Chapter 5: Causes of balance disorders

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Man has developed a very sophisticated system by which he maintains perfect equilibrium. Sensory information from the eyes and the vestibular apparatus, together with proprioceptive information from the neck and the limbs, passes into the central nervous system where, at the level of the vestibular nuclei, it is integrated and modulated by activity arising in the cerebellum, the extrapyramidal system and the cortex (Diagram 5.1). Pathology affecting the cardiovascular system, the central nervous system, the eyes, the ears, the locomotor system, the blood and the endocrine glands may all alter this fine balance of neural information and result in dys equilibrium.

Diagram 5.1. Diagram of the mechanisms involved in balance.*

1. Cortex, cerebellum, reticular formation, extrapyramidal system -->
2. Vision -->
3. Proprioception, superficial sensation -->
4. Labyrinthine activity -->

--> Integrating / data storage system -->

--> 1. Cortical awareness of head / body / motion
--> 2. Control of oculomotor activity
--> 3. Control of posture
--> 4. Control of motor skill.


Despite the prevalence of such disorders (Sheldon, 1948; Roydhouse, 1974), it is well recognized that most clinicians feel despondent when confronted with a patient complaining of a balance disorder; not least because the patient's complaints are often infuriatingly vague and non-specific. Some consideration of semantics is, therefore, of value. Dizziness is a lay term which encompasses a number of symptoms of dys equilibrium, including light-headedness, faintness, giddiness, sensations of 'swimming' or 'floating', vertigo, imbalance, ataxia, minor episodes of mental confusion or loss of consciousness. In the concise Oxford dictionary, it is defined as a 'feeling of being dazed, or in a whirl, or as if about to fall'. In medical terms, it is of little value in identifying a precise underlying pathological process. In contradistinction, vertigo is a specific symptom related directly to dysfunction of the vestibular system (Dix, 1973). By definition, vertigo is 'an hallucination of movement' (Cawthorne, 1952) or 'disagreeable sensation of instability or disordered orientation in space' (Agate, 1963). However, it is unreasonable to expect a patient, alarmed and confused by his unphysiological perceptions of movement, to define precisely his symptoms as vertigo and it is often helpful to ask if the sensation is similar to that experienced on a roundabout. It must be emphasized, however, that rotation, either subjective or objective, is not a necessary component of vertigo and, indeed, many patients complain of a sensation of instability that
is characterized by a rocking of the environment, or a feeling that the ground is not stable, for example as though they were on a boat.

A basic knowledge of vestibular function and the multiple connections of the vestibular system is essential if a systematic approach to balance disorders is to be undertaken. For a detailed explanation of the anatomy and physiology of the vestibular system, the reader is referred to Volume 1, Chapters 1 and 4. Briefly, the vestibular labyrinth is comprised of two parts: the semicircular canals, which respond to angular acceleration, and the otolith apparatus, which responds to linear acceleration. For practical purposes, the vestibular system may be considered in two halves, which are maintained in perfect balance, one with the other. Considering the semicircular canal system, upon turning the head, for example to the right, the right horizontal canal increases its firing rate, while activity of the left horizontal canal is reduced. It is the difference in activity between the functionally paired canals which is monitored by the central nervous system and allows awareness of head and body position in space, together with compensatory oculomotor and motor activity. During head movements, not only does vestibular input alter, but visual signals and the cervical proprioceptive input are also changed. From birth, this information is integrated and stored in a data centre (Roberts, 1967), which is though to be in the reticular formation of the brainstem. Afferent information is constantly compared with this data bank and under normal circumstances, there is a perfect match between the visual, cervical proprioceptive and vestibular inputs. If one system is functioning inadequately, or the integrating ability of the brainstem is impaired, there is a mismatch between the information generated by one sensory modality and that of another. This mismatch may give rise to symptoms of dysequilibrium.

### Table 5.1. Causes of dizziness

#### General medical

(1) Haematological
   (a) anaemia
   (b) hyperviscosity
   (c) miscellaneous

(2) Cardiovascular
   (a) postural hypotension
   (b) carotid sinus syndrome
   (c) dysrhythmias, including sick sinus and mitral leaflet prolapse syndrome
   (d) mechanical dysfunction: ventricular hypokinesia, aortic stenosis
   (e) shock

(3) Metabolic
   (a) hypoglycaemia
   (b) hyperventilation

#### Neurological

(1) Supratentorial
   (a) epilepsy
   (b) syncope
   (c) psychogenic
(2) Infratentorial
   (a) multiple sclerosis
   (b) vertebrobasilar insufficiency: subclavian steal syndrome, Wallenberg's syndrome, anterior inferior cerebellar artery syndrome
   (c) infective disorders: Ramsay Hunt, neurosyphilis, tuberculosis
   (d) degenerative disorders, including neuropathy
   (e) tumours, including acoustic neuroma
   (f) foramen magnum abnormalities

**Otological**

(1) Ménière's syndrome
(2) Post-traumatic syndrome
(3) Positional nystagmus
(4) Vestibular neuronitis
(5) Infection
(6) Otosclerosis and Paget's disease
(7) Vascular accidents
(8) Tumours
(9) Autoimmune disorders
(10) drug intoxication

**Miscellaneous**

(1) Ocular
(2) Odontogenic
(3) Orthopaedic, including cervical.

**Diagram 5.2. Diagnostic approach to dizziness.**

Dizziness:
1. Single episode
   1.1. Cochlear symptoms
      1.1.1. Vascular labyrinthine lesion
      1.1.2. Labyrinthine fistula
      1.1.3. Viral inner ear lesion
   1.2. No cochlear symptoms
      1.2.1. No associated symptoms
         1.2.1.1. Viral inner ear lesion
         1.2.1.2. Vestibular neuronitis
      1.2.2. Associated symptoms
         1.2.2.1. Vascular brainstem lesion
2. Recurrent attacks
   2.1. Cochlear symptoms
      2.1.1. No associated symptoms
         2.1.1.1. Endolymphatic hydrops
         2.1.1.2. Infection
         2.1.1.2.1. chronic suppurative otitis media
2.1.2.2. syphilis
2.1.3. Cerebellopontine angle lesion

2.1.2. Associated symptoms
2.1.2.1. Cerebellopontine angle lesion

2.2. No cochlear symptoms
2.2.1. No associated symptoms
2.2.1.1. Benign paroxysmal positional vertigo

2.2.2. Associated symptoms
2.2.2.1. Neurological
2.2.2.1.1. Temporal lobe epilepsy
2.2.2.1.2. Vertebrobasillary insufficiency
2.2.2.1.3. Multiple sclerosis
2.2.2.1.4. Migraine
2.2.2.2. Cervical vertigo
2.2.2.3. Medical
2.2.2.3.1. Hypoglycaemia
2.2.2.3.2. Hyperventilation
2.2.2.3.3. Cardiovascular

3. Continual imbalance
2.1. Cochlear symptoms
2.1.1. Associated symptoms
2.1.1.1. Brainstem lesion
2.1.1.2. Cerebellopontine angle lesion
2.1.2. No associated symptoms
2.1.2.1. Cerebellopontine angle lesion
2.1.2.2. Ototoxic drugs

2.2. No cochlear symptoms
2.2.1. Associated symptoms
2.2.1.1. Cardiovascular system
2.2.1.1.1. Postural hypotension
2.2.1.2. Central nervous system
2.2.1.2.1. Cerebellar disease
2.2.1.2.2. Multiple sclerosis
2.2.1.2.3. Vertebrobasillary insufficiency
2.2.2. No associated symptoms
2.2.2.1. Vestibulotoxic drugs.


A common example of such a mismatch occurs in peripheral vestibular lesions, such as Ménière's disorder, in which there is a change in the vestibular input to the central nervous system, but no change in the visual and proprioceptive inputs. Hence, a sense of vertigo is precipitated which is accompanied by spontaneous nystagmus. Alternatively, a sudden change in visual input, as occurs initially with the use of bifocal spectacles, may cause marked disorientation as a result of mismatch between visual signals and other sensory modalities required for balance. Although a derangement of one of the main sensory inputs may cause dysequilibrium, it is important to emphasize the multisensory deficit syndrome of dizziness identified by Drachman and Hart (1972). This is commonly seen in elderly patients and is
associated with minor impairment in two or more of the systems required for equilibrium: visual dysfunction (not correctable), neuropathy, vestibular deficits, cervical spondylosis and orthopaedic disorders interfering with ambulation. Furthermore, in the elderly it must be recalled that loss of neurons in the central nervous system may impair modulation of sensory activity. In addition, other multiple pathologies, such as arthritis and cardiovascular disease, may result not only in dysfunction of the vestibular system itself, but also of the multiple central connections.

**Ageing of the vestibular system**

There is a common misconception that episodes of dizziness are to be expected as a part of old age. Hence, many elderly patients with dizziness are not given the benefit of proper medical investigation. Recent work has demonstrated degenerative changes in the maculae (Johnsson and Hawkins, 1972), the crista ampullares (Rosenhall and Rubin, 1975) and in the vestibular nerve (Bergstrom, 1973). However, degenerative changes usually occur symmetrically and do not give rise to an imbalance of afferent information arising from the vestibular end organs. It is, therefore, unlikely that degenerative changes in the vestibular system themselves cause vertigo and it is more probable that this symptom is related to age-dependent changes in other systems, for example the vascular tree (Droller and Pemberton, 1953), the cervical mechanoreceptors (Arnold and Harriman, 1970), perceptual deficiencies of cutaneous and visual modalities (Bender, 1975), and cortical neurons (Brody, 1955).

**Clinical aspects**

Dizziness is an extremely common symptom, which may be consequent upon a diversity of pathologies. In approaching the diagnostic problem, it is helpful to ascertain whether the primary pathology is a vestibular, neurological or general medical disorder. To this end, a working knowledge of the differential diagnosis of dizziness is necessary (Table 5.1). It is essential to obtain a detailed and accurate history, to perform a full medical examination with special reference to the ears, eyes and neurological assessment, and to institute appropriate and specific special investigations. A simple diagnostic approach, as outlined in Figure 5.2, is of value. By considering the character of the complaint, the time course of the illness and the presence or absence of associated symptoms - cochlear, neurological or cardiovascular - it is often possible to glean valuable information upon which to base further investigation. Certain generalizations may be made.

(1) Vertigo is commonly associated with a vestibular disorder, whereas vague dizziness is more usually related to general medical disorders.

(2) Classically, a peripheral vestibular disorder is characterized by sudden, unprecipitated, short-lived (less than 24 hours) episodes of vertigo, but notable exceptions to this generalization include temporal lobe epilepsy and vertebrobasilar ischaemia. Central vestibular disorders are characterized by a gradual and insidious onset of continual imbalance, although anxious and depressed patients may also complain of constant dizziness, as may patients with bilateral vestibular failure.
(3) The duration of individual attacks is often a helpful diagnostic pointer: vertigo, secondary to cupulolithiasis may last only 30-40 seconds, whereas that associated with endolymphatic hydrops may last up to 24 hours and that secondary to labyrinthine failure may last for several days.

(4) Lesions of the labyrinth, or eighth nerve may produce associated auditory symptoms such as hearing loss, tinnitus, a sensation of pressure, fullness or pain in the ear.

(5) Vestibular symptoms and signs in the absence of cochlear abnormalities should indicate a careful search for other central nervous system abnormalities, with particular reference to oculomotor function.

(6) A thorough cardiovascular assessment is essential, particularly if a history of angina, intermittent claudication or palpitations, suggestive of an underlying dysrhythmia, is elicited.

Medical disorders

Cardiovascular disease

Vascular pathology is a major cause of morbidity and mortality in developed countries. Atherosclerosis affects predominantly middle-sized arteries, with a predilection for cerebral vessels, myocardial vessels and the peripheral vascular tree. Cardiovascular disease may give rise to impaired cerebral perfusion, resulting in dizziness and/or vertigo caused by systemic hypotension, cardiac dysrhythmia or mechanical cardiac dysfunction. Hyperlipidaemia, smoking and hypertension have been identified as risk factors in the development of arteriosclerosis.

Postural and systemic hypotension

The systemic arterial pressure is maintained by the baroreceptor reflexes which control peripheral resistance; the carotid sinus reflex and Bainbridge reflex which control reflex alterations in heart rate; and by alterations in cardiac output, consequent upon venous return. On standing from the lying position, systemic arterial pressure is maintained by reflex peripheral vasoconstriction and an increase in the heart rate. Carotid sinus disease, or pathology affecting the afferent pathways in the intermediolateral columns of the spinal cord, or in the efferent pathways of the reflex, may lead to dysfunction of reflex control of circulation with venous pooling, reduced venous return, no rise in heart rate and a resultant fall in cardiac output.

