dacay Test:- (TDT)

It is very useful for detecting retrocochlear pathology - eg. tumour of the eighth nerve. Patients having retrocochlear pathology have been observed to have abnormal adaptation. Adaptation is decrease of response to a persisting sensory stimulus. This phenomenon is common to all sensory systems. If a stimulus is presented continuously, then there is a decrease in response. When a tone is presented continuously at 5 db sl, normal hearing subjects can bear for at least 1 minute. Patients with retrocochlear pathology fail to hear the tone when the tone is presented continuously at 5 db sl for 1 minute. They need intensity increments for continuous audibility. Tone decay tests are based on this principle. The tone decay is the amount by which the intensity has to be increased in order to make the tone remain audible.

In pure cochlear otosclerosis, the results are those of end organ disease. There is moderate tone decay of upto 20 db at the

frequencies involved. In combined otosclerosis, there is a marked tone decay of up to 45 db (Morrison 1979).

The Short Increment Sensitivity Index:- (SISI)

In 1959, Jerger, Shedd & Harford described a test technique designed to differentiate subjects who were able to detect very small amplitude changes, presented periodically, in a pure tone signal. Having observed that many ears with a sensitivity loss due to abnormal cochlear function appear to have extremely keen discrimination for small changes in tonal intensity, these authors defined a relatively simple task for both the examiner and for the subject, which they called the short increment sensitivity index.

The SISI test consists of superimposing brief bursts of 1 db intensity increments on a sustained tone presented at 20 db SL at each frequency to be tested. The test is administered monaural3y through earphones. The patient is instructed to report any "jumps in loudness" he detects while listening to the sustained tone for a period of about 2 minutes. The size of the increment can be 0 db 1 db upto 5 db steps, but the test is scored only on the percentage of 1 db increments, correctly identified by the patient. During the test 20, 1 db increments are presented.

Results of the SISI can be reported as percentages scored at each frequency or they can be graphed on a SISI gram which has a frequency on the absiissa and percentages ascending from 0 to 100%.

Jerger (1962) presented a general discussion of the SISI test as one of the battery of useful techniques in otologic diagnosis He mentioned that this experience indicated scores between 0 and 20% for those with normal hearing, with conductive loss and with eighth

nerve involvement and scares between 60% and 100% for patients with cochlear pathology.

Accoriding to the revised SISI test, the test tone should be administered at 70 db spl or higher if required for audibility. In conductive or mixed deafness, the conductive barrier in db should be added to the 70 db SPL test signal to obtain a positive score.

A high SISI score near 100% at all frequencies, even at 250 and 500 cps has been reported by many investigators in patients with pure cochlear otosclerosis (Shambaugh 1966; Saunders 1965; Carhart 1966; Sheehy 1962; Causse et al 1977).

In combined stapedial and cochlear otosclerosis a low SISI score at some frequencies is reported.

Bqkesy Audiometry:-

In 1947, Bekesy introduced a new audiometer that subsequently gained wide acceptance and application in both research and clinical audiology. The instrument was continuously variable frequency, automatic recording unit. It placed the variables of test tone intensity and test tone duration under the control of the subject. The new instrument permitted the subject to trach his threshold by depressing and releasing a signal key, and allowed the measurement of absolute threshold all along the frequency range instead of at octave and midoctave points.

Bekesy audiometry is presently used for its diagnostic value in determining site of lesion as well as for threshold measurement. In the majority of papers concerned with the clinical application of Bekesy audiometry, measurement and description have been confine' almost exclusively to the width or amplitude of the audiometric

tracing. The distance or width may be expressed either in dbs or in number of threshold crossings over a given frequency span. It was observed that the width of the tracing differed for normals and for certain pathological ears.

The frequency range is from 100-10,000 Hz or 250-8000 Hz. The frequencies can be presented in two ways. We can present continuous variable frequency. It varies at 1 octave/min. It takes 7 min. to complete the frequency range 100-10,000 Hz. The frequency can be varied in ascending or descending order. This continuous variable frequency recording is known as sweep frequency tracing.

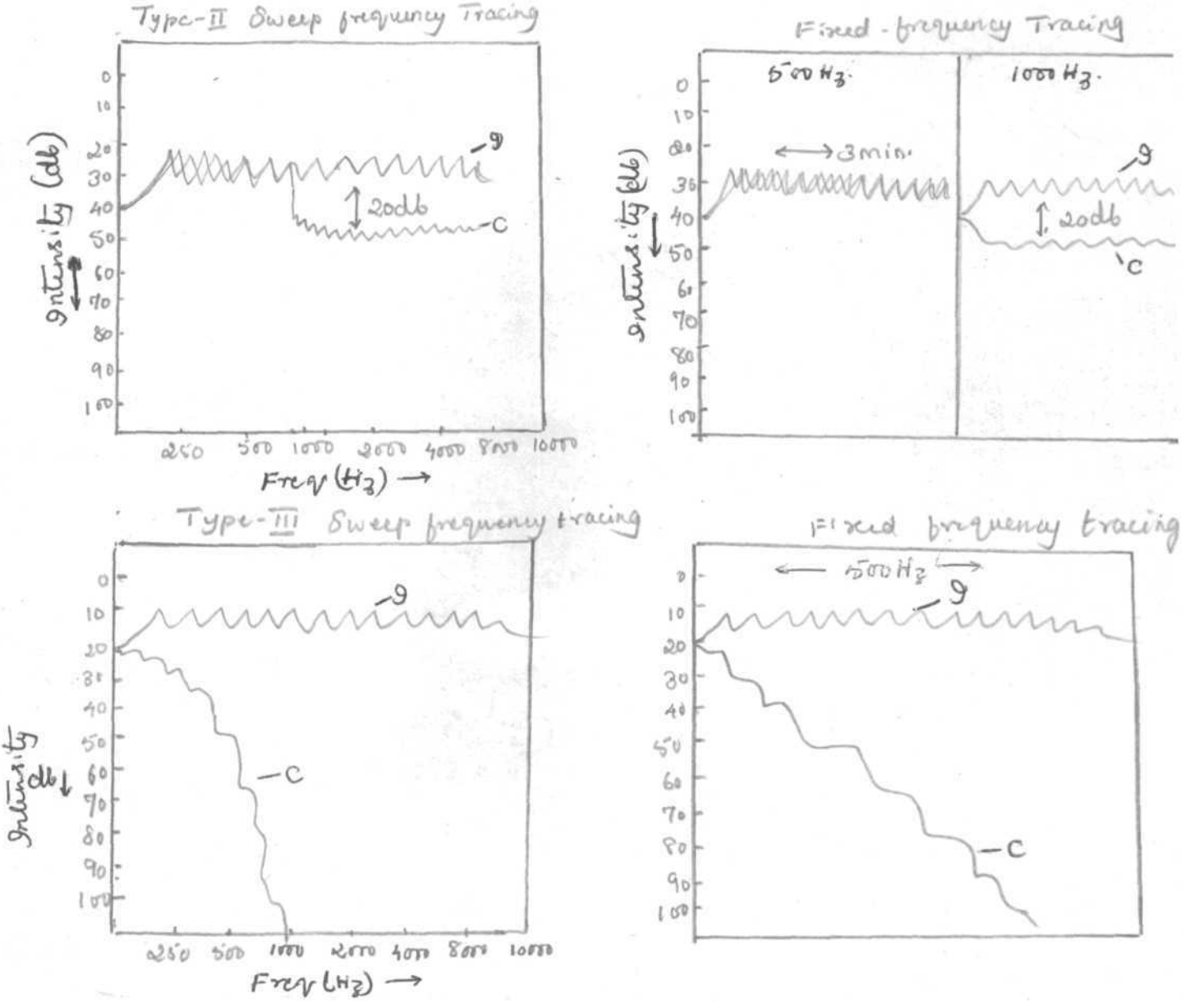
We can also have fixed frequency tracing. In this case, the instrument is set such that the frequency of the tone will not vary. The subject tracks his threshold for a particular duration.

There is also provision for continuous tone tracing and interrupted tone tracing. The tone is presented continuously in continuous tone tracing. The tone is interrupted in interrupted tone tracing. The tone will be on for 200 m.sec and off for 200 m.sec. The attenuation rate is 2.5 db/sec.

In pure cochlear otosclerosis, the patterns obtained are those of end organ disease. The Bekesy variable frequency tracing shows a tendency either toward a type 1 trace or toward a type 2 pattern in which the continuous tracing breaks away from the pulsed tracing at a middle frequency and shows little reduction in amplitude at the higher frequencies. On fixed frequency tracing, the two tracings usually show a gradually increasing degree of break from low to high frequency with minimal or a slowly increasing degree of narrowing of the continuous tracing at higher frequencies (Carhart

1966).

In combined stapedial and cochlear otosclerosis, Type III Bekesy pattern is observed.



Alternate Binaural Loudness Balance Test:- (ABLB)

It is a test for recruitment. The candidate for the ABLB procedure must have normal hearing in one ear for the frequencies to be tested. The hearing in the poor ear should be at least 28 db poorer than the hearing in the better ear.

The purpose of the ABLB test is to compare the growth of loud-

ness in an impaired ear with the normal growth of loudness in the opposite ear. In this way, the degree of recruitment, if present, can be demonstrated.

