

Peripheral Vestibular Disorders

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The peripheral vestibular system consists of the sense organs in the inner ear and the vestibular division of the eighth cranial nerve. Disorders in any of these parts will lead to peripheral vestibular dysfunction.

At the level of the brainstem central vestibular nuclei, the system is said to be in equilibrium when there is an equal electrical input coming in from both inner ears. If there is an imbalance or asymmetry to the input, this is then interpreted by brain as a head movement that requires compensatory postural and eye repositioning. When vestibular loss causing an asymmetry of input occurs in the acute setting an individual will typically experience a hallucination of movement better known as vertigo. Their eyes will demonstrate a rapid repetitive true and fro movement called nystagmus. If the acute loss of vestibular activity affects both ears symmetrically and at the same time (i.e. A bilateral vestibular loss from systemic aminoglycoside ototoxicity) vertigo does not typically occur. The individual will more likely complain of imbalance and oscillopsia (visual blurring with head movement) from impairment of vestibular ocular reflex function bilaterally

Following an acute unilateral vestibular loss there is gradual compensation by the brain. This is achieved by comparing the visual and sensory inputs to the vestibular inputs and resetting the balance point centrally. There are limits to the compensation process however. Although there is usually effective compensation for a low frequency vestibular loss, the brain unfortunately cannot compensate for a high frequency vestibular impairment.

As discussed previously, with a careful history and examination most of the time a diagnosis can be made. In the case of peripheral vestibular disorders they tend to fall into four major groups: benign paroxysmal positional vertigo (BPPV), endolymphatic hydrops (idiopathic (i.e. Meniere's disease) or secondary), recurrent vestibulopathy (RV) and vestibular neuronitis (VN).

Benign Paroxysmal Positional Vertigo (BPPV)

BPPV is by far the commonest peripheral vestibular disorder. It has an incidence reported to be from 10 to 20 per 100,000 although one suspects it is probably higher because of the self limiting nature of the condition. It can coexist with other peripheral vestibular disorders. Its association with Meniere's disease has been reported by several authors¹. The recognition of this condition is important as it is a benign condition and can usually be easily treated.

Pathophysiology

Most authors now agree that BPPV is caused by free floating particle in the semicircular canals (although the original theory of cupulolithiasis (i.e. particles attached to cupula) may still be the cause in few patients). The origin of these particles is thought to arise

from the otoconia found on the utricle. Displaced otoconia can enter into any of the three semicircular canals but are usually found in the gravity dependent posterior semicircular canal (superior canal BPPV is extremely uncommon and almost impossible to detect clinically). The majority of patients with BPPV therefore present with posterior canal involvement. In one study lateral canal BPPV was seen in 5 percent of BPPV patients². Clinically those with lateral (or horizontal) canal BPPV are more likely to recover spontaneously². In cupulolithiasis the patient often has a more persistent positional type of vertigo.

Etiology

Most patients presenting with BPPV have no obvious predisposing cause although several conditions have been associated with this condition. The most common cause of secondary BPPV is head injury. This can arise from a minor bump to the head. BPPV is also seen after vestibular neuronitis and as mentioned previously there is a higher incidence of BPPV in those with Meniere's disease. There are reports in the literature suggesting an association between migraine and BPPV³. BPPV has been also reported after inner ear (especially stapedectomy) surgery⁴.

History

By following the four by four method described in the previous chapter BPPV can be easily diagnosed on the basis of history alone. Patients usually give a crisp description of their dizziness. The nature of the dizziness in BPPV is that of a true rotatory vertigo typically lasting for 5-30 seconds at a time associated with certain head movements. It is most commonly provoked by turning in bed to the affected side or by suddenly extending the neck to look up. It is not unusual to have more than one attack of dizziness in a 24 hour period. Patients with BPPV do not usually have other symptoms of inner ear dysfunction such as hearing loss, tinnitus or aural fullness. Patients should also be questioned about recent head injury, vestibular neuronitis, migraine and recent ear surgery.

Examination

After the formal neurotological examination has been completed the diagnostic Dix-Hallpike's manoeuvre should be performed. With the patient sitting on the bed, the head is turned to either the left or right. The examiner looks at the patient's eyes for nystagmus when the patient's head is placed down in the supine head hanging position. The head should be held in this position for at least for 10 seconds as there is usually a latency of 1-5 seconds before any nystagmus might commence. When BPPV is clinically present a torsional geotropic nystagmus with the fast phase beating towards the affected (dependent) ear should be observed. The direction of nystagmus reverses (ageotropic reversal) as the patient sits up. With lateral canal BPPV a more horizontal and persistent nystagmus will be seen in the head hanging position of the affected side.

Management

BPPV is usually a self limiting condition and most cases resolve spontaneously with time. For persistent cases particle repositioning maneuvers should be tried before surgical intervention is considered.

