

RESISTANT CASES OF MENIERE'S DISEASE*

Treatment by Labyrinthotomy

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MUCH has been written about Meniere's disease and its treatment since Meniere's initial classic description in 1861. Many theories and explanations have been offered in an attempt to clarify the problems involved. This fact alone signifies that a certain amount of confusion, with regard to the etiology and pathology as well as the treatment of the disease, has existed. It has mainly been in the past decade that writers such as Day¹ have clarified the meaning of the term "Meniere's disease," and differentiated between it and the pseudo-Meniere's disease, which Dandy used to label those cases which had only vestibular involvement.

The term Meniere's disease is reserved for that group of patients presenting the syndrome comprising vertigo, tinnitus, deafness and sometimes nausea and vomiting, for which no definite systemic or local condition can be blamed. Heretofore, many patients with dizziness and nausea or vomiting were diagnosed as having Meniere's disease even though there was no cochlear involvement. Meniere's syndrome, or components, may be caused by a variety of conditions but now, it is generally agreed, only the patients belonging to the idiopathic group warrant a diagnosis of Meniere's disease.

Etiology

Investigators have attempted to prove the etiology of Meniere's disease for many years. Meniere's syndrome may have toxic origin such as poisoning from lead, arsenic in alcohol, mercury, salicylates, and other drugs. It is also present with chronic infections, the exanthemata, syphilis, tuberculosis, the anemias, leukemias, virus infections, purpuras, tumors of the cerebellopontine angle, trauma, and gastrointestinal conditions.² Among the simple local causes are impacted cerumen and blockage of the eustachian tube. The syndrome may be present in catarrhal or suppurative otitis media, vasospasm, hemorrhage or thrombosis of labyrinthine blood vessels, arteriosclerosis of cerebral vessels,

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neoplasms, labyrinthitis, multiple sclerosis and other degenerative central nervous system diseases.³ Some cases have been reported on the basis of allergy. Diet is thought to be a factor by some investigators. The basis of Meniere's syndrome or symptom complex may be summarized as vascular, inflammatory, degenerative or neoplastic lesion, which may be peripheral or central.

In 1938 Cairns and Hallpike⁴ gave a historic paper on the histopathology in Meniere's disease. They reported on sections of the temporal bones of three patients who had been previously operated upon for Meniere's disease by sectioning of the eighth cranial nerve. Their findings consisted of a dilated endolymphatic system, degeneration of the organ of Corti and stria vascularis and a pathologic elevation of the otolithic membrane of the utricle. The basic pathology appears to be the dilatation of the cochlear duct which encroached on the utricle and saccule. They postulated that the crisis in the disease did not occur until dilatation of the membranous canal was completed to the full extent of the bony canal. After this was accomplished, any further increase in pressure of the endolymph precipitated a crisis. It has since become accepted by many investigators that hydrops of the labyrinth is the pathologic change peculiar to Meniere's disease.

However, postulation continued regarding the endolymph,⁵ the decreased resorption of it, or possible chemical changes in the endolymph and perilymph. This point has not been proved. One theory is that a disturbed sodium metabolism with retention of sodium ions is the underlying causative factor. Another is that it is a vascular disturbance, probably a vasoconstriction. Williams⁶ feels it is basically an inherent allergic problem. Lindsay's⁵ work with monkeys showed that the volume of endolymph did not depend upon a functioning ductus endolymphaticus or even upon its existence. He also demonstrated that the hydrops of Meniere's disease could not be attributed to interference of absorption of endolymph from its dilated and convoluted middle portion.

Symptomatology

Meniere accurately described the symptoms of the disease that bears his name. The symptoms of tinnitus, deafness, vertigo, nausea and vomiting without neoplastic, vascular, or inflammatory cause associated, are the basis of the diagnosis. The disease occurs in episodes or attacks which are characteristically remittent. The symptoms may completely or only partially disappear between attacks.

The vertigo may take various forms. It is vertigo in its most accurate sense if the patient experiences a sense of motion, rotary or linear, and a disorientation in space. It may be objective, in which case the surroundings seem to move; or subjective, in which circumstance the individual feels as if he were moving. This may also take the form of tactile error in which the floor seems to tilt, or roll like the deck of a ship at sea. The vertigo may be of sufficient severity to cause staggering or even falling. Motion of the head aggravates this condition and so the patient tends to remain quiet and to assume the most comfortable position.

