



Invited Review Article

Vertigo and Imbalance

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Abstract

Vertigo, dizziness and imbalance are common complaints of patients seen in practice. Both of the terms are used interchangeably by the patients. Most of the patients seen in a balance clinic have recurrent vertigo. Recurrent vertigo is almost never due to a serious neurological problem. It is almost always due to; benign positional vertigo, Meniere's disease or migraine. The rest of the patients have an imbalance not vertigo. The clinicians should distinguish vertigo from imbalance, and do the proper tests for evaluating patient with vertigo. In this review, evaluating patients and conditions causing vertigo or imbalance are discussed in detail.

Key words: Vertigo, imbalance.

INTRODUCTION

Recurrent vertigo is almost never due to a serious neurological problem – it is almost always due to one of three basically aural conditions: benign positional vertigo, Meniere's disease or migraine. In Balance Disorders Clinic at Royal Prince Alfred Hospital, Sydney, about 150 new patients are seen each month; about 100 with recurrent vertigo and 50 with chronic or subacute loss of balance. Of the 100 or so patient with recurrent vertigo maybe one will have a neurological cause such as transient ischaemia or episodic ataxia, apart from migraine. Of the 50 patients with imbalance only about 10 will have a peripheral vestibular cause, i.e. unilateral or bilateral vestibular loss causing chronic vestibular insufficiency or benign positional vertigo (BPV) which causes imbalance and falls in the elderly. Of the other 40 only about half will have a single or principal identifiable neurological cause for the imbalance such as sensory neuropathy, hydrocephalus, spinal cord disorder; the other half, all elderly, will have several potential contributing factors such as poor vision, joint replacement, arthritis, maybe presbyopia.

In contrast a first ever attack of acute spontaneous vertigo, like a first ever headache, backache or loss of consciousness is due to serious problem until proven otherwise. While viral neurolabyrinthitis is the single most frequent cause of isolated acute spontaneous vertigo, the most important differential diagnosis is acute cerebellar infarction, a potentially lethal condition.

In general the 7 most common mistakes made by neurologists in evaluating dizzy patients are:

1. Not distinguishing vertigo from imbalance;
2. Not doing or not knowing how to do a positional test;
3. Not doing or not knowing how to do a head impulse test;
4. Not realizing that migraine is a frequent cause of vertigo without headache;
5. Not ordering or not being able to interpret an audiogram;
6. Not arranging to review the patient during a vertigo attack;
7. Ordering a magnetic resonance imaging (MRI) instead of examining the patient properly.

WHAT IS VERTIGO ?

a. Vertigo is an illusion of rotation. It is due to unequal neural activity between the left and right vestibular nuclei. Unequal activity can be produced spinning a normal person at constant speed for about 20 seconds and then suddenly stopping them, by heating or cooling one labyrinth as happens in the caloric test, or by moving otoconial particles along semicircular canal duct as happens in benign paroxysmal positional vertigo. It can also be produced by sudden unilateral destruction of a normal vestibular end-organ, nerve or nucleus or vestibulo-cerebellum, a structure that normally inhibits the ipsilateral vestibular nucleus. In contrast bilateral simultaneous vestibular destruction produces imbalance but not vertigo; in fact a patient with bilateral vestibular loss will never again experience vertigo, as would a normal subject, from unilateral vestibular stimulation.

b. Vertigo is always temporary. Even after the vestibular nerve on one side has been surgically severed, the vertigo and nystagmus that invariably follow will always abate within a few days. This is not because the vestibular nerve has rejoined but because neurochemical changes have taken place in the brainstem during the process of vestibular compensation which restores symmetrical vestibular nucleus resting neural activity (29).

c. Vertigo is always made worse by head movement so that patients who are dizzy all the time and are happy to move around when they are dizzy do not have vertigo.

Patients with aural vertigo should not lose consciousness but some patients can't give a convincing answer even to simple questions such as: "Did it feel like you were losing balance or like you were losing consciousness?" or "Did you feel like you were going to pass out or fall over?" Patients with vertigo might lose consciousness if they have been vomiting a lot, or if they had an otolithic drop attack (76, 77) and sustained a head injury on the way down.

Witness descriptions can be helpful in identifying other paroxysmal disorders such as seizure and syncope but are not in identifying vertigo. Panic attacks with hyperventilation, can cause dizziness, not vertigo, and patients with vertigo attacks can develop panic attacks and agoraphobia (78). "Phobic postural vertigo" is a variant of this problem in which patients, often with obsessive-compulsive

