Vestibular Neuritis and Labyrinthitis

Synonyms of vestibular neuritis: vestibular neuronitis; acute vestibular syndrome; idiopathic acute vestibular dysfunction

Vestibular neuritis and labyrinthitis are sometimes used interchangeably. However, experts in the field recommend that the term 'vestibular neuritis' be confined to cases in which the vestibular nerve only is involved, with the term 'labyrinthitis' being used in cases in which the vestibular nerve and the labyrinth are affected.

Vestibular neuritis is a very common cause of vertigo, labyrinthitis less so. Typically they produce disturbances of balance to varying degrees and may affect one or both ears. Essentially there is a sudden disruption of afferent neural input resulting in acute vertigo plus, in the case of labyrinthitis, hearing loss.

Aetiology

Vestibular neuritis is most likely a vestibular neuropathy caused by reactivation of latent type 1 herpes simplex virus in the vestibular ganglion, although autoimmune and microvascular ischaemic insults are also possible mechanisms. It most commonly affects the superior division of the nerve, which is much longer than the inferior division and travels through a very narrow bony passage, making it more vulnerable to the effects of swelling or ischaemia.

A prior upper respiratory tract infection has been reported to occur in as many as 100% of cases.

Labyrinthitis

The labyrinth consists of the peripheral sensory organs for balance and hearing, in a delicate membranous network (incorporating the utricle, saccule, semicircular canals and cochlea). Symptoms of labyrinthitis occur when there is inflammation of the membranous labyrinth and when there is damage to the vestibular and auditory end organs. Since the cochlea is invariably affected in labyrinthine inflammation, hearing loss is always present to some degree.

Many cases of labyrinthitis appear to be viral in origin and an upper respiratory tract infection precedes the onset of symptoms in about half of cases.

Bacterial labyrinthitis is a dangerous disorder in which bacteria gain access to the membranous labyrinth through anatomical connections:

- Between the central nervous system and subarachnoid space via the internal auditory canal and cochlear aqueduct; or
- Through congenital or acquired defects of the bony labyrinth.

Labyrinthitis may also be associated with systemic disease. Many factors can cause cochlear trauma, including vertebrobasilar ischaemia, meningitis, Ménière's disease and medication (eg, aminoglycoside).

It is worth noting that viral infections cause both congenital and acquired hearing loss (rubella and cytomegalovirus are viral causes of prenatal hearing loss). Postnatally, viral-induced hearing loss is usually due to mumps or measles. Viral infections are also implicated in idiopathic sudden sensorineural hearing loss (SNHL).

Epidemiology

There is a wide variability in reported prevalence of diseases causing vestibular dysfunction - from 3.1% one-year prevalence to 35.4%; however, in all studies the incidence increases with age.

Vestibular neuritis affects adults and children but has a peak age of onset of 40-50 years. This incidence is about 3.5 cases per 100,000.

Viral labyrinthitis is the most common form of labyrinthitis. It is usually observed in adults aged 30-60 years and is rarely observed in children. It is most common in the fourth decade with women outnumbering men by about 2:1.

Bacterial labyrinthitis is rare in the post-antibiotic era:

- Meningogenic suppurative labyrinthitis is most often seen in young children (under the age of 2 years), when children are at most risk of meningitis.
- Otogenic suppurative labyrinthitis can be observed in all ages and is almost always associated with cholesteatoma.

Presentation

History

Patients with vertigo may find it difficult to describe their symptoms but clarifying the timing of the vertigo and the triggers, or exacerbating factors if it is continuous, is crucial to making the correct diagnosis.
Characteristically, vestibular neuritis and labyrinthitis both present with sudden, spontaneous, severe and often incapacitating vertigo:

- Vertigo, the illusion of movement, is constant and ongoing.
- It is not triggered by movement but may be exacerbated by movement (vertigo due to benign paroxysmal positional vertigo (BPPV) is episodic and triggered by movement).

- Nausea and vomiting are frequent.
- Hearing loss occurs in labyrinthitis (although never in vestibular neuritis) but may not be complained of:
  - May be unilateral or bilateral, mild or profound.
  - A feeling of fullness in the ear is more typical of Ménière's disease.

- Tinnitus may occur in labyrinthitis. In combination with profound hearing loss and severe vertigo, it is found in suppurative labyrinthitis (tinnitus was universal in one study[9]).
- Upper respiratory tract infection symptoms (preceding or concurrent) are common and fever may be present, although high fever suggests a more serious cause such as mastoiditis or meningitis.
- 25% of cases have had a single brief prodrome in the week prior to the attack. More than one suggests transient ischaemic attack (TIA) or stroke.

