

## Introduction: how to use this book

Don't read this book — from cover to cover! This book has been written for the non-expert doctor who sees dizzy patients and who needs quick guidance to differential diagnosis and treatment. Conventional books are not always helpful in this situation as they are disease-oriented, and only after reading them from A to Z might you guess what your actual patient's problem is. This book is different in taking a symptom-oriented approach.

The book starts with two introductory chapters which deal with the essential anatomy and functions of the vestibular system and with the clinical assessment of the dizzy patient. These chapters are required reading as they prepare the ground for working with dizzy patients. From there on, you can turn directly to one of the problem-oriented chapters whenever you need to solve a particular dizziness problem. The clinical chapters are entitled according to common and easily identifiable clinical situations such as positional vertigo or recurrent dizziness. Each clinical chapter begins with a table for differential diagnosis with key features of the relevant disorders, followed by a concise text organised in the same sequence as the opening table. Numerous other tables facilitate the differential diagnosis. Common disorders are explained in detail, rarities are only briefly touched on. At the end of each of the clinical chapters you will find a section entitled 'What to do if you don't have a clue' that gives you some rescue ideas to manage impossible clinical situations.

The final chapter, 'Treatment of the dizzy patient', explains general aspects of treatment such as the use of vestibular sedatives and the principles of vestibular rehabilitation which are common to various disorders. The more specific aspects of treatment are dealt with in the dedicated chapters. The accompanying website material shows the clinical examination, the diagnostic and therapeutic positioning manoeuvres for benign paroxysmal positional vertigo, and exercises for vestibular rehabilitation, as well as examples of common clinical findings. Each chapter refers you to the corresponding video clips on the website, although you may prefer to see them all in one go by way of a 'hands on' introduction on how to deal with a dizzy patient.

The world of dizziness has changed completely in the last two decades, as new treatable syndromes have been identified, such as vestibular migraine, the variants of benign paroxysmal positional vertigo, and psychiatric causes of dizziness. We hope that this book will stimulate your interest in vertigo and balance disorders and that it will make you feel optimistic when facing your next dizzy patient.

## Chapter

## 1

# Essential anatomy and functions of the balance system

## Introduction

If you are reading this book you are almost certainly a busy clinician. We understand your apprehension at having to go back and review some basic anatomy and physiology concepts. All we can say is that everything you will see in this chapter will have a direct application in the diagnosis and management of your dizzy patient. One other preliminary comment: immediately after the three main headings in this chapter you will find *brief summaries* on pages 2, 11 and 15. You can read these summaries straight away, if you are conversant with the subject or wish to know what the content of this chapter is – but this may feel heavy going. Alternatively, you can come back to each of the summaries once you have read the whole section, to consolidate what you have learnt.

Normal balance is the consequence of continuous interaction between vestibular, proprioceptive and visual mechanisms, which in turn are integrated and modulated by all levels of the central nervous system (CNS). A lesion or dysfunction in any of these mechanisms can create balance problems per se or interfere with the recovery of otherwise straightforward vestibular disorders. For instance, a patient with an acute viral infection of the vestibular nerve (vestibular neuritis), even if it causes permanent unilateral absence of vestibular function, could recover quickly if the patient is a young, fit person. The same vestibular lesion can lead to permanent balance symptoms in an elderly person with age-related dysfunction of the visual, proprioceptive or central nervous systems.

## Anatomy and physiology of the vestibular system

### Summary

- The labyrinth is just one component in the balance system. However, most causes of dizziness are inner-ear problems.
- The semicircular canals (horizontal, anterior and posterior) sense angular head acceleration. The otoliths (utricle and saccule) sense linear head acceleration, including gravity.
- Each ear has three canals and two otoliths. Note that most vestibular tests examine only the horizontal canal, one-fifth of the vestibular labyrinth.
- The superior vestibular nerve contains afferents from the superior and horizontal canals and from the utricle. The inferior vestibular nerve carries the fibres from the inferior (posterior) canal and the saccule. This organisation explains why vestibular neuritis patients can have loss of horizontal canal function and posterior canal BPPV (benign paroxysmal positional vertigo).