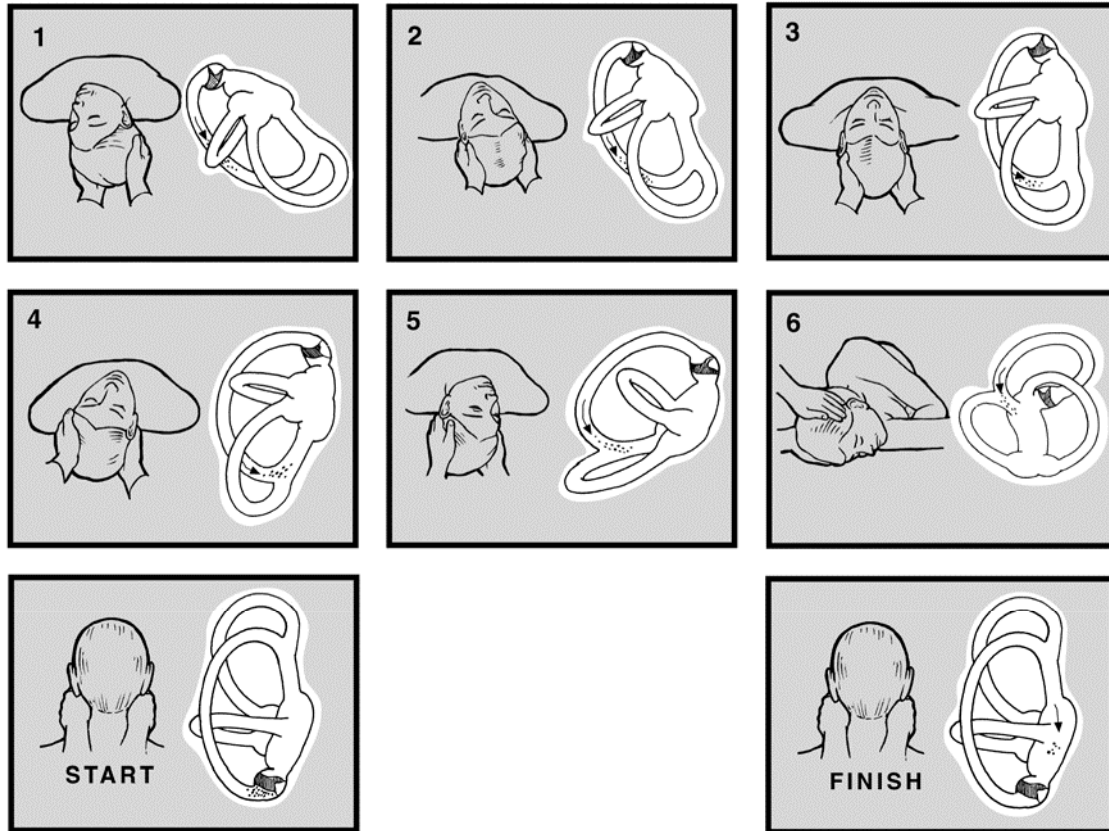
personalities, complain of a mild subjective disturbance of balance while standing or walking, with momentary illusions of motion (19). The symptoms usually occur in specific places or situations, and are associated with a distressing anxiety. Many cases follow a well-documented peripheral vestibulopathy. While not everyone likes the name "phobic postural vertigo"(22), typically patients like this improve with support and explanation (85).

POSITIONAL VERTIGO

Benign paroxysmal positioning vertigo (BPPV) is the most common cause of recurrent vertigo (93,121). The history can be unmistakable: "... whenever I turn in bed at night, or I hang the washing on the line or look under my car I am dizzy." In most patients the BPPV will occur in bouts lasting several weeks and will then spontaneously remit, only to return weeks, months, or even years later (26,131). Any patient with repeated bouts of vertigo over many years with no vestibular abnormalities on examination or testing is most likely has BPPV. BPPV is due to the movement of stray otoconial particles within the duct of one semicircular canal, usually the posterior and it is possible to remove these and so to put the patient into immediate remission basically by rolling the patient slowly by 180 degrees, from the most provocative position towards the normal side (43).

The Dix-Hallpike positioning test is the key to diagnosis (143). The aim of the positioning test is to make otoconia in the posterior semicircular canal move and so provoke vertigo and nystagmus. Brandt and Daroff (18) and then Semont et al (98) and Epley (34) showed that making the otoconia move within the duct allows them to be removed from the duct.

Consider a patient with left posterior semicircular canal BPPV, seated on a bed (Figure 1). In this position the posterior semicircular canal is gravitationally vertical so that its ampulla is its lowermost part; any otoconia in the duct will have come to rest next to the cupula (Fig. 1-start). The patient's head is now turned to the left and the patient is suddenly pitched backwards (in the plane of the posterior semicircular canal) until the head is hanging over the end of the bed so that the lowermost point becomes the midpoint of the posterior semicircular canal duct rather than the ampulla. The otoconia will now fall down from the cupula and come to rest at the midpoint of the duct. As they fall away from the



Left posterior semicircular canal BPPV. From an upright sitting position (**START**) the patient is rapidly placed into the left Hallpike position. (**1**). The head is then slowly rotated (**2-5**) to right. The patient then turns to his right side and the head is turned by another 90 degrees to the right. (**6**). After two or three minutes the patient slowly sits up again. (**FINISH**).

Figure 1. The Epley type particle repositioning manoeuvre in a patient with left posterior canal BPPV.

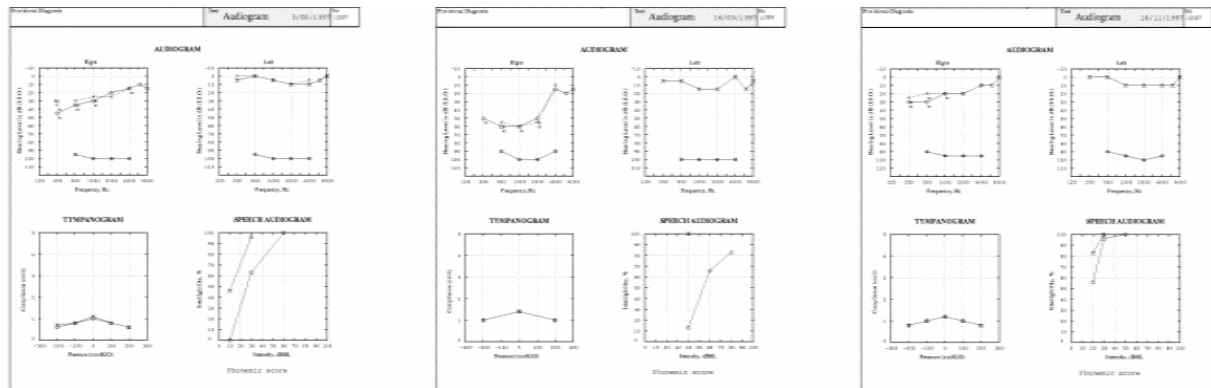


Figure 2. Serial audiograms in a patient with right Meniere's disease showing a fluctuating low-frequency unilateral sensorineural hearing loss with full recruitment – note the normal acoustic reflexes at about 100dB.