Symptomatic postural hypotension has been reported in 17-24% of elderly populations (Johnson et al, 1965; Caird, Andrews and Kennedy, 1973). A number of pathophysiological mechanisms have been postulated in the development of postural hypotension: Gribben et al (1971) have reported a marked diminution of baroreceptor sensitivity with increasing age; MacLennan, Hall and Timothy (1980) have documented structural abnormalities of the vascular tree, and Wollner (1978) has identified abnormalities in cerebral autoregulation. It would appear that, in any individual case, symptoms are precipitated by a combination of
these abnormalities, together with a superimposed cardiovascular stress such as: a fall in plasma volume (dehydration, haemorrhage); myocardial insufficiency (cardiac infarction, dysrhythmia); a rise in intrathoracic pressure (straining at stool, bouts of coughing); vasodilatation (hot weather, hot baths, exercise, micturition); a decreased venous return (hemiplegia, parkinsonism, varicose veins); electrolyte disturbance (vomiting, diuretics). Symptoms may vary from mild dizziness to sudden loss of consciousness on standing.

Secondary hypotension is associated with a number of conditions (Table 5.2). It is particularly important to exclude drug-induced hypotension; hypotensive drugs, especially if associated with diuretic administration, antidepressants, tranquilizers, antiparkinsonian agents, opiates and barbiturates should all be considered. Autonomic neuropathies are also of particular importance, and that associated with diabetes mellitus is probably the most common.

Table 5.2. Causes of postural hypotension

1. Idiopathic orthostatic hypotension
2. Secondary hypotension
   - Cerebrovascular disease
   - Parkinsonism
   - Holmes-Adie syndrome
   - Acute polyneuropathy
   - Chronic peripheral neuropathy
     - diabetes mellitus
     - alcoholism
     - amyloidosis
     - porphyria
     - carcinoma
   - Tabes dorsalis
   - Shy-Drager syndrome
   - Syringomyelia
   - Spinal cord neoplasia
   - Drugs.

The Shy-Drager (1960) syndrome is characterized by degeneration of the autonomic nervous system, associated with postural hypotension (Bannister, 1971). It is often possible to elicit other symptoms such as sweating, sexual impotence in the male, and fluctuating retention of urine, but in some patients there is no other involvement of the central nervous system. In certain cases, cerebellar ataxia, loss of tendon reflexes or signs of Parkinson's disease may develop. The natural history of the disorder is progressive, over many years and there is no curative treatment.

A hypersensitive carotid sinus reflex (Uesa, Eisenman and Stemmer, 1976) may be found in association with coronary heart disease, diffuse atherosclerosis, or hypertension. Sleight (1978) has reported that the carotid sinus is one of the most common sites for atheroma formation. In the normal subject, carotid sinus massage produces a bradycardia, but in a subject with a hyperexcitable carotid sinus, such massage, often accidental as a result of compression induced by turning the head, wearing a tight collar or shaving, may cause
symptoms of giddiness, faintness or loss of consciousness. Massage, with electrocardiographic (ECG) monitoring for up to 10 seconds, may confirm the diagnosis, but should be discontinued immediately if there is any ECG change. Although rare (Wayne, 1961), this is undoubtedly a cause of dizziness, particularly in the elderly, and relief may be obtained by denervating the sinus.

**Mechanical cardiac dysfunction**

Any pathological state interfering with cardiac output, and hence cerebral perfusion, may result in light-headedness, dizziness, vertigo or loss of consciousness. The most important cause, particularly in elderly men, is aortic valvular stenosis. The classic symptoms include breathlessness, chest pain and syncope but, in the early stages, dizziness may be the sole symptom. Commonly, the symptoms are related to exercise and on examination a slow-rising pulse of low amplitude with a sustained apex beat on palpation is found. On auscultation a single, second heart sound may be heard as the aortic component of the second sound is absent because of calcification of the valve. In addition, there may be a characteristic ejection systolic murmur at the base of the heart. A chest radiograph, electrocardiography, echocardiography and cardiac catheterization may be necessary to confirm the diagnosis. Surgical correction of this condition carries a low mortality and morbidity, even in the elderly, and the onus is therefore upon the clinician to identify this disorder during a full examination.

Other mechanical disorders which should be considered, but are beyond the scope of this chapter, include primary atrial myxoma, myocardial infarction, myocarditis, pericarditis and ventricular hypokinesia. It should be emphasized that dizziness is usually a minor manifestation in the presentation of these cardiac abnormalities.

**Dysrhythmia**

Cardiac dysrhythmias are associated with thyroid disease, intracranial pathology, cardiomyopathies, autonomic degeneration and a multiplicity of drugs including digitalis, quinidine, beta-blocking agents, antidepressants and potassium supplements. Nonetheless, the commonest cause of cardiac dysrhythmia must remain ischaemic heart disease, which has reached epidemic proportions in most developed countries. The effect of a cardiac dysrhythmia upon cerebral circulation is well established (Corday and Irving, 1960; Samet, 1973) and transient cerebral symptoms, including dizziness and syncope, may result (Goldberg,Raftery and Cashman, 1975). The detection of such dysrhythmias may prove difficult, especially if the resting electrocardiogram is normal, and prolonged ambulatory electrocardiographic monitoring may be essential to detect transient dysrhythmias as a cause of episodes of dysequilibrium (Harrison, Fitzgerald and Winkle, 1976; Luxon et al, 1980).

It should be emphasized that the complaint of dizziness and the finding of a transient dysrhythmia do not necessarily identify a cause-and-effect relationship. However, if a dysrhythmia is identified at the time of an attack, valuable positive information is obtained and, conversely, if a patient experiences a typical attack of dizziness and no dysrhythmia is found, valuable negative information is obtained (Swiryn, Rosen and Dhingra, 1980). If asymptomatic dysrhythmias are identified on prolonged electrocardiographic monitoring and full neurological investigation fails to reveal any other explanation for dizziness, a trial of antidysrhythmic therapy, or further electrophysiological cardiac investigation, may be
indicated. Treatment of asymptomatic dysrhythmias may require specific drug therapy or cardiac pacing but, in the first instance, it is important to exclude any causative disease process or medication.

The sick sinus syndrome (Editorial, 1977) is a clinical diagnosis characterized by episodes of tachycardia followed by sinoatrial block or sinus arrest (Ferrer, 1973; Kaplan et al, 1973), which may result in episodes of dizziness and/or Stokes-Adams attacks. Multiple pathological processes involving the sinoatrial node have been identified. The diagnosis may be confirmed by prolonged ambulatory electrocardiographic monitoring, or by electrophysiological studies of the conducting system of the heart. The treatment of choice requires insertion of a permanent cardiac pacemaker into the right ventricle.

**Haematological abnormalities**

Anaemia and polycythaemia are the two commonest haematological disorders giving rise to symptoms of dizziness. Severe anaemia may present with throbbing headache, dizziness, visual disturbance and fainting. All forms of anaemia should be considered including the deficiency anaemias and malignancy-related disease.

In the elderly, polycythaemia rubra vera is a diagnosis that should be considered, as one-third of patients with this disorder complain of dizziness or vertigo. Secondary polycythaemia, consequent upon acquired heart disease, respiratory disease, hyperventilation, obesity or cerebral lesions may produce similar symptoms. In the myeloproliferative group, myelomatisis and Waldenström's macroglobulinaemia may give rise to the hyperviscosity syndrome with associated dizziness or vertigo (Bloch and Maki, 1973).

**Metabolic conditions**

Hypoglycaemia is a recognized cause of dizziness and vertigo (Currier, 1971), usually resulting from overmedication with insulin, or an oral hypoglycaemic agent. In the primary care situation, any patient who is confused with light-headedness or dizziness should undergo a blood glucose estimation and then be given oral sugar. Rare endocrine disorders with associated hypoglycaemia include Addison's disease and hypopituitarism. Insulinomata are rare, but 30% of patients with this neoplasm of the beta-cells of the pancreas experience light-headedness (Crain and Thorn, 1949). Acute alcohol ingestion may precipitate hypoglycaemia, and cirrhosis, secondary to chronic alcoholism, must also be considered. The hyperventilation syndrome is a common cause of symptoms of faintness, light-headedness and disturbed balance, particularly in young women between the ages of 15 and 30 years (Pincus, 1978). It is important to identify this disorder, to avoid extensive investigation and greater anxiety. The patient is frequently reassured by the demonstration of her symptoms after 2 or 3 minutes of voluntary over-breathing at the request of the physician (see also Chapter 15).

**Iatrogenic dizziness**

Iatrogenic dizziness may be surgical or medical in origin. With improved public health measures and a reduction in chronic otitis media, together with the development of improved surgical techniques, the hazards of vestibular disturbances following middle ear surgery for otosclerosis and chronic otitis media has fallen dramatically. Nonetheless, a word of caution
is still of value. It cannot be overemphasized that great care and consideration must be exercised in the decision to perform destructive vestibular surgery. It is essential that the site of the lesion giving rise to the symptoms is conclusively demonstrated and it is established that bilateral pathology is not present; for example, Ménière's disease is frequently a bilateral condition (Stahle, 1976), but even in unilateral cases, the diagnostic problem may be compounded as nystagmus may occur in either direction (McClure and Lycett, 1978). The clinician may, therefore, easily be misled into thinking that the ear with auditory dysfunction is also the ear giving rise to vestibular symptoms. This may be incorrect, such that a labyrinthectomy results in a total hearing loss but no improvement in the vestibular symptoms. A further consideration, particularly in the elderly age group, is that compensation from total vestibular destruction may be incomplete, especially in the presence of impairment of other sensory modalities or a reduction in the integrating ability of the brainstem. These factors must all be taken into account before contemplating destructive surgery.

Despite these alarming surgical dilemmas, in this age of polypharmacy, drug-induced dizziness presents a much greater problem. Many, if not all, drugs may produce dizziness (Williamson and Chopin, 1980; Ballantyne and Ajodhia, 1984). The mechanism underlying dysequilibrium may be an unexpected side-effect of the drug, an unexpected result of a standard dose due to altered pharmacodynamics, accidental overdosage secondary to poor compliance or the result of a drug interaction.

The elderly are particularly at risk from iatrogenic dizziness as they frequently receive many drugs for multiple pathologies but are unable to comply with complicated schedules of treatment (Law and Chalmers, 1976; Wandless and Davie, 1977). Compliance must also be seriously considered in the very young and in any situation where there is a potential language barrier. Although absorption appears to be unimpaired with age, blood distribution and metabolism are reduced. Hence, drugs may produce dizziness in an elderly patient at a dose regimen which would not be expected to produce symptoms in a younger patient; Stevenson (1978) had reported increased plasma levels of propranolol, phenytoin and tricyclic antidepressants, following long-term treatment in elderly patients, compared with younger patients receiving the same dose regimen. The renal excretion of drugs is a function of renal performance, which may change suddenly and unexpectedly, for example as a result of myocardial infarction or acute infection associated with dehydration, and precipitate sudden toxicity, with dizziness, from drugs such as digoxin and streptomycin that are excreted by this route. A final consideration is that the target organ of any particular drug may be unexpectedly sensitive, for example the brain in elderly patients to psychotropic drugs such as barbiturates, morphine and nitrazepam (Castleden et al, 1977).

These brief considerations highlight the need to review carefully every drug regimen of a patient complaining of dizziness. Often, it is necessary, particularly in an elderly patient, to titrate the dose of a drug rather than commence with the standard dose. The main groups of drugs which give rise to dizziness are listed in Table 5.3. The various mechanisms by which drugs may produce dizziness may be summarized as:

(1) cardiovascular
(2) psychotropic
(3) hypoglycaemic
(4) anaemic
(5) ototoxic.
**Cardiovascular effects** may be further divided into:

(a) hypotensive effects  
(b) a reduction in cardiac output  
(c) dysrhythmias.

