OTOSCLEROSIS AND VERTIGO

By MICHAEL M. PAPARELLA (Columbus, Ohio) and
WERNER D. CHASIN (Boston, Mass.)

In recent years we have become increasingly impressed with the inner ear changes which can accompany otosclerosis. There are several examples in the literature which point to the clinical and histopathological features of sensorineural deafness associated with otosclerosis (Ruëdi, 1962; Schuknecht, 1962; Benitez and Schuknecht, 1962). There is very little reported on the vestibular changes in this common disorder.

An occasional patient with otosclerosis will also suffer from vertigo. This may or may not be associated with sensorineural deafness. It is possible that the mechanisms which cause sensorineural deafness in otosclerosis might also affect the vestibular labyrinth thereby producing vertigo.

When patients with otosclerosis have significantly reduced discrimination along with a sensorineural loss it is sometimes difficult to exercise the correct judgment in selection for stapedectomy. By the same token it may be difficult to elect to do a stapedectomy when vertigo is present. This decision is not made easier when a sensorineural loss and a discrimination loss exists in addition to the vertigo.

It is our purpose here to present the case histories of three representative patients with otosclerosis and vertigo in whom stapedectomy was performed. In each patient the diagnosis of otosclerosis was confirmed at the time of surgery. The vertigo was categorized as Ménière's disease in the first case, probable vestibular neuronitis in the second, and undefined but true vertigo in the third.

These three patients were selected from a somewhat larger group of otosclerosis patients with dizziness as part of their initial complaint. The majority of these patients had a poorly defined dizziness without objective vestibular findings which were not materially altered by stapedectomy.

In 1955 Cawthorne described momentary dizziness after head movements in 25 per cent. of cases of otosclerosis. Caloric tests of function were normal.

Otosclerosis With Ménière's Disease

Documented cases of otosclerosis combined with Ménière's disease are difficult to find in the literature. Understanding the mechanisms at

https://doi.org/10.1017/S0022215100065579 Published online by Cambridge University Press

work in this combination of disorders is difficult since the pathogenesis of neither otosclerosis nor Ménière's disease is fully understood.

Case Presentation—I.

Mr. T.M. is a 55-year-old engineer and supervisor whose job includes crawling through tunnels. His chief complaint was severe vertigo. The patient had no otological symptoms until 10 years prior to being seen when he began to notice a bilateral hearing loss which was insidious in onset and which progressed gradually throughout the following years.

Four years thereafter he developed his first episode of vertigo. This lasted approximately 1 hour and was associated with nausea. For the next 2 years he was symptom free with the exception of his progressive hearing loss. Then he began to experience intermittent episodes of vertigo which were associated with nausea and which occurred several times a year. During the past year his episodes of vertigo had increased to at least one attack per month, lasting approximately an hour and associated with nausea. During an attack the room spun about him from left to right. The attacks were ushered in by a visual aura. Each attack was accompanied by a sensation of fullness in his right ear. Although he had constant tinnitus in his right ear this seemed to intensify during an attack. During the preceding 5 months the frequency of attacks increased to the point where he lost 1 day from work each week. He felt that his hearing had decreased in his right ear in recent months. He had worn a binaural hearing aid for the past 3 or 4 years. He had no loudness intolerance or diplacusis.

The ear, nose, and throat examination was normal except for the auditory and vestibular findings. The Weber lateralized to the left. The Rinne was negative on the left side and positive on the right. There was no spontaneous nystagmus, but on right lateral gaze there was slight horizontal-rotatory nystagmus.

Audiometry revealed a bilateral severe hearing loss with surprisingly good discrimination in both ears (Fig. 1). There was primarily a conductive loss in the left side with a slight loss in bone conduction. There was a severe sensorineural loss in the right ear. Speech reception thresholds were 82 in the right and 58 in the left. Discrimination scores using *PB-Max* words were 88 per cent. in the right and 84 per cent. in the left ear. The Sisi response was 20 per cent. in the right ear at 2 kc. and 0 per cent. in the left ear at 2 kc. A consultation with otoneurology revealed a right canal paresis with the Hallpike-Fitzgerald Caloric test. The patient had no positional vertigo or nystagmus.