cupula they create a negative fluid pressure and so pull on the cupula producing an ampullofugal deflection which is excitatory for primary afferents of the posterior semicircular canal. As a result there is not only a brief (~20 sec) paroxysm of vertigo, but also of a nystagmus with upbeating and

counterclockwise fast phases, from the patient's point of view. That is, the rotation axis of the nystagmus is at 90 degrees to the plane of the stimulated semicircular canal, in this case the left posterior canal (8). If the patient is now slowly rotated by 180 degrees in towards his right, until the right side of his face is touching the bed, the posterior semicircular

canal will have been inverted (Fig. 1 F) so that the common crus, which joins the anterior and posterior semicircular canals, is now the lowermost point. At this stage the otoconia should move further along the semicircular canal duct and produce another, but this time less severe, paroxysm of vertigo and of counterclock-wise upbeat nystagmus. The patient, still face down, now stands up and the otoconia will continue along the common crus back into the vestibule. This is in essence the particle repositioning or perhaps better termed "liberatory" manoeuvre, as described by Epley. It will stop the BPPV attacks in about four out of five patients (143) although the condition usually remits by itself (75). Those who are resistant to repeated repositioning manoeuvres can be cured by surgical occlusion of the posterior semicircular canal (147). Post-traumatic cases in particular can be bilateral but it is sometimes difficult to tell bilateral BPPV from unilateral BPPV with a vigorous off direction (ampullopetal) nystagmus on turning toward the unaffected side (134).

In most patients with BPPV there are no other symptoms and there is no demonstrable abnormality of vestibular or auditory function. In a few it follows acute vestibular neuritis or occurs during the course of a progressive inner ear disease such as Meniere's disease or Cogan's syndrome (86). Very rarely typical BPPV occurs in a patient who turns out to have a posterior fossa disease such as a multiple sclerosis (40), tumour (32, 55), malformation, or degeneration (13) but BPPV is common enough for a patient to have both (39).

Lateral (or horizontal) semicircular canal BPPV is a variant in which the nystagmus is horizontal and usually beats toward the lowermost ear (geotropic) indicating that the otoconia in the duct are falling toward the cupula and sometimes beats toward the uppermost ear (ageotropic) indicating that the otoconia are attached to the cupula (37). Treatment of lateral semicircular canal BPPV consists of rotating the recumbent patient 360 degrees from the bad side towards the good side and then having the patient sleep only on the good side so that the otoconia can find their way out of the lateral semicircular canal back into the vestibule (117,142).

RECURRENT SPONTANEOUS VERTIGO

A patient with recurrent attacks of isolated spontaneous vertigo most likely has either Meniere's disease or migraine. The mechanism of Meniere's disease appears to

be episodic endolymphatic hypertension and distension which produces attacks of spontaneous vertigo with a low frequency hearing loss, tinnitus, and a sense of fullness or blockage in the affected ear (47, 109). The vertigo attacks usually last for a few hours, but the tinnitus and hearing loss might continue for days. The attacks might occur days, months, or even years apart. After the first few attacks of vertigo vestibular and cochlear function recover, so that the caloric test and the pure tone audiogram will both be normal. Later, after many more attacks of vertigo a permanent loss of auditory and of vestibular function becomes apparent even in between attacks. The key to the diagnosis is repeated audiometry to show a fluctuating loss frequency hearing loss (Fig 2). Improvement in hearing with glycerol or furosemide dehydration and transtympanic electrocochleography can help confirm the diagnosis (89). Meniere's disease can remit at any stage but if it does progress then in the late stages the patient is still subject to attacks of spontaneous vertigo but also has continual tinnitus in a deaf ear that distorts and recruits sound so that normal speech is unintelligible and loud sounds are painfully loud. Strict dietary sodium restriction aiming for a urinary sodium less than 50 mmol/day can be more effective and less troublesome than diuretics (<http://oto.wustl.edu/men/sodium.htm>).

Endolymphatic sac decompression can stop the vertigo attacks but can't restore the hearing and should not destroy any auditory or vestibular function. Low-dose intratympanic gentamicin, unlike labyrinthectomy, will not only stop the vertigo but should preserve hearing (154), probably as well as intracranial vestibular nerve section (49).

A common clinical problem is the patient who presents with repeated attacks of spontaneous vertigo, but is unaware of any temporary deafness, tinnitus, or fullness in one ear at the time of a vertigo attack, and who has no clinical abnormalities, a normal audiogram, and a normal caloric test. Such a patient could still have Meniere's disease. The patient might have had a temporary low-frequency hearing loss during the vertigo attack but would not have noticed it as the loss was below 1.5 kHz, the centre of the speech spectrum, and particularly they are too busy being sick to worry about a little deafness in one ear. There are also patients who have repeated spontaneous vertigo attacks for many years before they develop a unilateral tinnitus and hearing loss and the diagnosis of Meniere's disease finally becomes obvious. But a patient

with recurrent vertigo attacks and normal hearing might have vestibular migraine (125).

Some migraineurs will have vertigo as their aura (114), and will then develop a typical hemicranial headache with nausea and vomiting. Other migraineurs and their relatives will have repeated attacks of spontaneous vertigo, apart from the attacks of headache, typically lasting less than an hour, with nausea and even vomiting, but without any hearing disturbance or headache at the time (21). Attacks such as these have been called basilar migraine, although Bickerstaff was referring to a more dramatic clinical pattern (16). During attacks of migrainous vertigo there will be vestibular abnormalities that can look central, peripheral or both (145). Migrainous positional vertigo needs to be distinguished from benign positional vertigo due to vestibular lithiasis (144). Migraine can also cause a more chronic form of imbalance (41,150). Vertigo attacks in migraineurs can respond to medications used to treat migraine headaches such as an ergot, a triptan (115), or even aspirin and in some patients the vertigo attacks can be prevented by regular treatment with a beta-blocker, a calcium channel blocker, a tricyclic, valproate, acetazolamide, or methysergide (128).