- The vascular supply approximately follows the neural innervation – this is why vascular lesions can involve preferentially the cochlea or the vestibule. However, except when there is selective terminal branch arterial involvement, both organs (and the brainstem) are equally involved.
- Background discharge in the vestibular nerve (vestibular tone or tonus) explains why a unilateral vestibular lesion produces vertigo even in the absence of any head movements. It also explains why the remaining labyrinth can signal head movements in all directions: movements in the ‘on’ direction increase background discharge whereas movements in the ‘off’ direction reduce the background discharge.

Most readers of this book will be medical graduates and will have studied anatomy and physiology as separate subjects. Here we will try to combine these disciplines, and whenever possible, pathology as well, because an integrated approach will be more useful for clinicians.

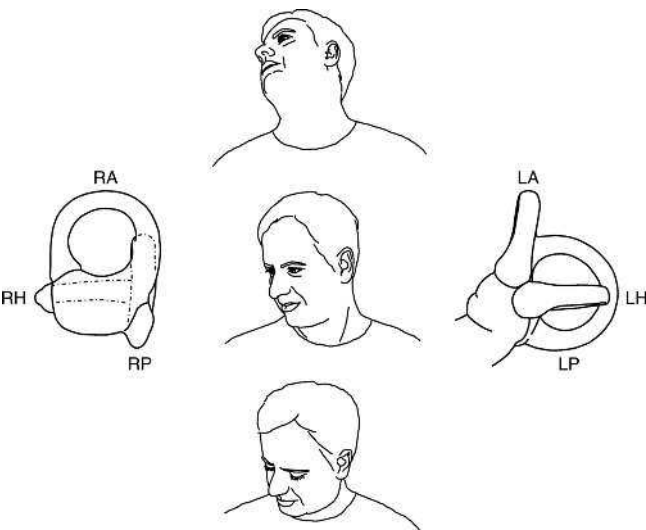
The actual symptoms (dizziness, vertigo, imbalance; see Chapter 2) in patients with various vestibular disorders are often similar. In many cases even the traditional distinction between vertigo and dizziness does not apply. The diagnosis often therefore depends on additional symptoms, which in many cases are due to extension of the causative lesion into neighbouring structures. For this reason it is important to know not only vestibular anatomy but also what structures are in the vicinity of vestibular structures or pathways.

The labyrinth consists of the bony labyrinth in the petrous section of the temporal bone and the membranous labyrinth contained therein. The sensory epithelium, which transduces sound (cochlear) and head motion (vestibular), is located within the membranous labyrinth. The membranous labyrinth contains the endolymphatic fluid which bathes the sensory epithelium; the perilymph is the fluid present between the bony and membranous labyrinths. The vestibular (or posterior) labyrinth comprises organs specialised to transduce angular acceleration, the semicircular canals, and organs specialised to transduce linear and gravitational acceleration, the otolith organs.

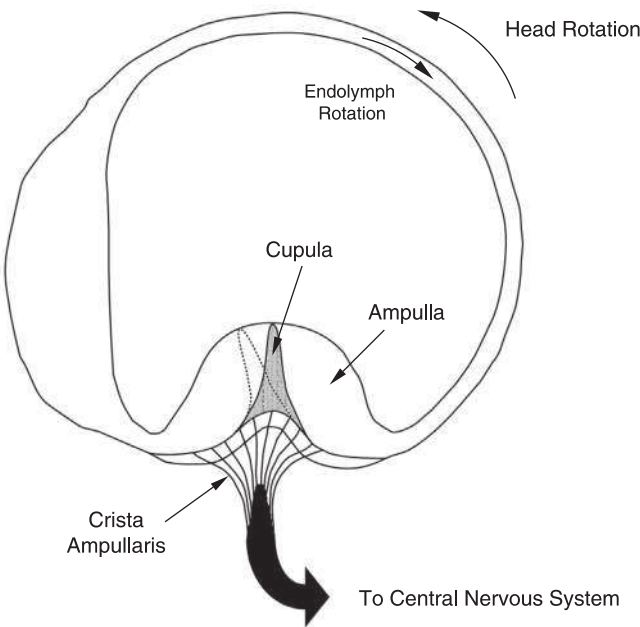
## Semicircular canals

There are three semicircular canals on each side, one horizontal and two vertical. They are positioned approximately orthogonally and so they can sense angular movements in any plane and direction (Figure 1.1). These semicircular canal planes are complementary on each side of the head so that head movements are complementarily signalled by a pair of functionally coplanar canals:

- The horizontal (or lateral) canals sense horizontal head rotation (‘no–no movements’).
- Diagonal or oblique head movements (e.g. first turn your head horizontally 45° to the right and then bend your neck up and down) are signalled by the combination of an anterior (or superior) canal on one side and a posterior (or inferior) canal on the other. In this example, the oscillation indicated will be sensed by the left anterior and right posterior canals (Figure 1.1).
- A purely sagittal head-down movement (‘yes–yes movements’) stimulates both anterior canals and inhibits both posterior canals; head-up movements do the opposite.
- A roll head movement, such as bringing the right ear down towards the right shoulder, stimulates both anterior and posterior canals on the right, and inhibits both vertical canals on the opposite side.



**Figure 1.1 Orientation of semi-circular canals.** The figure highlights pairs of canals that are activated by an oblique head movement. If you oscillate your head up and down, while your head is turned some 45 degrees to the right, the oscillatory movement is detected by the left anterior (superior) canal and the right posterior (inferior) canal (LA and RP, respectively – the LARP plane). If you were to turn the head 45 degrees to the left and repeat the up-down movement, you would be stimulating the right anterior and left posterior canal – the RALP plane.



**Figure 1.2 Semicircular canal activation.** As the skull rotates in one direction the endolymph is relatively displaced in the opposite. This endolymph motion exerts pressure on the cupula which thus activates the sensory epithelium. During a long-duration rotation (> 30 s) there is no longer relative endolymph displacement with respect to the head and thus canal stimulation gradually ceases.

The mechanism of activation of a semicircular canal is shown in Figure 1.2. Each canal has an open end, which communicates freely with the vestibule, and an enlarged or ampular end, where the sensory epithelium, the cupula, is housed. It is important to remember that each canal has an open and a closed (cupular) end – particle repositioning treatments for benign paroxysmal positional vertigo (BPPV) rely on you moving the patient’s head appropriately so that intracanalicular particles leave the canal through the open end.

The cupula is a gelatinous conglomerate of sensory hair cells – it is the bending of the cupula, and therefore of the hair or cilia, that generates bioelectrical activity and action potentials down the vestibular nerve. What makes the cupula bend is the pressure exerted by the endolymph during head rotation. As Figure 1.2 shows, head rotation to the left produces relative motion of the endolymph fluid in the opposite direction, which thus deflects the cupula.

In summary, the CNS knows *in what plane* the head has rotated by the pattern of activation of the various individual canals. The CNS knows *how fast* the head has rotated by the frequency rate of the action potentials in the vestibular nerve, which is in turn dependent on the magnitude of the endolymph-induced cupular deflection.

**Vestibular tone or tonus**

The concept of vestibular tonus is important as it has fairly immediate clinical significance. Essentially the term tonus is used because, even in the absence of any rotation, semicircular canal afferents in the vestibular nerve show a resting or ‘tonic’ discharge. Each canal has an angular direction in which cupular deflection increases the discharge in the vestibular nerve (the ‘on’ direction), and the opposite ‘off’ direction which decreases neural activity in canal afferents. The orientation of the ciliae is such that horizontal head acceleration to the right is ‘on’ for the right horizontal canal and ‘off’ for the left one. The brain knows that the head is turning because on one side neural activity increases whilst it decreases on the other.

The practical implications include the following:

1. The existence of a resting vestibular tone explains why a patient with unilateral hypofunction experiences vertigo even without making any head movement. The CNS detects a difference in discharge rate between the two sides and ‘assumes’ that the head is rotating.
2. Even in the presence of a total and permanent unilateral semicircular canal lesion, the brain is capable of sensing angular movements in all directions. The remaining labyrinth can signal both directions of motion due to the on–off arrangement. This bidirectional capacity of a single canal provides the basis for the phenomenon of *vestibular compensation* which underlies recovery of function and symptoms in patients with unilateral vestibular lesions.

