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It is nearly 150 years since Prosper Meniere described this condition accurately. His description of the disorder still holds good. This book is being released as a tribute to this man

Meniere's disease

Second Edition Incorporating
recent advances

Dr T Balasubramanian

Meniere's disease

By
Dr. T. Balasubramanian M.S. D.L.O.

History:

In late 19th century Prosper Meniere described a condition characterized by ear block, tinnitus, and vertigo. He even correctly identified the site of lesion to be labyrinth. It would be an understatement to say that precious little has been added to the knowledge and understanding of the disorder since then. Prosper Meniere in fact lived far ahead of his time. He was born in 1799 in France. In 1848 he began to translate the text book on hearing loss authored by Kramer. The book was written in German. This kindled his interest in otology.

In his classical seminal reports he goes on to describe a series of patients who presented with neural deafness, with hearing loss greater for low frequencies. Deafness was commonly unilateral in these patients. These patients usually present with tinnitus, vertigo, nausea and vomiting. He reported that these patients had a normal ear drum. He also reported that these symptoms were completely reversible.



Prosper Meniere

It was Galen during 130 - 200 AD who coined the term labyrinth because he had faced tremendous difficulties trying to fathom the functioning of inner ear.

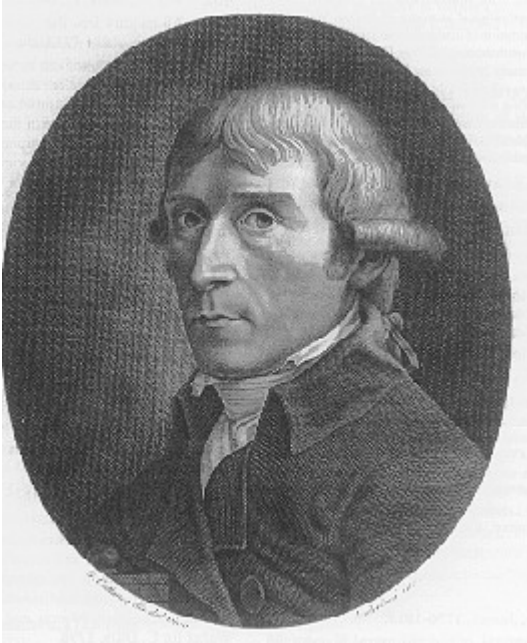


Galen

Studies of Egyptian Papyruses have demonstrated the emperic therapies devised by Greek physicians like Hippocrates to manage tinnitus / heaing loss etc.

The popular belief that inner ear is filled with ear was disproved by Domenico Cotugno (1736-1822). He successfully demonstrated the presence of inner ear fluid and christened it as endolymph. Cotugno was successful because he used fresh specimen in his dissection. He also demonstrated the presence of vestibulat aqueduct.

Antonio scarpa in 1747 described the correct anatomy of the membranous labyrinth. He named the fluid filling the membranous labyrinth as Scarpa's fluid.



Antonio Scarpa

It was Erasmus Darwin grandfather of Charles Darwin was the first to associate vertigo and tinnitus with inner ear pathology.

It was Georges Portmann who evolved the procedure "Endolymphatic sac decompression" as a treatment modality for Meniere's disease.



Georges Portmann

Walter Dandy was the first to treat Meniere's disease by sectioning the vestibular nerve. This procedure produced instant relief from tinnitus. The fact that Dandy was a trained neurosurgeon helped him in this procedure.

Incidence:

Incidence of Meniere's syndrome in the general population is highly variable. Crude estimates put the value to be 8 per 100,000 individuals.

Studies have demonstrated a significant female predominance.

Meniere's syndrome is rare in children. It is commonly seen in adults.

Etiology of Meniere's disease

The exact etiology of Meniere's disease is unknown, however various etiologies have been suspected.

Etiological factors of Meniere's disease:

- 1. Genetic**
- 2. Anatomical causes**
- 3. Traumatic**
- 4. Viral infection**
- 5. Allergy**
- 6. Autoimmunity**
- 7. Psychosomatic and personality disorders**

Etiological features of secondary endolymphatic hydrops:

- 1. Developmental insult**
- 2. Abnormal metabolic / endocrine states**
- 3. Syphilis**
- 4. CSOM**
- 5. Viral infection**
- 6. Autoimmunity**
- 7. Otosclerosis**
- 8. Abnormal fluid balance**
- 9. Leukemia**

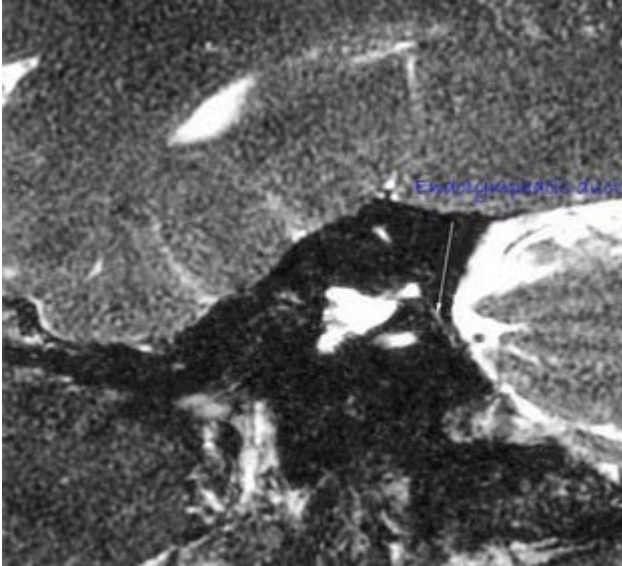
Genetic causes:

Familial tendency has been observed in nearly 20% of patients with Meniere's disease. Studies have demonstrated that Meniere's disease is attributable to a mutation on chromosome 6. Transmission is supposedly autosomal dominant in nature.

Anatomical causes:

1. Small vestibular aqueduct: Radiological studies of patients with Meniere's disease demonstrated a smaller vestibular aqueduct in nearly 10% of patients.

Considerable difficulty was experienced in visualizing the endolymphatic duct / sac in ears affected by Meniere's disease.



CT temporal bone showing narrow endolymphatic duct

2. Reduction in the rugose portion of the endolymphatic sac has been detected in a significant number of Meniere's disease patients.

Traumatic causes:

Association between Meniere's disease and trauma (physical / acoustic) has been implicated in Meniere's related symptoms. Trauma may cause biochemical dysfunction in the cells of the membranous labyrinth, or may simply cause release of debris into the endolymph causing obstruction of the endolymphatic duct / sac.

Viral infection:

Damage to the endolymphatic sac and duct by viral infection has been proposed as an etiological mechanism in Meniere's disease. Neurotrophic viruses have been implicated in this process. Researches in Sweden have identified a higher reactivity to herpes simplex virus type I in patients with Meniere's disease. DNA of herpes virus has been isolated from the endolymphatic sac of affected individuals.

Circulating levels of group specific proteins of enterovirus VP1 have been found to be elevated in patients with active disease. Absence of the protein can be correlated with remission.

Allergy:

Nearly 80% of patients with Meniere's disease have history of childhood allergy. Both food and inhalant allergens have been implicated. Treatment of allergy with immunotherapy caused a remission of the disease in majority of these patients. IgE changes have not been demonstrated in these patients causing a doubt regarding this etiological factor.

Autoimmunity:

Autoimmunity as an etiological factor has been considered in Meniere's disease. The endolymphatic sac has been shown to contain immunoglobulin and lymphocytes and is capable of generating immune response. Immunoglobulins have been found to be deposited in the walls and the luminal fluid of endolymphatic sacs of Meniere's disease. Elevated levels of immune complexes have been demonstrated in nearly 20% of patients with bilateral Meniere's disease. ESR has been found to be elevated. Circulating immune complexes have been found to be elevated in Meniere's disease. Antibodies directed against type II collagen have been found in the serum of these patients.

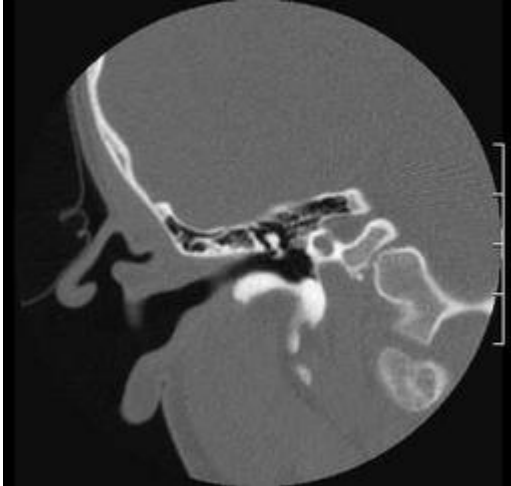
Psychosomatic features:

Patients with Meniere's disease have an increased incidence of personality disorders.

Secondary hydrops possible etiological factors involved:

Developmental insult:

Developmental insults can cause symptomatic endolymphatic hydrops. Mondini's deformity is commonly associated with secondary Meniere's disease. The true Mondini deformity occurs secondary to an arrest at the seventh week. Only the basilar turn of the cochlea has undergone complete development. Typically the interscalar septum or osseous spiral lamina is incomplete; resulting in a confluency of the apical and middle cochlea turns (incomplete partition). The vestibule and semicircular canal may or may not be normal.



CT scan showing cochlear aplasia (Mondini deformity)

Abnormal metabolic / endocrine states:

Certain abnormal metabolic and endocrine states predispose to the development of secondary hydrops. Both high and low blood glucose levels have been associated with dysfunction of inner ear. The hearing may fluctuate with blood glucose levels. It has been demonstrated that induced hypoglycemia resulted in a decrease in potassium concentration of endolymph associated with a rise in endolymphatic sodium levels. These changes resemble the changes seen in Meniere's disease.

Hyperlipoproteinemia has been associated with Meniere's like symptoms.

Endocrine disorders causing secondary hydrops:

1. Hypothyroidism
2. Nephrogenic diabetes insipidus
3. Adrenal insufficiency

Syphilis:

Syphilis is a known cause of endolymphatic hydrops. This could be caused due to the inner ear's reaction to syphilitic organism.

Chronic otitis media:

Endolymphatic hydrops have been observed in patients with CSOM. This could be caused due to the effect of otitis media on the inner ear due to percolation of infectious products / toxins and other associated enzymes. These substances could migrate into the inner ear through the round window membrane.

Otosclerosis:

Patients with otosclerosis could develop secondary hydrops. This could possibly be due to otosclerotic bone impinging on the vestibular aqueduct, or due to biochemical alteration of perilymph and endolymph. Paperella introduced the term otosclerotic inner ear syndrome to describe this problem.