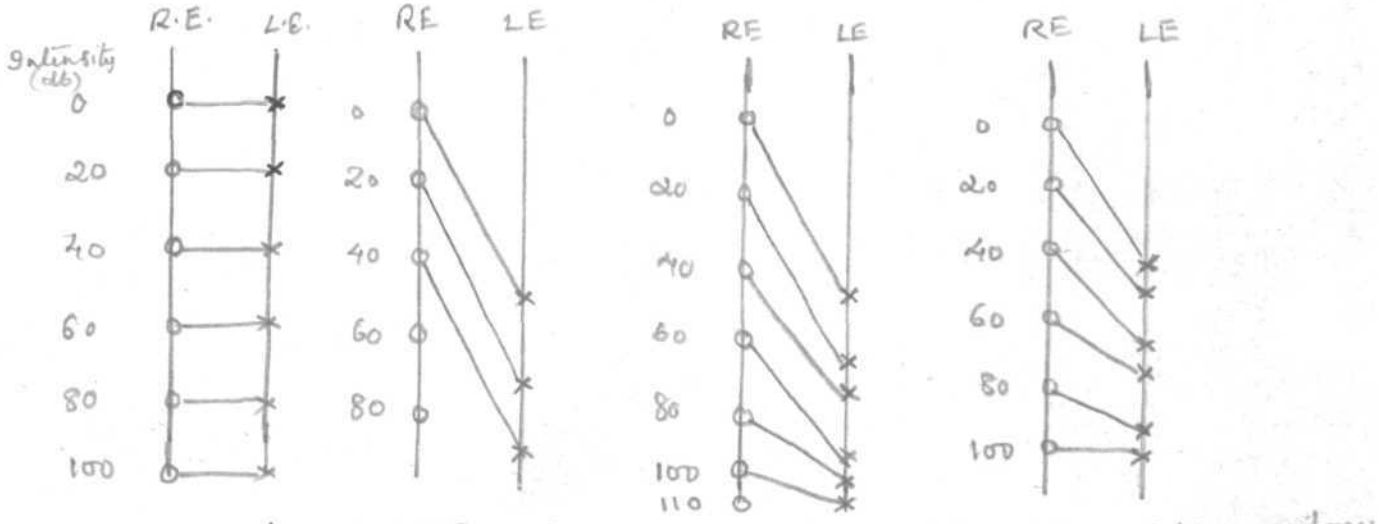
A tone is presented alternately and automatically to the two ears for brief intervals. The tone is presented at 20 db SL to the better ear. The intensity of the tone in the poorer ear is adjusted until the subject says that loudness is equal in both the ears. Equal loudness points are determined upto the maximum point en the audiometer.

The data obtained should then be recorded in a manner which wi] illustrate the growth of loudness in the impaired ear as it compare: with the reference ear. One of the two kinds of illustration can be used.

One is called a laddergram, it looks like an abbreviated audio-gram. The rung (of the ladder) at the top is equivalent to 0 db hearing threshold level. The successive rungs represent higher and higher hearing levels. Thresholds are plotbed and then the average variable ear loudness levels for the several trials at each reference level are plotted as on an audiogram. The variable plot is connecte by a straight line to its corresponding reference level plot.

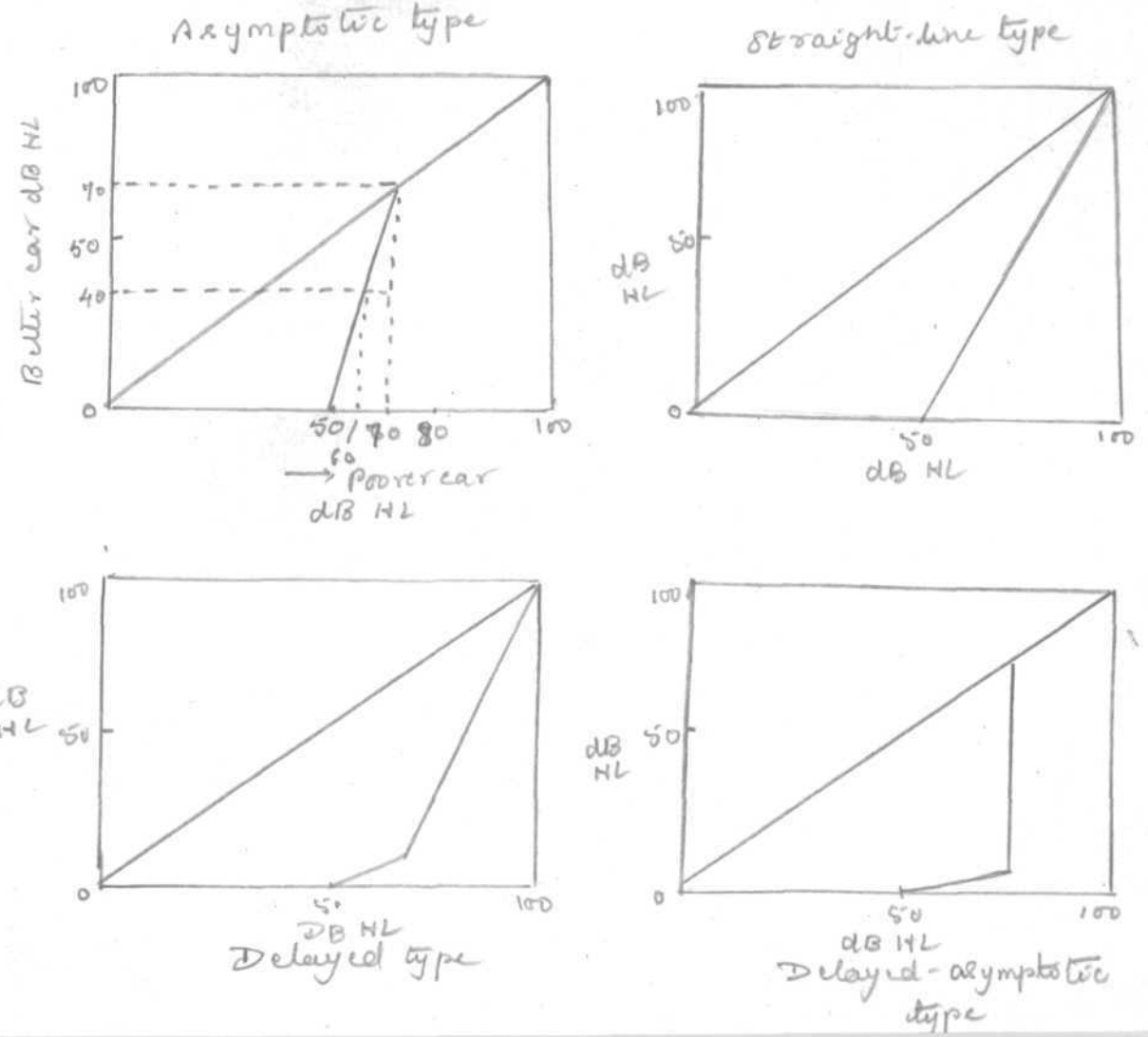
The other method used to illustrate loudness growth is a graph, where the poorer ear data are plotted from the abscissa and the better ear from the ordinate. Again the numbers represent bearing threshold level. The 45 degree line represents equal loudness growth for two normal ears. The points which are plotted are usually connected by a curve of best fit.

Laddergrams :-



Normal No Recruitment Partial Recruitment Complete recruitment

Graph :- Another method of explaining recruitment



In unilateral cases of pure cochlear otosclerosis, full recruitment is noticed. (Carhart 1966; Linthlcum 1972 & Morrison 19' In combined stapedial and cochlear otosclerosis, the results depend upon the degree of conductive and sensori-neural loss.

Impedance Audiometry:-

Changes in the acoustic impedance of the ear can be measured in the external meatus by the acoustic bridge method used by Metz (1961) and Zwislocki (1961), or by electro-acoustic techniques devised by Terkildsen and Thomson (1959) and other workers.

Using the impedance bridge, we can get three impedance measurements: 1. Tympanometry 2. Static compliance and 3. Acoustic reflex.

Tympanometry refers to pressure-compliance relationship. It gives us information about how the compliance changes as the pressure is varied in the external auditory meatus. In tympanometry a pure tone is fed into the ear canal and the static air pressure is continuously changed from -200 mm. water pr. to +200 mm. water pressure. Depending on the stiffness and mobility of the drum and the function of the middle ear, a portion of the tone will be reflec back to a microphone and recorded on a tracing. In tympanometry, we are interested in knowing the exact shape of the curve.

The compliance of the tympanic membrane is determined by the middle ear system. To know the compliance i.e. the equivalent volume of air we must know the large volume and the volume of air in the external auditory meatus. C_2 is the compliance value obtained when the eardrum is maximally compliant. C_1 is the compliance value obtained when the eardrum is maximally stiff. Equivalent volume of air is static compliance (C_S) which is obtained by subtracting

C_1 from C_2. (C_s . C_2-C_1)

Acoustic reflex measurement:-

This is also called as Reflexometry. This was the most applicable feature of Metz's pioneering efforts to measure the impedance of the ear in a clinical population. Based on the fact that loud signals presented to one ear elicit a bilateral contraction of the stapedius muscle, it became common practice to monitor the impedance change vhich occurs as a consequence of the muscle contraction. This monitoring has traditionally been performed in the contralateral ear (crossed reflex). In recent years instrumentation has become avails vhich permits both the eliciting signal and the impedance monitoring to take place in the same ear. This is referred to as ipsilateral (uncrossed) reflex testing. The contralateral acoustic reflex can be expected to occur at 70 to 96 db SL and 3 to 12 db lower SL for ipsilateral stimulation in case of normal ears.

In pure cochlear otosclerosis, A-type tympanogram is obtained, compliance is vithin normal limits (0.3 to 1.5). Stapedius reflex is present. (Morrison 1979). In combined otosclerosis, the results depend on the degree of conductive loss.

No single test provides selective enough data to be used alone in the diagnosis of cochlear otosclerosis. However, when the S.D. score, the SISI score, ABLB, and the response to Bekesy audiometry, TDT , are examined together, the resultant pattern will be of real value in the diagnosis of cochlear otosclerosis.

The otologist's diagnosis of cochlear otosclerosis is based on the presence of sensori-neural hearing loss accompanied by one or more of the following factors:- (Saunders 1965).