**Short and long rotations**

How the semicircular canals work during brief and long rotations also has to be understood as there are clinical implications here too.

**Brisk brief rotations**

The on–off arrangement described above is not perfectly symmetrical. During accelerations in the ‘on’ direction, canal afferents have almost no saturation; vestibular nerve activity increases linearly with the velocity of the rotation. Rotations in the ‘off’ direction, however, do reach a saturation point as the decrease in vestibular nerve activity can reach down only to zero – there is not such a thing as a negative discharge rate.

A clinical consequence is that during a very fast acceleration towards the side of a lesion, angular velocity will not be faithfully transduced by the remaining labyrinth as it is working in the ‘off’ direction. Patients can therefore report symptoms such as unsteadiness, dizziness or oscillopsia during fast head movements towards the lesion side.

This phenomenon also provides the basis of an important clinical finding, the positive ‘head-impulse’ or ‘head-thrust’ sign (see page 36). Essentially, during a fast head rotation towards the side of the lesion the vestibularly driven compensatory eye movement is insufficient. Instead of a smooth compensatory eye movement, the clinician observes a refixation saccade, indicative of a hypoactive labyrinth on the side of the head turn. This will become clearer later under ‘Vestibulo-ocular reflex’.

### Long-duration rotations

Cupular deflection during rotations occurs thanks to the endolymph inertia. Think of a soup (the endolymph) in a bowl (your skull). If you suddenly turn the bowl, the bowl moves but the soup doesn’t. This relative motion between the endolymph and the semicircular canal is what makes the cupula bend. This difference will be maximal at the onset of movement, i.e. if you place the bowl on a turntable, after a while the bowl and the soup will be turning at the same speed. For this reason – namely that cupular deflection will be maximal at the onset or accelerative phase of movement – one has to think of the canals as angular accelerometers, which measure the change in velocity rather than velocity itself.

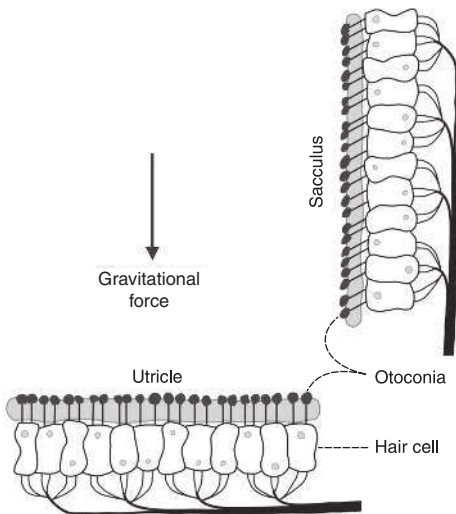
As in the culinary example given, if head rotation continues at constant speed there will be a point where skull and endolymph rotate at the same speed (i.e. there is no relative motion between endolymph and semicircular canal). During prolonged rotations, then, cupular deflection, and hence dynamic vestibular input, progressively decays and eventually stops altogether after 15–20 seconds into constant rotation. But then, if the body stops rotating, the inertia of the endolymph will deflect the cupula in the reverse direction – this explains why you feel as if you are turning when you stop a prolonged rotation such as stepping off a merry-go-round or after rolling downhill.

This is the basis of the ‘stopping’ response test during vestibular testing with rotatory Baranyi chairs. You can try the following experiment. Seat somebody on an office swivel chair, turn him round for 20–30 seconds and then stop him quite suddenly. The subject will feel vertiginous and, if you look at his eyes carefully, you will see a jerky beating of the eyes called *vestibular nystagmus*. If you time the duration of the nystagmus when stopping from right versus left rotation, you will have an indication of the degree of symmetry in vestibular activity – the essence of all vestibular tests.

## Otolith system

The otoliths sense head linear acceleration. Since gravity is linear acceleration, the otoliths also sense head tilt with respect to the gravity vector. There are two sets of otolith organs per side, the utricle and the saccule. These are shown schematically in Figure 1.3.