On the basis of these findings it would appear that the patient had developed bilateral otosclerosis with conductive hearing loss 10 years previously followed by the superimposed development of a sensorineural hearing loss more so in his right ear accompanied by vertigo 6 years before his admission.

Since the patient was incapacitated by his vertigo and since previous medical management had been to no avail it appeared that some form of surgical intervention should be employed. Partly because of the surprisingly good discrimination score in his right ear possible intervention

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by labyrinthectomy was deferred. Instead a stapedectomy procedure was elected for the right ear realizing that an improvement of hearing would probably not be forthcoming. Since the patient had otosclerosis first and Ménière's syndrome second in the involved ear a stapedectomy was advised on the basis of performing a conservative procedure which

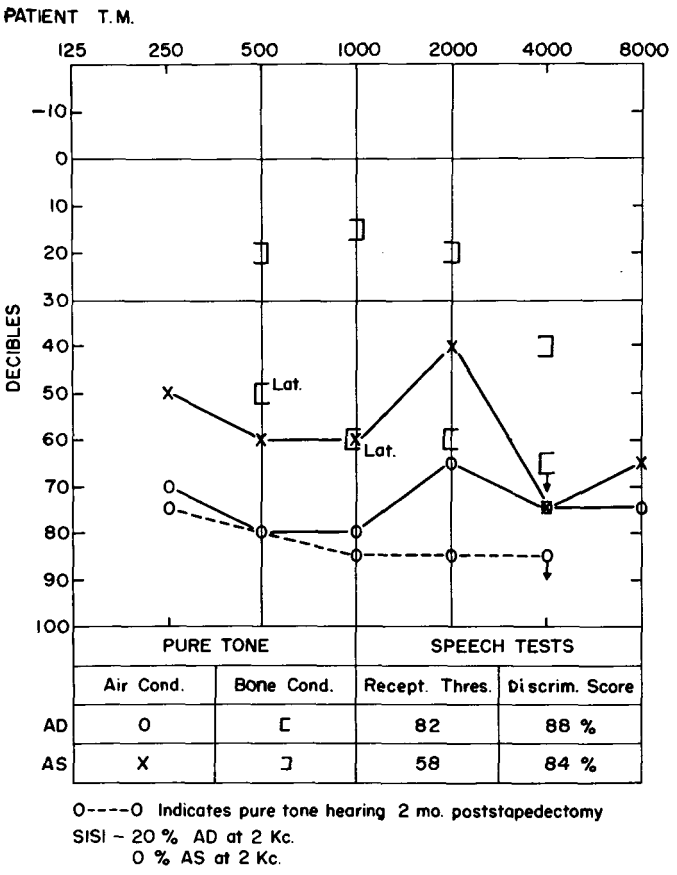


FIG. 1.

Patient with otosclerosis and Ménière's syndrome showing pre-operative and post-operative audiogram.

might benefit the vertigo. It was also felt that a conservative oval window procedure would provide less post-operative morbidity and easier recovery than a labyrinthectomy. The patient was informed that a subsequent labyrinthectomy might be necessary if this more conservative attempt was not successful.

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This patient was operated upon on October 30th, 1963. The stapes footplate was removed atraumatically but a small amount of blood entered the vestibule. A Schuknecht fat-wire prosthesis was employed. There was no vertigo during the procedure.

The patient's recovery from this stapedectomy was uncomplicated. The patient has now been seen periodically since surgery for more than two years. He has had one mild episode of vertigo occurring 2 months post-operatively which was mildly reminiscent of but not as severe as the attacks which occurred prior to surgery. Since that time he has not had further vertiginous attacks. The hearing has not changed substantially in the operated ear. He continues, of course, to wear a hearing aid in the unoperated ear with otosclerosis.

During this relatively short follow-up period there has been an unquestionable and remarkable improvement in the patient's problem with vertigo. However, we must wait longer and take into account a possible unrelated spontaneous remission such as frequently occurs in the natural course of Ménière's disease.