Abnormal fluid balance:

Haemodialysis has been reported to precipitate endolymphatic hydrops in contralateral ear of a patient with Meniere's syndrome. This could be explained by the fact that sudden changes in the plasma osmolality caused by dialysis affects the predisposed ear.

Pathophysiology of Meniere's disease

Meniere's disease by definition is idiopathic endolymphatic hydrops characterised by roaring tinnitus, vertigo, fluctuating hearing loss. Even though sometimes erroneously used interchangeably, meniere's disease is different from endolymphatic hydrops. It should be borne in mind that the term endolymphatic hydrops indicates the underlying pathophysiological mechanism of Meniere's disease. Endolymphatic hydrops can in fact be classified as primary and secondary according to the causative factors involved. Primary endolymphatic hydrops is in fact the classic Meniere's disease where in the underlying etiology is unknown. In

secondary hydrops the etiopathogenesis of the underlying disorder is clearly elucidable.



American academy of ophthalmology and otolaryngology committee on equilibrium defined Meniere's disease as follows: "a disease of the membranous inner ear characterized by deafness, vertigo and usually tinnitus which has as its pathologic correlate hydropic distention of the endolymphatic system".

Etiology and pathophysiology of Meniere's disease is still unclear. Most commonly accepted theory being overdistension of the membranous labyrinth due to excessive endolymphatic fluid volume. This accumulation of endolymph may be caused by impaired absorption of endolymph in the endolymphatic duct and sac, or excessive secretion of endolymph. These hypotheses are based on demonstrable presence of endolymphatic hydrops in patients with signs and symptoms of Meniere's disease. It has also been demonstrated that these patients have reduced vascularization and fibrosis of perisaccular tissue causing a reduction in the absorptive capacity of the endolymphatic sac. Experimentally also it has been clearly demonstrated that obliteration of endolymphatic sac will induce endolymphatic hydrops.

Which portion of the membranous labyrinth gets affected early in Meniere's disease?

Studies have clearly demonstrated that early increase in the volume of endolymphatic fluid (relative to the perilymphatic compartment) occurs in the pars inferior portion of the membranous labyrinth. This portion includes cochlear duct and saccule. In advanced stages of the disease the whole of the membranous labyrinth can be involved. Cochlear hydrops was seen in all patients of Meniere's disease and saccular hydrops was seen in most. Utricular hydrops was rarely seen.

The degree of endolymphatic space expansion is highly variable. The endolymphatic space bulged in the region of helicotrema in half of the cases, while saccule bulged against the foot plate in 60% of cases, into a semicircular canal usually horizontal in 1/3 of cases.

Fibrous adhesions can form between the saccule and the undersurface of the stapedial foot plate. This contact may explain Hennebert's sign (subjective vertigo, tonic eye deviation and nystagmus observed during a pressure induced excursion of the foot plate). It may also explain the "Tullio phenomenon" which is experienced by some meniere's patients.

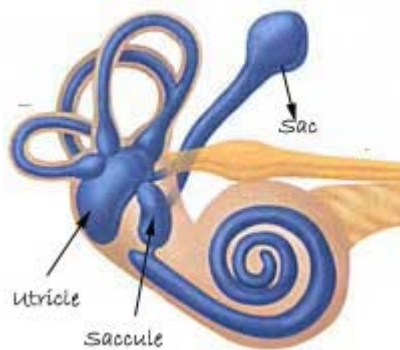
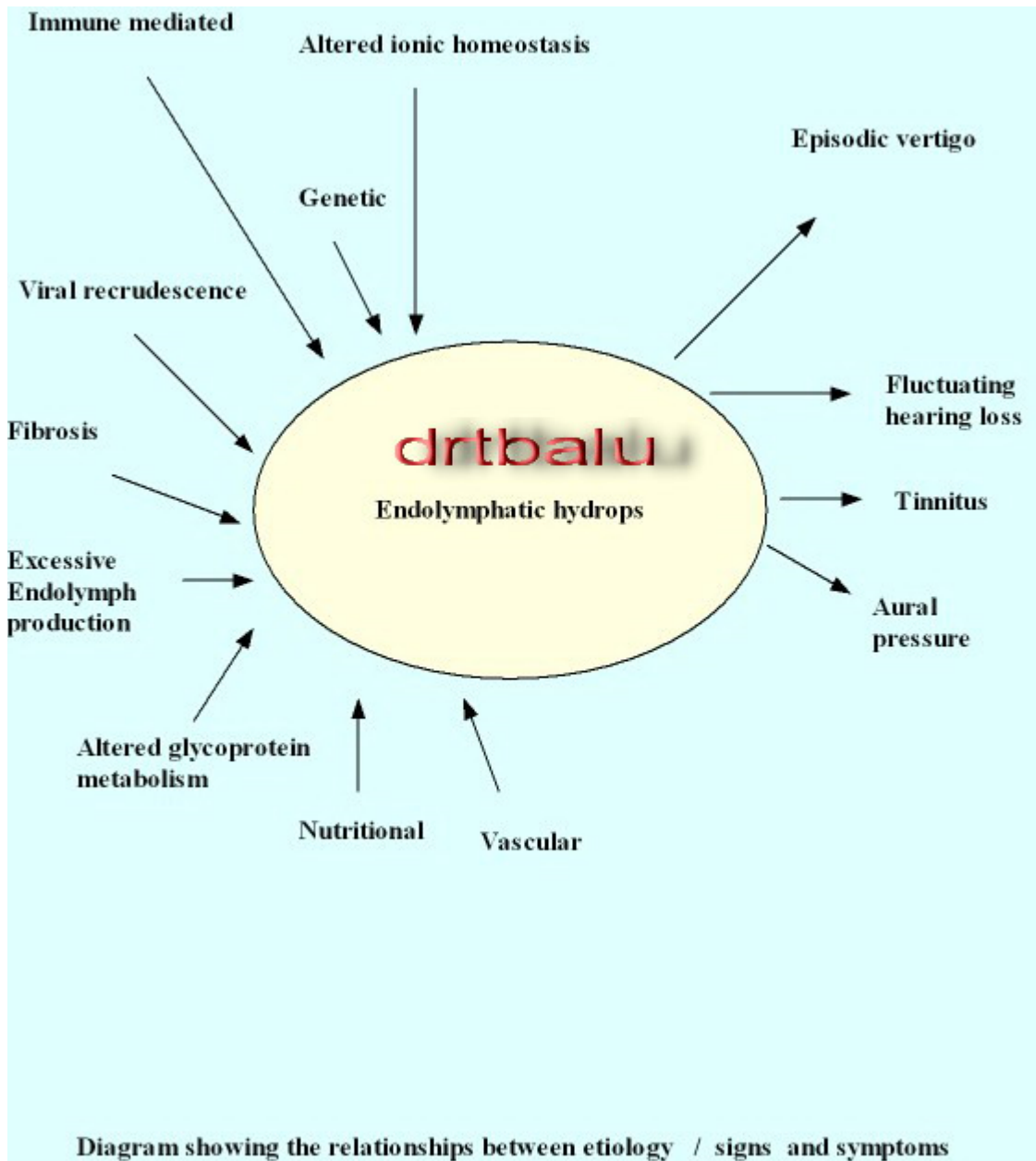


Diagram showing the membranous labyrinth

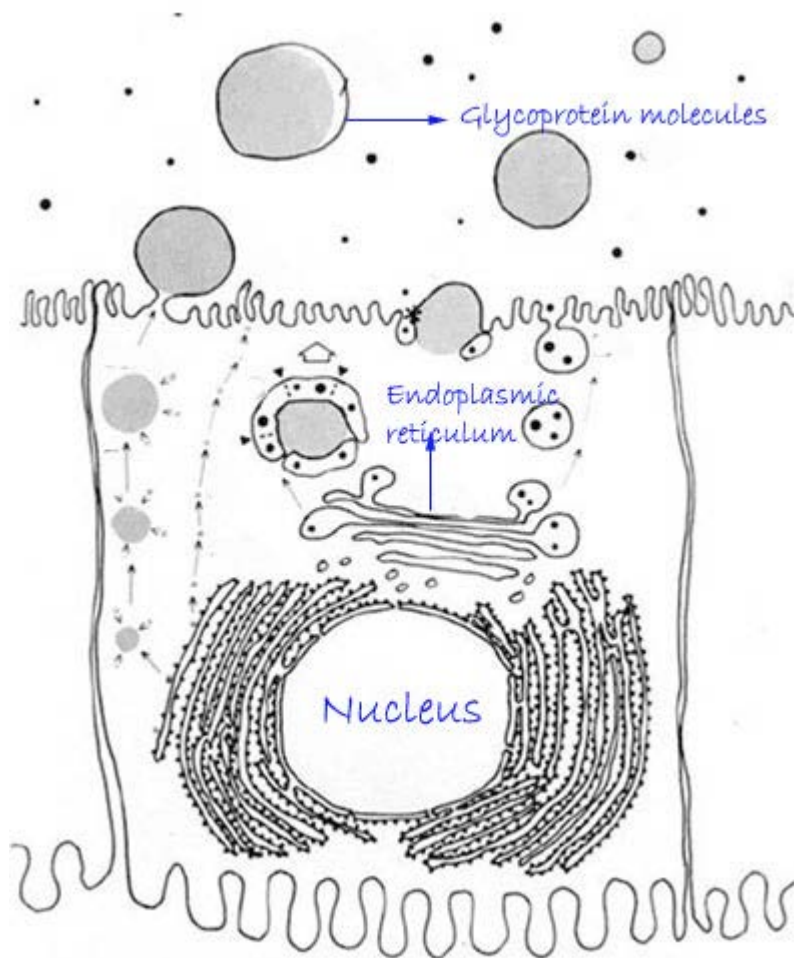
Anatomically, endolymphatic sac has two portions: Intraosseous and extraosseous. The intraosseous portion of the endolymphatic sac is found within the bony walls of vestibular aqueduct. Extraosseous portion of the sac is partially lodged in the duramater of the posterior cranial fossa. Involvement of the sac starts initially at the level of intraosseous portion since this area has very little space to expand.

Dix and Hallpike first proposed that dysfunction of endolymphatic sac as the contributing factor for the development of Meniere's disease. They demonstrated the absence of loose perisaccular connective tissue in these patients. Since this concept of perisaccular fibrosis has been given importance by various authors sac decompression and shunting procedures have been advocated as surgical modalities of managing Meniere's disease.