What makes the otolithic hair cells sensitive to linear acceleration is the fact that the gelatinous membrane embedding the ciliae of the hair cells is loaded with heavy calcium crystals called otoconia. As the head accelerates, the heavy otolith membrane is ‘left behind’, thus deflecting hair cells and generating action potentials in the vestibular afferents. The approximately horizontal orientation of the utricles makes them sensitive to linear accelerations in the horizontal plane. The sacculi are placed approximately in a parasagittal orientation and are therefore sensitive to linear accelerations occurring in the sagittal plane. It is apparent that the otolith organs also have some sensitivity to sound and this feature has some clinical relevance as new laboratory tests called VEMPs (vestibular-evoked myogenic potentials) use loud sounds, usually clicks, to activate them.



**Figure 1.3 Otolith organs.** Note the gelatinous membrane loaded with otoconia (calcium particles). In the normal upright position the utricle lies approximately horizontal and the saccule approximately vertical. In this position the force of gravity (arrow) will predominantly deflect the hair cells in the saccular organs. Note that linear head acceleration upwards will have the same effect.

It is important to bear in mind that normal head movements combine linear and angular accelerations in any plane and direction. The exquisite arrangement of four otolith organs and six semicircular canals is capable of transducing any such complex movements. At this point, we would like to remind readers that most of the traditional tests of vestibular function concentrate on horizontal semicircular canal function (caloric and rotational tests), namely 20% of the vestibular system! This is no doubt one of the reasons why so many patients have vestibular symptoms but normal vestibular function tests.

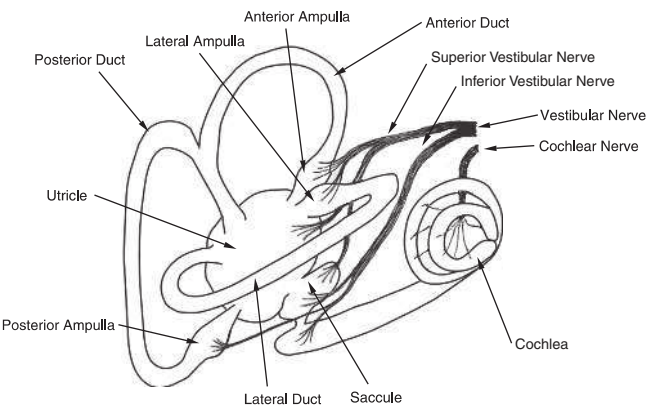
## Innervation and blood supply of the labyrinth

Each of the semicircular canals and otolith organs receives afferent innervation (singular nerves) from the vestibular nerve (Figure 1.4). Before reaching the Scarpa ganglion (where the neuronal body of these afferents is) the singular nerves are grouped into an inferior and a superior component. The superior vestibular nerve contains the fibres from the superior (anterior) and horizontal canals and from the utricle. The inferior vestibular nerve carries the fibres from the inferior (posterior) canal and the saccule. The vestibular nerve comprises the axons of the Scarpa ganglion neurons, which then enter the porus of the internal acoustic meatus, posteriorly to the cochlear nerve, forming the vestibulo-cochlear nerve or VIII cranial nerve.

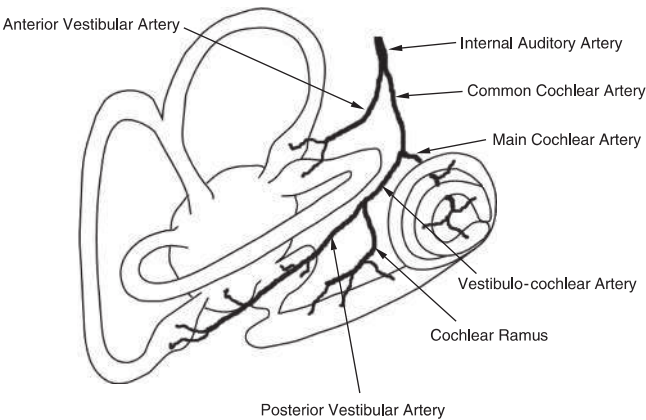
This anatomical distribution explains a few clinical points. For instance, viral vestibular neuritis or 'neuronitis' usually involves the superior vestibular nerve only. The loss of horizontal canal function explains the caloric canal paresis (see pages 36, 47 and 56). The utricular involvement can lead to degeneration of the otoconia and to the release of calcium crystals into the vestibule, from where they can fall into the lumen of the posterior semicircular canal. This is why some patients develop positional vertigo after an episode of vestibular neuritis: the spared inferior vestibular nerve and posterior canal are able to sense the abnormal endolymph currents created by the intraluminal otoconia (see Figure 5.1).