1. Gradual onset and slow progression of a hearing loss beginning in early adult life.

2. Clinical 0tosclerosis with stapes fixation ing one ear combined with cochlear degeneration in both ears.

3. Development of a conductive loss with stapes fixation, superimposed on the sensori-neural loss.

4. Strong family history of otosclerosis

5. Positive Schwartze's sign

6. Positive radiologic findings.

Shambaugh (1966), Sheehy (1962), Carhart (1966), Hildyard (1966 & Derlacki (1965) considered some more factors in addition to the above factors in the diagnosis of cochlear otosclerosis:-

1. The appearance of sensori-neural hypacusis in the presence of very good speech discrimination

2. The presence of recruitment, high SISI scores and a Bekesy type II audiogram

3. The patient's inability to assign a cause to the hearing los

4. A bilateral symmetrical configuration of the audiometric pattern

5. A grogressive sensori-neural hearing with an unusual audiometric pattern

6. Successful use of hearing aid prior to progression to severe limits

7. Paracusis willisiana in early history

8. A better speaking voice than one would suspect in the presen of a severe sensori-neural hearing loss.

9. A negative Rinne test

that of spasmophilia in the young and of the vascular recording in elderly patients;

- polytomographies of the bony cochlear capsule according to Valvassori's technique with two aspects: at the first otospongioti stage, a capsule generally thin or even interrupted in some areas corresponding with bone destruction portions; at the second rebuilt ing stage of otosclerosis an increased thickness of the walls of the bony structures without increase of the density.

3. Criteria of certitude:-

They are those which allows us to assert the existence of cochlear otosclerosis:-

- in Impedance audiometry, the appearance of an on-off effect (diphasic impedance change) on a slowly progressive sensori-neural loss reveals the beginning of stapedial fixation, even before the beginning of the a-b gap, and later the disappearance of the stapedial reflex.

- a secondary opening of the a/b gap, occurring in a slowly progressive pure sensori-neural loss, providing that it does not result from another associated factor for vhich impedance enables the differential diagnosis.

3. Radiologic Investigation:-

Radiologic investigation consists of the use of plain x-ray films, tomograms, CTT scans, myelograms, and air studies, and angiograms and venograms.

The radiographic changes of otosclerosis can be best demonstrated by hypocycloidal polytomography (Linthicum 1972). Linear tomography is adequate for many purposes but it does not show as

detail of the otic capsule. Polytomography of the cochlear capsul is found to be a very helpful technique in the diagnosis of cochler otosclerosis. The po3ytomographlc evaluation of cochlear otoscler is gaining increasing significance as more is learned about the relationship of cochlear otosclerosis to sensori-neural hearing impairment. Until polytomography became available, there was no way to confirm the diagnosis of cochlear otosclerosis in the living patient.

Derlacki and Valvassori (1965) have demonstrated graphically the usefulness of polytomograms in this area. In their study of otosderotic patients with combined stapes fixation and sensori-neural loss, otosclerotic changes in the labyrinthine capsule were demonstrated in 90% of the cases. When the diagnosis was based on the Shambaugh criteria for distinguishing sensorineural loss secondary to cochlear otosclerosis, changes in the labyrinthine capsule were found in 60% of the x-rays. Although, type, degree and extension of otosclerotic involvement can be determined by careful polytomography these findings are not always proportional to the functional impairment. Sites of cochlear involvement as demonstrate by the polytomograms are in order of decreasing frequency: basilar turn of the cochlea, remainder of the cochlea, semi-circular canals and internal auditory canal.

The interpretation of a radiogram consists of the detection of outlines and differences in densities of the two factors, the outline is the most important because it is more easily recognized by the human eye. This concept is applied to the interpretation of the tomograps of cochlear capsule. The normal cochlear capsule

appears as a sharply defined, homogenously dense although not homogenous, thick, bony shell outlining the lumen of the cochlea. The density of the capsule seen on profile enhances the clearness of the lumen of the enclosed cavity which appears, because of an optic phenomenon, more radiolucent then it would appear without the surrounding dense line (Valvassori 1969).

The abnormal cochlear capsule shows a more or less severe disruption, of this appearance. The observed radiographic changes in cochlear otosclerosls are characterized according to the extensi degree and type of the process by Valvassori (1966).

As far as the extension three groups are recognized:
1. Changes limited to the capsule of the basilar turn of the cochlea;
2. Changes diffuse in other portions of the cochlear capsule;
3. Changes widespread throughout the labyrinthine capsule.

The degree is classfied as minimal, moderate and severe according to the size of foci of involvement.

The type of involvement depends upon the maturation of process
1. Demineralizing or spongiotic changes correspond to the actively enlarging vascular otosclerotic foci. They are commonly observed ir young patients, in patients with rapidly progressive sensori-neural loss and in cases with a strongly positive schwartze's sign. The white line forming the outline of the capsule becomes thinner, intorrupted and finally, may completely disappear. As the changes progr the relation in density between the capsule and the lumen decreases and in severe involvement completely disappears so that the outline of the cochlea becomes undistinguishable from the surrounding bone

of the petrosa.

2. Scleroting changes are the result of the mature or maturing foci. They are formed by the opposition of mature otosclerotic bone increasing total thickness of the capsule and not by an increase in density of the capsule since the normal enchondral capsule offers already a mixed density. Such foci appear in the portion of the capsule seen on the end as areas of capsular thickening producing rougening and scalloping of the outer and inner aspects and in the portion of the capsule seen on face as areas of increased density superimposed upon the radiolucency of the lumen.

3. Mixed changes are characterized by the presence of dense foci of variable size within an area of demineralization. This feature may be the result of residual fragments of normal capsule in the spongiotic focus or may be produced by partial remineralization as portions of an active spongy focus progress toward the inactive recalcified foci.

Radiographically, Compere (1963) outline d three visible change which may be demonstrated in the petrous pyramid of patients with cochlear otosclerosis:

1. Local overgrowth of osseious tissue anywhere in the otic capsule
2. apparent sclerosis of the entire otic capsule which may almost obscure the labyrinthine system - in some instances, the opaque, are tends to have a nodular outline; and
3. hyperostosis of the entire petrous pyramid.

Radiographic changes in stapedial otosclerosis combined with cochlear otosclerosis was reported by Shambaugh (1966). 90% of patients with stapedial otosclerosis combined with cochlear otosclerosis, a spongifield, or partially spongy and partially recal-

cified focus in the cochlear capsule was found. In 40% of patient: with a pure sensori-neural hearing loss but a suspected cochlear otosclerosis, a spongy or partly spongy and partly recalcified focus of the cochlear capsule was found.

Applebaum and Shambaugh (1978) say that caution must be exercised in the interpretation of subtle polytomographic changes noted in the cochlear capsule and restraint used in the x-ray diagnosis of pure cochlear otosclerosis until there is evidence of correlation with pathologic material.

4. Histopathological Studies:-

The positive diagnosis of cochlear otosclerosis can be made by histologic section of the temporal bones after death. The polytomographic diagnosis of cochlear otosclerosis can be subsequently confirmed by histopathological studies of the temporal bone.

Histopathological studies from Ruedi (1962,1963), Altmann, Kornfeld & Shea (1966), Schuknecht & Gross (1966); Linthicum (1966) Nager (1966), Lindsay & Beal (1966), Holleman and Harrill (1967) Sando et al (1968), Myers and Myers (1968) and Schuknecht (1974) demonstrated the pathological changes in the otic capsule. The reports are in general agreement and can be summarized as follows: For sensori-neural loss to occur, the otosclerotic focus must spread to involve the endosteal layer of the otic capsule adjacent to the spiral ligament. The subsequent changes may be more pronounced if the scala tympani of the basal coil is implicated. Vascular shunts develop between the vessels of the spiral ligament and the vascular otosclerotic borne, and these in turn lead to degenerative changes in the spiral ligament, stria vascularis and hair cells. The process can spread from the basal to the apical coil or it can affect a

Causse et al (1977) considered three criteria for the diagnosis of cochlear otosclerosis.

1. Criteria of presumption
2. Criteria of probability
3. Criteria of certitude

1. Criteria of presumption:-

They are those which arous attention about the possibilities o: cochlear otosclerosis i.e. a pure, slowly, progressive, perceptive hearing loss in childhood, suddenly aggravated at puberty or at periods of endocrine activity, chiefly in a family of otospongiotic patients or with a family history including some progressive pure sensori-neural losses, previously called "degenerative labyrinthoses

- a sensori-neural loss aggravated by pregnancy, menstruation, contraceptive pill or treatment with estrogens.

2. Criteria of probability:-

They are those which lead to the conviction that the hearing loss is due to cochlear otosclerosis but without the possibility of proving it accurately, i.e. the existencp of a schwartze sign in one or both ears;

- the typical opposition between a perceptive loss with a 'cookiebite' curve, quickly distorted by the cochlear loss of high frequencies in stage III, and by an associated presbycusis in elderly patients, and a straight speech curve with good speech discrimination explaining the functional results obtained with a hearing aid.

- an irritative type of torsion sizing test ENG recording characterized by high frequency and low amplitude, different from

localized part of the cochlear duct. There is often partial spira ganglion cell loss, and occasionally a degree of labyrinthine hydro has been observed, and even, of the saccule or cochlear duct.

Thus based on the histopathologic examination of the temporal bone, a positive diagnosis of cochlear otosclerosis can be made.

B. DIFFERENTIAL DIAGNOSIS

Differential diagnosis of cochlear otosclerosis is very essential. It resembles many other diseases like Paeget's disease, osteogenesis imperfecta, syphilis, fibrous dysplasia, neurofibromatosis and presbycusis. In this group of diseases, the involvement of the labyrinthine capsule may occur as one of the pathological manifestations. Clinically and etiologically these diseases differ but histologically they are similar in many respects.