Altered glycoprotein metabolism:

Recently lot of interest has been focused on the altered glycoprotein metabolism as a causative factor of Meniere's disease. Ikeda and Sando demonstrated significant increase of intraluminal precipitate in the sac of patients with Meniere's disease. Glycoproteins found inside the sac is secreted by its lining epithelium. These cells have well developed rough endoplasmic reticulum indicating its secretory ability. It has also been shown that accumulation of endolymphatic sac glycoproteins could lead to dysregulation of inner ear fluid homeostasis.



Diagrammatic representation of secretory apparatus of endolymphatic sac

The inner ear is devoid of lymphatics. The functions of lymphatics are performed by endolymph circulating the membranous labyrinth. The endolymphatic sac has been demonstrated to secrete proteins. These proteins have been classified into two different classes i.e. immunoproteins and glycoproteins. The immunoproteins are responsible for the immunodefence of the inner ear. The glycoproteins on the other hand are very hydrophilic, and has been shown to suck out the contents of a cell when applied on it. The glycoproteins within the sac can relocate water from the perisac tissues to the interior of the sac causing an increase in the volume of endolymphatic fluid generated.

Role of Saccin in the pathogenesis of endolymphatic hydrops:

Qvortrup et al managed to isolate a natriuretic hormone (saccin) from the endolymphatic sac. It has been demonstrated that this hormone increases the amount of glycoproteins secreted by the endolymphatic sac lining epithelial cells. Qvortrup even postulates that saccin could be the driving force for movement and secretion of endolymphatic fluid.

Functions of endolymphatic sac:

Study of functions of endolymphatic sac will help us to understand the pathophysiology of Meniere's disease. Basic functions of endolymphatic sac include:

- 1 Resorption of water content from endolymph
2. Participates in ionic exchange with endolymph
3. Removal of metabolic and cellular debris which includes otoconia
4. Secretions of glycoproteins to attract extra fluid
5. Secretion of Saccin to increase endolymph production

Endolymphatic fluid circulation:

Studies pertaining to flow of endolymph have postulated three types of flow: 1. Radial flow and 2. Longitudinal flow, 3. Dynamic flow

Radial flow: In this type endolymph was postulated to be secreted and absorbed by stria vascularis. In this type of flow the turn over of fluid was supposedly very slow.

Longitudinal flow: This theory postulates that endolymph as being actively secreted and absorbed at the level of endolymphatic sac.

Dynamic flow theory: Lundquist combined both the theories of radial and longitudinal flow to come out with a new dynamic flow theory. He theorized that functioning of endolymphatic sac can be explained only by combining both these theories. He concluded that ionic exchange occurred during radial flow, and absorption of water content and removal of debris during longitudinal flow.

Endolymphatic flow from the apical region of the cochlea along the cochlear duct and down to the endolymphatic sac appears to be effected osmotically. The osmolality of endolymph gradually increases from the apex to the basal turn of the cochlea. This osmotic gradient is maintained by stria vascularis.

Role of endolymphatic sac in Meniere's disease:

Endolymphatic drainage into the sac appears to be active rather than a passive process. The osmotic gradient within the cochlea is associated with glycoprotein production within the endolymphatic sac . If the sac does not receive any endolymph and becomes relatively dry, it starts to secrete saccin. Presence of saccin increases the amount of endolymph in the cochlea promoting a faster longitudinal flow. It also secretes glycoprotein to attract fluid osmotically.

Stages of Meniere's disease and their possible pathogenesis:

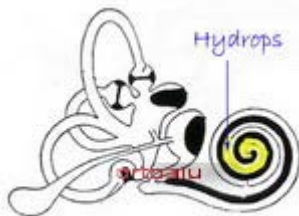
Any theory postulating the pathogenesis of Meniere's disease should be able to explain the clinical stages of Meniere's disease, Lermoyez's syndrome and Tumarkin attacks. It should also be able to explain the vertigo that occurs in congenital syphilis and secondary endolymphatic hydrops.

In early stages of meniere's disease, attacks of vertigo commonly predominate. Hearing is affected only transiently. During this stage the sac is said to be functioning well and has the ability to completely clear the duct. Hydrops occurs only briefly before each attack of vertigo and is completely cleared after each episode.

In later stages of Meniere's disease, glycoprotein secretion causes some amount of functional damage within the sac reducing its ability to reabsorb excess fluid. During this stage there is persistent endolymphatic hydrops within the cochlea. The attacks of vertigo persists in these patients and hearing also does not immediately improve since it is very difficult for all the excess fluid to drain through the narrowed duct.

In some ears (Lermoyez's syndrome) cochlear function improves even during the initial clearance of endolymph. These ears theoretically should have larger vestibular aqueducts.

Burnt out stage (Late stage) the endolymphatic sac is no longer capable of clearing the fluid. Once the duct is blocked completely, there can no longer be any acute vertigo. In these patients continuing secretion of saccin will increase the hydrops within the ear adversely affecting the patient's hearing.



Hydrops seen to start at the pars inferior portion



Hydrops seen progressing. Silt is seen inside the sac



Empty sac senses it is empty and secretes glycoproteins and saccin



Fully developed hydrops

Predisposing factors for development of Meniere's disease:

- 1. Fibrosis of endolymphatic sac and vestibular epithelia**
- 2. Altered glycoprotein metabolism**
- 3. Inner ear viral infection**
- 4. Tightly adherent dura in the region of endolymphatic sac**

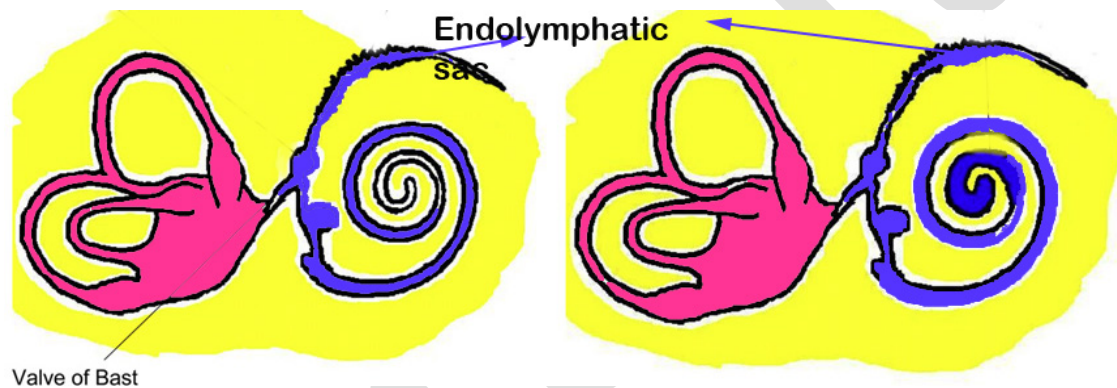
5. Lack of periaqueductal pneumatization

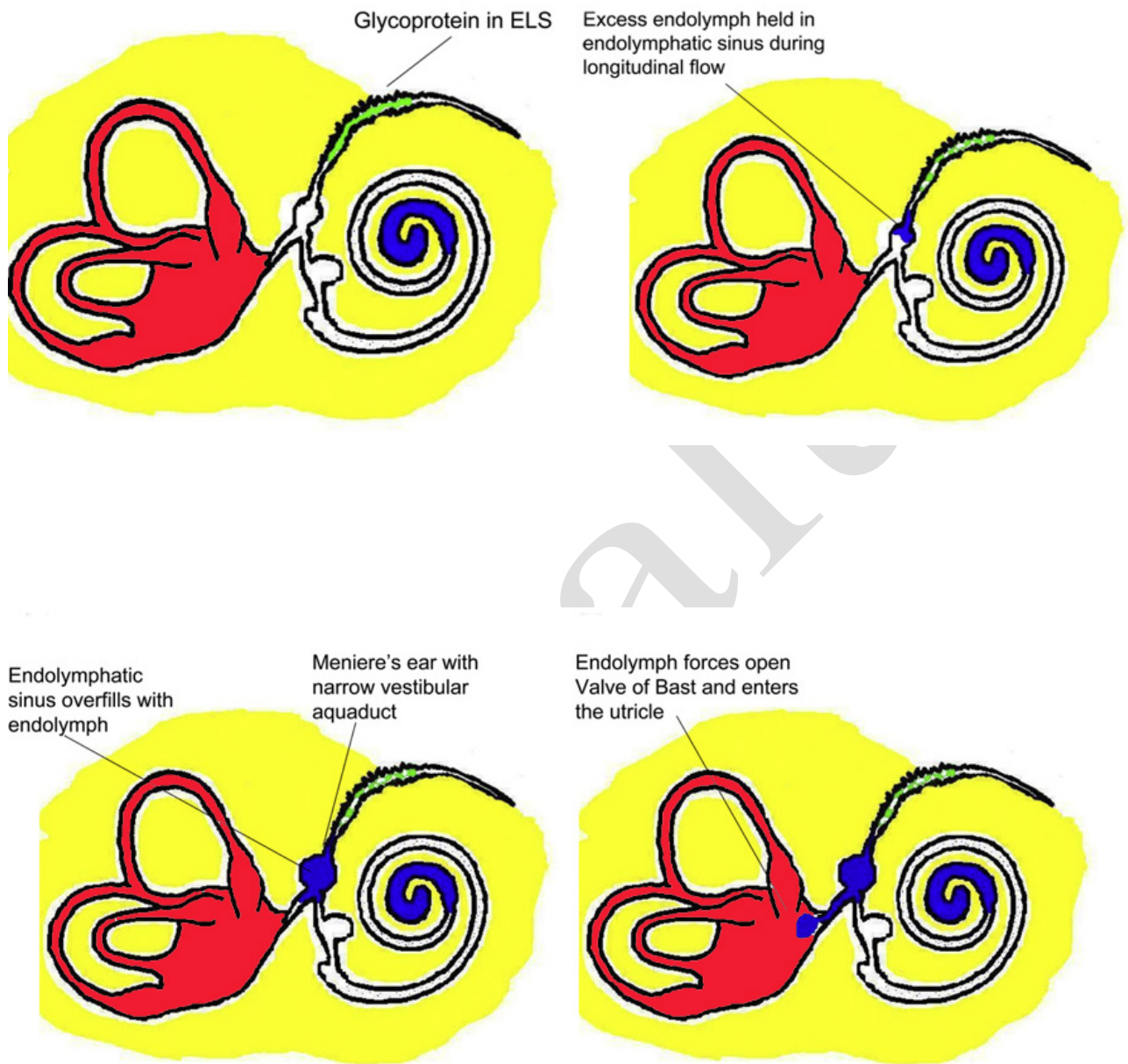
6. Anterior / medial displacement of lateral sinus causing a reduction in the size of Trautmann's triangle. This displacement also causes impediment to the venous drainage locally resulting in a disruption of hydrodynamics of the region.