### Table 5.3. Drugs commonly producing dizziness / vertigo

<table>
<thead>
<tr>
<th>Category</th>
<th>Drugs</th>
</tr>
</thead>
<tbody>
<tr>
<td>(1) Neurological disease</td>
<td></td>
</tr>
<tr>
<td>Anticonvulsants</td>
<td>Primidone, phenytoin, diazepam</td>
</tr>
<tr>
<td>Antiparkinsonian drugs</td>
<td>Artane, L-dopa</td>
</tr>
<tr>
<td>Trigeminal neuralgia</td>
<td>Carabamazepine</td>
</tr>
<tr>
<td>Hypnotics</td>
<td>Barbiturates, benzodiazepines</td>
</tr>
<tr>
<td>(2) Psychotropic drugs</td>
<td></td>
</tr>
<tr>
<td>Antidepressants</td>
<td>Tricyclics, monoamineoxidase inhibitors</td>
</tr>
<tr>
<td>Major tranquilizers</td>
<td>Phenothiazines (chlorpromazine), rauwolfia alkaloids (reserpine)</td>
</tr>
<tr>
<td>Minor tranquilizers</td>
<td>Benzodiazepines (diazepam)</td>
</tr>
<tr>
<td>(3) Cardiovascular disease</td>
<td></td>
</tr>
<tr>
<td>Sympathomimetics</td>
<td>Isoprenaline, salbutamol</td>
</tr>
<tr>
<td>Hypotensive agents</td>
<td>Methyldopa, propranolol, nifedipine, captopril</td>
</tr>
<tr>
<td>Antidyshrythmics</td>
<td>Atropine, digoxin, quinidine, procainamide, beta-blockers</td>
</tr>
<tr>
<td>Diuretics</td>
<td>Thiazides, frusemide, ethacrinic acid</td>
</tr>
<tr>
<td>(4) Antimicrobials</td>
<td></td>
</tr>
<tr>
<td>Antimalarias</td>
<td>Chloroquine</td>
</tr>
<tr>
<td>Aminoglycosides</td>
<td>Streptomycin, gentamicin, tobramycin</td>
</tr>
<tr>
<td>(5) Analgesics</td>
<td></td>
</tr>
<tr>
<td>Major analgesics</td>
<td>Morphine, pentazocine</td>
</tr>
<tr>
<td>Minor analgesics</td>
<td>Aspirin, phenylbutazone, indomethacin</td>
</tr>
<tr>
<td>(6) Miscellaneous</td>
<td></td>
</tr>
<tr>
<td>Cytotoxic drugs</td>
<td>Busulphan</td>
</tr>
<tr>
<td>Hypoglycaemic agents</td>
<td>Insulin, chlorpropamide, metformin</td>
</tr>
<tr>
<td>Antihistaamines</td>
<td>Chlorpheniramine, promethazine</td>
</tr>
<tr>
<td>Hormones</td>
<td>Corticosteroids, oestrogens.</td>
</tr>
</tbody>
</table>

Hypotension may result from the overvigorous use of diuretics; from autonomic side-effects of drugs such as L-dopa and phenothiazines; and with certain cardiovascular agents, for example prazosin or captopril. Cardiac output may be reduced by beta-blocking agents in the treatment of hypertension or ischaemic heart disease. Dysrhythmias may occur following treatment with tricyclic antidepressants and digoxin, particularly in the face of impaired renal function. Digoxin and diuretics are commonly prescribed simultaneously and the increased
risk of digitalis toxicity, in the presence of diuretic-induced potassium depletion, must be borne in mind.

**Psychotropic drugs** used in the treatment of neurological conditions may give rise to dizziness. Anticonvulsants exert a toxic effect on the central vestibular pathways, producing severe imbalance and nystagmus, indistinguishable from a posterior fossa syndrome (Nozue, Mizuno and Kaga, 1973). Sedatives and tranquilizers, such as nitrazepam and diazepam, may produce hallucinations, with distortion of both visual and auditory input. They may also impair psychomotor and coordination function (Goodman and Gillman, 1975) causing dizziness.

As noted above, overdosage with insulin or an oral hypoglycaemic agent may produce hypoglycaemia but, in a diabetic, drug interaction is of particular importance. Salicylates potentiate the effect of sulphonylureas, for example chlorpropamide and tolbutamide and may result in hypoglycaemia, despite a previous record of stable management. An even more serious problem may be encountered following the introduction of beta-blockers such as propranolol, which not only potentiate the effect of hypoglycaemic agents, but may mask the warning symptoms of the sympathetic nervous system, that is sweating and tachycardia.

Dizziness secondary to anaemia may be produced by the administration of analgesics and anti-inflammatory agents causing chronic gastrointestinal bleeding. In large doses, aspirin may produce dizziness by a central action, or by a direct ototoxic effect on the inner ear.

**Ototoxicity** is an important cause of permanent instability and is discussed under otological causes of dizziness.

**Otological conditions**

Otological conditions must be excluded in all patients complaining of symptoms of vertigo, dizziness or dysequilibrium. The large number of conditions to be considered may be divided into those involving:

(1) the inner ear  
(2) the eighth nerve  
(3) the central nervous system connections.

**Inner ear pathology**

**Congenital and hereditary lesions**

Konigsmark and Gorlin (1976) estimated that 2000-4000 children are born deaf each year in the USA. They suggested that up to one-half of these are of genetic origin, while probably over one-third represent a syndrome. Despite the size of the problem, there are relatively few papers documenting the relationship between vestibular abnormalities and associated congenital or inherited hearing loss (Shambaugh, 1930; Lidenov, 1945; Arnvig, 1955; Sandberg and Terkildsen, 1965; Diepeveen and Jensen, 1968). The last two groups of workers reported a correlation between the degree of hearing impairment and vestibular pathology. Vestibular abnormalities were documented in 20% of subjects with a hearing loss
of less than 90 dB, whereas in subjects with a hearing loss of more than 90 dB, vestibular abnormalities were found in 80%. Unfortunately, there was no clear information on the site of the vestibular abnormality with respect to the site of the auditory impairment.

In the various forms of isolated genetic hearing loss with no associated abnormalities in other systems, there are unfortunately few histopathological studies and little emphasis has been placed upon vestibular investigation. In general, the studies that have been carried out would suggest that vestibular function is normal, but vestibular investigations have often been rather crude and, in the absence of sufficient information, these results must be interpreted with some caution. In certain forms of isolated inherited hearing loss, abnormalities of vestibular function have been reported, for example in dominant congenital severe sensorineural hearing loss (Muller, 1936; Konigsmark and Gorlin, 1976); dominant unilateral sensorineural hearing loss (Everberg, 1960); and in recessive congenital severe sensorineural deafness (Konigsmark and Gorlin, 1976). Again, the lack of sufficient data and inadequate vestibular test procedures, do not allow any definitive conclusions to be made, but it has been suggested that adequate tests of vestibular function may help to separate various types of inherited deafness and allow characterization of specific subgroups.

Developmental ear abnormalities are found in the rare trisomies of group D and E chromosomes: the auditory labyrinth is consistently involved, whereas the older vestibular labyrinth is usually spared. Temporal bone studies in trisomy 13 and 18 have demonstrated vestibular abnormalities (Kos, Schuknecht and Singer, 1966).

In their excellent work on hearing loss, Konigsmark and Gorlin (1976) listed over 150 separate syndromes associated with hereditary deafness. In certain of these syndromes, vestibular abnormalities are well documented. Retinitis pigmentosa and congenital sensorineural hearing loss (Usher's syndrome) is characteristically associated with vestibular hypofunction (Hallgren, 1959; McLeod et al, 1971). Vestibular hypofunction has also been documented in association with a mixed hearing loss in the Kearns-Sayre (1958) syndrome, consisting of progressive external ophthalmoplegia, retinal pigment degeneration and cardiac conduction defects. In Wilderwanck's (1963) syndrome, vestibular responses are usually abnormal and in Waardenburg's (1951) syndrome, Marcus (1968) has reported that 15 out of 16 kindred had vestibular abnormalities, and more recent work by Hageman (1975) has supported this high incidence of vestibular dysfunction in this syndrome.

Trauma

In present day society, head injuries are extremely common as a result of simple falls, for example on ice, or downstairs; as a result of violent acts, for example, muggings; and in road traffic accidents. Experimental work has demonstrated that post-traumatic petechial haemorrhages may be found in the labyrinth, the eighth nerve, the joints and nerves of the neck, the brainstem and the cerebral hemispheres. Hence it is not surprising that symptoms of dysequilibrium may result.

Recognition of a post-traumatic syndrome, including dizziness and/or vertigo, after head and neck injuries is well recognized, but it is relatively recently that an organic vestibular basis, as opposed to a psychogenic aetiology, has been established (Hart, 1973; Pearson and Barber, 1973; Rubin, 1973). Confining the remarks in this section to the
labyrinth, there are two main post-traumatic vestibular syndromes: vestibular failure and benign positional vertigo of paroxysmal type. Severe head injuries may result in transverse fractures of the temporal bone, with sudden unilateral vestibular failure. There is severe vertigo, nausea and vomiting and these symptoms are associated with a loss of balance, with a tendency to veer towards the affected side and the development of spontaneous nystagmus directed towards the normal side. Symptoms are most severe with the affected ear downwards and are aggravated by any movement. Over the first 3 or 4 days the symptoms gradually resolve, but it is 6-12 weeks before the patient becomes asymptomatic. Although this clinical picture may be seen with or without a fracture following a moderately severe head injury, a fracture should be suspected in the presence of blood in the middle ear, the external auditory meatus or bruising behind the auricle. Total unilateral loss of vestibular and/or cochlear function is permanent.

**Table 5.4. Positional nystagmus**

<table>
<thead>
<tr>
<th></th>
<th>Benign paroxysmal type</th>
<th>Central type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Latent period</td>
<td>2-20 seconds</td>
<td>None</td>
</tr>
<tr>
<td>Adaptation</td>
<td>Disappears in 50 seconds</td>
<td>Persistent</td>
</tr>
<tr>
<td>Fatiguability</td>
<td>Disappears on repetition</td>
<td>Persistent</td>
</tr>
<tr>
<td>Vertigo</td>
<td>Always present</td>
<td>Typically absent</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>Rotational to undermost ear</td>
<td>Variable.</td>
</tr>
</tbody>
</table>

The most common clinical syndrome after minor head injury is that of benign positional vertigo of paroxysmal type (Table 5.4). The symptoms frequently develop after a symptom-free interval of some days or weeks. The patient then complains of brief, but severe episodes of rotational vertigo lasting less than a minute, upon sudden changes of head position, especially on lying down and turning towards the affected ear. The most valuable diagnostic test is a briskly performed Hallpike manoeuvre, with the affected ear undermost. A latent period of 2-20 seconds is observed, followed by severe vertigo lasting less than a minute, during which the patient feels extremely distressed, vertiginous and often nauseated. On repeated testing, the symptoms and signs classically fatigue. The symptoms are accompanied by a paroxysm of rotatory nystagmus, directed towards the undermost ear. The complaint may last for months or years and the natural history of the disorder is that of relapses and remissions with eventual complete recovery in the otherwise healthy patient. Spencer-Harrison and Ozshahinoglu (1972) have made the observation that 'a protracted recovery from positional vertigo is much more likely in the later years of life, when indeed the disability may be permanent'. In the absence of any other sinister symptoms or signs, it is invaluable to reassure the patient of the benign nature of this condition, particularly as the symptoms are so unpleasant. They should be advised against climbing ladders, or standing on the edges of cliffs, or station platforms, and should be instructed in an intensive course of Cawthorne-Cooksey exercises, in an attempt to expedite compensation (Cawthorne, 1945; Cooksey, 1945) (see Chapter 15). If the patient's symptoms are sufficiently severe that they find themselves unable to do the exercises, it is valuable to prescribe a short course of vestibular sedative, such as cinnarizine, but it must be emphasized that there is, as yet, no good controlled trial to show whether exercises are more or less efficient in expediting compensation, either alone, or with a vestibular sedative. However, there is a good scientific rationale for the use of head and neck exercises in peripheral vestibular disorders (Dix, 1979).
In the very rare instance when the symptoms are interfering with the patient's lifestyle to an intolerable degree, it may be of value to consider a destructive surgical procedure, such as labyrinthectomy, division of the vestibular portion of the eighth nerve (Barber, 1964) or denervation of the posterior semicircular ampulla by section of its nerve (Gacek, 1978).

Although trauma is the commonest cause of benign positional vertigo of paroxysmal type, it is frequently seen spontaneously, secondary to labyrinthitis, endolymphatic hydrops or so-called 'vestibular neuronitis'. Pathologically, Schuknecht (1969) has postulated the mechanism of cupulolithiasis underlying this condition. He proposed that there is degeneration of the utricular otolithic membrane with release of otoconia, which become deposited upon the cupula of the posterior semicircular canal, thereby provoking rotatory nystagmus on change of position. In the absence of any other satisfactory explanation, this hypothesis has been widely accepted, although there is very limited histopathological support.

The importance of benign positional vertigo of paroxysmal type lies in its differentiation from other forms of positional nystagmus. The features of the condition are outlined in Table 5.4 and, by applying these criteria strictly, it is possible to separate positional nystagmus into two broad categories: that which may be characterized as benign and another group, in which the nystagmus persists as long as the critical head position is maintained and in which vertigo is inconstant (Nylen, 1939; Hallpike, 1967a). The importance of distinguishing these two groups lies in the finding of Spencer-Harrison and Ozsahinoglu (1972), who demonstrated that 38% of patients with persistent positional nystagmus were found to have central nervous system pathology, compared with only 4% of patients with paroxysmal positional nystagmus as defined above. Nylen (1950) found that 22% of cases of cerebral tumour demonstrated positional nystagmus, but Schiller and Hedberg (1960) emphasized that only 2% of patients with positional nystagmus have intracranial tumours. This highlights the multitude of disorders which have been associated with positional nystagmus; various ear pathologies (Dix and Hallpike, 1952); cervical spondylosis (Sandstrom, 1962); vertebrobasilar ischaemia (Spencer-Harrison and Ozsahinoglu, 1972); brainstem disorders, such as demyelination, ischaemia (Lindsay and Hemenway, 1956); posterior fossa tumours (Nylen, 1939; Cawthorne and Hinchcliffe, 1961); alcohol (Aschan, 1964) and drug intoxication (Dix, 1973). It should be emphasized that the presence of persistent positional nystagmus indicates the need for extensive and thorough neurological examination and investigation (see below).