The deafness may be of varying degrees, but usually involves the entire scale. There is decreased bone conduction and the audiogram shows a "nerve deafness" curve. Diplacusis and distortion occur and may be annoying symptoms. It has been noted and emphasized that cochlear involvement is bilateral⁷ which may help to explain the distortion. It is also believed that the sound is carried or referred farther down the basilar membrane than is normal for its pitch, as a result of the increased pressure in the endolymph. The diplacusis and distortion tend to disappear as the deafness becomes more critical.

The tinnitus usually has two components which vary in intensity: One is a high-pitched hissing, ringing, or whistling; the other is a low-pitched roaring or throbbing and may be synchronized with the heartbeat. The latter is often the more troublesome and annoying.

Nausea and vomiting are not always present. These two symptoms indicate acuity and severity and may necessitate hospitalization for parenteral feeding to prevent prostration in extreme cases.

The symptoms at the time of onset are unilateral in approximately 90 per cent and bilateral in about 10 per cent of the cases.⁵ However, evidence of bilateral cochlear involvement has been shown in as many as 86 per cent of a series.⁷ Other authors state that bilateral involvement is more frequent than formerly presumed. There are varying degrees of the disease. Early in the course there may be symptoms of involvement of either the vestibule or cochlea, but eventually both are affected.

Caloric tests for vestibular function show a nonfunctioning or hypoactive labyrinth on the affected side. The supposedly unaffected side may show a similar diminution in response. Usually the more severe the deafness, the less the caloric response. The caloric response will simulate the spontaneous attacks.

Diagnosis

In arriving at a diagnosis one must take a careful history, do a thorough ear, nose and throat examination and eliminate general or systemic causes of the symptoms. The history is of such great importance that a diagnosis can almost be made from it alone. It is essential to discern the type of dizziness, whether subjective or objective, and distinguish between vertigo and a mere giddy or lightheaded sensation. Other salient points are to be considered such as: Is it associated with tinnitus, deafness, nausea or vomiting? Does it occur in attacks, or is it constant? Cochlear involvement must be present to make a diagnosis of true Meniere's disease.

Inasmuch as many general or systemic conditions or lesions outside the labyrinth can cause certain symptoms of Meniere's syndrome, it is necessary to exclude them by history, physical examination and the aid of the laboratory. A complete ear, nose and throat examination is done to eliminate any apparent local cause. Such examination may disclose blocked eustachian tubes, cerumen

impaction or local inflammation. Conversely, there may be no obvious cause for the symptoms and the physical examination will reveal nothing abnormal except a nerve deafness curve on the audiogram and diminished caloric response of the labyrinth. Spontaneous nystagmus of the rotary type is also an important indication of labyrinthine involvement but it is not always present. It is usually experienced during or shortly after an attack. The possibility of an acoustic neurinoma or other intracranial lesions should not be overlooked.

Having precluded inflammatory, vascular, neoplastic and general systemic causes; having localized the lesion to the vestibule with cochlear involvement; and having confirmed the syndrome of vertigo, tinnitus, and deafness with possible nausea and vomiting, one may arrive at a diagnosis of Meniere's disease.

Medical Treatment

Once the diagnosis is established, medical treatment should be the first alternative. This form of management affords definite improvement or controls the vertigo in 85 per cent, improves or relieves tinnitus in 50 per cent and improves hearing to some extent in 20 per cent.⁸ The estimates and affirmations vary depending upon author and treatment employed, but the foregoing figures are a general average.

The various forms of medical treatment depend upon the accepted etiology. One theory is that Meniere's disease is due to water imbalance with retention of fluid; thus, the therapy is limitation of fluid intake to induce dehydration. Another is that electrolyte imbalance is the causative factor and potassium is given as treatment. Still other investigators claim good results by nicotinic acid administered to relieve vasoconstriction.⁹ One of us first reported the beneficial effects of nicotinic acid in March 1940.

In our experience the nicotinic acid regimen and a hypo-allergic diet have brought favorable results in between 80 and 90 per cent of the cases treated. Occasionally, dramamine has been added when the patient did not respond satisfactorily; this has proved valuable in some of the more resistant cases. Other patients may obtain benefit from a mild sedative added to the regimen.