In this patient the history suggests the development of bilateral otosclerosis with superimposed Ménière's disease developing subsequently on the right side. The diagnosis of Ménière's disease was based upon presentation of typical symptoms along with a severe sensorineural hearing loss which had become worse of late plus a canal paresis on the involved side. These symptoms and findings appeared typical of Ménière's disease. The only missing factor was recruitment in the involved ear but it is possible that other tests might have demonstrated its presence. The diagnosis of otosclerosis was confirmed at time of stapedectomy.

More recently we have encountered two other patients who appeared to have the coexistence of the symptoms of Ménière's disease and otosclerosis in the same ear. These patients are also seemingly improved after stapedectomy and are currently being followed. During the stapedectomy in one of these patients the saccule was perforated with a very fine needle. The other patient had bilateral otosclerosis with excellent bone conduction thresholds and typical Ménière's symptoms and findings with the exception of sensorineural deafness in the right ear. At stapedectomy there was no apparent bulging of the utricle or saccule. There has been no vertigo in the post-operative period thus far and there has been a remarkable improvement in hearing.

In our cases we can only conjecture as to the effect the stapedectomy procedure had on the presumed endolymphatic hydrops of the inner ear. Two hypotheses seem possible. One that the placement of a soft tissue graft in the oval window served to produce a yielding membrane which might decompress or decrease the pressure effects of the hydrops at least within the vestibule. An alternative explanation appears more tenuous and would be that the graft itself or the small amount of blood which

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entered the vestibule might have a chemical, metabolic or toxic effect on the membranous labyrinth thereby altering permeability characteristics.

The following two patient presentations are other examples of true vertigo occurring with otosclerosis. Although definite diagnosis are difficult probable vestibular neuronitis was diagnosed in Case II and undefined vertigo in Case III.

Case Presentation—II.

Mrs. I.L. is a 37-year-old white female who was first seen in October of 1962 with a complaint of bilateral hearing loss and vertigo. Her vertigo began suddenly 1 year before but was relieved while she took Antivert. The vertigo sometimes was associated with headache and photophobia. Two days later she developed severe vertigo with nausea and vomiting which lasted 1 day followed by a marked unsteadiness for about 4 to 5 days. About 2 months later she noticed that sand between her toes on the beach had a very strange sensation. Because of progressive bilateral hearing loss of a conductive nature on both sides, worse on the left ear, the patient was advised to have a stapedectomy after the dizziness had subsided for 2 months. A stapedectomy using a fat-steel prosthesis was performed in April, 1963. The footplate was blue and fixed and the surgical procedure was atraumatic. Following the operation the patient had an excellent hearing result (Fig. 2). Approximately 2 months after the stapedectomy the patient noted unsteadiness again and became aware of a pins and needles sensation in her feet.

The patient was admitted to the hospital for a thorough medical and neurological evaluation. On sensory examination there was a dysaesthesia over both feet extending upward to both knees. Vibration and position sensation appeared normal. The Hallpike-Fitzgerald caloric test revealed a canal paresis of the right ear. Spinal fluid obtained by lumbar puncture was normal in pressures, dynamics, cells, protein, and colloidal gold curve.

The history of vertigo 1 year before followed by prolonged unsteadiness without evidence of cochlear involvement, the onset being accompanied by vomiting, headache and photophobia would be compatible with a diagnosis of vestibular neuronitis. The possible presence of peripheral neuropathy has not been described as part of this syndrome and raises other questions such as multiple sclerosis.

In 1952 a careful study was presented by Dix and Hallpike of the syndrome termed vestibular neuronitis. In contrast to Ménière's disease there are no cochlear symptoms or findings in this disorder. The onset of vertigo may be sudden and may become recurrent just as in Ménière's disease. In Harrison's (1962) 67 patients with vestibular neuronitis all demonstrated abnormal responses to caloric testing.

This patient's history and findings are typical of otosclerosis. This was confirmed at surgery. In Hallpike and Dix's description of vestibular neuronitis the absence of auditory findings was stressed, but of course the reference was to sensorineural changes. It is conceivable that the picture of vestibular neuronitis might be seen with the concomitant presence of

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otosclerosis in the same ear. In this patient the canal paresis was in the unoperated ear. This was the case both pre- and post-operatively. In explaining her findings it is possible that the unoperated ear might have vestibular neuronitis. This then accounts for the unaltered existence of her vestibular symptoms following the stapedectomy since the procedure was done in the ear with normal vestibular function by testing.