With the marked increase in commercial diving, barotrauma must be considered. Decompression sickness is a well-recognized cause of vertigo, known among divers as the 'staggers'. If decompression occurs too rapidly, gas bubbles form in many parts of the body, particularly periarticular sites and the central nervous system. The vestibular end organ, the eighth cranial nerve or the central nervous system connections of the vestibular apparatus may be involved. One per cent of divers experience decompression problems, and approximately 5% of these suffer hearing or balance disturbances (Head, 1984). Strict adherence to internationally recognized decompression tables is of the utmost importance in avoiding decompression sickness.

Various forms of trauma, including both head injury and inner ear barotrauma in diving, and minor physical exertion such as straining, sneezing, lifting or coughing may result in a perilymph fistula with perilymph leaking into the middle ear through a rupture of the oval
or round window. This condition is commonly associated with a sensorineural hearing loss and vertigo, and is notoriously difficult to diagnose as there are no specific, consistent symptoms or signs. Vestibular symptoms include benign positional vertigo of the paroxysmal type, episodic vertigo like that of Ménière's disease, and non-specific instability of gait. Although a positive fistula sign should be sought, it, too, is an inconsistent finding. The diagnosis must be considered in all patients with dizziness presenting with a history of trauma. The diagnosis may only be confirmed upon demonstration of a perilymph leak at tympanotomy, but even this may be difficult (Ludman, 1984). If a leak is identified, it may be plugged with ear lobe fat, temporalis fascia or tragal perichondrium. Vestibular symptoms are usually greatly improved by fistula closure, but hearing improvement is rare.

**Infection**

Viruses, bacteria, treponemes and fungi may give rise to infection of the inner ear.

Vertigo, associated with bacterial infection of the middle ear cleft, is more commonly a sequela of chronic destructive middle ear disease, than acute infection. Acute infective labyrinthitis is easily recognized by the development of severe rotational vertigo, together with sensorineural hearing loss. The condition demands immediate swabs for microbiological examination, followed by antibiotic therapy (see Volume 3, Chapter 12). It cannot be overemphasized that in any vertiginous patient, middle ear disease must be excluded before other avenues of investigation are pursued. If there is the slightest suspicion of middle ear disease, a fistula sign should be sought, but even if this is negative, it may be necessary to proceed to operative inspection, before finally excluding middle ear disease as a cause of labyrinthine erosion.

A spontaneous fistula of the labyrinth is almost always the result of bone erosion by cholesteatoma, but rarely it may occur in syphilitic osteitis, tuberculous otitis media, chronic perilabyrinthine osteomyelitis or neoplasia, for example a glomus jugulare tumour.

Two bacterial infections which may involve the inner ear and/or the eight cranial nerve deserve special mention. *Petrositis* is the result of perilabyrinthine infection which extends into the apical portion of the petrous bone. It may present as Gradengo's (1893) syndrome: otitis media with involvement of the trigeminal ganglion, giving rise to pain behind the ipsilateral eye and involvement of the abducens nerve, as it crosses the petrous bone, and consequent paralysis of the ipsilateral lateral rectus muscle. The syndrome is often associated with vertigo and hearing loss, either as a result of labyrinthine of eighth cranial nerve involvement. *Otitis externa* is a common benign disorder but, in debilitated patients, particularly diabetics, it may present in a more malignant form. An infection with *Pseudomonas aeruginosa* may invade the junction of the cartilaginous and osseous portions of the external auditory canal and spread through the adjacent bony structures, with consequent hearing loss and vertigo. Prolonged treatment with effective antibiotics, carbenicillin or gentamicin, has improved the previously poor prognosis.

Viral infections may reach the inner ear via the blood stream, the meninges, the eighth nerve or the middle ear. Four viral infections are known to affect the labyrinth by the blood-borne route - cytomegalovirus, mumps, measles and rubella. The virus of herpes zoster oticus enters the inner ear along the seventh and eighth cranial nerves (Blackley, Friedman and
Wright, 1967). Temporal bone studies in patients who have developed sudden, sensorineural hearing loss during the course of a head cold, pharyngitis or other symptoms suggestive of a viral infection, have identified changes similar to those found in the labyrinths of patients with hearing loss related to measles, mumps and rubella. It has been postulated, therefore, that these inner ear lesions are of viral origin (Lindsay, 1973; Sando et al, 1977). In measles and rubella, changes have also been observed in the utricle and saccule of the vestibular system, and it has been extrapolated, therefore, that acute episodes of vertigo may also be attributable to viral infection. However, viruses may only be definitively implicated in the aetiology of vertigo or deafness if a virus is isolated, either at surgery or autopsy, from the perilymph or endolymph of the inner ear, or viral particles, virus antigen or virus-infected cells are identified within the inner ear. Only one case of hearing loss in man with direct evidence of viral infection of the inner ear has been reported (Westmore, Pickard and Stern, 1979). Clinical studies which employ serological titre rises or nasopharyngeal virus isolates should be interpreted with caution.

Neurosyphilis may cause vertigo and all stages of the disease are to be found in the vestibular labyrinth (Eggston and Wolff, 1947). It is well recognized that both congenital and late acquired syphilitic labyrinthitis may relentlessly progress to result in profound deafness, but vestibular disturbances may be a presenting feature and almost always form an important part of the natural history of the disorder. The diffuse periostitic form of syphilis commonly involves the semicircular canals and the lumina may be completely obliterated. Vestibular manifestations may present as episodes indistinguishable from Ménière's disease, or there may be a progressive, bilateral destruction of vestibular end organs, resulting in imbalance and unsteadiness of gait, particularly in the dark. Morrison (1979) emphasized that the 'diagnosis of late acquired syphilitic ear disease is probably missed quite often, especially in the elderly male'. The recent resurgence of venereal disease highlights syphilis as a continuing problem, despite the widespread use of antibiotics. A high index of suspicion is still necessary if this disorder is not to be missed (Luxon, Lees and Greenwood, 1979). Specific serological tests for syphilis (Treponema pallidum haemagglutination test and fluorescent treponema antibody test) are required to establish the diagnosis, and treatment may require hospitalization to ensure adequate penicillin and steroid therapy (Pirozzi, 1973).

Neoplasia

Malignant tumours of the temporal bone are rare (Lodge, Jones and Smith, 1955), but secondary tumour involvement must always be considered. Direct malignant involvement of the labyrinth may be seen in nasopharyngeal carcinoma, neoplasia of the external ear, the parotid gland or the temporomandibular joint. Distant metastases are most commonly blood borne from kidney, lung, prostate, breast or uterus.

Vascular disease

The vertebrobasilar system supplies both the central vestibular connections and the peripheral labyrinth. The labyrinthine, or internal auditory artery, arises from the anterior inferior cerebellar artery or, more rarely, directly from the basilar artery (Sunderland, 1945). It is an end artery and therefore vascular disease, secondary to atheroma, or more rarely meningovascular syphilis, polyarteritis nodosa or giant cell arteritis, may precipitate profound ischaemia. Animal experiments and pathological examination of the cochlea in patients with
hearing loss have demonstrated end organ necrosis associated with secondary neural
degeneration (Gussen, 1976; Belal, 1980) and, indeed, Kimura and Perlman (1958) have
documented primary vestibular epithelial changes following arterial destruction of the
labyrinth. Lindsay and Hemenway (1956) reported a small group of patients with sudden
severe vertigo, associated with nausea and vomiting, which gradually subsided over a number
of weeks, but was replaced by the development of positional vertigo. A temporal bone
examination of one of these patients revealed degeneration of Scarpa's ganglion, together with
atrophy of the superior vestibular nerve. A mass of convoluted vessels in the external auditory
canal led the authors to hypothesize that these changes were of vascular origin. The majority
of patients in whom a vascular cause of vertigo is suspected are over the age of 60 and have
a clear history of cardiovascular disease, hypertension or vertebrobasilar insufficiency. It must,
of course, be emphasized that in the face of vertebrobasilar ischaemia (see below), it is often
difficult clinically to determine the precise pathological site of the ischaemic lesion (labyrinth,
eighth nerve or brainstem) giving rise to vertiginous symptoms.

Migraine is more fully considered below, but ischaemia of the peripheral labyrinth
secondary to migraine deserves special mention. A recent study by Kayan and Hood (1984)
identified vestibular and/or cochlear symptoms of disabling severity, requiring medical
intervention in 5% of 200 unselected patients with migraine. Full investigation of 80
migrainous subjects, referred because of vestibulocochlear symptoms, revealed that 77.5% had
objective abnormalities - one-half indicating peripheral, and the other half indicating central
pathology. Migraine should therefore be considered in the differential diagnosis of peripheral
vestibular disorders. It is of note that, in migraine, it would appear that the vertiginous
episodes may precede or occur simultaneously with the headache, but may also occur during
the headache-free periods. A family history is of particular important in the diagnosis of
migraine.

Drug-induced ototoxicity

Iatrogenic dizziness is discussed above and, in the present section, drugs causing
damage to the vestibular labyrinth are considered.

The use of streptomycin in the treatment of tuberculosis was first reported by Hinshaw
and Feldman in 1945, and three of their 34 patients developed vestibular disturbances.
Although ototoxicity is now a well recognized side-effect of the aminoglycosides (Lerner and
Matz, 1980), it remains impossible to predict which patients will be affected. There is clear
evidence that age, renal function, total dosage of drugs, duration of drug treatment, and the
particular aminoglycoside used, are all important factors in assessing the likelihood of
developing ototoxicity (Reeves, 1978). Recent work has demonstrated that the newer
aminoglycoside antibiotics are less ototoxic, while equally therapeutic in life-threatening
infections (Kahlmeter and Dahlager, 1984).

Aminoglycoside damage the hair cells of the inner ear, some affecting predominantly
the auditory system, while others have a predilection for the vestibular labyrinth. Berg (1949)
and Caussé (1949) documented the degenerative changes in the peripheral sensory epithelia,
and Lindeman (1969) documented clear regional differences in vulnerability to vestibular
damage. The sensory epithelia of the cristae ampullares of the semicircular canals are more
vulnerable than the macula utriculæ, which is more susceptible than the macula sacculæ.
The specific ototoxic effect of aminoglycoside antibiotics appears to be best explained by the high concentration of drug which accumulates in the inner ear fluid, and the prolonged half-life of the aminoglycoside antibiotics in perilymph compared with other body fluids (Federspiel, 1976). It has been postulated that the highly negatively charged aminoglycoside becomes bonded with the strongly positively charged mucopolysaccharides in the inner ear.

As mentioned previously, vertigo is consequent upon an imbalance in vestibular activity arising from the two vestibular end organs. Although ototoxicity may occur asymmetrically, more commonly both labyrinths are damaged simultaneously. In addition, the patient is usually gravely ill, in bed, and there is, therefore, very little vestibular stimulation. A very common history in this situation is that upon remobilization, the patient begins to notice a balance problem, and often complains of movement of the environment in association with head movements. This is known as ‘bobbing oscillopsia’ and is the result of a failure of vestibular activity to generate a normal compensatory eye movement in response to head movement. The tragedy of this situation is that a patient who was gravely ill makes an excellent medical recovery, only to find himself a vestibular invalid. The diagnosis may be confirmed by caloric or rotation testing, which demonstrates profound or total, vestibular failure bilaterally. The vestibular damage is permanent and, with the passage of time, the patient may adapt by using visual and proprioceptive information to maintain his balance. However, in elderly subjects, the symptoms are particularly intractable and the patient may be crippled by vestibular failure. Thick rubber-soled shoes may reduce the vertical oscillopsia on walking. A walking stick may provide additional proprioceptive information to aid balance, and a course of Cawthorne-Cooksey exercises may be of value, in order to help the patient to use maximally visual and proprioceptive clues. From the foregoing comments, it becomes clear that prevention is vital and infinitely preferable to rehabilitation.

Age-corrected normograms exist for calculating the correct dose of drugs such as aminoglycosides in the presence of renal impairment, but it cannot be overemphasized that this merely provides initial guidance in choosing a correct dose. Regular measurements of peak and trough serum levels of aminoglycosides are essential if a reasonable attempt to avoid ototoxicity is to be made, particularly in the elderly, gravely ill or renally impaired patient (Barza and Lauermann, 1978).

It is well documented that loop diuretics, particularly frusemide and ethacrynic acid, may produce hearing impairment, but whether or not they produce disturbances of balance has not been firmly identified. Ballantyne and Ajodhia (1984) have reported damage to the sensory vestibular cells in a patient who received very large doses of frusemide and ethacrynic acid.