Currently accepted theory explaining the pathogenesis of Meniere's disease is the drainage theory.

Drainage theory:

This theory makes a sincere attempt to encompass all the previously mentioned aspects of anatomy, physiology and pathophysiology.





According to drainage theory the excess endolymph volume which is present in endolymphatic hydrops accumulate in the apical end of cochlea, where the membranes are more lax than elsewhere. When the situation is normal mild increase in the volume of endolymph can be removed by radial fold whereas larger increase in its volume needs an intact longitudinal flow for efficient removal. When the excess volume of endolymph reaches the endolymphatic duct the sinus can temporarily accommodate the excess volume which the sac is not prepared to receive. This excess of fluid can usually be removed without causing any vestibular disturbance as the endolymphatic valve of Bast isolates the pars superior and prevents endolymphatic fluid draining out of the utricle. If the bony endolymphatic duct is narrow / occluded by accumulation of debris, endolymph may build up

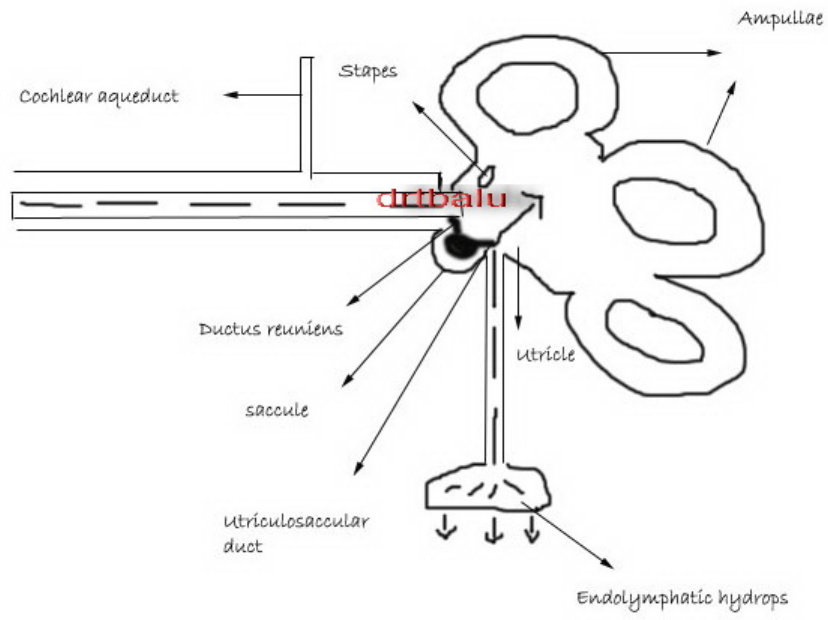
excessively in the endolymphatic duct during the longitudinal flow. Overflow begins to occur opening the valve of Bast so that endolymph enters the pars superior. This excessive volume of endolymph entering the sacculum distorts the crista in one direction causing vertigo to occur. As the excess endolymph is cleared, the amount of endolymph decreases and the stretched cristae reduces in size, thereby causing a reversal in the direction of nystagmus. Progression of the disease decreases the functionality of the sac due to damage to the cells lining the sac. During the late stages of the disease, the valve of Bast remains patent and during the longitudinal flow a sudden drainage of endolymph from the utricle causing drop attacks to occur (Tumarkin's crisis).

Endolymphatic fluid circulation

A clear understanding of circulation of endolymph will help us in understanding the pathophysiology of Meniere's disease. Endolymph is predominantly derived from striavascularis. The planum semilunatum and dark vestibular cells contribute a small amount. Endolymph can also be produced as an ultrafiltrate from perilymph fluid across the labyrinthine membranes.

Lawrence hypothesis of endolymphatic fluid circulation:

According to Lawrence endolymphatic fluid circulation is both radial and longitudinal. Longitudinal flow starts with the production of endolymph in the stria vascularis of the cochlea, circulation occurs in the scala media through the ductus reuniens to the saccular duct, from where it proceeds into the vestibular labyrinth. Elimination of endolymph occurs via circulation through the vestibular aqueduct and onto the endolymphatic sac, where its absorption takes place. Radial flow results from the production of endolymph in the dark vestibular cells and planum semilunatum with local absorption. Lawrence also suggested that both longitudinal (slow process) and radial (rapid process) circulations occur concurrently in a subject. These circulations are subject to both hydrostatic and osmotic pressure gradients.



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Role of endolymphatic sac in the pathophysiology of Meniere's disease current concepts:

The role of endolymphatic sac in the pathophysiology of Meniere's disease has been extensively studied. Studies have shown that endolymphatic sac is lined by uneven epithelium characterized by the presence of crypts and folds. These crypts and folds tend to increase the surface area of the mucosal lining of the sac. Endolymphatic sac contains a variety of ion transporters and aquaporins thus stressing its role in the maintenance of the electrolyte composition of the endolymphatic sac.

Experiments have shown that the endolymphatic sac is highly sensitive to endolymph volume manipulations. When the volume of endolymph increases the concentration of luminal potassium increases and sodium concentration is decreased. When the volume of endolymph is decreased luminal potassium concentration is decreased and the luminal sodium concentration undergoes corresponding increase. Endolymphatic sac is capable of bidirectional response being capable of secretion and absorption of endolymph under necessitating conditions.

Morphological studies have demonstrated that the appearance of the sac differs between its two functional states.

Normal sac: Demonstrates a stainable homogenous substance could be seen filling the distal regions of the sac lumen.

When the volume of endolymph increases the homogenous substance within the lumen of the sac disappears, the dark cells present in the epithelium appears to be activated and in some areas appear to cover the apical surfaces of light cells. This is known as "veiling effect".

When there is reduction in the volume of endolymph either by osmotic dehydration or by endolymph withdrawal the luminal substance becomes darker and is present throughout the lumen of the sac and in addition the light cells become enlarged and activated.

How exactly the sac is able to perceive the volume changes in the endolymph has been bothering us for nearly 150 years. Studies have failed to demonstrate the presence of mechanoreceptor cell within the sac. The location of the sac is not conducive to sensitive mechanoreception, as it is directly influenced CSF pressure fluctuations and vascular pulsations of sigmoid sinus. Attempts to measure intraluminal pressure within the sac have demonstrated it to be a noisy place due to these pressure fluctuations. Hence detection of endolymphatic volume occurs not at the level of endolymphatic sac but at the level of endolymphatic sinus. The endolymphatic sinus is a small membranous bulb located where the endolymphatic sac enters the vestibule.

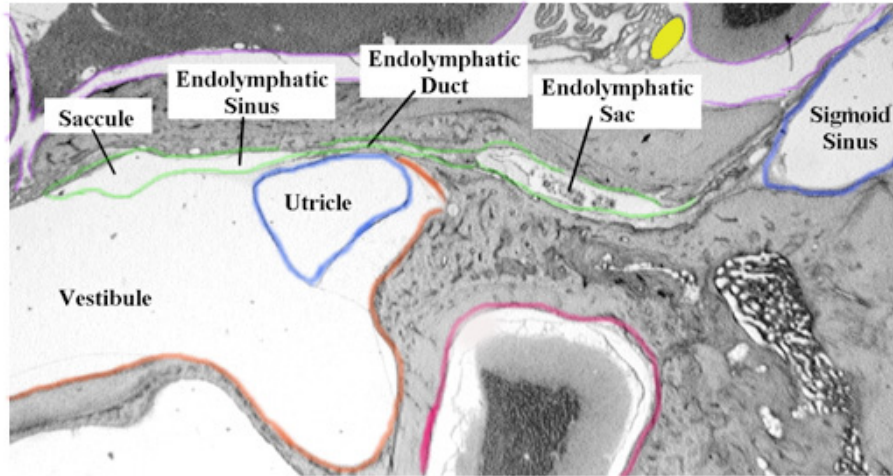
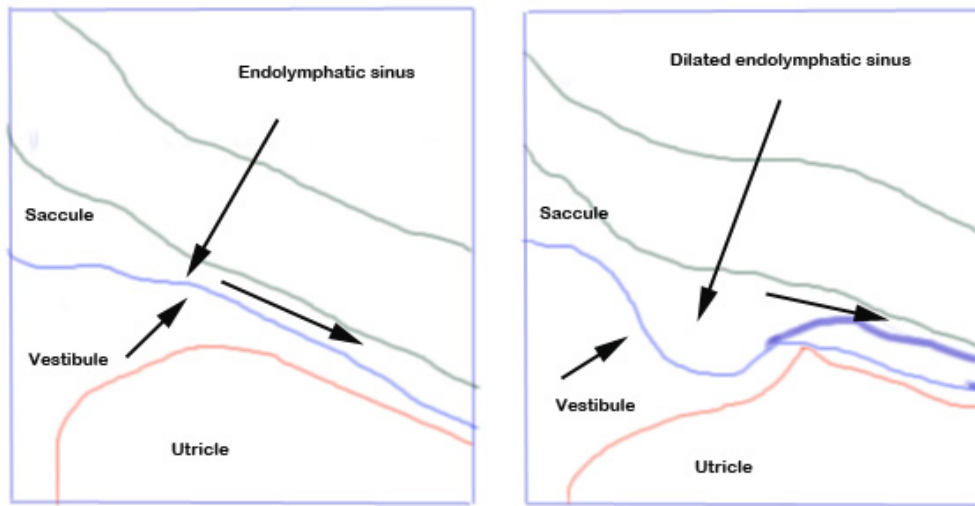


Figure showing endolymphatic sinus



Schematic representation showing how endolymphatic sinus perceives changes in the endolymphatic volume.

When the volume of endolymph is normal, pressure elevations in the vestibule produce only small movements of endolymph into the sac before the endolymphatic sinus membrane occludes the endolymphatic duct. On the contrary if the endolymph volume is elevated and the sinus is dilated, a pressure elevation in the vestibule results in larger volumes of endolymph forced into the sac before the duct is occluded. This counteracts the increased volume of endolymph in the system. Since the membrane constituting the endolymphatic sinus is highly compliant in nature it would result in greater distention of the sinus. In this state positive pressure applied to the vestibule would drive greater amounts of endolymph into the sac before the flow gets occluded by the dilated endolymphatic sinus membrane.

Sources of pressure fluctuations in the inner ear fluids are numerous. For this to occur there should be pressure differential between the CSF and the endolymphatic fluid. Changes in CSF pressure occurs during:

- 1. Breathing**
- 2. Heart beat**

3. Postural movements
4. Coughing
5. Sneezing

These changes in CSF pressure are faithfully transmitted to the endolymphatic system via the cochlear aqueduct. If the volume of endolymph exceeds the capacity of the sac to reabsorb then the endolymphatic sinus becomes over dilated and is not in a position to function as a regulator of the volume of endolymph. This scenario causes irreparable damage to the inner ear.