Arterial irrigation to the labyrinth comes from the internal auditory artery, which is usually a branch of the anterior inferior cerebellar artery (AICA) or, less frequently, of the basilar artery. The internal auditory artery gives off a branch called the anterior vestibular





**Figure 1.4 Innervation of the labyrinth.** The two divisions of the vestibular nerve, the superior and inferior vestibular nerve, are shown.



**Figure 1.5 Arterial blood supply to the labyrinth.** The internal auditory artery is a branch of the anterior inferior cerebellar artery, or AICA (not shown). The structures irrigated by the anterior vestibular artery correspond to those innervated by the superior vestibular nerve (Figure 1.4), principally the anterior and horizontal semicircular canals.

artery, which irrigates the anterior and horizontal canals and the utricle (the same structures innervated by the superior vestibular nerve) (Figure 1.5). The internal auditory artery continues as the common cochlear artery and it divides into two terminal arteries: (a) the vestibulo-cochlear artery, irrigating the inferior (posterior) canal, the sacculus (just what the inferior vestibular nerve innervates) and the basal turn of the cochlea, and (b) the main cochlear artery serving the bulk of the cochlea.

It can be seen that, as in other body regions, neural and vascular supply converge considerably. Accordingly, an acute selective lesion of the anterior/horizontal canals, sparing hearing, could be equally produced by vascular ischaemia of the anterior vestibular artery or neuritis of the superior vestibular nerve (as in vestibular neuritis).

Another applied concept explains the ‘AICA syndrome’, which combines unilateral deafness, canal paresis and cerebellar dysfunction, all on the same side. This is due to the fact that the internal auditory artery usually branches off the anterior inferior cerebellar artery. In view of the known anatomy, we would argue that the likelihood should be very small of recurrent vertigo in isolation (i.e. no cochlear or cerebellar–brainstem–occipital lobe symptoms) being due to vertebrobasilar ischaemia or a transient ischaemic attack (TIA). Based on this anatomical fact, clinicians should refrain from overdiagnosing



**Table 1.1** Central vestibular projections and their symptoms

Projections	Symptoms
Vestibulo-cortical	Vertigo
Vestibulo-ocular	Nystagmus
Vestibulo-spinal	Unsteadiness
Vestibulo-autonomic-limbic	Nausea, sweating, anxiety

**Table 1.2** Localisation and structures

Neighbouring structures as the basis of topographic diagnosis	
Inner ear/temporal bone	Cochlea and cochlear nerve
Internal auditory meatus	Cranial nerves V, VI, VII and cochlear nerve
Brainstem	Cranial nerves III, IV, V, VI, VII, IX, X and cerebellum

‘vascular vertigo’ due to ‘vertebro-basilar insufficiency’ when there are no accompanying auditory or CNS symptoms.