Ménière’s disease

In 1861, Prosper Ménière drew attention to the inner ear as the source of a complex of auditory and vestibular symptoms and signs which have subsequently become known as Ménière’s disease. This is a common condition that appears to be determined by hereditary and environmental, rather than racial factors (Kitahara, Futaki and Nakano, 1971), but the aetiology remains obscure, as does a cure. The literature abounds with controversy on all aspects of the condition, although most clinicians agree that the triad of symptoms of hearing loss, tinnitus and vertigo is a prerequisite of the diagnosis.
Ménière's disease affects all age groups from childhood to old age, but is most commonly seen in the third and fourth decades of life. The commonest presenting symptoms are tinnitus and hearing loss, which characteristically fluctuates, but diplacusis, an intolerance of loud sounds and a fullness or pressure sensation in the ear, are also common early complaints. In 60% of those affected, vestibular symptoms develop within the first 6 months (Morrison, 1984). Vertigo may occur in the absence of cochlear involvement, although this is rare (Harker and McCabe, 1980). It must be emphasized that the vestibular symptoms vary from mild light-headedness to acute rotatory vertigo, with prostration, nausea, vomiting and occasionally diarrhoea. The acute vestibular attacks, in whatever form, rarely last more than 24 hours. The natural history of the disorder is characteristically that of relapses and remissions, with clusters of episodes lasting weeks or months. With the passage of years, the vertiginous attacks abate in severity and frequency, while the early fluctuant hearing loss becomes established and, although it may continue to fluctuate, normality is never regained. The frequency of bilateral disease has been reported to be as high as 60% (Jongkees, 1971), and as low as 10% (Cawthorne, 1969), Morrison (1975) and Stahle (1976) have documented that the presence of bilateral involvement increases with the passage of time.

Ménière's disease is primarily a clinical diagnosis, but certain investigative findings facilitate the diagnosis. Tuning fork tests reveal a positive Rinne test bilaterally and, in unilateral cases, the Weber is referred to the better ear. Diplacusis can commonly be elicited.

Classically, pure-tone audiometry reveals a unilateral, low-tone hearing loss which fluctuates from attendance to attendance. Later in the disease, a plateau hearing loss develops which progressively deteriorates. Tests of recruitment (uncomfortable loudness levels, alternate loudness balance, stapedial reflex threshold measurements) are characteristically positive. Morrison, Moffat and O'Connor (1980) have advocated the diagnostic and short-term prognostic value of the glycerol dehydration test. Transtympanic electrocochleography may also be a useful diagnostic and prognostic indicator. The technique was developed by Portmann, Le Bert and Aran (1967) in Bordeaux and depends upon electronic averaging and separation of the three main electrical potentials generated in the inner ear in response to acoustic stimuli (see Chapter 8). One of these potentials, the negative summating potential, is significantly enhanced in the majority of ears affected by Ménière's disease (Morrison, Moffat and O'Connor, 1980).

Vestibular function is frequently abnormal and, in particular, the caloric test has been reported to be abnormal in 94% of cases (Dix and Hallpike, 1952). The dynamic nature of Ménière's disease makes changes in vestibular function and test results commonplace (Stahle, 1968). The 'recovery nystagmus' documented by McClure and Lycett (1978) is worthy of mention. Following a severe attack of Ménière's disease, a secondary nystagmus develops towards the side of the lesion. A knowledge of this phenomenon is of particular importance in attempts to identify the side of the lesion and, as mentioned previously, the novice in the field may interpret this nystagmus incorrectly.

The underlying pathological state giving rise to Ménière's disease is now well established - endolymphatic hydrops (Dix and Hallpike, 1952) - but the underlying pathophysiological mechanisms remain poorly understood (Dohlman, 1983).
In the literature, there are diverse reports of disorders such as diabetes mellitus, hyperlipidaemia, myxoedema and allergy giving rise to secondary endolymphatic hydrops. A recent review by Moffat, Booth and Morrison (1981) found no significant association between Ménière's disease and these metabolic disorders compared with a normal control population. A clear association between Ménière's disease and migraine (Hinchcliffe, 1967a; Fedorova, 1970; Kayan and Hood, 1984) has been established and Morrison (1984) has reported that 'up to one third of Ménière's disease sufferers have migraine, usually prior to the onset of neuro-otological symptoms, which frequently replace the migraine'. Syphilitic labyrinthitis with associated endolymphatic hydrops may mimic Ménière's disease (see above). Vertiginous symptoms, associated with otosclerosis must be differentiated (see below). In young adult life, vestibular symptoms associated with developmental dysplasia (see above) should be differentiated.

The condition of delayed endolymphatic hydrops (Nadol, Weiss and Parker, 1975; Schuknecht, 1978) should be mentioned. In this disorder, a Ménière-like syndrome develops in a patient in whom there has been total, or subtotal, cochlear damage many years previously. such cases are relatively common and are identified by the presence of a previous history of long-standing unilateral hearing loss. The rare, but well documented occurrence of paroxysmal vertigo secondary to an acoustic neuroma may mimic Ménière's disease. Characteristically audiometric assessment may reveal a retrocochlear type of hearing loss with no loudness recruitment, abnormal auditory adaptation and poor speech discrimination. The development of auditory brainstem evoked potentials has allowed the two conditions to be readily differentiated (Selters and Brackmann, 1977).

The relationship between Ménière's disease and emotional factors remains obscure. Psychological features of anxiety and depression are particularly prominent in Ménière's disease (Cawthorne, 1957; Morrison, 1981) and the role of these factors in the aetiology of attacks has been highlighted by a number of authors (Hinchcliffe, 1967b; Harrison and Naftalin, 1968; House, Crary and Wexler, 1980).

The therapeutic options in Ménière's disease are legion (Arenburg and Bayer, 1977), but it must be recalled that in Ménière's disease a 60-80% success rate has been reported, regardless of the treatment modality (Torak, 19770. The fluctuant nature of the disease and the high rate of placebo effect makes assessment of any treatment regimen extremely difficult. By necessity, treatment is symptomatic and encouraging results have been reported using a low sodium diet, vasodilators, antihistamines, including betahistine, and many surgical procedures. Hydrochlorothiazide (Klockhoff and Lindblom, 1967) and chlorthalidone (Klockhoff, Lindblom and Stahle, 1974) have been convincingly demonstrated in double-blind, cross-over trials to improve hearing loss, vertigo and general condition. There is no similar convincing data for other treatments such as prochlorperazine (Stemetil), betahistine (Serc) and cinnarizine (Stugeron). Various surgical drainage operations have been advocated and would again appear to be helpful in individual cases, but lack definitive confirmation in trial (Bretlau et al, 1983). In the face of severe disabling attacks, labyrinthine ablation or vestibular nerve section may be considered (Glasscock et al, 1980). As mentioned previously, it is imperative that sophisticated investigations confirm the abnormal side and take into account the possibility of bilateral disease, before any destructive vestibular procedure is undertaken.
Diseases of the temporal bone

Otosclerosis is a common, hereditary condition, in which there is localized disease of the bone derived from the otic capsule. Donaldson (1976) has reported vertigo in 4% of a series of cases and Paparella and Chasin (1966) have reported a variety of vestibular symptoms in otosclerosis. Morrison (1984) has documented that 4% of patients with otosclerosis demonstrated benign positional vertigo of paroxysmal type and 1% of patients gave a history of paroxysmal attacks of vertigo, similar to Ménière's disease. The pathological mechanism producing these symptoms is unclear, but may be consequent upon involvement of the vestibular labyrinth, by disease of the otic capsule, or by encroachment upon the eighth cranial nerve, within the internal auditory canal. The presence of a conductive hearing loss in a patient with dysequilibrium should raise the possibility of this diagnosis.

Paget's disease may also affect the otic capsule and give rise to hearing loss and vertigo (Davies, 1968). In the presence of characteristic skeletal changes (enlarged skull, short stature, kyphosis, bowing of the legs), the diagnosis is obvious, but osteitis deformans may be symptomless apart from the otological features of a mixed or conductive hearing loss. Skull radiographs together with an elevated serum alkaline phosphatase and increased total urinary hydroxyproline confirm the diagnosis.

Metabolic disorders

There are many reports in the literature of auditory and vestibular disorders in association with diabetes mellitus (Jorgensen and Buch, 1961). The early studies made little attempt accurately to establish the precise site of auditory or vestibular derangement, but the most common histological abnormality demonstrated has been a PAS-positive thickening of the capillary walls, most prominent in the stria vascularis of the cochlea (Makishima and Tanaka, 1971). In a recent unselected study of 200 cases of adult-onset diabetes mellitus, Harner (1981) concluded that hearing loss and vestibular disturbances did not occur with increased frequency in this group of patients. Reviewing hearing loss in diabetic subjects, Axelsson, Sigroth and Vertes (1978) came to the same conclusion.

The development of inner ear symptoms in patients with renal disease was emphasized by Beaney (1964), when he identified six patients with auditory and/or vestibular symptoms in a series of 262 chronically haemodialysed patients. These auditory and vestibular symptoms are undoubtedly multifactorial in aetiology and may, in part, be explained by coincidental ear disease, infection, inflammatory disorders, immunosuppression, electrolyte disturbances, ototoxic drugs and genetic factors. A number of histological abnormalities have been documented, including endolymphatic hydrops, degeneration of the stria vascularis and fibrosis and calcification of the membranous labyrinth. Temporal bone studies in eight patients with chronic uraemia (Oda, 1974) identified abnormal connections in the stria vascularis of the cochlea and in the subepithelial connective tissue of the maculae and cristae. The precise histopathological relationship between various forms of renal disease and inner ear pathology remains to be identified. It is of interest that reversal of the auditory impairment associated with renal failure has been reported following successful renal transplantation (Mitschke et al, 1977).
Autoimmune disorders

Autoimmunity is defined as the state in which immune reactions of the host are directed against his own antigenic structures. Such immune reactions are mediated by sensitized lymphocytes and/or antibodies and can lead to destruction of the antigens involved and consequently to disease. Depending on the distribution of the autoantigens, the autoimmune disease may be either organ specific, such as thyroiditis, or unrelated to a single organ, like systemic lupus erythematosus.

The aetiology of sudden deafness and sudden vestibular dysfunction remains unidentified in many patients. Viral and vascular pathologies are frequently suggested, but not proven. In the last decade, considerable attention has focused on the immunological aspects of many disorders, including cochleovestibular pathology. As early as 1958, Lehnhardt reported on autoantibodies in patients suffering from sudden deafness, but in 1979 McCabe hypothesized a new disease entity called 'autoimmune sensorineural hearing loss'. He reported 18 patients with cochleovestibular symptoms demonstrating immunological findings and suggesting an autoimmune phenomenon. All patients responded to long-term cortisone and cyclophosphamide therapy. By 1985, McCabe had added a further 38 patients to his series and reported that 'vestibular spells or crises were not a feature of the disease, but ataxia in the dark was a feature of all patients who had moderate to severe disease'. In addition, he noted that vestibular symptoms and caloric test results generally paralleled the hearing loss. Furthermore, in patients with severe to profound hearing loss, all ocular counter-rolling is lost and he therefore suggested that the autoimmune process involves not only the cochlea, but the entire membranous labyrinth including the otolith organs.

In a study of patients with chronic progressive sensorineural hearing loss of unknown aetiology, Kanzaki and Ouchi (1981) established the presence of immune complexes in the serum of some of their patients. Yoo et al (1982a) identified cochlear and vestibular deficits in rodents after injection of collagen type II antibodies. The same author, with other coworkers (1982b), found an increased collagen antibody titre in patients with Ménière's disease and otosclerosis. It should be emphasized that autoimmunity of the inner ear would now be recognized as a cause of endolymphatic hydrops (Hughes et al, 1985) and in 1982, Shea and Yoo reported a good response to cortisone treatment in patients with Ménière's disease and immunopositive findings.

In 1982, Stephens, Luxon and Hinchcliffe found a high incidence of cochleovestibular symptoms in patients suffering from a variety of autoimmune disorders including autoantibody-mediated disease such as the Vogt-Koyanagi-Harada syndrome and immune-complex-mediated disease, such as systemic lupus erythematosus, polyarteritis nodosa, Wegener's granulomatosis, Cogan's syndrome and Behçet's syndrome.

In the Vogt-Koyanagi-Harada syndrome (bilateral inflammation of the eye with granulomatous uveitis and systemic pigmentary disturbances), meningitic symptoms and sensorineural hearing loss, vertigo and vestibular hypofunction are common findings (Maxwell, 1963; Seals and Rise, 1967), McCrae and O'Reilly (1957) reported vertigo in four out of a series of 55 patients with systemic lupus erythematosus. The site of vestibular dysfunction was not identified. A recent temporal bone study in a patient with polyarteritis nodosa (Gussen, 1977) revealed hydropic changes in the cochlea, suggesting the likelihood
of vestibular involvement. Similar changes of hydrops together with ossification in the scala vestibuli have been reported upon histological examination of the temporal bones in patients with Cogan's syndrome (Fischer and Hellstrom, 1965; Wolff et al, 1965; Zechner, 1980). In Behçet's syndrome there are a number of reports of vestibular involvement (Hughes and Lehner, 1979; Brama and Fainaru, 1980). In the latter study, on the basis of audimetric and vestibular examinations, the site of involvement was identified as the labyrinth.