Although neither vestibular neuritis nor labyrinthitis is life threatening, it is essential to distinguish each from other disorders, such as a TIA, stroke or brain tumour. The following should be sought, as their presence suggests an alternative diagnosis:

- Otorrhoea is associated with middle ear disease but may also occur following head trauma.
- Otalgia suggests herpes zoster oticus, especially if the tympanic membrane is not inflamed.
- Neck pain/stiffness suggests meningitis or vertebral artery dissection.
- Facial weakness is not a feature but may occur in herpes zoster oticus and stroke.
- Cardiovascular risk factors including smoking, diabetes, hypertension and previous stroke all increase the likelihood of the symptoms being due to a TIA or a stroke.
- Drugs may cause acute vertigo[10]:
  - Aminoglycosides and other ototoxic medications.
  - Antihypertensives, such as amlodipine.
  - Antidepressants. NB: abrupt discontinuation of a selective serotonin reuptake inhibitor (SSRI) may cause vertigo.
  - Tranquilisers, including benzodiazepines.
  - Anti-epileptics.

- A family history of migraine or Ménière's disease increases the likelihood of these conditions.
- Carbon monoxide exposure is a rare cause of acute vertigo.
Examination

Clinical examination should include:

- Assessment of the external ear and tympanic membrane, looking for cholesteatoma or vesicles suggestive of herpes zoster oticus.
- Cranial nerve examination for evidence of palsyies and hearing loss.
- Check for mastoid tenderness, nuchal rigidity and high fever.
- Assessment of gait:
  - Patients tend to fall towards the affected side when standing or walking.
  - Because the brain can still process information from the visual and somatosensory systems, they should still be able to sit and stand unaided.
  - Inability to stand or walk unassisted is suggestive of ischaemia.

- A simple hearing test using a 256 Hz (middle C) tuning fork or 512 Hz (top C):
  - Weber's test involves placing a vibrating tuning fork on top of the forehead and asking if it is heard louder in either ear. There should be no difference but in nerve deafness, it is quieter in the affected ear, whilst in conductive deafness, the sound transmitted through the skull is louder in the affected ear.

- Head impulse test - a sensitive test of peripheral vestibular function - is easy for the non-expert to interpret:
  - Always start by asking the patient to sit upright and to turn their head to either side to assess any limitation of movement and ensure it is safe to proceed.
  - Advise the person to fix their gaze on your nose.
  - Using your hands, turn the head 10-20° and then rapidly turn it back to face you and watch the eyes for saccades. Repeat several times randomly to both sides.
  - If the vestibulo-ocular reflex is intact the patient will be able to keep their gaze approximately on your nose; this is normal but will also be the case in central causes of vertigo.
  - If the reflex is impaired, as it is in vestibular neuritis and labyrinthitis, a ‘catch-up’ reflexive saccade will occur at the end of the head thrust.

- Nystagmus type:
  - Nystagmus is spontaneous.
  - It is usually fine horizontal but may be mixed horizontal-torsional.
  - It may be easier to see if sclera are exposed and the movement of scleral blood vessels can be followed.
  - Keeping their head still, ask the patient to look to the right and then to the left. Holding up a blank piece of white paper to the side of their face will help by preventing them from being able to fix their gaze on something.
  - The nystagmus is consistent and unidirectional, even when the head is turned; it will be most obvious when looking towards the direction of the fast phase. It is reduced when the vision is fixed on a point - eg, your nose.
  - Nystagmus that reverses direction with gaze position, ie fast phase to the left when looking to the left that then changes to fast phase to the right on looking to the right, suggests a central cause such as stroke or TIA.

- Skew deviation:
  - This is tested by using the cover/uncover test.
  - Ask the patient to look at your nose and use your hand to cover one eye and then the other. Observe for any vertical movement of the eye as it is uncovered. Movement of the eye suggests a central cause of vertigo.
The HINTS examination, refers to the combination of Head Impulse test, Nystagmus Type and Skew and is used in patients presenting with acute, ongoing vertigo and spontaneous nystagmus, to differentiate vestibular neuritis or labyrinthitis from stroke:

- An abnormal head impulse test, unidirectional nystagmus and no vertical skew are sensitive markers of vestibular neuritis and of labyrinthitis.
- The combination of a normal head impulse test, the presence of bi-directional nystagmus and vertical skew is more sensitive than an initial MRI for ischaemic stroke detection when patients are seen in the first 48 hours of symptom onset.
- Including new hearing loss to the HINTS examination makes it an even more sensitive test for stroke; there is increasing evidence that new hearing loss in people with acute vertigo syndrome is more likely if the cause is ischaemic.[12]

A patient with labyrinthitis or vestibular neuritis is dizzy at rest, feels worse with any head motion and already has spontaneous nystagmus. As such, the Dix-Hallpike test, used to confirm posterior canal BPPV, is not indicated.

Investigations

- Routine blood tests are not helpful; neither are viral antibody tests. However, if a systemic infection is suspected, FBC and blood cultures are indicated.
- Perform culture and sensitivity testing of middle ear effusions if present.
- Most patients do not require imaging. However:
  - A CT scan can help rule out mastoiditis.
  - A temporal bone CT scan may help in patients with cholesteatoma and labyrinthitis, although gadolinum MRI is more useful in the early stages of suppurative labyrinthitis.[9]
  - If a sinister cause is suspected, MRI scan can be helpful. Compared with CT, it provides much better pictures of the posterior fossa and VIII nerve course, although still has a low sensitivity for ischaemic stroke in dizziness, especially in the first 24-48 hours.[6]
- Pure tone audiometry may be indicated in hearing loss.
- Vestibular function testing:
  - Caloric testing and an electronystagmogram may help in diagnosing difficult cases and in determining the prognosis for recovery.
  - Vestibular-evoked myogenic potentials have been developed to assess vestibular activity.[13]
  - Portable video-oculography is available which measures eye movements so as to quantify the vestibulo-ocular reflex.