Cochlear otosclerosis should be differentially diagnosed from the following conditions:-

a. Capsular changes in normal ear
b. Paget's disease
c. Osteogenesis imperfecta
d. syphilis
e. Fibrous dysplasia
f. Neurofibromatosis
g. Presbycusis

Apart from these conditions, cochlear otosclerosis should also be differentiated from dominant hereditary deafness which present in adult life. In the absence of a Schvartze sign or a family history, of stapedial otosclerosis, or definite polytomographic evidence of cochlear otosclerosis, It must be assumed that a progressive sensor.

neural hearing loss is due to one of the dominant inner ear diseases. Some of these are very rare such as Alport's syndrome, the Flynn-Aird syndrome or Norrie's disease.

a. Capsular chaises in normal ear:-

Capsular changes may be demonstrated in persouns without hear loss. Valvassori (1969) studied a control series of 100 ears from 50normal individuals. He found no abnormality in 94 ears but in 6 ears one or more small foci were recognized in the capsule of th basilar turn of the cochlea. Their figure is certainly consistent with the study of Guild (1944) who found foci of so-called histological otosclerosis in approximately 10% of unselected routine autopsy specimens.

b. Paget's diseasa:-

Paget's disease affecting the skull may cause obstruction of the external auditory meatus, inducing conductive deafness. Obliteration of the labyrinth and perceptive deafness are less common. Lindsay and Pearlman (1936) described 5 cases of Paget's disease and pointed out some of the features which distinguish it from otosclerosis. These include the later age of onset, no family
$ history, sensori-neural hearing loss showing rapid deterioration, tinnitus, and radiological evidence of Paget's disease of the skull.

The radlographic diagnosis of Paget's disease can be usually made without difficulty. Even in the absence of typical changes vhich usually occur elsewhere in the skeleton the appearance of the petrous pyramids is pathognomic. The haversian bone of the pars petrosa is affected first, and because of its demineralization, the cochlear capsule and the ossicles become more prominent than usual. Then the otic capsule becomes involved. The progression

of the involvement follows a path from the apex to the base of the petrous pyramid. The internal auditory canal is involved first followed by the cochlea and the vestibular system.

C.Osteogenesis imperfecta:-

It is a relatively uncommon inherited, generalized connective tissue disease. The characteristic syndrome of blue sclerae, brittle bones vith a tendency to fracture and bearing loss of a conductive type is generally related to the names of Vander Hove an dekleign who were the first to suggest in 1918, that the hearing loss was due to otosclerosis. The hearing loss is usually conductive and symmetrical, but may be mixed or purely sensorineural.

Weber (1930) found in the callus of a healing fracture (in a patient with osteogenesis imperfecta) bony changes highly reminiscent of the so-called blue mantle in typical otosclerotic foci.

Simson, Hall and his associates (1961,1962) as well as Wullstein (1960) have studied the relationship between osteogenesis imperfecta and otosclerosis. They arrived at the conclusion that the two diseases are due to a common genetic anomaly in the osteoblasts, otosclerosis being interpretable as a localized form of osteogenesis imperfecta, differing from it only in degree, extent and localization.

The changes in the labyrinthine capsule are indistinguishable from the changes of otosclerosi. However, the appearance of the long bones which are abnormally thin and slender, and the presence of multiple fractures often leading to a gross distortion, with the blue colour of the sclera are unmistakeable findings leading to the diagnosis.

d. Syphilis:-

The lesions are manifestations of tertiary congenital or acquired syphilis and caused by gumma formation or by leutic endarteritis. Because of the microscopic size of the lesions, the radiographic findings remain negative until the leutic foci become confluent and form large areas of involvement. At this point spongiotic changes are detectable similar to the ones seen in active cochlear otosclerosis. A sclerotic component may be present due to proliferation of the surrounding bone. Patients usually present in the fourth or fifth decade with bilateral (or occasional unilateral) progressive or fluctuating hearing loss. A serological test for syphilis should be performed in order to accept or rule out the diagnosis of cochlear otosclerosis during life.

e. Fibrous Dysplasia:-

The involvement of the skull by this is quite characteristic whereas the involvement of calcarium and mandible consists of expansion of the affected portion by cystic lesions, the changes in the base of the skull including the temporal bone are almost always of the proliferative type. The affected petrous pyramid becomes extremely dense and thick Kith consequent asymmetry between the two sides. The outline of the labyrinthine capsule becomes first poorly distinguishable from the surrounding bone and finally may disappear as the lumen of the inner ear structures becomes partially or totally obliterated. On conventional radiograms there is a superficial resemblance to Paget's disease. However, in Paget's diseases the increased density of the petrous pyramid is merely the result of an increase in thickness of the bone which is actually demineralized, whereas in fibrous dysplasia there is an addition to the increase in thickness a real diffuse sclerosis of the bone with obli

iteration of the lume n of the air cells and of the inner ear structures.

f. Neurofibromatisis:-

In addition to the typical pressure erosion from a neurofibro which may involve any portion of the labyrinthine capsule, generalized neurofibromatosis may produce less common and less specific changes in the petrous pyramid and labyrinthine capsule. These consist of disturbances in the growth which may lead to hypoplasia to hypertrophy. In the latter case the bone is unusually thick and the picture becomes similar to the one described in fibrous dysplasia. In addition, areas of demineralization may be detected in the labyrinthine capsule. The definite diagnosis of neurofibromatosis is based on the detection of the typical skin tumours, "cafe au lait" spots and skeletal deformities.

g. Presbycusis:-

The atrophic changes in ears with cochlear otosclerosis are identical with those observed in aging individuals without otosclerosis. Hence cochlear otosclerosis should be differentiated from presbycusis.

Schuknecht (1965) describes a common type of sensorineural hearing loss which has its onset in middle age, is slowly progressive and is characterized by a flat audiometric pattern. It is associated with degenerative changes of the vascular stria of the middle and apical turns.

The effect of aging on the organ of corti consists of loss of hair cells and supporting cells. This is most pronounced in the basal coil and is associated with an abrupt high tone hearing

hearing loss.

The basilar membrane becomes progressively narrower and thicker from apex to base.

Schuknecht (1S65) noted that aging may lead to an atrophy of both the spiral ligament and basilar membrane, rupturing the latter and causing profound deafness.

A descending audiometric curve as Nager (1920), Schuknecht and Igarashi (1964) and others have suggested, might be due to progressive alterations in the basilar membrane, which may reduce the resilience of its filaments and fibres and alter the structure of the interfilamentous substance. Similar histopathological changes are found in cochlear otosclerosis.

Due to the similarity of cochlear otosclerosis with the dif ferent conditions mentioned above, differential diagnosis of cochlear otosclerosis is very essential. The advancement of cochlear Otosclerosis can be halted with effective fluoride treatment if it is identified early.

In all patients with a sensori-neural or vestibular loss of unknown origin and positive tomographic findings, serologic tests for syphilis, determination of serum calcium, phosphorus, alkaline phosphatase and a radiographic bone survey should be performed. This additional information would be of great help in accepting or ruling out the diagnosis of cochlear otosclerosis during life (Valvassori 1969).

CHAPTER-VI

COCHLEAR OTOSCLEROSIS AND SENSORI-NEURAL BEARING LOSS

The question of sensori-neural hearing loss occurring in patient with cochlear otosclerosis has attracted the attention of many invest gators. There is less agreement among the authorities concerning sensori-neural hearing loss due to cochlear otosclerosis without an associated stapedial involvement. The strongest advocate for the frequent existence of this eneity is Shambaugh (1969), and against it Schuknecht (1971,1974).

Shambaugh (1965) listed the floowing clinical manifestations suggestive of a sensori-neural hearing loss due to cochlear otoscle-rosis:

1. bilaterally symmetrical sensori-neural loss without specific etiologic history but with unilateral ankylosis;
2. a pure progressive sensori-neural loss beginning in early or middle adult life combined with a strong family history of clinical otosclerosis and
3. a pure progressive sensori-neural loss beginning in early or middle adult life combined with a strongly positive Schwartze sign.

Altmann et al (1966) made an extensive review of the histological findings described in the literature as possible causes of sensori-neural hearing loss in patients with cochlear otosclerosis. These included:

1. Venous stasis in the veins draining the cochlea;
2. irritation of the endosteal layer with production of fibrous tissue and bone in the scala tympani;
3. disintegration of the Organ of Corti followed by ascending retrograde degeneration of the corresponding nerve fibers and ganglionic cells; this in turn being possibly due to toxic substances released by the oto-sclerotic bone.
4. Increase in the vascular loops in the stria and pro-liferation the epithelium of the stria near the active otosclerotic focus;
5. atrophic changes in the spiral ligament adjacent

to the foci;

6. invasion of the cochlear scalae by otosclerosis: and
7. interference of foci in close proximity to the spiral ligament with the circulation in the stria vascularis and the upper portion of the spiral ligament vith subsequent changes in the composition of endolymph.

Glorig and Gallo (1962) on the basis of audiologic test data stated, in general, otosclerosis does not increase sensori-neural los above that to be expected in the general population.

On the other hand, otologists almost vithout exception have the clinical impression that progressive sensori-neural hearing loss is frequently associated with cochlear otosclerosis. Lending support to this probability are findings of Kelemen and Linthicum (1969). Based on an extensive histological study, they state that "...the majority of individuals with otosclerotic involvement of the cochlear endostea layer have an elevated bone conduction threshold. Contrarily only a few individuals in whom otosclerosis remained restricted to the footplate or the vestibulum had a significant increase of the thresho

In 1966, at its annual meeting, The American Otological Society sponsored a symposium on sensori-neural loss in otosclerosis. Convincing evidence was presented to show that severe involvement of the cochlear bony labyrinth by otosclerosis frequently causes sensori-neural loss; however, no evidence that would withstand statistical analysis was presented to show that milder degree of involvement causes sensori-neural loss.

Unfortunately, some of the deliberations of the meeting gave great impetus to a new concept "cochlear otosclerosis" which implies that otosclerosis may cause sensori-neural hearing loss without an

associated conductive loss.