The entity of cochleovestibular dysfunction on an autoimmune basis appears to be well established, although it should be emphasized that different authors have adopted various criteria for confirming the diagnosis of autoimmun ear disease. In certain instances, a positive 'antigen-specific' test result has been required, that is positive lymphocyte transformation and/or migration inhibition testing, using crude inner ear membrane antigens, while other workers have based the diagnosis of autoimmune hearing loss and/or vertigo on cerebrospinal total protein content and immunoglobulin concentrations, together with a screen for tissue autoantibodies. Tissue autoantibodies were determined by the indirect immunofluorescent technique and sera were incubated with various heterologous antigens such as heart, liver, kidney and smooth muscle, but also with preparations of laboratory animal sliced labyrinth and basilar membrane.

*Eighth cranial nerve pathology*

In the last decade, the introduction of electrophysiological investigations, including electrocochleography and brainstem evoked potentials, has facilitated the clinical feasibility of diagnosing eighth nerve lesions with confidence. The clinical problem of differentiation of cochlear from eighth nerve pathology is compounded by the unusual pathophysiological degenerative changes exhibited by the cochleovestibular nerve. In the auditory division, sensory and neural degeneration are interdependent (Johnsson, 1974), whereas the vestibular nerve behaves rather differently. Transection of the vestibular nerve in the internal auditory meatus, or destruction of the peripheral vestibular nerve fibres or end organ, does not result in neural degeneration (Gacek, 1960; Igarashi, Watanabe and Maxian, 1970). Like the cochlear end organ, the vestibular end organ is independent of normal primary neurons. Sections of the vestibular nerve removed from a large number of patients with neurootological disorders revealed four cases of severe degeneration of the nerve fibres and ganglion cells, with severe fibrosis (Ylikoski, House and Hernandez, 1981). The authors interpreted these findings as indicative of a primary neuropathy. It is therefore of value to consider the pathologies which have been identified as affecting the eighth cranial nerve.

*Congenital and hereditary disorders*

In *Michel aplasia*, a complete absence of the eighth cranial nerve accompanies the absence of the otic capsule. This condition has been reported secondary to thalidomide ingestion, raising the suspicion that aplasias may be environmentally, rather than genetically, induced (Jorgensen, Kristensen and Buch, 1964). In the aplasia of Scheib, there is membranous cochleosaccular dysplasia, with atrophy of the cochlear and vestibular nerve.

Atrophy of the eighth nerve has been documented in a number of syndromes where deafness has been associated with a skeletal abnormality and undoubtedly vestibular involvement may occur:
anencephaly (Lindsay, 1973)
Treacher Collins syndrome (Lindsay, 1973)
preauricular (Fitch, Lindsay and Srolovitz, 1976)

The occurrence of hearing loss and vestibular abnormalities in the spinocerebellar degenerations is well recognized, but only recently have studies of the temporal bones indicated eighth cranial nerve involvement. Spoendlin (1974) studied the temporal bones of two sisters with Friedrich's ataxia and concluded that 'the most striking features are extensive and selective damage to the neurones of the VIIIth nerve, whereas the peripheral receptor organs, the organ of Corti and vestibular sensory epithelia remain unaffected'. A subsequent clinical study of auditory function in four patients with Friedreich's ataxia, without overt hearing dysfunction, revealed speech audiometry and brainstem evoked potential abnormalities, supporting eighth nerve pathology (Satya-Murti, Cacace and Hanson, 1980).

Clinically and pathologically, there is considerable overlap of the varying syndromes of inherited ataxia. In 1951, Denny-Brown reported 'thin auditory nerves' at a postmortem examination of a patient with an hereditary sensory neuropathy with deafness. Hallpike (1967b) reported a similar case in which the caloric responses were also absent, and depletion of the fibres of the vestibular nerve was documented. It would seem, therefore, that in a proportion of patients with vestibular abnormalities in spinocerebellar degeneration, there is evidence of eighth nerve involvement.

The Arnold-Chiari malformation is a congenital deformity which may become manifest in the first few months of life and is associated with hydrocephalus and other central nervous system malformations. The brainstem and cerebellum are elongated downwards into the cervical canal, and auditory and/or vestibular symptoms have been identified in 20% of patients with this disorder (Rydell and Pulec, 1971). It is postulated that these symptoms may be the result of stretching of the eighth cranial nerve, with changes in brainstem position with age, or may be the result of compression of the eighth nerve as it is bent over the edge of the porus acusticus. There are no pathological data to conform or refute these hypotheses.

Trauma

Transverse fractures of the temporal bone may involve the roof of the internal auditory meatus with total deafness and acute vestibular failure, as outlined above. The prognosis of vestibular symptomatology depends not only upon the damage to the vestibular nerve, but also upon any other central nervous system dysfunction. There is no indication for surgical decompression of the eighth nerve in terms of any cochleovestibular deficit, but decompression of the eighth nerve for plugging of a persistent cerebrospinal fluid leak may be required.

Infection

Bacterial infection

Bacterial meningitis may involve the eighth cranial nerve and, by extension into the inner ear, may result in total destruction of the sensory and neural components (Lindsay,
Profound bilateral meningitic hearing loss is usually encountered in small children and neonates. The most common infecting organisms are *Streptococcus pneumoniae*, the meningococcus and *Haemophilus influenzae* (Baloh and Honrubia, 1979). The prognosis is poor; any auditory and/or vestibular impairment is permanent. If total vestibular destruction has occurred, there may be oscillopsia and severe imbalance. If residual activity remains, in a young person good compensation occurs. In many cases of eighth nerve involvement, secondary to meningitis, there are other sequelae of involvement of the central nervous system, especially in cases of tuberculous meningitis.

*Syphilitic labyrinthitis* has been discussed above, and eighth nerve involvement secondary to syphilitic meningitis occurs in only about 0.2% of patients with syphilis (Jerger and Jerger, 1981).

**Viral infection**

As mentioned previously, viral infections are often involved in the aetiology of vertigo and sensorineural hearing loss, but the objective scientific evidence for this is slim. However, Lindsay (1973) demonstrated marked infiltration of the eighth cranial nerve neural remnants in the modiolus of the cochlea and branches of the vestibular nerve by lymphocytes and histiocytes. These findings were attributed to viral infection of the eighth nerve.

The *Ramsay Hunt syndrome*, or herpes zoster oticus, would appear to be an example of a mononeuritis of the eighth cranial nerve. The patient experiences a deep burning pain in the ear, which is followed within a few days by a vesicular eruption in the external auditory canal and on the concha. The patient develops hearing loss and vertigo, with or without seventh nerve involvement. Blackley, Friedman and Wright (1967) have documented marked perivascular, perineural and intraneural round cell infiltration in both divisions of the eighth and the seventh cranial nerves, in this condition.

A brief consideration of *Bell's palsy* is of interest at this point. A limited number of patients with Bell's palsy experience vestibular symptoms and vestibular abnormalities have been documented in this condition (Rauchbach and Stroud, 1975). Furthermore, a small percentage of patients with idiopathic facial palsy show an elevation in complement fixation antibodies to herpes zoster antigen (Petersen and Anderson, 1967). Hence, although it may be hypothesized that Bell's palsy is a mononeuropathy of unknown cause limited to the facial nerve, the overlap between seventh and eighth nerve findings in the Ramsay Hunt syndrome and Bell's palsy has led to two alternative aetiological hypotheses. First, it has been suggested that the swollen seventh nerve in Bell's palsy may compress the eighth nerve with which it is intimately related, or vice-versa, in the Ramsay Hunt syndrome. Alternatively, the same disease process may affect both the seventh and eighth cranial nerves. Adour and colleagues (Adour, 1976; Adour, Hilsinger and Byl, 1980) have postulated that Bell's palsy is one expression of a cranial polynueuritis associated with the herpes simplex virus. In this respect, it is known that rheovirus, herpes simplex virus and rubeola virus may infect the ganglion cells of the vestibular ganglia.

Other studies in Bell's palsy have identified abnormalities of auditory brainstem evoked potentials in some 25% of patients with this condition (Uri, Schuchman and Pratt, 1984). This abnormalities documented indicate prolongation of brainstem conduction time and
were not specifically related to the side of the Bell's palsy. These findings led the authors to suggest that Bell's palsy may be a clinical sign of a diffuse central nervous system disorder. It is, however, valuable to recall that a lower motor neuron seventh nerve palsy, merely reflects pathology affecting the seventh nerve nucleus in the brainstem, or the efferent pathway thereof. Many pathologies may produce such involvement and it is therefore not surprising that, in any series of Bell's palsy, certain patients will be found with vestibular, auditory or indeed central nervous system signs.

No discussion of the viral pathology of the eighth nerve is complete without some consideration of the controversial subject of vestibular neuronitis. The precise diagnosis and underlying pathology of vestibular neuronitis remains as elusive today as when the vestibular syndrome was initially outlined by Ruttin in 1909. Diagnostic criteria were described in the classical paper of Dix and Hallpike (1952). The clinical syndrome is characterized by vestibular symptoms and signs unassociated with any cochlear or neurological deficit, and frequently preceded by an upper respiratory tract infection. These authors postulated that focal sepsis produced toxic damage to the eighth cranial nerve.

Recent work has cast doubt on the homogeneity of the original 100 patients studied by Dix and Hallpike; although there would appear to be a group of patients who present with sudden vertigo usually involving one ear, which lasts between 1 and 3 weeks and which follows a benign course with recovery occurring over a number of months. Certain cases may suffer recurrent attacks. In the acute phase, there may be marked spontaneous nystagmus, beating away from the affected ear, with no hearing loss and no neurological signs. Upon brief consideration, it is apparent that these findings merely represent those of a peripheral vestibular upset. Reviewing the literature, some interesting clinical aspects may be established.

First, the clinical course of the disorder is far from clear; the vertiginous symptoms may vary from sudden dysequilibrium to vague unsteadiness (Dix and Hallpike, 1952). Certain authors feel that the label 'vestibular neuronitis' should be confined to a single attack of vertigo (Coates, 1969), while others have expressed the view that there are two types of the disorder - a single attack form and a multiple attack form (Schuknecht and Kitamura, 1981). It is unclear whether the disease affects one ear, or both ears, and different authors disagree on the association with upper respiratory tract infections (Pederson, 1959; Lumio and Aho, 1965; Coates, 1969; Wilmot, 1973). There is no specific sign or investigation which confirms the diagnosis, although Sekitani (1983) placed considerable importance upon the galvanic test.

Follow-up of patients with this disorder is also relevant. Wallace and Barber (1983) found that at a 5-year follow-up, 23.5% of their patients with this disorder required correction of the diagnosis, most commonly to Ménière's disease or benign positional vertigo. Furthermore, Wennmo and Pyykkö (1982), who carried out a full neurological, neuro-ophthalmological and neuro-otological assessment in 30 patients with vestibular neuronitis, identified that 20% of cases had evidence of intrinsic nervous system disease and a further 20% exhibited central visuovestibular abnormalities on electronystagmography. A number of other studies, which have carefully assessed the neurological status of patients with vestibular neuronitis have identified transient EEG abnormalities (Anttinen et al, 1983), abnormal brainstem evoked potentials (Silvoniemi and Aantaa, 1983), and abnormal stapedial reflex
thresholds (Bergenius and Borg, 1983). All of these findings add weight to the heterogeneity of the group of patients labelled as suffering from vestibular neuronitis.

Regrettably, there are few histopathological studies in this condition (Morgenstein and Seung, 1971; Schuknecht and Kitamura, 1981) as it is essentially a benign condition and post mortem examination may not be carried out until some considerable time after the onset of symptoms. Furthermore, it is clear that certain findings may reflect the end result of previous indeterminate pathology. The patients studied by Schuknecht and Kitamura (1981) were all elderly and suffered from a number of other disorders, which may have given rise to coincidental vestibular pathology; hence, definitive conclusions cannot be drawn. Nonetheless, these authors felt that the pathological findings reflected an isolated neuropathy and, therefore, preferred the term ‘vestibular neuritis’ to that of ‘vestibular neuronitis’ in more common usage.

Despite the lack of clear evidence of such a disorder, certain authors still believe in an entity of isolated viral mononeuritis of the vestibular nerve (Ylikoski, House and Hernandez, 1981), while others prefer the concept that vestibular vertigo may be a focal manifestation of a more widespread cranial polyneuritis (Adour, Hilsinger and Byl, 1980). It is the view of the present author that vestibular neuronitis represents a mixed bag of vestibular conditions of various pathologies, and is an unhelpful term, in that it implies some understanding of the vestibular system and its pathology, which unfortunately is incorrect in our present state of knowledge.