Clinical manifestations of Meniere's disease

Clinical manifestations of Meniere's disease:

1. Episodic attacks of rotatory vertigo
2. Ipsilateral hearing loss
3. Aural fullness
4. Roaring tinnitus

Episodic vertigo: Is always associated with vegetative signs such as nausea and vomiting. This is supposed to be the most debilitating symptom manifested by the patient.

The vertigo begins all of a sudden in a otherwise normal individual. It is accompanied by pallor, sweating, nausea, diarrhoea and vomiting. During the attack the patient is fully conscious, oriented in time and space. The patient suffers no residual neurological symptoms after the attack is over. If there is diplopia then it could be due to acute vertigo causing it. The general rule of the thumb is that attacks of vertigo in Meniere's disease last somewhere between 24 minutes and 24 hours. The frequency of these attacks are also highly variable. Patients with severe hydrops suffer attacks on a daily basis while others have long quiescent periods in between attacks.

After the acute phase is over, the symptoms gradually subside, and the patient invariably falls asleep. Some patients may complain of dysequilibrium and motion intolerance within the first 24 hours after the initial attack.

Meniere's disease variants:

In variants of Meniere's disease like Lermoyez syndrome the vertiginous episode is preceded by increasing levels of tinnitus and hearing loss. Unlike in classic Meniere's disease, the hearing loss and tinnitus dramatically resolve during or shortly after the onset of vertigo.

In another variant of Meniere's disease like Tumarkin's crisis (Drop attacks) sudden unexplained falls occur without vertigo or loss of consciousness. These patients describe the sensation as being pushed, or thrown to the ground. Tumarkin who first described this variant postulated that this could be caused due to acute dysfunction of otolith organs. Sudden changes in the output of gravity reference information from the otolith organs cause this condition.

Meyerhoff described another variant of Meniere's disease (abnormal oculovestibular response). These patients experienced vertigo, with all its accompanying vegetative symptoms like nausea when exposed to optokinetic stimuli such as riding in a train or car. Most of these patients have aural fullness, tinnitus, fluctuating hearing loss. Most of these patients had abnormal electronystagmography.

Cochlear Meniere's disease is another variant. This disorder is characterised by fluctuating hearing loss. There is no vertigo in this variant.

Nystagmus: These patients manifested nystagmus. To start with they had nystagmus beating towards the affected side (irritative nystagmus). This lasts for 20 seconds. After a short time the nystagmus changes its direction towards the healthy ear (Paralytic nystagmus). Hours after the attack, the auditory and vestibular symptoms subside, the nystagmus reverses again beating towards the affected ear (recovery nystagmus). This recovery nystagmus may be horizontal, or rotatory. Since rotatory nystagmus are not visible in ENG a careful observation of the eye movements should be made in these patients. Recovery nystagmus may constitute an important localizing sign in these patients.

These changing types of nystagmus can be accounted by the membrane rupture theory of Meniere's disease pathophysiology. Developing hydrops cause distention of the whole of the endolymphatic system causing the membrane to rupture. As soon as the membrane ruptures, the perilymphatic potassium starts to rise initially. This initially has an excitatory effect on the first order vestibular neurons causing irritative nystagmus. Thereafter the concentration of potassium keeps on increasing causing a blockade of action potentials at the level of these first order vestibular

neurons causing paralytic nystagmus. This is seen within minutes of the attack. The recovery nystagmus may be the result of vestibular adaptation.

Shea's symptomatic classification of Meniere's disease:

Stage I: The patient has solely cochlear symptoms

Stages II - IV: These patients have progressively more cochlear and vestibular symptoms

Stage V: End stage Meniere's disease.

Hearing loss:

This is sensorineural in nature and is a cardinal feature of Meniere's disease. The hearing loss is typically fluctuating and progressive. Hearing may in fact fluctuate significantly during the early phases of the disease. The deafness is classically known to involve lower frequencies as compared with s/n loss caused by noise exposure which involves higher frequencies. End stage Meniere's disease is characterised by profound sensorineural hearing loss.

Diplacusis is the common complaint in a majority of Meniere's disease patients. Here the same frequency sound is perceived to be different by both the ears.

Tinnitus:

Tinnitus in a Menier's patient is highly variable. It is commonly roaring in nature. It could in fact be the first symptom of the attack. It could be continuous / intermittent. It is invariably non pulsatile in nature. The pitch of the tinnitus usually corresponds to the region of cochlea having the most severe hearing loss.

Aural fullness:

This is one of the most important symptoms of Meniere's disease. This is mostly caused by enlarging membranous labyrinth. This pressure symptom is limited usually to one ear.

Diagnostic criteria for Meniere's disease

Meniere's disease is diagnosed only with a high degree of suspicion. The following are the pointers that could help in the diagnosis of Meniere's disease.

Meniere's disease can be diagnosed by:

1. Vertigo: Vertigo is spontaneous, lasting minutes to hours. It could be recurrent, and if recurrent the patient must have at least 2 episodes within 20 minutes. These episodes should be accompanied by nystagmus.

2. Hearing loss: In frequencies (200, 500, 1000 Hz) 15 dB. Hearing loss is sensorineural in nature covering the lower frequencies. When compared with the other ear, it should be less by 25 dB in all the frequency ranges studied audiometrically.

3. Tinnitus: Roaring in nature

4. Aural fullness.

Criteria for diagnosis of Meniere's disease:

Possible Meniere's disease:

1. Episodic vertigo of Meniere's type without documented hearing loss

2. Fluctuating hearing loss with dysequilibrium but without definite episodes

Probable Meniere's disease:

1. One definitive episode of vertigo

2. Audiometrically documented hearing loss at least on one occasion

3. Tinnitus / aural fullness in the treated ear

Definite Meniere's disease:

1. Two or more definitive episodes of spontaneous vertigo one at least lasting for 20 minutes

2. Audiometrically documented hearing loss at least on one occasion

3. Tinnitus and aural fullness in the treated ear.

Department of otolaryngology University Hospital Groningen evolved the following definition of Meniere's disease:

- **A sensorineural (cochlear) hearing loss combined with**
- **Tinnitus present now or in the past and**
- **Vertigo attacks (atleast two, present now or in the past) and**
- **Exclusion of other pathology following Groningen protocol**

Hearing loss: Groningen criteria lays down that hearing loss when present should be sensorineural in nature. The conductive component should be absent and this fact should be proved by a puretone audiogram and impedance audiometry. Sensorineural loss being defined as hearing loss of 20 dB or worse at one of the 6 measured thresholds (0.25 KHz, 0.5 KHz, 1 KHz, 2 KHz, 4 KHz, and 8 KHz) ranges.

Vertigo: Is characterized by paroxysmal attacks of dizziness with a sense of rotation. These attacks are usually accompanied by nausea and vomiting. At least two episodes of vertigo / dizziness should be reported by the patient during the course of the illness. One of the attacks should have lasted for more than 5 minutes. Inbetween these attacks there may be associated periods of unsteadiness.

Affected / Unaffected ears: When sensorineural deafness and h/o tinnitus is present in an ear then the ear is considered to be affected by Meniere's disease. It is not required that all three finding should be present at the time of investigation. But deafness should be present at the time of investigation to consider the ear to be an affected one. When sensori neural hearing loss and tinnitus is absent then the ear is considered to be unaffected.

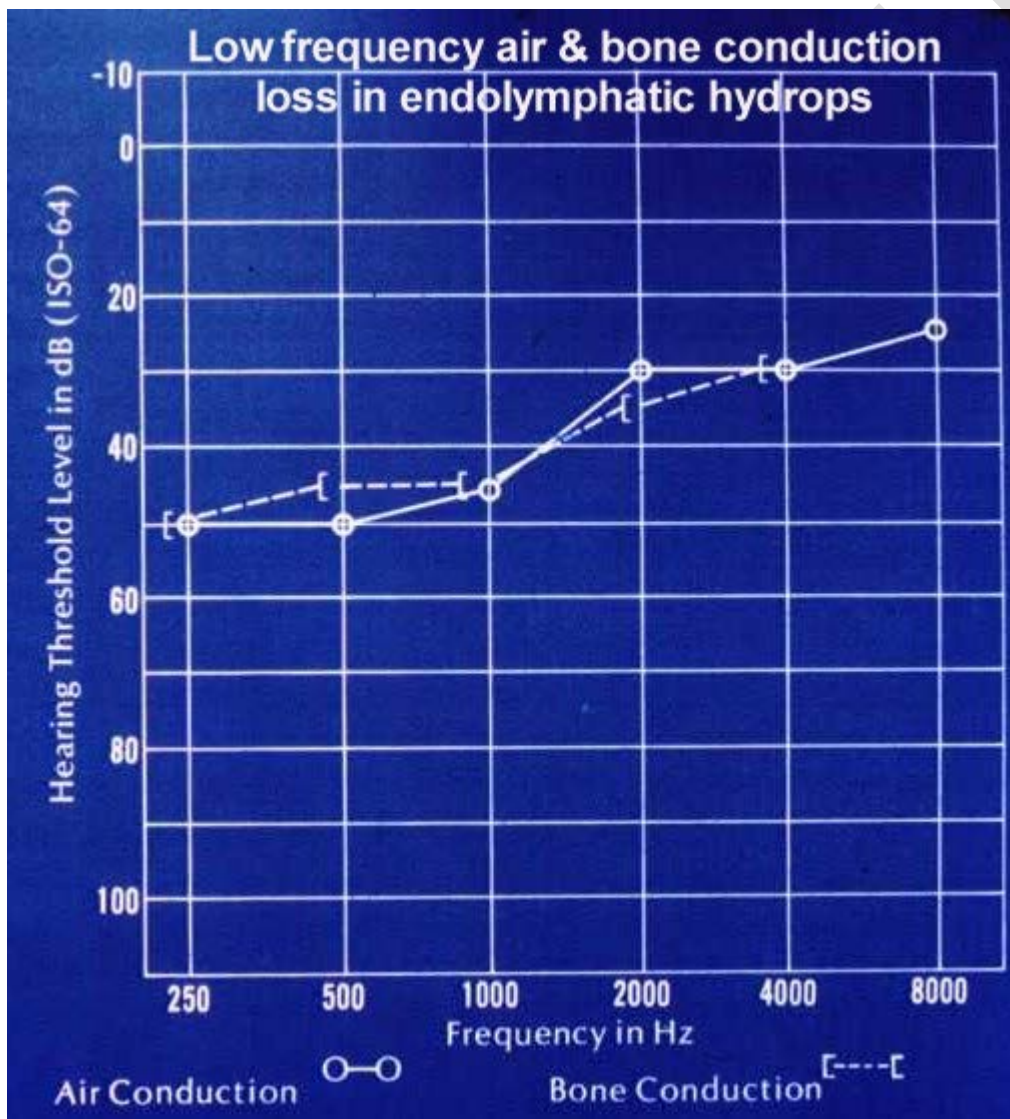
Modern definition of Meniere's disease in its diagnosis helps in:

- 1. Guiding the optimal treatment modality**
- 2. To ascertain the prognosis of the disease**

Currently 2 critical feature of Meniere's disease have been used to identify whether the patient is suffering from Meniere's disease or not. These features include instability of hearing and balance and involvement of both these system. The old nomenclature cochlear and vestibular Meniere has been abandoned since 1985. The pattern of involvement can range between auditory dominant and vestibular dominant symptoms.