The concept was bom of indirect and circumstantial evidence. Carhart (1966) on the basis of audiological studies, stated that "sufficient parallelism exists between a group of cochlear otosclerot and a large sample of patient with sensori-neural loss of indefinable etiology, to support the hypotheses that cochlear otosclerosis is a common etiology within the latter group.

Freeman (1979) examined the patients presenting the clinical pro blem of progressive sensori-neural loss in depth including diagnostic audiology and po3ytomography of the petrosa. He found that a high percentage of such diagnostic problems are due to cochlear otosclerosis

Gross (1969), Schuknecht (1971,1974) and many other investigators disagree with the concept that sensori-neural loss is due to cochlear otosclerosis. Gross (1969) analysing the temporal bones of patients with unexplained deafness has not been convinced that the histologic otosclerotic foci seen were of significant incidence or size to explain the inner ear changes.

Schuknecht and Kirchner (1974) also say that sensori-neural loss occurring in pure form without a conductive loss cannot be attri-buted to cochlear otosclerosis, for when the otosclerotic lesion is sufficiently severe to cause atrophy of the supporting, sensory and neural structures within the cochlea, it invariably also fixes the stapes*

Lindsay and Beal (1966), Sando, Hemenway et al (1968) and Linthicum, Bredy and Filipo (1975) expressed that light microscopy has failed to unravel the problem of sensori-neural hearing loss associated with cochlear otosclerosis. Antoli-Candela Jr. et al (1977)

failed to show abnormal changes in the cochlear hair cell population and stria vascularis. Hyalinization, and decrease in the width of the spiral ligament, they found, could not be correlated with the sensori-neural hearing loss.

Various mechanisms by which the cochlear otosclerosis can produce sensori-neural hearing loss has been reported by many investigators (Carhart 1963; Altmann et al 1966; Frost 1967; Linthicum, Fillpo & Bredy 1975). They are as follows:

1. Toxic substances produced by the otospongiotic focus
2. Invasion of the basilar turn of the scala tympani
3. Rupture of the basilar partition
4. Other involvements within the upper cochlear turns
5. Vascular shunts between the inner ear vessels and the otospongiotic focus
6. Atrophic changes in the spiral ligament adjacent to the otosderotic foci
7. Atrophy of the stria vascularis
8. Atrophy of the Organ of Corti
9. Narrowing of the lumen of the cochlea
10. Distortion of the Basilar Membrane
11. Damage to the auditory nerve in the Internal auditory meatured by pressure of an ingrowth of otosclerotic bone
12. Enzymatic concept of otospongiosis

1. Toxic substances produced by the Otospongiotic focus:-

Siebenmann (1899) suggested that there might be toxic substances liberated by the otospongiotic focus into the inner ear fluids, that impaired the function of the cochlea. According to his teaching, the course of events can be reconstructed in the following way: when the otosclerotic focus reaches the endosteum, a series transdudate occurs within the perilymphatic cochlear spaces. The metaplastic bone libe-

rates metabolites which by way of the transudate reach the peri- and endolymph spaces of the cochlea with damage to the Nervous apparatus

Using electron microscopy, Bretlau et al (1971) and Chevance et al (1970) have demonstrated lysosomes at the margins of the advan. cing otospongiotic focus. They suspected that these lysosomes might produce a toxic substance that caused absorption of the normal bone and if, liberated into the inner ear fluids, could produce a sensori-neural loss.

Gussen (1975) suggested that cochlear otosclerosis brings about sensori-neural loss by inducing changes in the composition of the inner ear fluids or also by altering the blood supply in the cochlea or both. There has been speculation that toxic substances arising from the otosclerotic focus might reach the cochlea r fluids resulting in degeneration. This might well be so, but it is also conceivable that the decrease in the perilymph-vascular tissue of the spiral ligament signifies a decreased resorbing capacity of the tissue and that certain metabolic products normally present in the cochlear fluid may new increase in concentration to a degree detrimental to the cochli.

2* Invasion of the basilar turn of the scala tympani:

The otosclerotic foci can bring about an inner ear conductive disorder. Here, the cause is the invasion of the scala tympani by a mass of new tissue which interferes mechanically with vibration of the Basilar Membrane and the perilymph(Fig 1). Nager and Fraser (1938) say that lesions of this type originate at the round window and extend for varying degrees apical ward.

Lindsay and Beal (1966) reported 16 ears with sensori-neural impairment and associated otosclerosis. In 12 of these ears, histo-pathological factors were noted which distinguished them form other

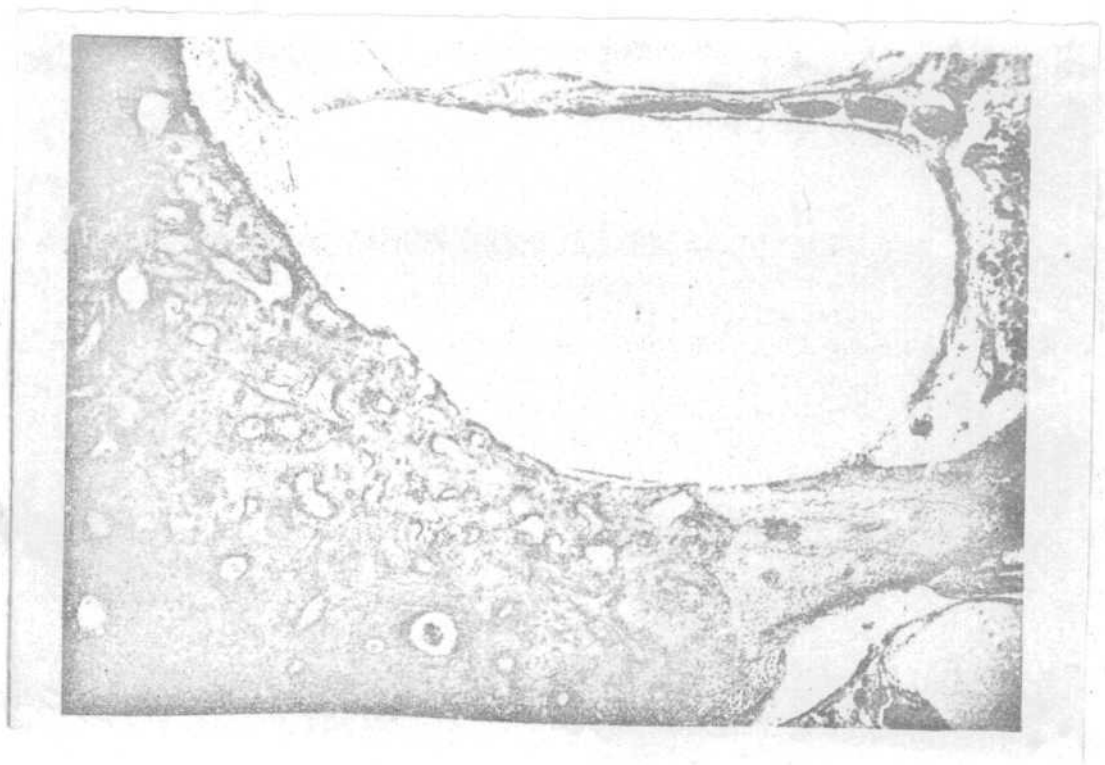


Fig.1: Diffuse otosclerosis spreading into scala tympani of middle coil of the cochlea (Friedmann, 1974).

cases of sensori-neural deafness. They raise the question of a possible relationship. Cochlear reconstructions were presented in two ears and they felt that the degree of impairment of hearing was not explained on the basis of the hair cell and ganglion cell count There was some decrease in the extent of the stria vascularis but did not feel that this was a sufficient explanation for the impaired function. In those ears with a single focus at the oval window region and sensori-neural deafness this focus consistently extended through the endosteal layer of the capsule at the attachment of the spiral ligament. According to that the endosteum of the spiral ligament and scala tympani in the basal turn appears to be more susceptible to this irritating effect of an active advancing focus as compared to areas where the focus borders on the large perilymphatic cystern.

Altmann, Kornfeld and Shea (1966) concluded in their paper that specific sensori-neural hearing loss is encountered in otosclerosis in those cases in which the focus extends to the endosteum of the cochlear scalae. They felt that the damage to the snesori-neural elements is produced by the release of substances into the labyrinthine fluids either by the focus or abnormally functioning stria vascularis.

Changes in the labyrinthine capsule especially in the cochlear capsule are found to be diffuse and involve the entire capsule or localized to a segment of it, especially to the basilar turn. They consist of foci of demineralisation, foci of sclerosis, by a combination of the two, producing a more or less severe disruption of the normal pattern of the capsule. These changes were found in cases of pure sensori-neural loss diagnosed as cochlear otosclerosis

(Valvassori, 1966).

Applebaum and Shambaugh (1978), in one of their patient with cochlear otosclerosis observed moderate mixed changes in the capsule of the basllar turn of the cochlea which appears dimineralized although irregularly thickened. This patient had a symmetrical, moderately severe loss in the high frequency.

3. Rupture pf the Bpsilar partition and Other Involvements within the Upper Cochlear Turns:-

Benitez and Schuknecht (1962) described a patient in whom an otosclerotic focus in each temporal bone extended to the endosteum bounding the corresponding cochlear space. Severe atrophic change in the spiral ligament occurred in association with each lesion. Further lesions appeared in one ear. One feature was a "rupture of the Basilar Membrane in the regions corresponding closely with the area of atrophy of the Spiral Ligament". This rupture was mos pronounded in the upper basal turn, but it also extends through most of the middle turn and into the apical turn. In addition, the tectorial membrane and the organ of corti of this ear are markedly shrunken in these same regions, although the various cell types in the organ of corti were distinguishable at most points. They speculated that the mechanical effects due to the ruptured membrane may be combined here with sensori-neural malfunction.