Differential diagnosis[1]

There are many causes of vertigo that include:

- Serous labyrinthitis:
  - Is associated with acute or chronic middle ear disease and is a common complication of otitis media.
  - An audiogram reveals mixed hearing loss when a middle ear effusion is present.
  - Vestibular symptoms may occur but are less common.
  - The hearing loss is usually transient but may persist if the otitis is left untreated.
  - Treatment is aimed at the underlying infection and clearing the middle ear effusion.

- BPPV:
  - The characteristic nystagmus and vertigo are brief and triggered by changes of position but between movements they may have few or no symptoms.
  - Onset is sudden and severity very variable.
Vestibular migraine[14, 15]:
- Vestibular migraine is thought to be the most common cause of recurrent spontaneous vertigo attacks. In the general population the lifetime prevalence is about 1% and one-year prevalence 0.9%.
- Vertigo lasts from 5 minutes to 72 hours.
- Vertigo can precede, accompany or occur after the headache but there will be one or more migraine features with at least 50% of episodes.
- However, in about 6% the symptoms alternate between episodes.
- Hearing is only mildly and transiently affected.
- During an attack patients may develop either central or peripheral vestibular dysfunction.
- Interictal ocular motor abnormalities may be present and appear to increase over time.
- The cause may be enhanced vestibular excitability inducing interactions in the vestibular and pain pathways from the inner ear to the thalamus and cortex.

Herpes zoster oticus (Ramsay Hunt syndrome):
- Occurs when varicella-zoster virus is reactivated in the facial nerve.
- It causes facial paralysis, loss of taste, vestibulocochlear dysfunction and pain.

Perilymph fistula:
- Due to a breach in the barrier between the middle and inner ear.
- Typically vertigo presents after direct trauma or barotrauma but can occur from a cholesteatoma.

Menière's disease.
- Stroke, especially posterior inferior cerebellar artery (PICA) syndrome.
- TIA.
- Meningitis.
- Multiple sclerosis.
- Subarachnoid haemorrhage.
- Tumours of the brain or acoustic neuroma.
- Cervical spondylosis.
- Verteobasilar occlusion and vertebral artery syndrome, including vertebral artery dissection.
- Disequilibrium of ageing: premature ageing of the vestibular apparatus, similar to age-related hearing loss.
- Drug-induced vertigo, hearing loss, or both.
- Carbon monoxide poisoning.
- Autoimmune inner ear disease.
- Wernicke-Korsakoff syndrome (thiamine deficiency).

Management[7, 8]
- Consider admission to hospital or an emergency referral to an ENT specialist:
  - If the patient presents with sudden-onset unilateral hearing loss. Hearing loss can be indicative of acute ischaemia of the labyrinth or brain stem.
  - Emergency treatment in such cases can restore the patient's hearing if seen within 12 hours of the onset of symptoms.
- Otherwise, reassure the patient; they can usually be managed at home. During an acute attack the patient will want to lie still with their eyes closed.
- Encourage the patient to be active as soon as they can, even if it worsens the vertigo, as this is thought to speed up the development of vestibular compensation.
- Patients should be advised to seek further medical care for worsening symptoms - especially neurological symptoms (such as diplopia, slurred speech, gait disturbances, localised weakness or numbness)[16].
Further reading & references

7. Vertigo; NICE CKS, April 2010 (UK access only)
8. Vestibular neuritis; NICE CKS, February 2011 (UK access only)

NB: always advise patients not to drive and not to operate machinery when experiencing symptoms of vertigo or when taking medication for symptoms.

Complications

Vertebral neuritis and labyrinthitis are generally benign and self-limiting.

- Falls.
- Unilateral hearing loss.
- BPPV.

Chronic or recurrent cases merit referral to exclude sinister aetiology.

Prognosis

- The acute symptoms of vertigo, nausea and vomiting resolve after several days to a few weeks in the majority of cases of both vestibular neuritis and viral labyrinthitis, with or without symptomatic treatment.
- Labyrinthitis: recovery of hearing loss is more variable:
  - Suppurative labyrinthitis usually leaves permanent and profound hearing loss.
  - Hearing loss associated with viral labyrinthitis may recover. Disequilibrium or positional vertigo may be present long-term following resolution of the acute infection.
- Anxiety disorders and depression have been shown to be associated with self-reported vestibular vertigo, as has cognitive impairment[19]. A study of 68 patients with organic vertigo syndromes found[20]:
  - Psychiatric comorbidity rates for those with vestibular neuritis corresponded roughly with those in the general population.
  - Anxiety, phobic disorders and depression are increased in people with vestibular migraine and those with Ménière’s disease; the reasons for this may be neuro-anatomical and neuro-biological.

Further reading & references

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