DISEASES THAT DON'T USUALLY CAUSE RECURRENT VERTIGO

Certain diagnoses are almost certain to be wrong in a patient who has recurrent isolated vertigo attacks, a normal clinical examination and no objective loss of auditory or vestibular function. Acute otitis media does not cause vertigo unless there is a suppurative labyrinthitis. Chronic otitis media can, rarely, produce vertigo due secondary endolymphatic hydrops or cholesteatoma causing a perilymph fistula can but not without a hearing loss. Probably all cases attributed to spontaneous perilymph fistula are in fact a dehiscence of the superior semicircular canal into the middle cranial fossa (107) and not of the lateral semicircular canal into the middle ear. Superior canal dehiscence can also produce an air-bone gap on the audiogram that can, superficially at least, mimic otosclerosis (59,105,108). Acoustic neuroma, (vestibular schwannoma), unless intralabyrinthine, rarely produces attacks of spontaneous vertigo, and maybe never in a patient who has no fixed unilateral or asymmetric abnormalities of auditory or vestibular function (113). Microvascular loop compression is a validated cause of paroxysmal symptoms related to the trigeminal and facial nerves, but the evidence

that microvascular compression of the vestibular nerve causes paroxysmal vestibular symptoms, or any symptoms at all, is unconvincing (11). The anterior inferior cerebellar artery (AICA) normally loops into the internal auditory canal and is not a bona fide cause of symptoms. Frequent brief attacks of vertigo accompanied by unilateral hyperacusis or tinnitus can respond to treatment with carbamazepine and have been called "vestibular paroxysmia" (20) there is scant evidence for symptomatic microvascular compression in these patients.

Fisher is still correct: transient posterior circulation ischaemia is unlikely to be correct in a patient who has recurrent attacks of isolated vertigo (38). Nonetheless it could be suspected in a patient in whom some of the vertigo attacks are accompanied by other symptoms of brainstem dysfunction such as diplopia (50, 53,118). In the absence of any simultaneous brainstem symptoms, a short history, of days rather than months, of frequent brief vertigo attacks lasting minutes rather than hours several times a day, should raise the suspicion that the attacks are posterior circulation transient ischaemic attacks (TIA). Hearing symptoms, tinnitus and deafness if unilateral and occurring at the same time as the vertigo attacks suggest an aural rather than a brainstem problem. In contrast sudden, temporary bilateral hearing loss does suggest brainstem ischemia (97). Vestibular function tests are expected to be normal in patients with posterior circulation ischaemia and are only of help in a negative sense: if they show a definite unilateral abnormality this will suggest an aural rather than a central cause for the vertigo.

Rarely isolated vertigo attacks can be produced by stenosis or impending thrombosis of one vertebral artery or more seriously of the basilar artery; in such cases intervention, thrombolysis, angioplasty and stenting might be required, sooner rather than later. (122, 157).

With the widespread availability of non-invasive computed tomography (CT) and MRI vascular imaging many patients with non-specific neurological symptoms, including dizziness, perhaps even vertigo, end up having CT or MR angiography (MRA), both intracranial and extracranial. Some of these patients have abnormalities in the posterior circulation, but many of these will be asymptomatic and not dangerous. Proximal stenoses such as those at the origin of one

vertebral or subclavian artery are potentially symptomatic or dangerous only if the other

patients will have normal variants which need to be recognized as such; examples include one large dominant and one small, even vestigial, vertebral artery; one vertebral artery terminating as a posterior inferior cerebellar artery (PICA), not joining the basilar; fenestrations of the basilar; AICA loops in the internal auditory canal adjacent to the vestibulo-cochlear nerve; apparent absence of, or rather failure to image, one PICA or one AICA or one of each. (120),

FIRST ATTACK OF ACUTE SPONTANEOUS VERTIGO

In a patient who suddenly develops, for the first time ever, isolated spontaneous vertigo, nausea, and vomiting the two main diagnoses to consider are vestibular neuritis and cerebellar infarction. Sudden, spontaneous, isolated, unilateral, total, or subtotal loss of peripheral vestibular function is usually ascribed to viral "vestibular neuritis", also called "labyrinthitis", "vestibular neuronitis" and "neuro-labyrinthitis". There evidence for viral infection is slim (6) and some prefer to call it "acute unilateral peripheral vestibulopathy"

vertebral artery is vestigial or occluded. Other

(17). Corticosteroids given early help recovery of peripheral vestibular function whereas antivirals (i.e. valacyclovir) do not (136).

In patients with combined superior and inferior vestibular neuritis the clinical signs are the same as those that occur after a labyrinthectomy or a vestibular neurectomy. There is a horizontal-torsional spontaneous nystagmus with the slow phases towards the affected ear - that is, quick phases towards the unaffected ear. The nystagmus is always strictly unidirectional - bidirectional gaze-evoked nystagmus excludes the diagnosis. The nystagmus is, to some extent, always suppressed by visual fixation, and for that reason it might be missed on the standard clinical examination. When some means are used to view the eyes in the absence of visual fixation such as ophthalmoscopy with the other eye covered (159) or Frenzel glasses, the nystagmus will be evident in the primary position. The head impulse test (Figure 3) is invariably positive and shows impaired lateral semicircular canal function on the affected side (60).