4. Vascular shunts between the inner ear vessels and the otospongiotic focus:-

Ruedi (1965) and Ruedi and Spoendlin (1966) demonstrated most convincingly a vascular basis for the sensoi-neural loss in cochle otosclerosis. Normally no anastomoses exist between the vascular system of the otic capsule and the vascular system of the membranou labyrinth. However, when active otosclerosis encroaches on the liga-

mentum spirale, venous communications or shunts spring up between the capillaries of the cochlea and the venous spaces of the otosclerotic focus. The flow of blood is from the spaces to the spiral capillaries of the cochlea and vessels of the stria vascularis. The radical change in circulation causes venous stasis, congestion, and hypertrophy in the vascular loops in the stria vascularis as well as intense vasodilation of other veins in the inner ear. In a series of otosclerotic patients with severe sensori-osural deafness, degeneration of neuroepithelial elements in the inner ear and lamellar bone formation in the scala tympani were traced to the preceding vascular disturbances

5. Atrophic changes in the Spiral Ligament adjacent to the otosclerotic foci:-

Histologic evaluation of 32 temporal bones with cochlear otosclerosis has revealed that the amount of sensori-neural hearing impairment is directly related to the amount of involvement of the endosteum of the spiral ligament by the otosclerotic process and not to age (F.H. Linthicum 1972)

A study of changes apparent in the spiral ligament on the light microscopy has been made on 151 human temporal bones by Wright & Schuknecht (1972). Of 42 otosclerotic ears studied 20 showed involvement of the cochlear endosteum with spiral ligament atrophy. Audiograms available on 11 of these ears showed a sensori-neural loss with a descending pattern.

Their Observation indicate that atrophy which has progressed to the stage of shrinkage which may occur as a result of otosclerosis or aging, is associated with snesori-neural hearing loss which is characterized by the descending audiometrlc threshold pattern. Possibly these morphological alterations in the

ligament are accompanied by similar changes in the basilar membrane and supporting cells of the organ of corti, all of which might alter the physical response characteristics of the cochlear duct and cause an inner ear conductive deafness.

Gussen (1975) observed spiral ligament changes adjacent to the stria vascularis in the a case with cochlear otosclerosis. There was a decrease or loss of the capillary-perilymph system. It is suggested ed that this results in increased concentrations of metabolic products in the cochlear fluids that can no longer be absorbed effectively. This might result in reduced auditory acuity.

6. Atrophy of the Stria Vascularis:-

The stria vascularis an integarl part of the spiral ligament, has a selective absorptive function rather than secretory and absorptive (Ruedi 1965). Should this be the case, interference of the circulation, by the otosclerotic focus could result in a build up of metabolites in the inner ear fluids and decrease its efficiency. This interference would result in a build up of carbon-di-oxide and other products of metabolism and interfere with the normal function of the hair cells in those cases in which there was found, by light microscopy to be no abnormality.

Manasse (1912) saw at the lower end of the stria, thick, round, fibrous, prominences of the dubious vascular origin, occasionally jutting far into the lumen of the cochlear duct. Mayer (1917) reported engorgement, venous stasis in the stria vascularis and thought it responsible for the functional damage.

Hollemann & Harrill (1966) presented a case of cochlear otosclerosis in which there was retrogressive changes in the striae and

spiral ligaments from endosteal otosclerotic invasion. They also demonstrated membranous adhesions in the scala tympani of one ear. Venous shunts, were noted, but they concluded that they were not sig nificant in this case.

The temporal bones revealed mild distortion of the involved cochlear coils, atrophy of the spiral ligament, atrophy of the stria-vascularis with a corresponding loss of hiar cells and ganglion cells in those areas with endosteal involvement by the otosclerotic process They found a relationship between cochlear otosclerosls and sensori-neural hearing loss based on these histopathologlcal findings.

7. Atrophy of the Organ of Corti:-

Lawrence (1S66) declared the arcade vessels beneath the basilar membrane to be the nutrient vessels of the organ of corti. According to this concept, it is not the stria system which feeds the hair cells and therefore stria damage by the focus, approaching through the capsular wall, would not cause sensori-neural deafness. On the other hand, warping of the arcade vessles themselves might endanger the function of the organ of corti.

8. Narrowing of the lumen of the cochlea &

9. Distortion of the Basilar Membrane:-

The propagation of the traveling wave along the basilar membrane was analyzed by Bekesy (1960). Later, Tonndorf (1960) demonstrated that increased tension on the basilar membrane, as occurs with the endolymphatic hydrops, could account for the phenomenon of diplacusis in patients with this condition.

Analysis of the width of the basilar membrane in the posterior basal turn of 12 cochleas with cochlear otosclerosis involving the endosteum indicated a median width of 0.78 mm. The median vidth of

of the basllar membrane in 16 cochleas without cochlear otosclerosis was 0.85 mm. anstatistically significant difference (Linthicum, Eili & Bredy, 1975).

This difference suggested the possibility of the cochlear focus enroaching upon the spiral ligament and narrowing the width of the basilar membrane and the diameter of the cochlea. This distortion could possibly alter the amplitude of the traveling wave.

A model of the cochlea was constructed by Linthicum et al (1975) in such a manner that the basilar membrane could be narrowed in the simulated basal turn of the cochlea. It was found that the amplitude of the traveling wave was decreased with the exception of the area where the membrane had been narrowed. In this area, the amplitude increased. This increase was felt to allow energy to be transmitted directly from the scala vestibuli to the scala tympani via the cochle duct and basilar membrane, thus decreasing the amplitude of the traveling wave along the remainder of the basilar membrane.

These findings suggest that part of the sensori-neural hearing loss in patients with cochlear otosclerosis could be due to basilar membrane distortion.

10. Damage to the Auditory Nerve in the Internal Auditory Meatus by pressure of an ingrowth of otosclerotic bone:-

The inner meatus frequently is a veritable showplace of otosclerotic proliferation resulting in coarse alterations of the lumen. Concentric narrowing or excrescences into the lumen impinge on the nerve. Here lies possibly the core of the problem of cochlear deafness.

Manasse (1917) considered constriction of the lumen of the inner meatus by otosclerotic bone a serious influence on the function

of the eighth nerve. The normally smooth wall of the inner meatus becomes frayed, torn up; over the otosclerotic process the dura is engorged, producing a "Schwartze sign" in the inner meatus.

Nager (1947) confirmed that observations by Grey (1932) and Bruht (1929) that constriction in the lumen of the cochlea occur. He felt that the eighth nerve may thereby be compressed with consequent disturbance to hearing.

Histological examination of the temporal bones was made by J.O.Frost (1967) in one of his patient with otosclerosis. The otoscl rotic focus was situated medial to the cochlea, anterior to the Inter Auditory meatus and projected from the anterior wall into the canal impinging on the cochlear part of the trunk, but no damage to the ner fibers. This focus also extended into the floor of the internal audio tory canal. The author said that this was not the cause af any hearing loss in this patient as the nerve fibers are intact. But the history hearing loss and the histological findings makes it almost certain the otoeclerotic foci projecting to the internal auditory meatus was either the cause or a major contributing factor in the production of sensori neural hearing loss. (Fig. 2)

11. Enzymatic concent of otosclerosis:-

The enzyme concept proposed by Causse et al (1977) envisages an upset to the normal equilibrium of trypsin/antitrypsin in the inner fluids, disturbing the relative metabolic isolation of the cochlea. higher ratio of trypsin causes the hair cell loss and other changes. Certainly their analytical studies show a definite relationship betwen this enzymatic ratio and the clinical evidence of sensori-neural loss in cochlear otosclerosis.

Shambaugh (1978), also explains the mechanism of sensori-neural

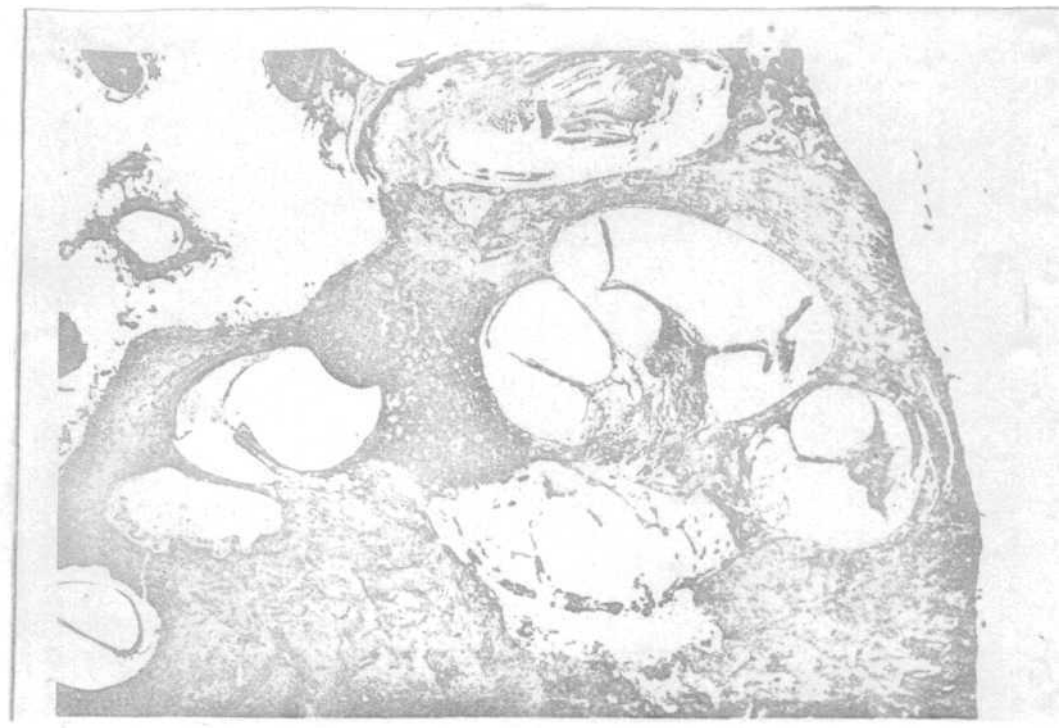


Fig.2: Diffuse otosclerosis spreading into the cochlea and internal auditory meatus (Friedmann, 1974).

loss due to cochlear otosclerosis by demonstrating the presence of proteolytic enzymes in the perilymph in such cases.