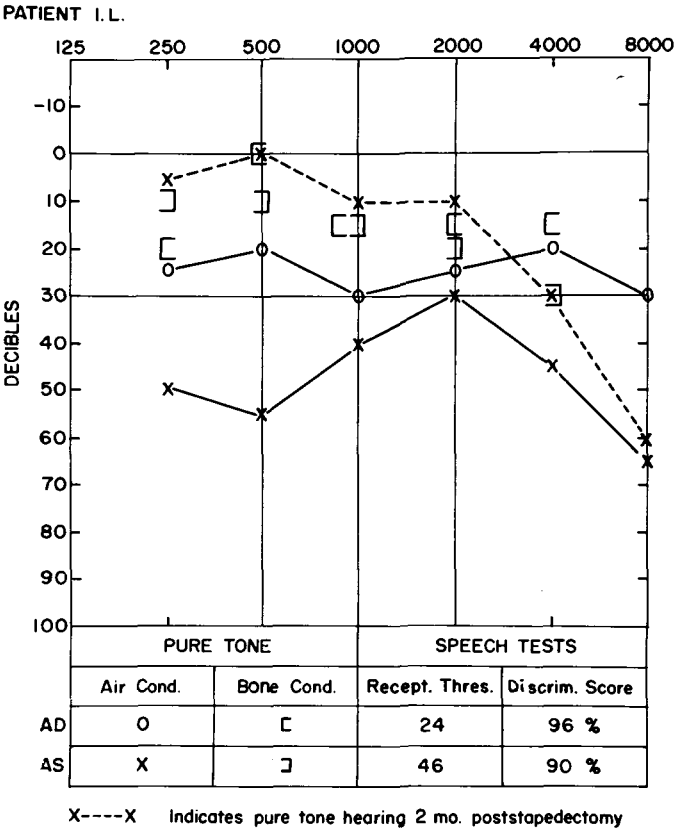


FIG. 2.

Patient with otosclerosis and vestibular neuronitis showing pre-operative and post-operative audiogram.

Vestibular neuronitis is a clinical syndrome and the causative mechanisms at work are not understood. A diagnosis of vestibular neuronitis is not precluded because of the presence of a conductive hearing loss and otosclerosis. Providing the conductive loss in the ear with vestibular findings was substantial enough to warrant surgery one wonders what effect this might have on the patient's vertigo.

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Otosclerosis and Vertigo

In most patients with otosclerosis and dizziness it is difficult to categorize the vertigo into one or another of the accepted syndromes. Many patients will complain of lightheadedness, giddiness, swimming sensations but few will present the picture of true vertigo. The following case demonstrates just such a patient—that is one with otosclerosis and definite vertigo, but without vestibular or neurological findings.

Case Presentation—III.

Mrs. M.O. is a 59-year-old female who gave the history of progressive hearing loss of the right ear for 30 or more years, and of the left ear for the past 10 years. She also had tinnitus and vertigo. The vertiginous episodes occurred several times a year and would last several hours to 1 day and would be true vertigo associated with nausea and vomiting. Approximately once every 4 or 5 years the patient had a more severe episode of vertigo. The hearing did not fluctuate but she did notice a pressure sensation in her ears associated with the vertigo. A careful pre-operative medical evaluation found her to be in excellent health. Examinations of the ear, nose, and throat including caloric testing indicated normal results. The hearing test showed a bilateral conductive loss worse in the right ear with a superimposed high tone sensorineural loss (Fig. 3). Discrimination was excellent in both ears. Accordingly the patient was advised to have a right stapedectomy in March of 1964. Because of a severe nervous tic of the right shoulder which would not allow the procedure to be done under local anæsthesia, the patient was placed under general anæsthesia. There was also a history of tachycardia and a cardiac monitor was utilized. There were no problems during the procedure. The posterior fragment of the footplate was extremely thick and was removed separately from the anterior fragment which was thinner. Some bleeding occurred into the vestibule. The stapes was then replaced with stainless steel wire and adipose tissue. Post-operatively the patient developed an initial excellent hearing result with a subsequent drop of discrimination to 66 per cent. in the operated ear (Fig. 3). A complete closure of the bone air gap was achieved without any significant decrease in the bone conduction thresholds.