The Particle Repositioning Manoeuvre (PRM) for posterior canal BPPV has been shown to be an effective method of treatment². The manoeuvre starts with the patient in the head hanging position on the affected side. After the nystagmus fatigues and vertigo stops the patient is rolled away towards the unaffected side keeping head in full extension. The patient is kept in this position for 30 to 60 seconds or until any secondary nystagmus disappears. The patient is then asked to sit up. If the procedure is successful retesting should not induce any nystagmus.

For lateral canal BPPV a “barrel roll” type procedure has been described by Eply⁵. In this procedure the patient is rolled away from the affected side in 90 degree increments until a full roll is achieved.

For patients whom cupulolithiasis is suspected Brandt-Daroff exercises may be used⁶. In this physical therapy manoeuvre the patient is asked to sit in the middle of the bed and move rapidly from sitting to lying onto the affected side with the head turned half way up staying in this position for 30 seconds or until the vertigo disappears. The patient then returns to sitting position and stays in this position for another 30 seconds. The same manoeuvre is performed on the other side. The patient performs five repetitions three times daily for two to three weeks.

Surgical management should be reserved only for patients with severe recurrent and disabling BPPV. Singular neurectomy historically was a very effective procedure initially popularized by Gacek⁷. This procedure however is technically demanding and has a relatively high complication rate with regards to vestibular and sensorineural hearing loss.

Posterior semicircular canal occlusion on the other hand has been now shown by many authors to be a safe and highly effective treatment for posterior canal BPPV^{2, 8, 9}. In this procedure following a cortical mastoidectomy, the labyrinth is identified and the posterior canal is blue lined creating a small window (fenestra) into the canal. The membranous labyrinth is then compressed using previously harvested periosteum to formally occlude the canal. Care should be taken not to suction on the opened canal as this could lead to a sensorineural hearing loss.

Meniere’s disease (Idiopathic endolymphatic hydrops)

In 1861 Prosper Meniere described a symptom complex which later became synonymous with recurrent attacks of episodic vertigo, fluctuating sensorineural hearing loss and tinnitus. Later the sensation of aural fullness (or pressure) was added to the definition. Not all patients with Meniere’s disease present in a classical manner. As a result the diagnosis of Meniere’s disease can be very difficult. Unfortunately many patients are probably inappropriately labeled as having Meniere’s before they demonstrate the full symptom complex.

The incidence of the disease varies in different countries but it has been reported between 7 to upwards of 150 per 100,000 persons. There is no racial or sex difference. The incidence of bilateral disease from one report was about 47 percent¹⁰.

Pathophysiology

Most authors agree that pathologic basis for Meniere's disease is that of idiopathic endolymphatic hydrops thought to be due to excess endolymph accumulation (or possibly overproduction). Although most patients with Meniere's disease have been identified to have endolymphatic hydrops at necropsy not all hydrops will lead to Meniere's disease. Hydrops has also been demonstrated after labyrinthitis, head trauma, otitis media, mumps and meningitis¹¹. This questions whether hydrops is the cause for Meniere's disease or a pathologic reaction to injury at the level of the inner ear.

In a typical attack of Meniere's it is believed that when a critical pressure is reached the membranous labyrinth ruptures leading to the admixing of potassium (K⁺) rich endolymph with sodium (Na⁺) rich perilymph (the so-called "Na⁺ - K⁺ intoxication" theory). This in turn leads to reduced neuronal activity resulting in auditory and vestibular dysfunction^{12, 13}. As the membrane ruptures repair themselves auditory and vestibular function recovers. Repeated ruptures however lead to an eventual permanent loss of function.

History

Diagnosis of Meniere's disease is most often made from history alone. The vertigo usually lasts minutes to hours. It can happen at rest but is typically exacerbated by head movement. The usual pattern is one of acute vertigo associated with hearing loss and tinnitus; the hearing loss, tinnitus and any aural pressure improving as the dizziness resolves. In general it is very unusual for the vertigo to last more than 24 hours although a feeling of imbalance can last for many days. Lermoyez described a variant in a Meniere's attack in which the tinnitus and hearing loss preceding the vertigo resolved with the onset of acute dizziness¹⁴.

Examination

Meniere's patients are rarely seen by physicians during acute attacks. If acutely symptomatic a horizontal nystagmus would be expected away from the involved side and a sensorineural deafness might be detected. When in remission the neurotological examination tends to be unremarkable. In advanced cases however there might be evidence for a vestibular loss on the Halmagyi and head shake tests (both high frequency clinical tests of horizontal canal vestibular function).