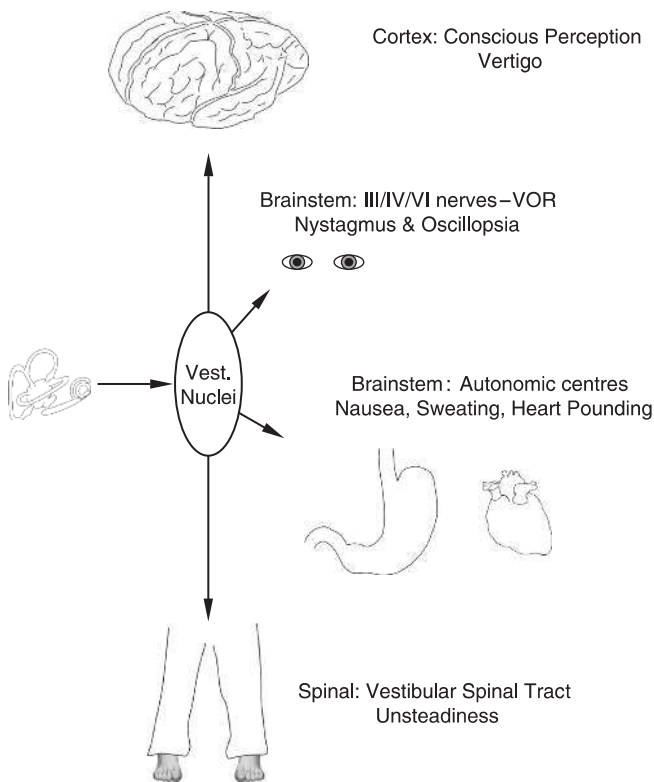
Central pathways

The vestibular pathway starts with the primary vestibular neurons in Scarpa’s (vestibular) ganglion in the temporal bone. These neurons project on to the secondary vestibular neurons in the vestibular nuclei of the brainstem. From here axons project to (a) thalamo-cortical structures, (b) oculomotor nuclei via the medial longitudinal fasciculus (MLF), (c) the spinal cord, (d) the cerebellum and (e) autonomic medullary centres (Figure 1.6). This is a perfect example of anatomic–clinical correlation because these projections explain why patients with vestibular lesions have, respectively, (a) a conscious illusion of spinning (vertigo), (b) nystagmus, (c) lateropulsion, (d) gait ataxia, and (e) autonomic symptoms such as nausea, vomiting and sweating (Table 1.1).

Topographical diagnosis

There are two important aspects of clinically applied anatomy. One is the function that is lost when a structure or pathway is damaged. The other is the structures that are near a pathway of interest; in our case, what is near the vestibular pathway at various anatomical regions. The first point is easy for us – vestibular lesions at all levels, from the ear to the cortex, will produce dizziness, vertigo or imbalance. For the second point, we need to remember the neighbouring structures and this is the basis of topographical diagnosis. Within the temporal bone, these are the organ of Corti and the cochlear nerve; as we leave the internal auditory meatus, the V, VI and VII cranial nerves are neighbours of the vestibulo-cochlear (VIII) nerve (Table 1.2).

Vestibular and cochlear pathways separate soon after entering the brainstem at the pontomedullary junction, as they proceed to their respective vestibular (medial) and cochlear (lateral) nuclei. This explains two facts:



**Figure 1.6 Central projections of the vestibular nerve**, explaining the symptoms in acute vestibular disorders: vertigo (cortex), nystagmus and oscillopsia (ocular motor nerves), unsteadiness (vestibulo-spinal tracts) and autonomic symptoms. VOR: vestibulo-ocular reflex.

- There is a high frequency of associated ipsilateral hearing problems in labyrinthine, temporal-bone and extra-axial brainstem pathology.
- There is a rarity of clinically obvious hearing problems in central vestibular disorders. This is due not only to the separation of vestibular and cochlear pathways in the CNS but also to the multiple crossings and bilateral representation of central auditory pathways.

Recall from anatomy lessons that the brainstem is a small structure comprising many vital nuclei and pathways. And yet, within this tightly packed formation, vestibular representation is quite large (no doubt indicating the evolutionary weight placed on balance function). For these reasons, almost any structure in the brainstem is a 'neighbour' to the central vestibular system, and this explains why vestibular symptoms and signs are so common in brainstem lesions. In the vicinity of vestibular pathways are cranial nerve structures accounting for symptoms of diplopia (III, IV, VI), facial numbness (V) or weakness (VII), and swallowing or speech difficulties (IX, X) (Figure 1.7). The strong functional interaction between the vestibular and cerebellar systems explains the presence of vertigo in cerebellar lesions. Reciprocally, there is frequently cerebellar ataxia in central vestibular lesions, owing to the anatomical closeness of vestibular structures and the three cerebellar peduncles.

- Long-tract symptoms such as hemianaesthesia and hemiparesis are relatively less common in central vestibular disorders than cerebellar/cranial nerves ones. This is due to the fact that the cortico-spinal (pyramidal) and somatosensory (medial lemniscus) pathways are in the ventral (anterior) segment, whereas vestibular pathways are located in the dorsal tegmentum of the brainstem, towards the floor of the IV ventricle (Figure 1.7).