Surgical Treatment

There are two sites of surgical attack for operative treatment. One is the acoustic nerve, which is partially or completely sectioned, and the other is the end organ in the inner ear which is destroyed by various means. Both have their proponents. More recently a third site, the autonomic nervous system, has been interrupted, based on the belief that the etiology of Meniere's disease is vasoconstriction.

Ray¹⁰ and others have used the intracranial sectioning of the acoustic nerve, either partial or complete. The partial sectioning was done on the anterior one-half to five-eighths of the nerve in an attempt to interrupt the vesti-

bular fibers and preserve the cochlear fibers and the hearing.¹¹ Better results in relief of the vertigo are obtained by complete sectioning of the nerve as no vestibular fibers are missed. The vertigo has been relieved or improved in 95 to 100 per cent of these patients⁸ and the tinnitus has been partially relieved or improved more decidedly by total section than by partial section of the nerve. Hearing has been preserved in some with partial section, but these patients are also more likely to have some persistent vertigo and tinnitus. The hearing is usually not serviceable in these patients who are selected for surgical treatment; consequently little is gained by preserving it.

A number of operations on the end organ have been performed with varying results. It was hoped that a means of relieving the vertigo and tinnitus could be devised at this site and still preserve the hearing. In England investigators^{12,13,14} injected alcohol through the oval window or a trephine in the lateral semicircular canal into the labyrinth after a mastoid approach was effected. This always destroys hearing but frequently results in a facial paralysis. The drainage of the saccus endolymphaticus affords only temporary relief and others have not had as much success with this procedure as Portmann who introduced it. Experiments with monkeys⁵ showed that the drainage and destruction of the saccus endolymphaticus was followed by sealing and healing without effect on the labyrinth beyond; and, therefore, could only give temporary relief of vertigo though it preserved hearing.

It has also been demonstrated that ablation or avulsion of the membranous semicircular canal destroys the function of the canal but has no effect on hearing if the ampullary end remains intact. Cawthorne removed the membranous canal and ampulla, and though the vertigo was cured, hearing was lost.

One operator used the intracranial route to apply a coagulating current within the superior semicircular canal in an attempt to destroy the vestibule and preserve the cochlea. Day^{1,3} admitted this method was plausible, but not the best approach. Subsequently, he devised the following method: Through a partial, simple mastoidectomy by the postauricular route, he opened the horizontal semicircular canal near the ampulla and inserted a coagulating needle into the vestibule. A light coagulating current was applied two or three times for about one second each. He was successful in relieving vertigo with little postoperative morbidity or risk to the patient. At first he reported the effect on hearing varied from complete loss to return to normal. More recently he and others report loss of hearing as the more frequent result. The low-pitched tinnitus is usually relieved; the high-pitched type persists but it is not often a serious complaint.

The success of this procedure led to further investigation as to its effect on the end organ and the possibility of controlling the current so as to preserve the cochlea and destroy only the vestibule. Experiments on rabbit ears treated by coagulation and trauma showed pathologic change in the vestibule but none in the auditory apparatus. Thus, the cause of deafness following the coagulation procedure was unexplained. Simple decompression of the perilymph was also shown to be valueless.⁵

Schlandler¹⁵ reported that it is not the coagulating effect of the current that destroys the labyrinth, but that hemorrhage into the perilymphatic space at the time of surgery is followed by a serofibrinous labyrinthitis which destroys the inner ear. Schall and Rambo,¹⁶ working with monkeys, concluded that where optimum current is used, the damage to the vestibular mechanism is unpredictable and that retention of serviceable hearing is improbable. The facial nerve was not damaged in any of these animals. Lempert¹⁷ states that tinnitus and vertigo should both be relieved by aseptic decompression of the cochlea and vestibule. He reported success in relieving tinnitus in nine of ten, and vertigo in all ten cases by opening the oval and round windows.

Passe and Seymour² reported on a series of operations in which the autonomic nervous system was interrupted. They pointed out that the blood supply to the inner ear is from the internal auditory artery, from the anterior inferior cerebellar artery, or from the basilar artery which is formed by the union of the vertebral arteries. After the internal auditory artery goes through the internal auditory meatus it divides into the cochlear and vestibular arteries which may be equal or unequal in size. The variability of distribution could account for the predominance of vestibular or cochlear symptoms in some cases by accepting the vasoconstriction theory as the cause of Meniere's disease. Also a smaller sized vessel could have greater vasoconstrictive power by virtue of its size.