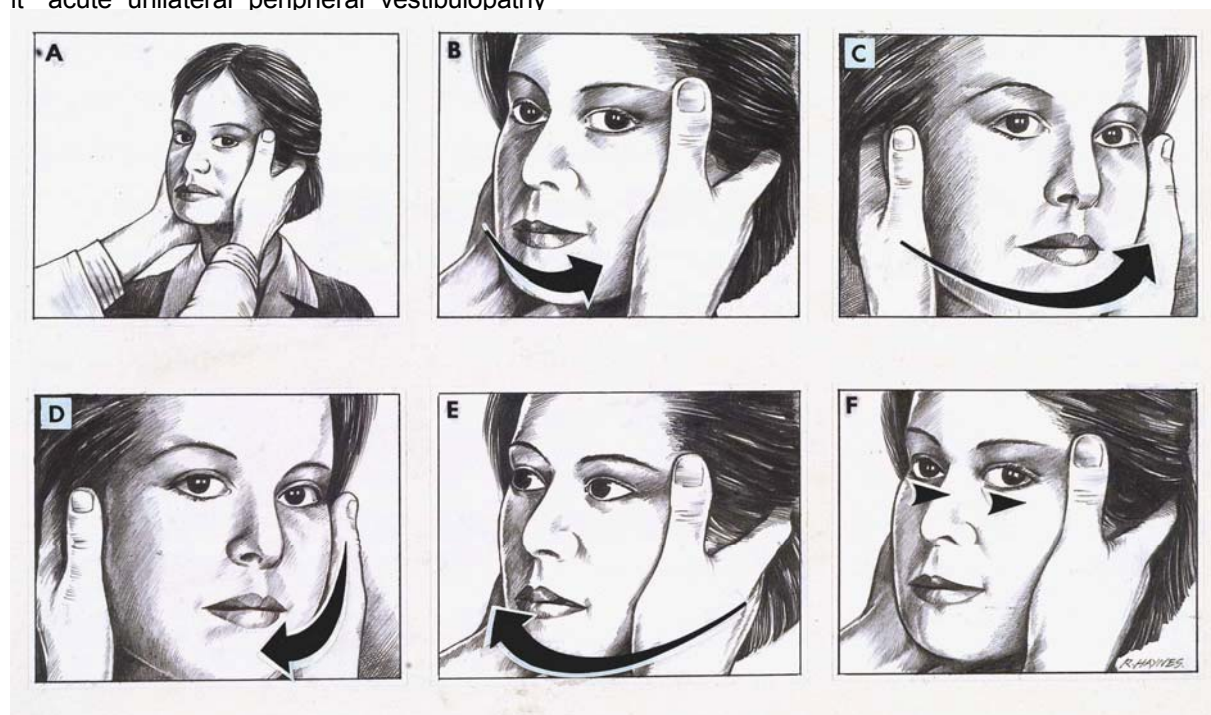


Figure 3. The head impulse test. The examiner turns the patient's head as rapidly as possible about 15 degrees to one side and observes the ability of the patient to keep fixating on a distant target. The patient illustrated has a right peripheral vestibular lesion with a severe loss of right lateral semicircular canal function. While the examiner turns the patient's head to toward the normal left side (top row) the patient is able to keep fixating on target. By contrast, when the examiner turns the patient's head to the right the vestibulo-ocular reflex fails and the

was normal. **(b)** A transoesophageal echocardiogram (showed a small atrial septal defect with spontaneous right to left shunting).

If the patient has had vestibular neuritis, there is a 1 in 5 chance that he will present later with attacks of typical posterior semicircular canal BPPV or with imbalance due to inadequate vestibular function. If the patient has had a small embolic infarct in the cerebellum, he might not present until he has had another one, this time perhaps not in the cerebellum but elsewhere.

IMBALANCE

Imbalance is not often due to a peripheral vestibular cause. About 20% of patients with imbalance will have chronic vestibular insufficiency due to unilateral or bilateral vestibular loss (127) or due to BPV (94). About 40% will have a single identifiable neurological cause for the imbalance most often: a sensory ataxia due to peripheral neuropathy or myelopathy, an extrapyramidal disorder especially progressive supranuclear palsy (PSP), a cerebellar ataxia either acquired or hereditary, a posterior fossa tumor, normal pressure hydrocephalus or orthostatic tremor. The other 40%, mainly old will have many possible factors such as poor vision, joint replacement, arthritis, maybe presbyopia (10).

Bilateral vestibulopathy. Bilateral vestibular loss causes ataxia and oscillopsia, not vertigo (129) and in the absence of any significant and relevant hearing loss it can cause diagnostic difficulties because an aural cause might not be considered in the differential diagnosis of imbalance. The patient will be able to walk perfectly well heel to toe and the only easily demonstrable abnormality will be an inability to stand, with the eyes closed, but only when trying to do so on a soft, yielding surface such as a mattress or two pillows—a sort of Romberg's test. The head impulse test will be positive to the left, right, up, and down and caloric and rotational tests will show bilaterally absent or severely impaired lateral semicircular canal vestibulo-ocular reflexes. Sometimes patients with severe unilateral loss of vestibular function will present with the same symptoms (149). The most common known cause of bilateral vestibular loss without hearing loss is gentamicin toxicity (61). Systemic gentamicin

is not cochleotoxic in human – it does not cause deafness or tinnitus, but as far as the vestibular system is concerned there is no safe dose, and any patient who notices imbalance after a hospital admission has gentamicin vestibulotoxicity until proved otherwise. As the patient might not be aware of having been given gentamicin it might be necessary to requisition the hospital's records. Other causes of isolated bilateral vestibular loss with normal hearing include hereditary vestibulopathy (81) superficial siderosis (87) and meningeal carcinomatosis (124).

Hereditary and episodic cerebellar ataxias

Patients with dominantly inherited spinocerebellar ataxia (SCA) develop, in some combination, progressive limb and gait ataxia, dysarthria and abnormal eye movements. Although horizontal gaze-evoked nystagmus and impaired smooth pursuit can occur in any SCA, in SCA6 there is downbeat nystagmus (50,138,155) or periodic alternating nystagmus (64) but, considering the abnormal smooth pursuit, VOR-suppression is sometimes surprisingly normal (139). Saccades are slow in SCA1 and SCA2 (23, 25). The vestibular function is bilaterally impaired in SCA1 (23,25) and SCA3 (52, 156) and in typical Friedreich's ataxia but unlike in SCA, it is accompanied by bilateral deafness (33, 44). While the functions of the SCA1, 2 and 3 genes are unknown, it is known that the gene underlying SCA6 codes for the alpha1 sub-unit of a P/Q-type voltage-gated calcium channel, CACNA1A.