Investigations

Pure tone audiometry classically demonstrates a low frequency sensorineural hearing loss on the affected side although many different audiometric configurations can be present. Electronystagmography (ENG) is still the most commonly performed vestibular test. A significant caloric reduction helps to identify the involved side. Electrocochleography (ECOG) is often used for confirmation of the diagnosis. The usual finding in hydrops is that of an increase in the summating potential (SP) relative to the

action potential (AP). The SP/AP ratio is typically increased by more than 0.50 (SP/AP ratios > 0.30 being somewhat suspicious for hydrops).

The glycerol dehydration test typically involves a loading dose of glycerol followed by serial audiograms over the next three hours. A threshold shift of 25 decibel over three consecutive frequencies or a 16 percent improvement in the speech discrimination indicates a positive result suggestive for hydrops.

Management

Success of medical and surgical treatments in Meniere's disease must be tempered by the fact that there is a significant placebo effect in this condition. This problem and the many different ways of reporting results has lead to the development of formal guidelines by the American Academy of Otolaryngology- Head and Neck Surgery (AAO-HNS) for reporting results in the treatment of Meniere's disease.

Medical Management

Most treatments aim to reduce endolymphic hydrops. Patients are advised about dietary modification which includes reduction of salt and caffeine intake. Diuretics have been used for Meniere's patients with variable efficacy. Some studies have shown control of vertigo in up to 58% of the patients¹⁵. Nevertheless there have been few proper double blind randomized control trials for this condition^{16, 17}.

Betahistine has also been used extensively for this condition. Vasodilatation caused by this drug is believed to be responsible for its effect. The result from this treatment has not been consistent. Calcium channel blockers (flunarizine, verapamil etc) have been tried as well.

Surgical Management

Surgery can be divided into functional and deafferentative procedures. Before attempting any surgical procedure one must keep in mind that not all patients might be able to compensate centrally for an iatrogenic peripheral vestibular loss. Some patients might be left with a more severe disability after such a procedure. The other consideration for a more conservative approach is that over a patient's lifetime there is upwards of a 50% chance they might develop the disease in the contralateral ear.

Endolymphatic sac decompression is both a functional and a hearing preservation procedure that was first described by George Portmann in 1926. Initially a popular procedure, its effectiveness was seriously questioned by the so-called Danish Sham Study¹⁸. In this double blinded study patients were treated either with sac decompression or simple cortical mastoidectomy. The results were similar in the 2 groups at the 1 year follow-up mark. Long-term follow-up interestingly has demonstrated more favorable results in those who had a simple cortical mastoidectomy (i.e. the sham procedure).

The operation of a vestibular nerve section (vestibular neurectomy) represents an intracranial hearing preservation though deafferentative procedure which was popularized by House in 1961¹⁹. The operation is performed via a middle fossa approach with selective section of the vestibular nerve. In good hands there appears to be a 95 % control of the vertigo. Others have tried different access routes including retrosigmoid and retrolabyrinthine approaches^{20, 21, 22}. Complications of these operations include CSF leakage, facial palsy, hearing loss, meningitis and subdural haematoma.

Chemical vestibular ablation (intratympanic therapy) appears to be a less invasive method for the deafferentation of vestibular function. Aminoglycosides such as gentamicin are known to be ototoxic but appear preferentially vestibulotoxic. These drugs are used either as drops through ventilation tubes or they are injected through the tympanic membrane into the middle ear directly. In one study Nedzelski et al using transtympanic injections of gentamicin demonstrated vertigo control in 83% of the patients. A profound hearing loss however occurred in 10%^{23,24}.

When hearing preservation is not a concern then a labyrinthectomy should be considered in those with incapacitating vertigo. Various methods have been described but a transmastoid total osseous labyrinthectomy has been favored by the senior author. Vertigo control is achieved in more than 95% of patients.

Bilateral Meniere's disease

Mycostatin suspension (100,000 U PO QID) has been shown to be effective in a small group of patients treated at the House Institute. The mechanism of action however is not known.

Injection of streptomycin parenterally has also been used for control of incapacitating bilateral Meniere's disease.

Transtympanic local overpressure treatment with the Meniett device has been shown to be an effective and safe method of treatment in Meniere's disease. In one study from Sweden, Densert and Sass reported vertigo improvement in 34 of 37 patients followed for two years²⁵. In a more recent study Gates et al showed a significant difference between treatment and placebo groups although follow up was only four months²⁶.

Secondary endolymphatic hydrops

Otologic Syphilis

Treponema Pallidum infection can certainly lead to a vestibulocochlear loss. It has been reported to account for 6.5% of unexplained sensorineural hearing loss and 7% of patients initially considered to have Meniere's Disease in few series^{27,28}. Symptoms result from otic capsule involvement. Gummatous osteitis and obliterative endarteritis leads to fibrosis of the membranous labyrinth and subsequent endolymphatic hydrops. Osteitis involving the ossicles can also lead to conductive hearing loss. In congenital syphilis a triad of sensorineural hearing loss, interstitial keratitis and notched incisors is an exclusive feature of Hutchison's triad.