Since, it has been shown that vasoconstrictor fibers are exclusively sympathetic ganglionic; that there are intramural fibers in some vessels, and that vasospasm recurs after periarterial sympathectomy; it was considered necessary to destroy the cell station in the stellate ganglion and to strip and cut the vertebral artery to interrupt all vasoconstrictor impulses. A neck approach was used; the vertebral artery was cut and ligated; the stellate ganglion was excised, and the preganglionic fibers of T1 and T2 were cut.

These patients develop a Horner's syndrome immediately which gradually improves over a period of months. The authors claim relief of vertigo and improved hearing with varied effects on tinnitus. Arteriosclerotic patients are never subjected to this operation because of the danger of thrombosis of the posterior inferior cerebellar artery; nor is the operation ever performed bilaterally. This is a radical procedure and further investigation is necessary to prove its worth.

Selection of Cases

Every case presenting the symptoms of Meniere's disease should be observed for a sufficient period of time to confirm the diagnosis of Meniere's disease according to the criteria mentioned previously in this paper. Medical treatment should then be given an adequate trial as it will control about 80 to 90 per cent, and the disease characteristically goes through remissions which may last for many months or years.

If medical treatment fails after a four to six months' period, surgical relief

should be advised. Before deciding to operate upon these patients, one must carefully determine the presence and extent of general systemic diseases which might offer a contraindication to surgery. Life expectancy should be such that the procedure is practicable. The economic position of the patient is also a factor in determining the extent of his incapacity. If he has a hazardous occupation or cannot afford to stay away from work and spend time and money for prolonged medical treatment, he should have the operation. The amount of hearing loss is also a factor in deciding upon surgery, as the procedures which relieve vertigo with the best results also destroy hearing. Therefore, we feel that hearing should be below the serviceable level on the side to be operated upon. Furthermore, the operation should be performed on one side only as those who have the vestibular function destroyed bilaterally do not compensate for this loss. The symptoms are usually due to the disease in one ear and operation gives relief. If the other ear causes symptoms, the attacks are well controlled by medical management as a rule, especially if treatment is started when the symptoms originate.

While there are valid points in favor of each of the various procedures, we feel that the labyrinthotomy with electrocoagulation of the labyrinth is the treatment of choice in the proved cases resistant to medical management. Our reasons for this conclusion are that the results are excellent for relief of vertigo, the procedure is relatively short, requiring about 30 minutes, is fairly simple, and the risk to the patient is minimal; a craniotomy is avoided. This method attacks the site of pathologic change in the labyrinth, and is therefore logical. It requires only a short period of hospitalization of five to seven days, recovery is rapid, and no facial paralysis has occurred in our cases.

Summary

The vertigo was relieved in all these 12 patients observed by us and the hearing, which was below the serviceable level in the ear operated upon, was further diminished by the procedure. The effect on the tinnitus was variable, but for the most part the condition was not improved. These patients were definitely benefited and rehabilitated economically. Each case is considered as having been successful. No facial paralysis occurred; there was no post-operative infection, and healing was rapid. Antibiotics were used. The hospital stay averaged six days.

The etiology, symptomatology and diagnosis of Meniere's disease were reviewed. It is emphasized that medical treatment gives good results in the vast majority of cases and should be given a thorough trial before surgical treatment is considered. The various surgical procedures are discussed with emphasis on labyrinthotomy and destruction of the membranous labyrinth by electrocoagulation. Factors in selection of patients for surgical treatment are reviewed and reasons advanced for choosing this procedure in 12 cases treated over the past two and a half years.

Conclusions

1. Patients with Meniere's disease who cannot be controlled by medical treatment should have the benefit of surgery.

2. Labyrinthotomy and destruction of the membranous labyrinth has proved effective in the control of vertigo of labyrinthine origin.

3. The procedure should be reserved only for the few carefully selected patients that defy good medical management and who have hearing in the involved ear that is below the serviceable level.

4. This is not the final answer to this problem even though the patients are rehabilitated economically, since hearing is not restored to normal.

5. A plea is made for the early diagnosis and recognition of the true nature of this disease, as vasodilators such as nicotinic acid (especially when given early) will nearly always control the symptoms and prevent or delay its progress, thus preserving hearing at serviceable levels.

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