Various CACNA1A mutations can also cause an episodic rather than a progressive ataxia - EA2. CACNA1A mutations causing EA2 have been nonsense or splice site or frame-shifts which disrupt the open reading frame, leading to truncated mutant protein products that might be non-functional (80). Several missense mutations that alter single highly conserved amino acid residues can cause EA2 (56, 137) or severe progressive ataxia (158). The episodes of ataxia lasting hours begin before the age of 20 and are typically triggered by exercise and stress and are relieved by treatment with acetazolamide. Between attacks EA2 patients can have gaze-evoked nystagmus, rebound nystagmus and positional downbeat nystagmus in the head-hanging position that, over time, becomes a spontaneous downbeat nystagmus. Later a mild truncal ataxia develops, along with impaired smooth pursuit and saccadic dysmetria. During attacks, EA2 patients often have some additional spontaneous nystagmus that is not seen during the interictal

examination. Some SCA6 patients, like EA2 patients, experience vertigo attacks which also respond to acetazolamide (82).

Patients with EA1 have shorter attacks of ataxia than EA2 patients, have interictal myokymia rather than nystagmus and are usually acetazolamide unresponsive (35, 96). The mutations in EA1 involve a potassium channel gene, KCNA1.

Sporadic and acquired cerebellar ataxia

In some patients late-onset, progressive, sporadic cerebellar ataxias might also have a genetic basis. Some of patients with this type of ataxia also have a progressive bilateral vestibular loss with the characteristic impairment of all smooth compensatory eye movements due to combined loss of vestibulo-ocular reflex and smooth pursuit eye movements (104). These patients might have Multiple System Atrophy (MSA), which can present as a cerebellar ataxia, with or without evidence of autonomic involvement, especially orthostatic hypotension and extrapyramidal features such as bradykinesia and rigidity (152).

The most likely cause of an inexorably progressive, subacute cerebellar ataxia is a paraneoplastic cerebellar degeneration. Some of these patients also have vertigo (88). Anti-neuronal antibodies can sometimes be found (54, 132) and when present these can not only help make the diagnosis, they can also narrow the site of the usually occult primary tumor which might be evident on whole body CT or positron emission tomography (PET) scanning (5, 12); both these tests can find non-specific lesions or give false-positive results. Creutzfeldt-Jakob disease can present with a similar clinical picture of inexorably progressive subacute cerebellar ataxia (84)

Cerebellar ataxia can occur in patients with gluten intolerance (coeliac disease) (1,58). Although doubts have been expressed about the causal relationship (28, 153) a gluten-free diet can improve balance. Testing for anti-gliadin antibodies is recommended in patients with imbalance and a positive result is an indication for a small bowel biopsy.

Other rare possible causes of progressive cerebellar ataxia in adults include glutamic acid decarboxylase (anti-GAD) antibodies (70, 141) and vitamin E deficiency (71).

In patients with cerebellar ataxia as part of the Wernicke-Korsakoff syndrome there should also be memory and eye movement disorders especially dysarthria and gait ataxia rather than limb ataxia; vestibulo-ocular reflexes can also be abnormal (42). Wernicke-Korsakoff syndrome is due to Vitamin B1 (thiamine) deficiency and is not always due to alcoholism. Acute Wernicke-Korsakoff syndrome is a medical emergency (160).

Extrapyramidal and basal ganglia disorders

Extrapyramidal disorders particularly Progressive Supranuclear Palsy can present with progressive deterioration of gait and balance, axial rigidity and falls (48,112). Later there is dysarthria and bradykinesia and then the characteristic eye movement abnormality: loss of vertical, at first of downward, saccades so that the patient has problems when trying to read or eat (15). Vestibulo-ocular reflex eye movements are unaffected and the patient acquires the characteristic unblinking stare. Later all saccades are lost and swallowing becomes impaired. Life expectancy is less than 5 years. Balance, posture and gait are also involved in typical Parkinson's disease (110) and in diffuse Lewy body disease (24) with characteristic small rapid steps, difficulty starting, turning and stopping. If the examiner, from behind, quickly pulls the patient back by the shoulders a patient with an extrapyramidal disorder might topple backwards, unless stopped. This is a positive pull test (111). Gait apraxia, lower body Parkinsonism or rather "Primary progressive freezing of gait" refers to elderly patients with profound difficulties in starting and stopping walking and in turning with, at least at first, no motor difficulties moving their legs while supine or with upper limbs or cranial nerves (36). Most develop other extrapyramidal features and are chair-bound within 5 years. Patients with MSA (152) can present with freezing of gait (57) or with dizziness and balance problems (148) due to extrapyramidal or cerebellar dysfunction or orthostatic hypotension.

Balance and gait normally deteriorate with age and deterioration in sensory input, especially vestibular, and the appearance of subcortical white matter changes will only account for about 30% of this deterioration (10).

Sensory ataxia in spinal cord and peripheral nerve diseases

Diseases affecting the dorsal columns of the spinal cord produce proprioceptive impairment

in the lower limbs and consequently poor balance on standing and walking – a sensory ataxia. Diseases affecting the dorsal root ganglia, called sensory neuronopathies (92), the dorsal roots themselves or their large myelinated axons in the peripheral nerves can also produce sensory ataxia. In the good old days of syphilis tabes dorsalis was the usual cause. These days vitamin B12 deficiency producing subacute combined degeneration of the spinal cord (66), copper deficiency (91) radiation damage to the spinal cord paraneoplastic sensory neuropathy, Sjogren's syndrome, coeliac disease, paraproteinemia, chemotherapy especially with platinum compounds and HIV/AIDS all need to be considered in patients with subacute or chronic sensory ataxia. Acute sensory ataxia is common to three eponymous syndromes: Guillain Barre, Miller Fisher and Bickerstaff's "brainstem encephalitis". While there are clear clinical differences in these three syndromes, they are all characterised by the presence of serum anti-ganglioside antibodies and all respond to immunotherapy with intravenous gammaglobulin or plasmapheresis.