Post-operatively the patient had vertigo persisting intermittently for a period of 2 weeks. This finally subsided. The drop in the discrimination has been disconcerting to the patient post-operatively.

In considering otosclerosis and vertigo this patient represents an example of true vertigo with otosclerosis but without vestibular findings. The role which otosclerosis plays in the production of vertigo is possibly through a medium of biological alteration of perilymph producing a disturbance but not a measurable dysfunction in the vestibular labyrinth. Of course it is impossible to estimate what specific effect stapedectomy has in this type of case particularly because we cannot be certain as to which ear or both the vertigo relates. It is possible that there is bilateral disturbance or that a disturbance exists in the unoperated ear.

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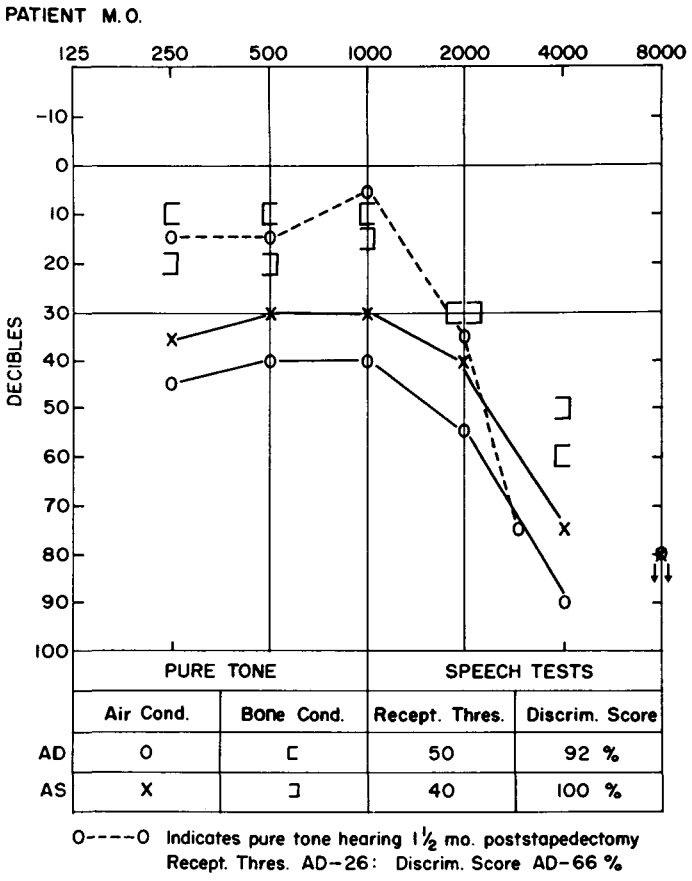


FIG. 3.
Patient with otosclerosis and undefined true vertigo showing pre-operative and post-operative audiogram.

Conclusions and Summary

In all three patients presented it is impossible to state with any certainty whether or not their vertigo is a manifestation of a disease process separate from otosclerosis or if the vertigo is a result of the influence of otosclerosis on the vestibular labyrinth. Certainly it is feasible for a patient to develop in the same ear both peripheral vestibular disease and otosclerosis with unrelated ætiologies. However, it seems more reasonable to assume that the vertigo is in some way related to otosclerosis and develops as a result of alteration of the membranous vestibular labyrinth. When one considers the propensity for otosclerosis to involve the fissula ante fenestram and oval window area overlying the

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otolithic structures it seems that on the basis of proximity alone the vestibular structures may be involved just as we know the cochlear structures are.

Our objective here has been to present three patients with otosclerosis and vertigo in whom Ménière's disease was diagnosed in the first, probable vestibular neuronitis in the second, and undefined true vertigo in the third. It is difficult to categorize with any specificity vertigo occurring in combination with findings of otosclerosis.

It is our hope that further interest in this combination of disorders will be aroused so that a better understanding of these patients will be possible and a better criterion for selection of patients for stapedectomy will be established.

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Department of Otolaryngology,
Ohio State University,
Columbus, Ohio.

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