Mycotic infection

Cryptococcosis and coccidioidomycosis may produce a basilar meningitis, with involvement of multiple cranial nerves including the eighth nerve. The clinical picture resembles tuberculosis or sarcoid meningitis with an insidious febrile illness. Cerebrospinal fluid findings reveal a lymphocytic pleocytosis, an elevated protein level and a decreased glucose concentration, but the diagnosis may be confirmed by the identification of cryptococcal or coccidioidal antigen in the cerebrospinal fluid, using complement fixation, latex agglutination or immunofluorescent techniques.

Vascular disease

The vertebrobasilar circulation supplies the eighth cranial nerve, as well as the peripheral labyrinth and the brainstem. Ischaemia of the nerve may occur, but clinical identification may prove impossible. Aneurysms of the anterior inferior cerebellar artery may compress the eighth cranial nerve and give rise to vestibular and auditory symptoms clinically indistinguishable from any other cerebellopontine angle lesions (Porter and Eyster, 1973; Mori, Miyazaki and Ono, 1978). The correct diagnosis is made at angiography or air meatography (Phelps and Lloyd, 1982).

Neoplasia

As in all sites, neoplasia may be divided into malignant and benign tumours. Malignant disorders, involving the eighth cranial nerve, are rarely of temporal bone origin and are more usually the result of metastases (see above).
Acoustic neuromata (acoustic neurinoma or, more correctly, vestibular schwannoma) are fully discussed in Volume 3, Chapter 21. The most common mode of presentation is that of hearing loss or tinnitus (Erickson, Sorenson and McGavran, 1965; Pulec et al, 1971; Morrison, 1975). Nonetheless, a discussion of vestibular symptoms arising from the eighth cranial nerve would be incomplete without some consideration of this relatively rare, but frequently considered tumour.

The majority of acoustic neuromata arise from the Schwann cells (Luse, 1960; Ramondi, Mullan and Erans, 1962) of the superior division of the vestibular nerve in the internal auditory meatus. Occasionally, they may arise from the inferior division, or indeed even the cochlear nerve (Schuknecht, 1974).

Although acoustic neuromata must be excluded in a patient with dizziness, it should be emphasized that the frequency of occurrence is very low (0/125) in a random group of dizzy patients (Drachman and Hart, 1972). Frequently, vestibular symptoms are absent, as the slowly progressive nature of the disorder allows compensation to occur. Nonetheless, in a large series of acoustic neuromata, Morrison (1975) reported that 10% of patients complained of vertigo, dizziness or unsteadiness as an initial symptom. More specifically, Morrison (1984) reported that 5% of patients with eighth nerve tumours suffer paroxysmal attacks of vertigo, which may mimic Ménière's disease, while 16% suffer momentary vertigo or dizziness on sudden head movements and 8% may suffer acute vestibular failure, with or without hearing loss during the course of the disorder.

With increasing size, the tumour extends out of the internal auditory canal and into the cerebellopontine angle. The fifth and seventh cranial nerves become involved and this is followed by encroachment upon the cerebellum and displacement of the brainstem. Clinical examination reveals an ipsilateral trigeminal impairment, together with a facial palsy and cerebellar ataxia, and contralateral pyramidal spasticity and weakness. If the patient is left untreated, the prognosis is generally poor, with death from brainstem compression and hydrocephalus. Recent work has refuted the alleged slow rate of expansion of these tumours and, in view of the increased risk of surgical mortality and morbidity associated with operations of medium and large sized tumours (Yasargil and Fox, 1974), early diagnosis is essential (Morrison, 1975; House, 1978).

Any patient with vestibular symptoms, with a unilateral sensorineural hearing loss or an asymmetrical sensorineural hearing loss, must be carefully investigated to exclude an eighth nerve neuroma. Characteristically, a sensorineural hearing loss with no evidence of recruitment, marked tone decay and a poorer speech discrimination than would be expected from the pure-tone audiogram, are associated with this disorder. In addition, there may be a marked 'roll-over' phenomenon on speech audiometry (Jerger and Jerger, 1971). However, it must be emphasized that reviewing 200 cases of acoustic neuromata, Johnson (1968) reported that only 42% showed a classic retrocochlear hearing loss and it is now well recognized, that in the early stages of an acoustic neuroma, the auditory findings may mimic an end organ disorder, such as Ménière's disease. The addition of auditory brainstem evoked potentials to the audiological test battery has greatly improved the certainty with which eighth nerve pathology may be identified (Selters and Brackmann, 1977).
Caloric testing almost invariably reveals an ipsilateral canal paresis (Dix, 1974), with or without a directional preponderance, depending on cerebellar or brainstem involvement. Electronystagmographically, nystagmus is directed away from the side of the lesion, but bidirectional nystagmus is most commonly observed (Dix and Hallpike, 1966). Nystagmus to the contralateral side results from involvement of the eighth nerve, while ipsilaterally directed nystagmus reflects involvement of the cerebellar connections in the brainstem. In the absence of optic fixation, vestibular nystagmus of both end organ and eighth nerve origin is enhanced. Recurrent positional vertigo is an early feature in 6% of acoustic neuromata (Morrison, 1984), but no characteristic features of nystagmus have been identified with eighth nerve lesions.

Conventional radiography (Towne's and Stenvers' views) and transorbital views may show erosion and/or funnelling of the internal auditory meatus, while anterior posterior linear tomography allows greater detail to be visualized. Nonetheless, it must be emphasized that normal plain or tomographic studies do not exclude the presence of a tumour (Pulec et al, 1971). Until recently, posterior fossa myelography was the definitive diagnostic investigation, but recent work has established the value of thin section computerized tomography, with small quantities of air introduced by lumbar puncture. This technique has been shown to differentiate vascular anomalies from eighth nerve pathology and to delineate clearly the seventh and eighth nerves (Phelps and Lloyd, 1982). Magnetic resonance scanning will also undoubtedly prove to be an efficient diagnostic aid.

The treatment of acoustic neuroma is fully discussed in Volume 3, Chapter 21, and suffice it to say that one of the roles of the otologist is to identify the patient with an acoustic neuroma from the many patients with sensorineural hearing loss of cochlear origin.

Although acoustic neuromata constitute some 70-80% of cerebellopontine angle mass lesions (Gonzalez-Revilla, 1948), neuromata may also arise on the fifth, seventh, ninth, tenth and eleventh cranial nerves. These latter tumours may compress the eighth cranial nerve and give rise to a very similar clinical picture to that associated with acoustic neuroma. Other pathologies deserving consideration are meningiomata, identifiable on angiography as vascular tumours with major feeding vessels arising from the external carotid circulation, and epidermoid cysts, which classically cause erosion of the petrous apex, visible on routine skull radiographs.

Rarely, non-metastatic carcinomatous neuropathy may affect the eighth cranial nerve. McGill (1976) described a case of carcinomatous encephalitis with auditory and vestibular manifestations, secondary to an oat-cell carcinoma of the lung. Both divisions of the vestibular nerve were affected, but the vestibular nuclei were normal.

Carcinomatous meningitis, secondary to systemic cancer, is a well documented cause of both hearing impairment and vestibular symptomatology. Eighth nerve symptoms have been reported in 10% of patients with this condition (Alberts and Terrence, 1978). The hallmark of carcinomatous meningitis is the simultaneous occurrence of symptoms and signs in more than one area of the neuraxis. Olson, Chernik and Posner (1974) emphasized that neurological signs are much more prominent than the symptoms. The clinical diagnosis is confirmed by the presence of malignant cells in the cerebrospinal fluid. In addition, Paparella
et al (1973) have reported that 11 of a series of 25 patients with various forms of leukaemia revealed infiltration of the eighth cranial nerve on temporal bone examination.

**Disorders of the temporal bone**

The occurrence of vestibular symptoms in otosclerosis has been discussed previously. Sando, Hemenway and Miller (1974) studied the temporal bones of two patients with otosclerosis with vertiginous symptoms. They documented otosclerotic foci, in apposition to the superior vestibular nerve, with distal neural degeneration. Paget's disease may cause vestibular dysfunction by encroachment upon the eighth cranial nerve in the internal auditory meatus and, rarely, in fibrous dysplasia, compression of the eighth nerve has also been documented (Nager, Kennedy and Kopstein, 1982).

**Toxic disorders**

Thalidomide has been documented to produce aplasia of the eighth cranial nerve (Jorgensen, Kristensen and Buch, 1964). Poisoning with both lead and mercury is known to produce auditory and vestibular symptoms, but the pathophysiological mechanisms of these symptoms are poorly understood (Mizukoshi et al, 1975). Animal studies have demonstrated segmental demyelination and axonal degeneration of the eighth cranial nerve (Gozdzik-Zolnierkiewicz and Moszynski, 1969). A recent report by Ylikoski, House and Hernandez (1981) documented extensive degeneration of the vestibular and cochlear divisions of the eighth cranial nerve, in a patient suffering from chronic alcoholism and a marked peripheral neuropathy. It is not possible to state whether this degeneration was a toxic effect of alcohol ingestion or secondary to a nutritional vitamin deficiency.

**Immunological disorders**

There are a number of reports in the literature of retrocochlear hearing loss consequent upon immunological disorders, but reports of vestibular symptoms are rare. Morrison (1975) reported the case of a 17-year-old girl who developed retrocochlear deafness with vertigo after a hypersensitivity reaction to ampicillin. A good recovery was made following treatment with corticosteroids and it was postulated that an autoimmune mechanism was responsible.

Auditory and vestibular abnormalities are well documented in many autoimmune disorders, as outlined above, but there is no evidence of vestibular nerve involvement. Five per cent of patients with sarcoidosis developed a granulomatous meningitis which directly infiltrated the cranial nerves. The eighth cranial nerve is the fourth most frequently affected (Jahrsdoerfer et al, 1981) and vestibular symptoms may result.

**Neurological causes of balance disorders**

A particular concern for the otologist is the differentiation of peripheral vestibular disorders from central nervous system disease. A clear understanding of central vestibular physiology (see Volume 1, Chapter 4) is necessary if the correct interpretation of vestibular signs and investigations is to allow this differentiation to be made. An examination of the ocular movements, as part of the neuro-otological examination, is invaluable in locating
vestibular pathology. Although didactic, the following generalizations may prove helpful in suggesting the presence of neurological disease:

1. impaired smooth pursuit, or abnormal saccadic movements
2. gaze paretic, bidirectional, vertical or disconjugate nystagmus
3. positional nystagmus with no latent period, no adaptation, no fatigue and little associated vertigo
4. deranged optokinetic nystagmus
5. failure of suppression of vestibulo-ocular responses by optic fixation.

It is valuable to note that bilateral derangement of pursuit and bidirectional nystagmus, in the absence of optic fixation, are frequently observed in patients taking psychotropic drugs such as tranquilizers, sedatives, anticonvulsants and anti-depressants. These non-specific abnormalities may also be observed if the patient is tired, systemically unwell or has been drinking alcohol, shortly before testing. It is therefore sensible to enquire about current medication, sleep pattern and alcohol intake before undertaking vestibular investigation. Psychotropic medication should be discontinued, if clinically feasible, for 1 week prior to undertaking vestibular investigation.

Table 5.5 outlines the auditory, vestibular and oculomotor signs that might be reasonably sought at various levels in the central nervous system.

Cerebrovascular disease

The cerebral vessels are a site of predilection for atherosclerosis, together with the myocardial and peripheral vasculature. The specific risk factors identified with cerebral ischaemia are diabetes mellitus, an elevated haematocrit and hypertension.

Vertebrobasilar ischaemia

Vertebrobasilar ischaemia may give rise to end organ or eighth nerve dysfunction as described previously, but frequently gives rise to vestibular symptoms, as a result of ischaemia of the vestibular nuclei, which occupy a large area in the lateral zone of the brainstem (Gillilan, 1964) and are particularly susceptible to a reduction in the blood flow of the main basilar artery. Williams (1964) has documented that vertigo and/or dizziness is the first and most frequent symptom of vertebrobasilar ischaemia, whereas cochlear symptoms are rarely found (Millikan, Siekert and Whisnant, 1959).

Vertebrobasilar insufficiency is defined as 'a state of transient decrease in the cerebral blood flow, without actual infarction, resulting in transient inability to meet the metabolic requirements of the brain' (Millikan and Seikert, 1955). While the pathophysiology of vertebrobasilar insufficiency remains to be fully explained, the primary underlying pathology is that of occlusive vascular disease (Williams and Wilson, 1962). Although this pathology alone may compromise the vertebrobasilar system and produce intermittent brainstem symptoms including vertigo, a number of precipitating factors have also been identified in the aetiology of recurrent transient ischaemic attacks in the vertebrobasilar territory: recurrent thromboembolism; postural and/or systemic hypotension; compression of the vertebral arteries and haematological disorders, for example anaemia and polycythaemia.
Classically, vertebrobasilar ischaemia gives rise to the following symptoms, in order of frequency: dizziness and/or vertigo, dysarthria, numbness of the face, hemiparesis, headache and diplopia (Fisher, 1967). Other common, although less frequently encountered, symptoms include oscillopsia, dimness of vision, field defects, transient blindness, dysphasia, drop attacks, alternating weakness of opposite sides of the body, dysaesthesiae and cerebellar ataxia. The diversity of symptoms and signs reflects the close proximity of cranial nerve nuclei with motor and sensory tracts within the small confines of the brainstem. Transient ischaemic attacks vary in duration but, by definition, must be less than 24 hours. They occur randomly and may or may not be stereotyped. The occurrence of similar symptoms on opposite sides of the body is characteristic of this disorder (Millikan and Seikert, 1955). Classical attacks of vertebrobasilar ischaemia with the symptoms outlined above do not present a diagnostic dilemma, but isolated episodes of vertigo, particularly in an elderly patient with other cardiovascular symptoms and signs, may give rise to diagnostic difficulties.