Investigations

Audiological assessment: It is very important to assess cochlear function in a patient with Meniere's disease. Cochlear function can easily be assessed by pure tone audiometry. Patients with meniere's diseased usually manifest with a flat audiometry curve. Most of these patients have low frequency sensori neural hearing loss.



Audiogram showing low frequency s/n loss

Other pure tone audiometric features seen in Meniere's disease include:

1. Peaked pattern

2. Downward sloping pattern

Serial audiometry performed over a period of time may demonstrate fluctuating hearing loss. Fluctuations are often seen in the frequency range between 250 - 1000 Hz. Special audiometric tests need to be performed to ascertain whether the hearing loss is a cochlear or hair cell related disease. Presence of recruitment can be demonstrated by alternate binaural loudness balance test in unilateral disease, or SISI test in unilateral / bilateral disorders.

Stapedius reflex thresholds: Are within normal limits in these patients.

Speech discrimination thresholds: Closely resemble pure tone thresholds in most patients. Poor speech discrimination out of proportion to the pure tone thresholds should arouse suspicion of retro cochlear lesion. The phenomenon known as 'roll over' a marked decrease in discrimination is seen in retrocochlear lesions.

Evoked response audiometry:

Evoked response audiometry has been found to be instrumental in the diagnosis of Meniere's disease. This test determines the electrical activity occurring in the cochlea and central auditory pathways in response to sound stimuli.

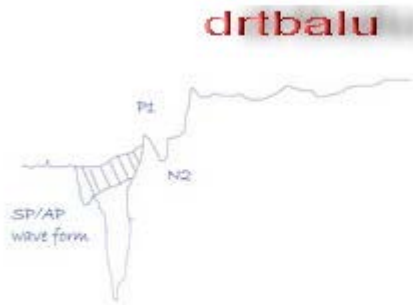
Electrocochleography: Belongs to the battery of tests under evoked response audiometry. It evaluates the evoked potential activity of the cochlea and 8th cranial nerve. Electrocochleography is the best existing objective test for Meniere's disease. This test measures the electrical events generated either within the cochlea or by primary afferent neurons. The recorded potentials include: Cochlear microphonic potential and summing potential from cochlea, and the whole nerve action potential from the cochlear division of 8th nerve.

Cochlear microphonics is an alternating current, the polarity of which is identical to that of the auditory stimulus. It is in fact thought to be the sum of the individual hair cell intracellular potentials. Most of the cochlear microphonic potential is produced by outer hair cells within the first few millimeters of the basal turn of cochlea. In patients with Meniere's disease these cochlear microphonic potentials are small and distorted. In some patients a marked 'after ringing' (a sinusoidal wave) of the cochlear microphonic is seen.



Cochlear microphonic recording from a patient with Meniere's disease

Summating potential: This potential is of short latency (0.3 msec) and is usually present at high stimulus intensities. It is actually a DC shift from the base line of response, generally in a negative direction. This potential occurs for the entire duration of the stimulus. Major component of summating potential is derived from the asymmetry in the vibration induced deflection of the basilar membrane. In normal ears, at high stimulus intensities, the basilar membrane vibrates more upwards towards the scala media than down wards generating a negative summating potential. Endolymphatic hydrops accentuates this asymmetry by stretching and stiffening the basilar membrane, limiting its downward vibration. This mechanical deformity of the basilar membrane is greatest at its basal end and this is the region where the majority of summating potentials are generated from. The normal upgoing asymmetry is enhanced, leading to a negative summating potential of increased amplitude and width.



Normal summing potential

Evoked action potential: This is a compound action potential representing the synchronous firing of multiple cochlear neurons derived mainly from the basal turn of the cochlea. A click stimulus because of its faster rise time, will stimulate more of the basilar membrane than frequency specific tone bursts.

In Meniere's disease, the common findings on electrocochleography include:

1. **Increased summing potential and action potential ratio:** A summing potential /action potential ratio of up to 1:3 is within normal range, a higher ratio is suggestive of hydrops.
2. **Widened summing potential and action potential complex:** The normal width of the summing potential / action potential complex is 1.2 - 1.8 ms and a widening of greater than 2ms is usually significant.
3. **Small distorted cochlear microphonic.**



Widened action potential / summing potential wave form

Glycerol dehydration test:

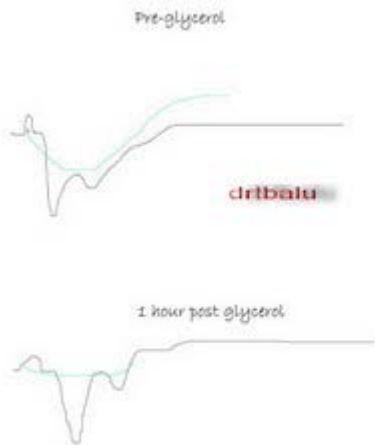
This test was originally introduced by Klockhoff and Lindblom in 1966. The drug initially used to cause dehydration was chlorthalidone which promoted sodium excretion without appreciable potassium loss. Pure tone audiometry was performed before and after the administration of the diuretic. A rise in threshold of atleast 10dB in three consecutive octave bands were considered diagnostic of Meniere's disease. This test became sensitive when it was combined with transtympanic electrocochleography. Glycerol was later substituted for chlorthalidone. During glycerol dehydration the marked negative summing potential is seen to decrease. Positive result to glycerol testing can occur if the patient has a fluctuating hearing loss due to endolymphatic hydrops. Glycerol is administered orally in doses of 1.5 mg /kg body weight in the fasting state, and the test can only be considered positive only if there is an increase in serum osmolality of atleast 10 mOs/kg to verify the effectiveness of the dehydration process. After one hour the amplitude of the action potential appeared to diminish by 12%.

Side effects of glycerol administration:

1. Headache

2. Nausea / vomiting

3. Drowsiness

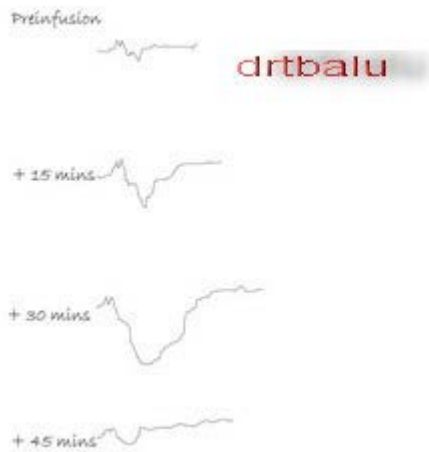


EcocG performed before and after glycerol administration

Glycerol can also be administered parenterally to shorten the duration of the test. Intravenous administration is performed using 200 ml of 10% glycerol solution.

Acelazolamide test:

This is also another one of the dehydration tests used in the diagnosis of Meniere's disease. This drug is carbonic anhydrase inhibitor. It has been used to increase the cochlear hydrops. This test is hence also known as "reverse glycerol test". Azetazolamide 500 mg in aqueous is injected intravenously over one minute, and electro cochleogram is recorded continuously for 45 minutes. Pure tone audiometry and speech audiometry are also performed. Ecocg showed an enhanced negative summating potential within 10 - 15 minutes of drug infusion, reversing towards the pre-infusion base line level and 45 - 60 minutes. No change was seen in normal individuals or in those with other cochleo vestibular pathologies. This test is useful in patients who have intense vomiting when glycerol is administered.



Ecocg recording before / after acetazolamide injection

It must be accepted there is currently no test is available which could diagnose Meniere's disease 100% of the time. The tests described above can at best be considered presumptive.

Caloric test:

This demonstrated directional preponderance, labyrinthine weakness, or labyrinthin asymmetry. This test should atmost be considered as a nonspecific test. This test infact probes the functioning of the lateral semicircular canal ignoring the other portions of the labyrinth which are commonly involved in Meniere's disease. The sensitivity of caloric test may also be somewhat compromised by central compensatory mechanisms. Caloric test in patients with Meniere's disease will show a loss only when the amount of loss is too high to be compensated by central mechanisms. In caloric tests the stimulus used is low frequency in nature.

VEMP:

Vestibular evoked myogenic potentials are believed to be generated by sacculo collic reflex. In this reflex the afferent limb of the reflex pathway is caused by

stimulation of acoustic sensitive cells in the saccule that responds to loud, brief monaural stimuli which stimulates the inferior vestibular nerve. The efferent limb terminates in the fibers of sternomastoid muscle causing it to contract. Myographic recordings from sternomastoid muscle in response to stimulation of the saccule reflect saccular function. The greatest sensitivity to VEMP occurs in the frequency range of 200 – 1000 Hz. Since Meniere's disease is associated with cochleo saccular hydrops presence of VEMP reflexes indicate the saccular function. VEMP responses in individuals with Meniere's disease show altered frequency tuning, such that the greatest sensitivity of the sacculocollic reflex seems to occur at higher frequencies and across broader frequency ranges compared with normal subjects

Differential diagnosis of Meniere's disease

The diagnosis of Meniere's is by exclusion. There are many disorders that could mimic this condition. Before considering the differential diagnosis the variants of Meneire's disease should be excluded.

Various differential diagnosis of Meniere's include:

Central causes:

1. Acoustic neuroma
2. Multiple sclerosis
3. Vascular loop compression syndrome
4. Aneurysm
5. Arnold chiari malformation

6. Brain stem tumors

7. Cervical vertigo

8. TIA

Peripheral causes:

1. BPPV

2. Labyrinthitis

3. Autoimmune ear disease

4. Perilymph fistula

5. Otosclerosis

6. Migraine induced vertigo

Metabolic causes:

1. Diabetes

2. Hyper / Hypothyroidism

3. Syphilis

4. Cogan's syndrome

5. Anemia

6. Autoimmune disorders

It is very easy to make the diagnosis when all the four classic features of Meniere's disease is present in a patient. Unfortunately it is usually not the case. Many variants of Meniere's should be considered.