Hennebert's sign (pressure induced nystagmus and vertigo) and the Tullio phenomenon (nystagmus produced by loud noise and vertigo) often presents in patients with otologic syphilis.

Investigations

Serologic tests for syphilis are the main diagnostic tools. These include the venereal disease research laboratory (VDRL), rapid plasmin regain (RPR) and flocculation treponemal antigen absorption antibody (FTA- Abs) tests.

Management

This is usually involves high dose penicillin intramuscular or intravenous (2.4 million units) for at least two weeks plus steroid therapy (Prednisone 40-60mg per day).

Delayed Endolymphatic Hydrops

Hydrops sometimes develops in patients who have lost their hearing in one or both ears previously^{29,30}. The cause of hearing loss can be viral, from head injury, meningitis or any other etiology. Patients subsequently develop attacks of vertigo similar to that seen in Meniere's disease in a delayed fashion.

If one ear is affected then labyrinthectomy provides for excellent results. In patients with bilateral or involvement in their only hearing ear then management should be conservative at all times.

Cogan's Syndrome

This syndrome is characterized by interstitial keratitis, bilateral endolymphatic hydrops and nonreactive tests for syphilis. It is believed to be an autoimmune vasculitic disorder. It is divided into typical or atypical variants according to the eye findings. In the typical form there is an interstitial keratitis involving the eye. In the atypical form there might be a scleritis, episcleritis, papilledema or retinal detachment³¹. There is usually multisystem involvement which can include the central nervous system, cardiovascular system, gastrointestinal tract, lungs and kidneys.

Otologic symptoms appear at times similar to Meniere's disease. Because of bilateral involvement an affected individual usually experiences a progressive hearing loss with oscillopsia, ataxia and imbalance if left untreated.

Management

Treatment is with high dose steroid (prednisone 2mg/kg/day) which might help prevent permanent hearing loss if administered early. Cyclophosphamide can be used for the systemic disease and as a prednisone sparing agent.

Recurrent Vestibulopathy (RV)

Recurrent vestibulopathy (RV) refers to recurrent attacks of vertigo similar to Meniere's disease but without the cochlear symptoms (i.e. hearing loss, tinnitus and aural fullness). In one study patients with recurrent vestibulopathy were followed for 8.5 years; 60% of the patients had spontaneous resolution of their vertigo, 15-20% developed Meniere's disease, 10% experienced benign positional vertigo and 10% continued to have similar disease symptoms³². The etiology of recurrent vestibulopathy remains unknown.

Management

Symptomatic treatment is usually sufficient but the medical treatment options used in Meniere's disease (diuretics or Betahistine) can sometimes help to control the attacks.

Vestibular Neuronitis

Patients with vestibular neuronitis (VN) usually present with sudden severe onset vertigo which lasts for days with gradual improvement. There is often a history of preceding upper respiratory tract viral infection. There should be no hearing loss or symptoms of focal neurological dysfunction. The nystagmus is directed away from the involved ear³³. When the acute vertigo subsides patients often have a sense of imbalance made worse with quick head movements which may last for months. Some patients go on to develop benign positional paroxysmal vertigo (BPPV).

Investigations usually demonstrate a caloric reduction on the affected side. Audiometry is normal although some investigators have shown a high frequency sensorineural hearing loss.

Management

Symptomatic treatment with vestibular suppressants helps reduce the vertigo and nausea an individual with acute VN experiences. Vestibular rehabilitation therapy may also prove helpful.

Inner Ear Fistula

By definition a fistula is an abnormal communication between two epithelial lined surfaces. In an inner ear fistula there is communication between perilymph of the inner ear and the middle ear space.

Etiology

Surgery is the most common cause especially post stapedectomy³⁴. Other causes include chronic infections, cholesteatoma, head trauma and barotrauma.

History

Patient's symptoms are very variable and may mimic Meniere's disease. An increase in the CSF pressure for example after straining, coughing and lifting may lead to vertigo and imbalance (Hennebert's phenomenon). Loud noise can also cause vertigo (Tullio's phenomenon).

Examination

After a full neurotological examination the diagnostic fistula test should be performed. With the pneumatic otoscopy air is insufflated into the ear and eyes are observed for nystagmus.

Investigations

Useful investigations include an audiogram looking for sensorineural hearing loss. Electronystagmography may show a vestibular loss on caloric testing but this is not

always reliable. Electrocochleography findings are said to demonstrate larger summing potentials especially with straining. This however is not very specific for this condition as similar findings are said to occur in Meniere's disease.

Management

In acute situations this should include bed rest, head elevation and laxatives to reduce the risk of increased intracranial pressure. Surgical exploration is required in those with persistent symptoms. Both oval and round windows and other suspected sites should be closed with fat or fascia.

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