CHAPTER-VII

TREATMENT

Any treatment which can arrest the progression of deafness in cochlear or combined otosclerosis must be given serious consideration. It has been found that surgery has no place in the management of pure cochlear otosclerosis. Hence, other methods of treatment like medical treatment and non-surgical measures have been tried with patients of cochlear otosclerosis.

MEDICAL TREATMENT

Fluorides in Treatment:-

Increased intake of fluoride is regarded as "nature's method" of preventing and combatting osteoporosis either generalized or localized form in the labyrinthine capsule (Shambaugh 1973).

In its early and actively expanding stage, otosclerosis consists of one or more areas of localized osteoporosis with a spongy vascular type of bone poor in calcium and rich in connective tissue , cells and blood vessels, replacing the ivory-dense bone of the labyrinthine capsule. The spongy vascular otoporotic focus may spontaneously recalcify and again assume a density comparable to the normal capsular bone. Recalcified inactive otosclerosis is a little more common in older individuals, while active otoporotic lesions are somewhat more common in young persons. Thus there appears to be a slight natural tendency for otosclerosis to spontaneously mature and inactivate itself, but only a slight tendency. It is to encourage this natural tendency, that sodium fluoride in large doses was proposed in 1966.

Theoretical and Laboratory Research Considerations for the Use of Sodium Fluoride

A. Epidemiologic Studies:- (Bernstein D.S. et al (1966); & Daniel 19

1. In communities with very low fluorine content in the water supply, dental decay among school children is twice as prevalent as in communities with one part per million of fluorine in the water supply. This fact is being utilised by adding fluorine to the water supply in areas where it is deficient.

2. Investigations of generalized osteoporosis in low and high fluoride areas indicate the beneficial effect of increased intake of fluoride in adults to prevent post menopausal otosderosis.

3. A similar study of the incidence of stapedial otosclerosis indicated a significant decrease in a higher fluoride area.

4. Calcification of the aorta is more frequent in low fluorine areas than in areas with high fluorine content in the drinking water. Thus fluorine appears to be necessary to keep calcium where it belongs: in the teeth and bones rather than the arteries.

B. Invivo Studies:- (Petrovic and Shambaugh Jr.1966)

The favourable effects of moderate dosage sodium fluoride therapy in promoting recalcification and reducing bone remodeling activity of active otosclerosis have been demonstrated by experiments on otospongiotic bone.

1. By means of tetracycline labelling of new bone it was found that there is an optimum dose of sodium fluoride that promotes calcification of bone in the baby rat. Smaller doses have less effect or even a negative effect retarding calcification if the dose is further increased.

2. Optimum dosage sodium fluoride accelerates healing of

fractures in the adult rat by retarding the early decalcification at the site of fracture and then accelerating calcification of the callus.

3. Optimum dosage sodium fluoride prevents osteoporosis by experimentally induced heparin or cortisone.

4. The promotion of calcification of bone by fluoride is partly by decreasing osteoclastic bone resorption and partly by increasing osteoclastic bone formation. Their studies vere the first to demonstrate this feature of sodium fluoride therapy.

C. Invitro Studies:- (Petrovic and Shambaugh 1966)

Organ culture studies on otospongiotic bone indicate the mechanism of sodium fluoride effect.

1. Otospongiotic bone from the stapes footplate of patients who have been treated with sodium fluoride, when grown in organ culture takes up radioactive calcium more rapidly and shows a significant decrease in acid phosphatase as compared to otospongiotic bone from untreated patients. These findings indicate recalcification of the focus and reduced bone remodeling activity after fluoride therapy.

2. When fluoride is added to the culture medium of otospongiotic bone in organ culture, radioactive calcium is taken up more rapidly than without fluoride. This indicates the calcification-promoting effect of fluoride invitro.

3. Chick and mouse embryo bone in organ culture lose calcium. When fluoride is added to the culture the loss of calcium is reduced, indicating diminished osteoclastic bone resorption invitro.

D. Enzyme Studies:- (Causse J. & Chevance 1973)

1. Cytotoxic bone dissolving enzymes can be demonstrated at the

border of an expanding focus of otosclerosls. These are produced by histiocytes and osteoclasts.

2. These enzymes readily diffuse into the cochlear fluids when an active otospongiotic focus reaches the endosteum or less readily via bone canaliculi. When an active focus approaches but does not yet reach the endosteum. Samples of perilymph removed during stapedectomy show a high correlation between sensori-neural progression prior to stapedectomy and enzyme activity. A number of cytotoxic enzymes have been identified: collagenase, phosphatasic acid, alpha chymotrypsin, ribonuclease, lactate dehydrogenase and trypsin.

3. After 6 months or more of fluoride therapy, 37 patients with preoperative sensori-neural loss failed to show qytotoxic enzymes in the perilymph with one doubtful (slightly positive) result, indicating either an inhibition of enzyme activity by fluoride or reduced bone remodeling activity of the focus as a result of fluoride therapy or both.

4. Causse et al (1977) HxplahmR experimented with the drag sodium fluoride on patients with cochlear otosclerosis and found that this drug acts as an enzymogenesis inhibitor which would create the balance enzyme/antlenzyme by suppression of the enzymatic activity into the otospongiotic microfoci. The low molecular weight 42 of this antienzyme enables it to pass through the otic capsule. It acts by presence more than by weight, but it is necessary to reach an action threshold beyond which its cativity does not appear more obvious, regardless of the dose.

E. Fluoride assay of otospongiotic bone:- (Petrovic & Shambaugh 1936)

1. Otospongiotic bone from the footplate of untreated patients

has a considerably higher content of fluoride than enchondral bone from the stapes superstructure or skeletal bone from the meatus. This is explained by the increased vascularity of otospongiotic bone enhancing the opportunity for contact between the blood circulating fluoride normally absorbed from drinking water and food in trace amounts.

2. After moderate dosage sodium fluoride for 6 months to a year or more, the fluoride content of spongiotic bone is increased about threefold over untreated cases, while the fluoride content of enchondral bone and skeletal bone is increased only slightly. This is due to the vigorous bone remodelling activity and vascularity of otospongiotic bone compared to the virtual absence of bone remodeling in adult endochondral bone and the scant remodeling and vascularity of adult skeletal bone.

3. The experiment reported by Linthicum et al (1973) substantiates the above observations. They found significant decrease in radioactive strontium 85 uptake by otospongiotic bone after sodium fluoride therapy indicating reduced bone remodeling activity of the focus.

Thus there is ample experimental evidence that sodium fluoride in optimum dosage promotes maturation and reclacification of an active immature focus of otosclerosis, thus reducing its enzyme activity and the resultant diffusion of cytotoxic enzymes into the perilymph. (Shambaugh 1977)

The Mode of Action of Fluoride in the Body:-

This was reported by Shambaugh and Causse (1974). Sodium fluoride is rapidly resorbed from the intestinal tract, 90% appearing in the bloodstream within a few hours. It is fairly rapidly excreted by the kidneys, about 50% appearing in the urine within 24 hours, the other

50% remaining in chemical union with bone where the fluoride ion replaces the hydroxy ion of hydroxyapatite bone crystals. Fluoro-apatite crystals are harder and less soluble than hydroxyapatite and resist osteoclastic enzymatic bone resorption. The fluoride content of the skeleton increases with age, more rapidly. When the intake of fluoride is increased in water, food, by inhalation or by medication.

Clinical Experience with Sodium Fluoride Treatment:-

A total of 157 patients with clinical and audiometric findings suggestive of cochlear otosclerosis and tomographic evidence of capsular changes were treated with sodium fluoride by Valvassori (1969) and then reexamined by tomography. A total of 111 or 71% of the patients showed no change in the degree, extension, and type of involvement following sodium fluoride therapy. In 9 cases (5.5%) worsening of the capsular involvement was noticed due either to increase in size of the focus or foci or extension of the involvement to other portions of the cochlear capsule. In 30 (19%) of the cases a significant improvement of the changes in the cochlear capsule was observed. This improvement ranged from a complete disappearance of the focus or foci of involvementso that the capsule had returned to a normal density to a partial recalcification usually characterize by the evolution from a spongiotic to a mixed focus. In an additions group of 7 (5.5%) of the cases there was suggestion but not certainty of improvement or disappearance of the capsular changes.

Freeman (1980) found sodium fluoride therapy to be beneficial not only in controlling the sensori-neural hearing loss but also the symptoms of vestibular dysfunction in patients with cochlear otoscle-rosis. Sodium fluoride therapy for control of progressive sensori-neural hearing loss was found to be effective in 80% of the patients

who had cochlear otosclerosis.

There is a critical level of fluoride dosage for optimum calcification-promoting effect. Too little has less effect and too much has less effect.

At first Shambaugh's (1969) philosophy was to use sodium fluoride 40-60 mg. daily in patients with stapedial otosclerosis and a Schwartze sign. Following the development of polytomography with visualization of the otic capsule, evidence of capsular disease whether alone or combined was taken as an indication for therapy. Progression of the cochlear component of the bearing loss is also used as a criterion. Efforts were made to achieve radiological improvement in mineralization of the otic capsule and to this end 40-60 mg. sodium fluoride (sometimes higher doses) were administered daily for 1-2 years. X-ray improvements were reported in 23.5%, no change was noted in 70% and deterioration seen in 5.5%.

Of over 4000 patients treated between Chicago and Beziers by Shambaugh and Causse (1974), only a handful have experienced some recovery of the sensori-neural component of the hearing loss, but it has stabilized in 80%. In about 20% the sensory loss has progress In a much, smaller group which was used as a control, however, progression of sensori-neural deafness was noted in a much higher percentage. Among those who stabilized on fluorides were some who replaced 2-7 years later. Daily maintenance of 20 mg. is now advised in these patients.