Medical management

Medical management of Meniere's disease includes:

1. Dietary management
2. Physiotherapy
3. Psychological support
4. Pharmacologic intervention

Dietary management:

This includes reduction of sodium in the diet. Infact it was Frustenberg in 1934 who introduced a low salt diet for patients with Meniere's disease. Pathophysiology of Meniere's disease is enlargement of membranous labyrinth due to excess accumualtion of endolymphatic fluid. Any attempt to reduce this fluid level will help in alleviate the symptoms of the patient.

Medical management is mainly used to treat patients during the acute phase of the attack. Vestibular suppressants are commonly used. Drugs used to control attacks of vertigo have varying levels of anticholinergic, antiemetic and sedative properties. Drugs used to alleviate symptoms include phenothiazines (prochlorperazine and perphenazine), antihistamines like (cinnarizine, cyclizine, dimenhydrinate, and meclizine hydrochloride), benzodiazepines like (lorazepam and diazepam).

Vestibular suppressants:

Diazepam: when used acts as vestibular depressant. It also alleviates the anxiety associated with this disorder. The beneficial effects of diazepam on the vestibular system is presumed to be due to an increase in the cerebellar GABA-ergic system. Stimulation of cerebellar GABA-ergic system mediates inhibition on the vestibular response. This drug is very useful in alleviating vertigo especially when associated with anxiety. Usual dose is 5 mg administered orally every 3 hours. The initial dose may also be administered intravenously.

Antiemetic drugs:

Drugs belonging to this group help to alleviate vomiting in Meniere's disease.

Anticholinergic drugs:

Glycopyrrolate an anticholinergic drug when combined with diazepam is helpful in controlling inner ear symptoms of nausea and vomiting. In adults it is administered in doses of 1-2 mg. It may also be administered as intramuscular injection (0.1 - 0.2 mg) every 4 hours. Side effects (reversible) of this drug include dry mouth, distortion of visual acuity, exacerbation of symptoms in patients with prostatic hypertrophy. This drug is contraindicated in patients with glaucoma and prostatic hypertrophy.

Antidopaminergic drugs:

Droperidol: This is an antidopaminergic drug used to alleviate the symptoms of Meniere's disease. This drug is administered in doses of 2.5 - 10 mg orally in adults. If administered intravenously it is given as 5 mg bolus. This drug has fewer incidences of side effects like extrapyramidal symptoms / sedation / hypotension.

Prochlorperazine: This drug belongs to the phenothiazine group. It is used as an antiemetic and a potentiator of analgesic and hypnotic drugs. Usual recommended dose is 10 mg given orally or intramuscularly every 4 - 6 hours in adults. This drug has an excellent antiemetic effect.

Antihistamines:

Dimenhydrinate: is useful in preventing and treating vertigo associated with Meniere's disease. It is also very effective in controlling nausea and vomiting. Only side effect of this drug is its propensity to cause drowsiness. It is administered as 50 - 100 mg doses thrice a day. This drug can also be administered intramuscularly / intravenously.

Diphenhydramine: This drug is not useful in treating acute vertigo. It may be useful in prevention of vertigo. The usual duration of action is 4-6 hours. Usually this drug is administered as an initial loading dose of 50 mg orally.

Meclizine: This drug is one of the most useful antiemetics to prevent / treat nausea and vomiting associated with vertigo of vestibular origin. It has a slower onset and a longer duration of action (24 hours). For vertigo the usual dose administered in adults is 25 - 100 mg daily in divided doses. Side effects of this drug include: drowsiness, blurred vision, drowsiness.

Promethazine: This drug has pronounced antihistaminic activity in addition to its strong central cholinergic blocking activity. It is effective in the treatment of vertigo and motion sickness. It is administered usually in doses of 25 mg every 4 to 6 hours. One major advantage of this drug is that it can be administered rectally, when severe vomiting prevents its effective oral administration. Most common side effect of this drug is sedation.

Maintenance therapy:

The goal of maintenance therapy is

1. To prevent acute attacks of vertigo
2. To maintain hearing in Meniere's disease

This therapy usually includes dietary modifications combined with pharmacological intervention.

Dietary modifications: The mainstay of diet modifications is to reduce sodium intake. A very low sodium intake or low sodium diet is usually recommended. A strict low sodium diet means a daily allowance of 1500 mg. This is a very stringent diet and patients find it very difficult to comply with this diet. A more practical approach would be to advise the patient to avoid excessively salty food. Restrictions are also imposed on the intake of caffeine, nicotine and alcohol.

Diuretics:

The use of diuretics in the maintenance therapy is based on the supposition that these drugs can alter the fluid balance of inner ear, leading to a depletion of endolymph and a correction of hydrops. In 1934 Furstenburg demonstrated that the

symptoms of Meniere's disease were due to retention of sodium. He went on to recommend a low sodium diet / use of diuretics to control Meniere's disease. Boles in 1975 demonstrated that most patients had their vertigo controlled with an 800 - 1000 mg of sodium diet / day.

Hydrochlorthiazide: This diuretic causes natriuresis and kaliuresis by blocking sodium reabsorption in the loop of Henle. Potassium supplementation is required in patients using this drug. Side effects of this drug include: hypokalemia, hyperglycemia, hypotension, and hyperuricemia. It is usually administered as 50 mg tabs orally / day in adults. Potassium supplements is usually required in these patients.

Dyazide: Is a potassium sparing diuretic. It can be conveniently administered as a single daily dose.

Fursemide: This is a loop diuretic. It is a very potent diuretic. It can cause electrolyte and volume depletion more rapidly than other diuretics. It usually causes hypokalemia. Usual adult dose is 10 - 80 mg/day. The duration of action lasts for about 4 hours.

Amiloride: This is a potassium sparing diuretic acting on the distal tube of Henle. Its diuretic potency is highly limited. It is usually used in combination with other diuretics in order to minimize potassium loss.

Carbonic anhydrase inhibitors:

Acetazolamide: Is a carbonic anhydrase inhibitor. It causes a decrease in the sodium - hydrogen exchange in the renal tubule inducing diuresis.

Methazolamid: Is another carbonic anhydrase inhibitor shown to be effective in controlling symptoms of Meniere's disease. This drug is usually administered in doses of 50 mg / day, 5 days a week for 3 months.

Medical ablative therapy:

Aminoglycosides: Ototoxic effects of aminoglycosides are well documented. Streptomycin and gentamycin are predominantly vestibulotoxic. Intramuscular injections of streptomycin administered twice daily for periods of days to weeks have been used in patients with debilitating bilateral disease / unilateral disease in the only hearing ear. Complete ablation causes disabling oscillopsia. Many authors have suggested lower doses and fewer injections to achieve partial ablation, thereby reducing the incidence of severe ataxia. Currently the recommended daily dose is 1 g of streptomycin intramuscularly 5 days a week until vestibular ablation occurs as

manifested by absence of ice water caloric test. Intratympanic injections of these drugs have also been used with success.

Vasodilators:

The use of vasodilators is based on the idea that Meniere's disease results from ischemia of the stria vascularis. Betahistine has been used with varying degrees of success. This drug can be used for short term control of vertigo and for maintenance therapy.

Nicotinic acid is another vasodilator which when administered 30 minutes before meals in doses of 50 - 400 mg helps in resolving the acute crisis associated with Meniere's disease.

Calcium channel blockers:

Nimodipine a highly lipophilic drug is very useful in the medical management of Meniere's disease. It readily crosses the blood brain barrier. This drug is useful in patients who have failed diuretic medical therapy.

ACE inhibitors:

These are very effective vasodilators. These drugs block the renin-angiotensin-aldosterone system. They produce vasodilatation by blocking angiotensin II induced vasoconstriction.

Lipoflavins and vitamins:

Combination of lipoflavins and vitamins have been tried as a management modality with varying degrees of success.

Restricting tea and coffee intake to once daily will help these patients in reducing endolymphatic fluid volume. Ingestion of excessive amounts of caffeine and alcohol cause enormous fluid shifts in the physiological fluid compartments.

Middle ear effusion of dexamethasone and streptomycin

There is a common conception that Meniere's disease could be an immune mediated disorder. Dexamethasone injection hence is supposed to play an important role in

the management of this disorder. Dexamethasone in the normal course does not cross the blood labyrinth barrier in significant quantities. Perfusion around round window membrane is a must for adequate doses of dexamethasone to reach the inner ear in significant quantities.

Technique of injection: The ear drum is anesthetised with a topical anesthetic cream (2.5% xylocaine and 2.5% prilocaine). Two holes are made in the inferior quadrant of the ear drum either with a myringotome / argon laser. Care is taken to make one of the openings in the area corresponding to the round window niche. This opening should be large enough to remove any adhesions that could block access to the round window membrane. Approximately 0.5 cc of hyaluron, containing 16 mg of dexamethasone / ml is injected into the round window niche filling the middle ear cavity. The patient is made to remain lying with the perfused ear up for 3 hours. This injection is repeated during the course of next two days. After three perfusions the ear drum holes are covered with a moist gelfoam. The patient also should receive concurrent doses of 0.25 mg of dexamethasone by mouth for atleast a month.

If dexamethasone perfusion proves ineffective then streptomycin is combined with it. About 120 mg of streptomycin / ml of hyaluron plus 16 mg of dexamethasone for intratympanic medication protocol. This protocol is also continued for 3 days consecutively.

Intratympanic injections of gentamycin has also been tried out with varying degrees of success. This drug is preferred because of its greater degree of vestibular selectivity.

Drug delivery modalities include:

1. Intratympanic injections
2. Gelfoam delivery
3. Silverstein micro wick (This wick is inserted in the the middle ear cavity via a myringotomy opening). This wick is expected to tranmit the medication administered in the external auditory canal into the middle ear cavity.
4. Microcatheter delivery for continuous infusion of gentamycin

Surgical Management

Surgical management of Meniere's disease is reserved for those patients who fail to respond to conservative medical management. They constitute 10 - 20% of the overall number.

Surgical therapy in the management of Meniere's disease has a long history.