With sensory ataxias there will be some impairment of distal position and vibration sense. With spinal cord diseases the tendon reflexes in the lower limbs will be brisk and the plantar reflexes extensor, if the pyramidal tracts are also involved. With diseases of the dorsal root ganglia, dorsal roots or peripheral nerves the tendon reflexes will be absent. The traditional Romberg test, standing feet together eyes open and then eyes closed should be positive if the ataxia is to be called sensory. If the patient passes the standard Romberg test, the test can be made more difficult by having the patient stand on a thick foam mat. While this will detect a subtle sensory ataxia it will also detect a vestibular ataxia or rather astancia/astasia and needs further information for correct evaluation.

Nerve conduction studies to look for abnormalities especially of sensory conduction, somatosensory evoked potentials to look especially for abnormalities of spinal cord conduction, cerebrospinal fluid (CSF) examination to look especially for high protein levels, spinal MRI to look for compression or high T2 signal in the dorsal columns and sural nerve biopsy to look for axonal degeneration, demyelination or vasculitis are often needed in addition to blood testing for the diseases mentioned above.

In the elderly, compressive spinal cord and cauda equina problems can present with imbalance and falls and a whole of spine MRI is worth doing in an elderly patient with imbalance and falls for no obvious reason. Nitrous oxide anaesthesia or abuse, usually in the young but sometimes in health professionals, can block vitamin B12 utilization and produce a clinical pattern similar to subacute combined degeneration (74).

Vestibular schwannoma

These days patients with vestibular schwannomas ("acoustic neuromas") are more likely to have balance problems from the treatment than from the disease. Vestibular schwannomas often grow slowly; the unilateral vestibular loss occurs over years so that patients compensate well and are unlikely to have any symptomatic vestibular ataxia. With MRI scanning of just about everyone with unilateral hearing loss or tinnitus, and those without, small asymptomatic vestibular schwannomas, are, like small pituitary adenomas (27), found incidentally (100). When symptomatic, vestibular schwannomas present with unilateral or asymmetrical bilateral, sensorineural hearing loss or tinnitus (30), or both, to an otologist rather than to a neurologist; however the otologist will not order vestibular function tests, even after MRI shows a vestibular nerve tumor, since the patient has no balance problem (101). If the tumor is then removed, while the patient actually has normal vestibular function, the operation will be like a vestibular nerve section of a normally functioning nerve. There will be an intense post-operative unilateral vestibular deafferentation syndrome with vertigo, vomiting, nystagmus etc and a 25% chance of a mild but permanent ataxia due to chronic vestibular insufficiency (127) needing ongoing vestibular rehabilitation (14,73).

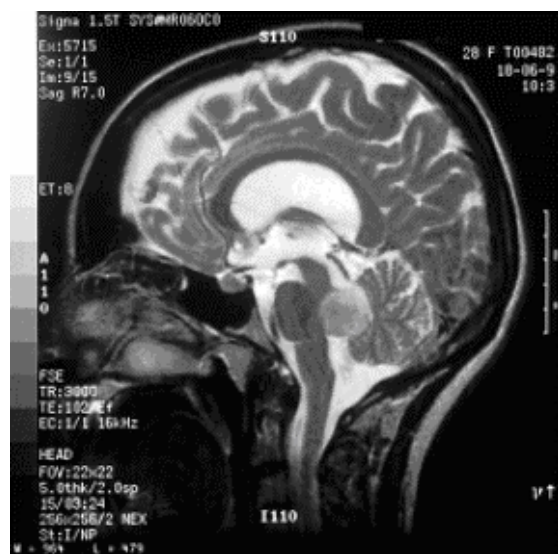
Intracanalicular vestibular schwannomas are best managed with MR monitoring since surgery will inevitably produce total unilateral loss of vestibular and cochlear nerve function. On the other hand letting the tumor grow too far into the cerebello-pontine cistern puts the facial nerve at risk during subsequent surgery (68). Other cerebellopontine angle tumors such as meningiomas and epidermoids cause imbalance only when they large enough to compress the brainstem and cerebellum

Intralabyrinthine vestibular schwannomas are rare but seem to have a propensity to cause vertigo, perhaps through a secondary Meniere-like disorder (113). Hyperventilation-induced

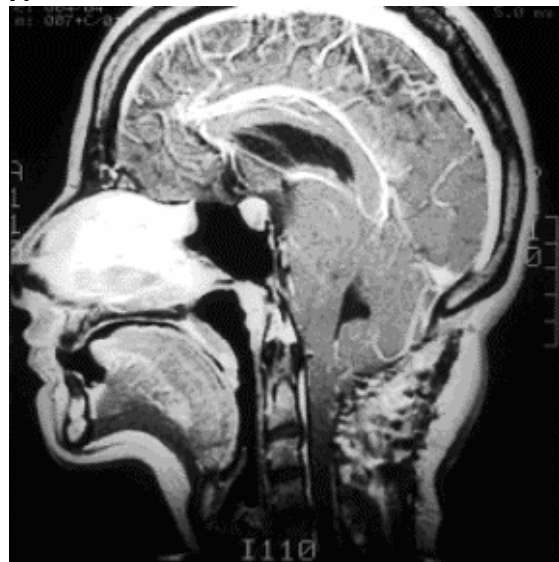
nystagmus is a rare but characteristic sign of vestibular schwannoma (106)

Cerebellar tumor

Increasing imbalance with headache and positional vertigo is the classic presenting syndrome of cerebellar tumor (67). The positional vertigo is usually persistent rather than paroxysmal (32, 55). Patients 4th ventricular tumors such as ependymomas (Figure 5) (102) can have positional vomiting, without any vertigo (31) as well as complex eye movement abnormalities, especially after surgery, including vertical strabismus due to skew deviation rather than a cranial nerve palsv (126).



A



B

Figure 5. MRI from two patients presenting with positional vertigo and progressive ataxia and headache. (a) a 4th

ventricular ependymoma and (b) a Chiari malformation.