Shambaugh's recent practice is to prescribe calcium gluconate 0.5 g. and vitamin D 400 units thrice daily before meals, together with enteric-coated sodium fluoride 20 mg. twize or thrice daily after meals. Therapy is continued for two years and four years if

necessary. A skeletal survey for evidence of fluorosis is conducted every two years.

Morrison (1975) using 20-60 mg. sodium fluoride daily reported the results of a double-blind trial in 40 patients with cochlear otosclerosis followed from two to five years. The treated group vere statistically better than the control group with a probability of 0.02. The hearing gains, however, were only of the order of 15 db at three or more frequency. The present practice is to prescribe 20 mg. once daily after meals for three months,to repeat the course of therapy as indicated at yearly intervals; this is used for cochlea and combined otosclerosis and for stapedial disease when surgery is not indicated.

Progression or reactivation of otosclerotic disease as shown by tomography, despite moderate dosage sodium fluoride therapy, is due to poor intestinal absorption of the drug in its enteric coated form. A work conducted by Deka et al (1978) on the absorption of the sodium fluoride enteric coated tablet (20 mg.) showed that the drug that were used for the last 16 years had wide variation in absorption as indicated by urinary fluoride analysis. This fact led them to think whether or not it was the poor absorption of the "enteric coated tablet of sodium fluoride" taken by those cases who showed poor therapeutic results to sodium fluoride and showed evidence of progression or reactivation of the disease on tomographic evaluation.

Indications for Sodium Fluoride Treatment:

In the present state of our knowledge it seems that sodium fluoride therapy has a place in the management of otosclerosis. The indications for moderate dosage sodium fluoride therapy of otosclerosis are four:-

1. Positive Schwartze sign indicating an active type of focus.

2. Progressive sensori-neural hearing loss disproportionate to the patient's age, in a patient with stapedial otosclerosis or diagnosed as pure cochlear otosclerosis.

3. Polytomographic demonstration of an active type of lesion of the cochlear capsule, and

4. Vertigo or severe tinnitus in a patient with otosclerosis. (Morrison 1979).

Side effects of sodium fluoride Treatment:-

A reluctance among some physicians to prescribe moderate doses of sodium fluoride for generalized osteoporosis as well as for otosclerosis is due partly to unknown effects of prolonged medication with fluoride and partly to the known cases of severe crippling skeletal fluorine seen in certain parts of India, Japan and China where the drinking water has a very high fluoride content and manual labourers in the hot sun consume large quantities of water.

According to two publications, one by the WHO in Geneva, Switzerland, entitled "Fluorides and Human Health" and the other "Fluoride in Medicine", there has not been a single case reported of permanent harm to a patient from moderate dosage sodium fluoride therapy. Prolonged treatment should be monitored by a bone survey including the spine made at the onset of therapy and repeated every two years to detect early fluorosis.

Gastric irritation is the most common side effect; increase in arthritic symptoms can occur; rarely there may be a typical drug-induced skin rash. There may be chronic nephritis with nitrogen retention where impaired excretion of fluoride could result in toxic levels in the bloodstream although no such case has been reported. There is no evidence that fluoride in moderate dosage harms

the kidneys or any other parenchymatous organ.

The only precautions in giving moderate dosage sodium fluoride are not to give it in severe chronic nephiitis and to obtain a radiologic bone survey every two years to detect the earliest evidence of skeletal fluorosis. When there is a gastric distress, omitting the tablets for a week or two, then resuming them gradually, develop: a tolerance. (Shambaugh & Causse 1974).

Osmotic Drugs:-

Osmotic drugs certainly cannot modify the bone changes of otosclerosis. Nevertheless, it is possible to hypothesize that they influence the cochlear symptoms of the illness by their action on labyrinthine fluids. The validity of this hypothesis was supported by Celesteno & Orofino (1978). They presented two cases.

The first case is that of a 56 old woman affected by post stapedectomy labyrinth degeneration of the right ear with continuous tinnitus not responding to symptomatic drugs. The administration of a dose of g3ycerol provoked in the space of a few hours the reduction of tinnitus to a bearable level. The result was maintained for a period of two years by taking a tiazide diuretic twice a week.

The second case regards a 32 year old woman whose mother and brother both suffer from otosclerosis. At the end of a pregnancy, the patient had a dizzy crisis with vomiting, acute tinnitus and hearing loss in the left ear. Since then the dizziness had not recurred but the hearing loss remained unvaried in the left ear and after four months deafness occurred in the right ear as well. The audiometric examination showed a bilateral hearing loss of a sensori-neural loss type with some doubt about recruitment. The almost flat tracing and the previous dizzy attacks induced them to

experiment with the osmotic effect of glycerol. After the first administration, the audiometric tracing changed radically and after the second dose it took on the definite features of a typical otosclerosis with carhart's notch and negativity in Bing's and Gelle's tests.

In a case of cochlear otosclerosis, after glycerol administration, the audiometric pattern of end-organ disease whowed a change into a classical otosclerosis audiogram. This proves the diagnostic utility of osmotic drugs.

NON-SURGICAL MEASURES

The use of a transistorized hearing aid with an insert a/c receiver gives excellent results in almost all patients with deafness whether stapedial, cochlear or combined otosclerosis. Speech discrimination scores are often high and even patients with moderate sensori-neural loss obtain considerable benefit. Insert hearing aids are indicated post operatively in a group of patients with combined otosclerosis when stapedectomy can produce only a moderate threshold improvement.

SUMMARY

Cochlear otosclerosis implies that otosclerosis may cause sensori-neural hearing loss without an associated conductive loss. In this case, the otosclerotic focus involves the endosteum suffieien to cause degenerative changes in the membranous labyrinth while not causing footplate fixation.

Estimation of the frequency of 'cochlear' otosclerosis is more difficult. Shambaugh (1969) considers thpt pure cochlear otosclerosi Mill prove to be as frequent as stapedial otosclerosis. At first acquaintance this figure seems staggering, but increasing awareness of the entity and of its clinical and radiological manifestations makes the idea more acceptable. His personal records indicate that pure cochlear otosclerosis was diagnosed once for every 20 cases of clinical otosclerosis and the proportion is increasing with experience

Many theories have been advanced concerning the pathogenesis and etiology of cochlear otosclerosis. Constitutional, local and general activating factors are considered. Constitutional factor accounts for the heredity of cochlear otosclerosis. Under local factors, developmental, vascular and mechanical factors are considered General activating factors or the biochemical concept considers the metabolic disturbances. The biochemical concept of cochlear otoscle-rosis has been accepted by majority of the investigators. This concept explains the progressive sensori-neural hearing loss due to cochlear otosclerosis.

The otosclerotic focus which reaches the endosteal layer of the otic capsule may produce degenerative changes in the spiral ligament, stria vascularis, organ of corti, and cochlear neurons. There is often partial spiral ganglion cell loss, and occasionally a degree of labyrinthine hydrops has been observed and even rupture of the saccule

or cochlear duct.

The patient with cochlear otosclerosis complains of a progressive hearing loss which begins in early adult life and which is usually bilaterally symmetrical. There is usually a dominant family history (70%)and relatives, may have proven stapedial disease (12%). Tinnitu is the presenting symptom in pure cochlear otosclerosis arising in pregnancy. Transient episodes of vertigo often positional occur in 20% of patients with cochlear otosclerosis, while the symptoms of hydrops are found in 6%.

The diganosis of cochlear otosclerosis has been made possible by case history, auditory tests, radiologic investigations and histo-pathological studies. The correct identifiecation of cochlear oto-sclerosis is stressed by the possibility that fluoride treatment may effectively halt its advance.

Differential diagnosis of cochlear otosclerosis is very essen-tial as it resembles many other diseases which involve the labyrin-thine capsule. It should be differentiated from the capsular changes that takes place in normal ear, Piaget's disease, osteogenesis imper-fecta, syphilis, fibrous dysplasias, neurofibromatosis, presbycusis and other dominant hereditary deafness in adult life.

In all cases with a sensori-neural or vestibular loss of unknown origin and positive tomographic findings, serological tests for syphilis, determination of serum calcium, phosphorus, alkaline phos-phatase, and a radiographic bone survey should be performed. This additional information would be of great help in accepting or ruling out the diagnosis of cochlear otosclerosis during life (Valvassori 1965)

Cochlear otosclerosis can produce sensori-neural hearing loss

various mechanisms (Carhart 1963; Altmann et al 1966; Frost 1967; Linthicum, Filipo & Bredy 1975). They are as follows:-

- Toxic substances produced by the otospongiotic focus
- Invasion of the basilar turn of the scala tympani
- Rupture of the basilar partition
- Other involvements within the upper cochlear turns
- Vascular shunts between the inner ear vessels and the otospongiotic focus.
- Atrophic changes in the spiral ligament adjacent to the otosclerotic foci
- Atrophy of the stria vascularis
- Atrophy of the organ of corti
- Narrowing of the lumen of the cochlea
- Distortion of the basilar membrane
- Damage to the auditory nerve in the internal auditory meatus by pressure of an ingrowth of otosclerotic bone
- Enzymatic concept of otospongiosis.

Progression of snesori-neural hearing loss due to cochlear otosclerosis can be baited by administration of fluorides. Increased intake of fluoride is regarded as "nature's method" of preventing and combatting osteoporosis either generalized or localized form in the labyrinthine capsule (Shambaugh 1973).

The rationale for the administration of fluorides is based on a number of studies like epidemiologic studies, Invivo studies, Invitro studies and enzyme studies.

There is ample experimental evidence that sodium fluoride in optimum dosage promotes maturation and recalcification of an active immature focus of otosclerosis, thus reducing its enzyme activity and the resultant diffusion of cytotoxic enzymes into the perilymph Shambaugh 1977)

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