- 1. In 1877 Gowers reported that simple blistering behind the ear caused a marked reduction in the vertigo in patients with Meniere's disease.**
- 2. Babinski in 1903 proposed lumbar puncture as a management modality in treatment of Meniere's disease.**
- 3. Crockett in 1903 removed stapes as a treatment modality of Meniere's disease**
- 4. Lake in 1905 described labyrinthectomy as treatment of vertigo associated with Meniere's disease**
- 5. Parry first performed the first intracranial 8th nerve section to treat Meniere's disease via middle cranial fossa approach**
- 6. In 1907 T.W. Parry described the use of Seton to treat Meniere's disease. Seton is a thread / tape placed in a subcutaneous tract fashioned over the nape of a neck on the side of the affected ear.**
- 7. Cervical sympathectomy was performed as a treatment modality by Seymour**

Surgical procedures can be classified as:

- 1. Procedures involving hearing / vestibular preservation**
- 2. Procedures involving hearing preservation and vestibular ablation**
- 3. Procedures involving ablation of 8th nerve**
- 4. Procedures involving chemical ablation of the vestibular end organ**
- 5. Procedures involving non chemical ablation of the vestibular end organ**
- 6. Procedures involving hearing and vestibular ablation**

Procedures involving hearing and vestibular function:

Surgeries described under this head attempt to reverse the pathology involved in the hydrops and restore normal endolymphatic volume and pressure.

These include endolymphatic sac procedures, Cody's surgery and Fick cochleosacculotomies.

Procedures on endolymphatic sac:

In 1926, Portmann performed the first surgery on the endolymphatic sac decompression to treat Meniere's disease.

Endolymphatic sac enhancement: This procedure involves just exposing the endolymphatic sac and duct. Patient's symptoms improve by this simple surgical procedure. The effectiveness of this procedure can be accounted by increased blood supply to the periductal tissues surrounding the sac. This increased blood supply flushes out endolymph and debris from the sac reducing its volume.

Endolymphatic sac decompression:

Portmann's operation is otherwise known as endolymphatic sac decompression. This procedure involved opening the sac. Drainage of endolymph from the sac drains into the subarachnoid space. Shambough advocated wide decompression of the sac by removing the bone overlying it and the adjacent posterior fossa dura. Shea inserted a teflon film through the opening in the sac to keep it patent.

Extended mastoidectomy is performed. Care is taken to skeletonize posterior fossa dura, sigmoid sinus and posterior semicircular canal. The endolymphatic sac is distinguished from the posterior fossa dura by differences in its color and texture. The sac looks whiter and thicker than the surrounding dura. Anatomical landmark for location of the sac is Donaldson's line. This is an imaginary line passing from the centre of the horizontal canal cutting through the posterior canal. The sac usually lies below this line.

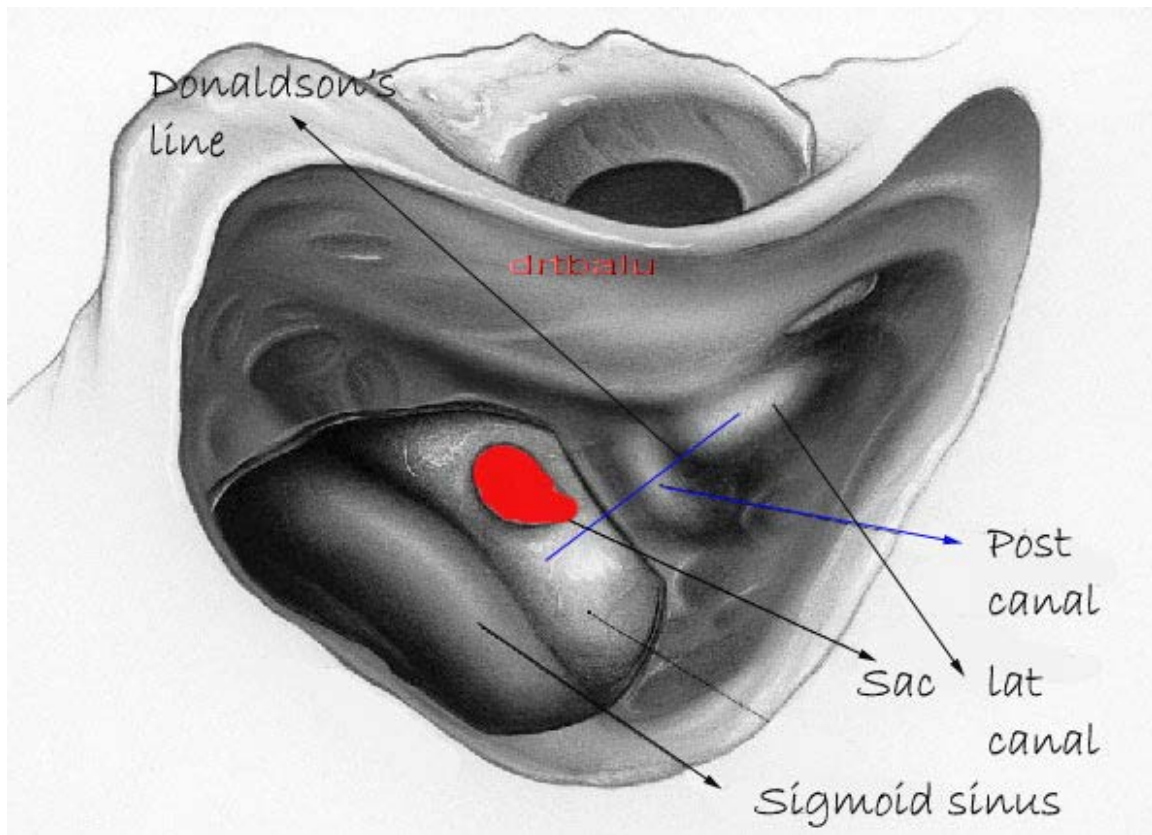


Figure showing the position of the sac

Lateral leaf of the sac can be incised. Teflon shunts can be inserted to keep the fistula patent.

One way Arenberg valve can also be introduced in place of teflon shunts.

This surgical procedure is safe. Hearing and balance mechanisms are preserved.

Critics of this procedure question the ability of low volume low pressure sytem to decompress through the fistula created. Proponents of this surgical procedure say that it works by producing surgical trauma which causes an increase in blood flow. This increased blood flow clears the sac from debris and excess endolymph.

Cody's tack procedure:

This procedure again is designed to create a fistula in the saccule via the oval window in the hope of decompressing the sac. In this procedure a sharp tack is placed through the membranous attachments of the foot plate. This tack will perforate the saccule when it starts to enlarge during acute phase of Meniere's

disease causing it to drain its secretions thus decompressing it. This surgical procedure has been abandoned because of its inconsistent results.

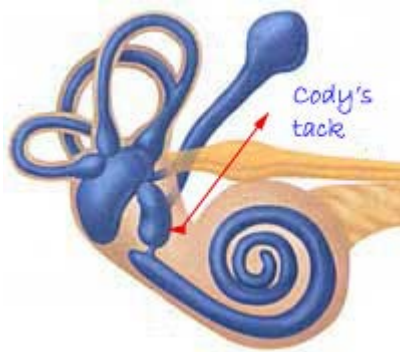


Figure showing Cody's procedure

Non chemical ablation of vestibular end organ:

Ultrasonic radiation when applied to the labyrinth ablates the vestibular function while preserving hearing. Ultrasound energy applied to the inner ear must be applied by placing the probe over the thinned out portion of the lateral canal or on the round window. When ultrasonic probe is applied to the thinned out lateral canal wall, is associated with irritative nystagmus for the first half hour, then there is a period of no nystagmus. This is followed by paralytic nystagmus.

Vestibular ablation could also be performed by using a cryoprobe. The cryoprobe is cooled to -160 degrees centigrade and is applied to the lateral canal which has been exposed after mastoidectomy. Cryo probe is applied for three cycles of 2 minutes each. These procedures have not found favour because of the high incidence of recurrent vertigo.

Ablation of 8th nerve:

In 1904 R.H. Perry performed the first 8th nerve division for persistent aural vertigo. In 1928 Dandy used suboccipital approach to section the 8th nerve. McKenzie in 1936 popularized selective sectioning of the vestibular component of 8th nerve. He also showed that by selectively sectioning the vestibular component vertigo could be controlled with excellent preservation of hearing. House in 1961 exposed the internal acoustic meatus through middle cranial fossa. He sectioned the nerve medial to Scarpa's ganglion. Sectioning of the nerve medial to the ganglion reduces the incidence of neuroma formation.

Fisch in 1984 proposed a variation of middle fossa approach called the transtemporal supralabyrinthine approach. This procedure involves removal of the root of the zygoma and roof of epitympanum, in addition to as much bone as possible from above the labyrinth itself. This approach provides better exposure to the internal acoustic meatus, with minimal degree of temporal lobe retraction.

Labyrinthectomy:

This procedure is a highly destructive surgery. It is used in patients with Meniere's disease associated with severe degree of hearing loss. The labyrinth is exposed after performing mastoidectomy. All the three semicircular canals are drilled out exposing the membranous labyrinth. The membranous labyrinth can be opened up and its inner contents sucked out using suction.

Vibrator therapy

Vibrator therapy has been approved by FDA as a treatment modality for Meniere's disease. Vibrations are produced by Meniett Device. This device is a low pressure pulse generator whose vibrations are used as a treatment modality for Meniere's disease.



Meniett device

Major advantages of this treatment modality are:

- 1. Non destructive**
- 2. Non invasive**
- 3. Safe**
- 4. Portable**
- 5. Does not require post therapy rehabilitation**

The ear plug of the Meniett device is used to plug the external auditory canal. The device then performs a leakage test to ascertain whether the external canal has been sealed properly by the ear plug. Once the leakage test is performed and no leakage has been detected, the device will transmit the vibrations through the external auditory canal. Patient must undergo grommet insertion prior to this treatment. The vibrations of the device gets transmitted to the middle ear cavity through the ventilation tube. These vibrations then influence the inner ear fluid mechanism via oval and round windows.

The actual mechanism of this vibrator therapy is still unknown. One possible theory is that vibrations reaching the membranous labyrinth agitates and pushes endolymph out of the endolymphatic sac.

Treatment plan:

- 1. Patient should be confirmed of having Meniere's disease**
- 2. Ventilation tube should be inserted prior to treatment**
- 3. Initially patient undergoes training to use the device**
- 4. The treatment is self administered by the patient.**
- 4. It is administered thrice a day, 5 minutes each time**
- 5. The treatment schedule is continued for 5 weeks**

Who should receive vibrator therapy?

- 1. Classic unilateral Meniere's disease**
- 2. Intense vestibular and cochlear symptoms**
- 3. Failed medical therapy**
- 4. Bilateral Meniere's disease**
- 5. Over 65 years of age**
- 6. Imbalance, aural fullness and tinnitus after gentamicin treatment**

Contraindications for vibrator therapy:

- 1. Perilymph fistula**
- 2. Acoustic neuroma / brain tumor**
- 3. Retrocochlear damage**
- 4. Low pressure hydrocephalus**