In adults, a metastasis is the most common cerebellar tumor and can grow to a considerable size in a cerebellar hemisphere before it causes any problem apart from headache. Acute spontaneous vertigo can occur when there is bleeding into a tumor such as a metastasis or when a cystic tumor such as a hemangioblastoma suddenly enlarges. In such cases everyone gets a nasty surprise once the scan is done. Sometimes it is hard to tell a cerebellar tumor from a cerebellar infarct on CT or MRI; in such cases a PET scan can help. Hydrocephalus, which eventually occurs when cerebellar tumors grow large also, also causes ataxia (135). Recurrent subarachnoid bleeding from cerebellar tumors can produce superficial siderosis with progressive bilateral loss of vestibular and cochlear function (87). Other potential causes of progressive bilateral vestibulo-cochlear loss in cerebellar tumor patients is radionecrosis of the temporal bone (83) and malignant meningitis (72,116, 123,146)

Chiari malformation

In adults Chiari malformation can cause vestibular and balance problems (133, 151) such as cerebellar ataxia, pressure-induced vertigo, positional vertigo, positional nystagmus, often downbeating (13), nystagmus-induced oscillopsia and positional vomiting. Minimal Chiari malformations, like tiny vestibular schwannomas are discovered in patients having MRI for vertigo or non-vestibular symptoms such as headache and it can be hard to decide whether the Chiari in such cases is symptomatic or incidental (2). Herniation through the foramen magnum of the cerebellar tonsils, the human equivalent of the paraflocculus, a part of the vestibulo-cerebellum, appears to be the cause of the balance and vestibular disorders in Chiari malformations. Cerebellar tumors and lumboperitoneal CSF shunts can also cause tonsillar herniation and produce a symptomatic *acquired* Chiari malformation. http://nyneurosurgery.org/chiari_intro.htm

Normal pressure hydrocephalus

Hydrocephalus of any cause can produce imbalance in children and adults. The classical picture of low or normal pressure hydrocephalus is an elderly person presenting

with gait difficulties imbalance, urinary incontinence and failing memory. Imaging shows enlargement of all 4 ventricles with little or no cortical atrophy. CSF pressure is normal although there is increased resistance to CSF inflow with an infusion test (65). CSF diversion with a ventricular shunt can produce a dramatic improvement in balance, memory and continence, although it is hard to predict whether a particular patient will or will not improve (103). The risk of shunting is infection and subdural hemorrhage due to overdraining. If gait and balance improve after 3 days lumbar CSF drainage the patient is more likely to benefit from permanent CSF shunting (135). http://www.neurosurgery.medsch.ucla.edu/Diagnoses/Adult/AdultDis_1.html

Orthostatic tremor

Patients with orthostatic tremor have shaky legs when they stand but not when they walk or sit. Typically the patient can walk around the supermarket normally but cannot stand in the checkout queue. It is a ~16Hz tremor which is present in all the leg muscles when the patient stands but not when contracting the same muscle while sitting. Unlike essential tremor which is an ~ 8Hz tremor affecting the hands and arms and not the legs. Not only can the tremor be recorded with a simple surface EMG (Figure 6) it can also be heard with a stethoscope (sounds like a distant helicopter). Mostly there is no other neurological problem; maybe about 25% also have Parkinsonian features (46). When the tremor becomes disabling treatment with clonazepam, levodopa, etc can be tried, more in hope than expectation. <http://orthostatictremor.org/>

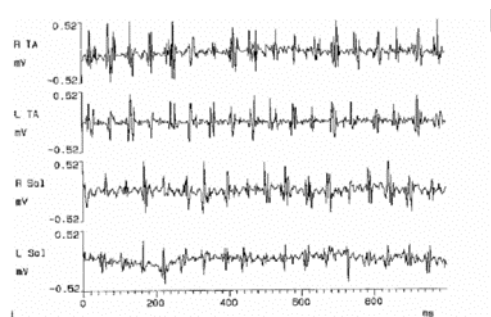


Figure 6. Surface EMG of quadriceps during standing in a patient with orthostatic tremor. The typical bursts of activity at 16 Hz are seen.

SUMMARY

In the patient with repeated attacks of isolated vertigo

- (1) Always do a positional test.
- (2) Learn to do the particle repositioning manoeuvre.
- (3) Always order an audiogram.
- (4) Try migraine treatment.
- (5) Put vertebrobasilar insufficiency at the bottom of the list.

In the patient having the first ever attack of acute spontaneous

- (1) Learn to do the head-impulse test.
- (2) Always think of cerebellar infarction.

In the patient who is off-balance

- (1) Think of gentamicin vestibulotoxicity.
- (2) Think of normal pressure hydrocephalus.
- (3) Beware of the posterior fossa tumour or malformation.
- (4) Think of orthostatic tremor.
- (5) Consider spinal cord or peripheral nerve pathology and do a serum B12.

Vertigo ve dengesizlik

ÖZET

Vertigo, dizziness ve dengesizlik günlük pratikte başvuran hastaların yaygın yakınmalarındandır. Bu terimlerin hepsi hastalar tarafından birbirlerinin yerine kullanılır. Denge kliniğinde görülen hastaların çoğunun tekrarlayan vertigosu vardır. Yineleyen vertigo hemen hiçbir zaman ciddi bir nörolojik sorun oluşturmaz. Bu durum hemen hemen her zaman; benign pozisyonel vertigo, Meniere hastalığı veya migrene bağlıdır. Hastaların geri kalanında vertigo değil dengesizlik vardır. Klinisyenler vertigo ile dengesizlik ayırımını iyi yapmalıdırlar ve vertigolu hastalarda uygun testleri yaparak değerlendirmelidirler. Bu derlemede, vertigo ve dengesizliğe neden olan durumlar, ve vertigolu hastaların değerlendirilmesi ayrıntıları ile tartışılmaktadır.

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