Certain points are worthy of note in this respect. Fisher (1967) reported that 77% of a large series of patients with basilar occlusion experienced dizziness but under one-quarter of these gave a history of true rotational vertigo. In 25% of these patients with vestibular symptoms, dizziness or vertigo occurred alone as the presenting symptom, whereas in a further 50%, a diagnosis was established on the basis of other neurological symptoms or signs. In differentiating peripheral vestibular dysfunction from vertebrobasilar episodes, Barber and Dionne (1971) have emphasized that tinnitus and deafness are unusual manifestations of vertebrobasilar ischaemia and, if present, are almost always accompanied by other symptoms and signs of brainstem involvement. Conversely, dizziness or vertigo accompanied by only eighth nerve manifestations are unlikely to be of vascular origin. Fisher (1967) has emphasized that vestibular symptoms unaccompanied by other brainstem symptoms or signs within 6 weeks, make a diagnosis of vascular pathology extremely unlikely. Despite the presence of vestibular and oculomotor abnormalities in vertebrobasilar ischaemia, no characteristic pattern of neuro-otological findings has emerged in this disorder (Covera et al, 1980).

In the presence of brainstem symptoms and signs, neurological investigation is required. Great emphasis has been placed upon the early diagnosis of vertebrobasilar disease, on the assumption that therapy may avert the possibility of an impending stroke. There is, however, no good evidence to support this contention and Marshall (1964) has drawn attention to the good prognosis of patients with ischaemia in the posterior circulation, as opposed to the middle and anterior cerebral circulations.

In addition to atherosclerosis, certain other rare conditions, such as syphilitic endarteritis and the autoimmune vasculitides, for example systemic lupus erythematosus, giant cell arteritis and polyarteritis nodosa, may give rise to transient ischaemic episodes in the vertebrobasilar territory. Anatomical abnormalities may also produce such symptoms: anomalous origin of the vertebral artery (Sheehan, Bauer and Meyer, 1960); mechanical compression of the vertebral arteries by skeletal structures (cervical spondylosis) and/or muscular structures (fibrous band formation) (Powers, Dirslane and Nevins, 1961).

The subclavian steal syndrome is found in 3% of patients with vertebrobasilar symptoms. There is occlusion of the subclavian artery such that use of the involved upper limb causes reversal of blood flow in the vertebral artery on that side, so that it acts as a
collateral to the upper limb. Blood is syphoned from the vertebrobasilar system into the distal subclavian artery. This diagnosis should be considered when claudication or fatigue of the upper limb is accompanied by vertebrobasilar symptoms. A systolic bruit in the supraclavicular fossa with a disparity of blood pressure between the two arms is the characteristic sign of this disorder.

**Completed strokes**

Completed strokes in the vertebrobasilar territory are associated with a number of well-recognized syndromes. The Wallenberg, or lateral medullary, syndrome may be the result of occlusion of the posterior inferior cerebellar artery, or primary disease of the vertebral artery (Fisher, 1967). The syndrome is characterized by vertigo with ipsilateral dissociated sensory loss in the distribution of the facial nerve and contralateral truncal loss, together with ipsilateral cerebellar ataxia, bulbar palsy and Horner's syndrome. Baloh, Yee and Honrubia (1981) have documented specific derangements of visuo-vestibular interaction including spontaneous rotatory nystagmus, with the fast phase directed towards the normal side; tonic deviation of the eyes towards the side of the lesion, with loss of fixation; voluntary and involuntary saccades of larger amplitude in the direction of the lesion and asymmetry of smooth pursuit; optokinetic and vestibular responses as a result of the interaction between spontaneous nystagmus and all slow eye movements. Vertigo is also a feature of the less common anterior, inferior cerebellar artery syndrome.

**Pontine and cerebellar haemorrhage**

These conditions may also present with dizziness. In the former, there are multiple brainstem signs, and dizziness is usually a fleeting event before the patient becomes unconscious. Cerebellar haemorrhage is of particular important as, if diagnosed early, it may be surgically corrected. The patient presents with acute vertigo, vomiting and an inability to stand. Without rapid intervention, the patient dies from brainstem compression. Magnetic resonance scanning, computerized axial tomography and/or angiography facilitate diagnosis.

**Supratentorial vascular lesions**

Vertigo frequently accompanies supratentorial vascular lesions, but the pathophysiological mechanism remains unclear. In Fisher's excellent review of vertigo and cerebrovascular disease (1967), it is of interest that 8% of patients with internal carotid artery and/or middle cerebral artery ischaemia, complained of dizziness, but no such complaint was found in 13 cases of anterior cerebral infarction.

**Migraine**

Migraine is the most commonly encountered neurological disorder in the UK, affecting 5-10% of the population (Lance, 1969). It is often familial and is usually characterized by headache. Although benign, migraine can produce catastrophic disruption of the patient's lifestyle. There are two main forms: classical migraine, which is preceded by an aura consisting of neurological phenomena such as visual, sensory or speech disturbances; and non-classical, or common migraine, which may best be described as a 'sick headache' unassociated with any specific neurological deficit.
Reviewing 200 migrainous subjects, Kayan and Hood (1984) documented vestibular symptomatology in 39%, cochlear pathology in 4.5% and combined auditory and vestibular disturbances in 15.5%. Full investigation of 80 migrainous subjects, referred for assessment because of vestibulocochlear symptoms, revealed that 77.5% had objective abnormalities and one-half of these suggested central pathology. These findings would support the many reports in the literature of an association between dizziness and/or vertigo and migraine (Fedorova, 1970; Kayan, 1973). It is now clear that vestibular symptoms may occur as part of the aura, in the headache phase, or during the headache-free periods (Kayan, 1984). As mentioned above, there would appear to be a clear association between migraine and Ménière's disease (Hinchcliffe, 1967a; Fedorova, 1970; Kayan and Hood, 1984).

Basilar artery migraine, which was first described by Bickerstaff (1961), is particularly common in adolescent girls and is closely related to the menstrual cycle. The condition consists of an aura lasting 2-45 minutes, of vertigo, ataxia, dysarthria, tinnitus and sensory disturbance in the distal limbs and around the lips, together with visual disturbances, characteristic of dysfunction in the territory of the posterior cerebral circulation. The diagnosis of basilar artery migraine is clinical and established by the characteristic history together with the exclusion of other pathology.

**Multiple sclerosis**

Multiple sclerosis is primarily a disease of young adults, and is one of the most commonly encountered conditions in neurological practice. The condition is characterized by lesions of demyelination which occur randomly in time and space. The diagnosis is usually based on clinical criteria supplemented by characteristic investigative findings on examination of the cerebrospinal fluid (elevated protein, lymphocyte count and the presence of oligoclonal bands), abnormalities of electrophysiologic potentials (visual evoked potentials, auditory brainstem responses) and magnetic resonance scanning.

Vertigo and dizziness are common complaints at some time during the course of the disease in patients who have definite multiple sclerosis (Rudge, 1983) and vertigo is the initial symptom in approximately 5% of cases (McAlpine, Lumsden and Acheson, 1972). Neurologically, as the disease is characterized by disseminated lesions, the presentation is infinitely variable, but certain features deserve special mention because of the consistency of their occurrence. Retrobulbar neuritis, with blurring and/or loss of vision and pain in or behind the eye, is associated with demyelination of the optic nerve and is an initial presenting symptom in approximately 20% of cases. Diplopia, limb weakness, sensory disturbances and ataxia also occur early in the course of the disease. In long-standing cases, examination frequently reveals involvement of the pyramidal tracts, with hyperreflexia and extensor plantar responses; cerebellar signs of intention tremor, dysdiadochokinesis, ataxia and dysarthria; sensory involvement and visual disturbance.

Neuro-otologically, multiple sclerosis is characterized by derangement of eye movements. Pursuit has been reported to be almost always deranged (Solingen et al, 1977; Mastaglia, Black and Collins, 1979) and the same authors have also reported a high incidence of saccadic abnormalities. Bentzen, Jelnes and Thygesen (1951) reported spontaneous nystagmus in 80% of a population of patients with multiple sclerosis, over the age of 40 years. They also noted the common occurrence of positional nystagmus. Acquired pendular
nystagmus and dissociated nystagmus are particularly valuable diagnostic pointers in multiple sclerosis, as they are relatively unusual findings in other disease processes (Cogan, 1970; Aschoff, Conrad and Kornhuber, 1974).

Vertical nystagmus is another frequent finding, in particular upbeat nystagmus. Cerebellar and brainstem lesions may result in abnormalities of nystagmus induced by rotation or caloric testing. Caloric responses in the presence of optic fixation are commonly symmetrically enhanced (Huygen, 1983) and, characteristically, removal of optic fixation does not enhance the vestibular response. This is most probably the result of disruption of the cerebellar pathways to the vestibular nuclei. Brainstem involvement at the level of the vestibular nuclei may give rise to a canal paresis, with or without a directional preponderance.

Despite the controversy in the literature with respect to the audiometric findings in multiple sclerosis, it is clear that auditory brainstem evoked potentials are a sensitive test in establishing the presence of occult brainstem plaques (Robinson and Rudge, 1977, 1978). Stockard, Stockard and Sharborough (1977) have suggested that auditory brainstem evoked potential findings in multiple sclerosis may provide additional evidence of disseminated disease and thus avoid potentially dangerous procedures such as myelography and angiography.

**Neoplasia**

Dizziness and/or vertigo are early or initial symptoms in 25% of cases of brainstem tumours (Barnett and Hyland, 1952). Primary brainstem gliomata are rare in adult life, but metastases, pinealomata, haemangiomata and haemangioblastomata may occur (White, 1963). Typically, the history and examination reveals progressive development of multiple cranial nerve palsies, together with long tract signs, but confirmation of the diagnosis may prove difficult despite computerized tomographic scanning and vertebral angiography. Magnetic resonance imaging may prove to be a valuable diagnostic tool in this condition.

Although auditory symptoms are not prominent in brainstem neoplasia, a bilateral symmetrical hearing loss may occur (Luxon, 1980). More sophisticated auditory testing, using stapedial reflex threshold measurements (Jerger and Jerger, 1975; Jerger, Neely and Jerger, 1980) and auditory brainstem evoked potentials (Nodar, Hahn and Levine, 1980; Prasher, 1981), have been shown to be of value in the diagnosis and siting of brainstem tumours.

Cerebellopontine angle lesions and, in particular, acoustic neuromata, have been discussed previously. It is of note that Hard, Gardner and Howieson (1982) have recently emphasized that one-third of patients with acoustic neuroma seek medical attention for non-audiological symptoms, such as unsteadiness and headache.

Neurological examination may help to differentiate acoustic neuromata from other cerebellopontine angle lesions. Fifth nerve neuromata are characteristically associated with pain and facial nerve symptoms, which usually precede other symptoms including hearing loss. The presence of an associated fourth nerve palsy, or other extraocular palsy, may point to this diagnosis. Seventh and lower cranial nerve neurofibromata are rare, but the latter may be distinguished from eight nerve neuromata by the early development of bulbar signs, which are a late feature in large acoustic neuromata. The presence of trigeminal neuralgia, in
association with a cerebellopontine angle mass lesion, should raise the suspicion of a cholesteatoma (Revilla, 1947), a menigioma or, more rarely, a fifth nerve neuroma. Meningiomata usually present with a less severe hearing loss than one would expect with an acoustic neuroma of a similar size, and are often associated with headaches.

Primary tumours of the cerebellum are rare, but as primary brainstem gliomata, they occur more commonly in childhood. In adults, a cerebellar metastasis is a much more common diagnosis. Midline cerebellar lesions give rise to truncal ataxia and oculomotor abnormalities such as impaired pursuit, saccadic dysmetria and rebound nystagmus. Hemisphere lesions cause ataxia of the ipsilateral limbs, with little truncal ataxia. In addition, there may be lateralized oculomotor abnormalities such as an ipsilateral derangement of pursuit, together with a directional preponderance of optokinetic nystagmus and failure of suppression of nystagmus.

Considering supratentorial tumours, Spiegel and Alexander (1936) reviewed a large series of patients, many of whom complained of symptoms of dysequilibrium, which were not specifically termed 'vertigo'. It appeared that symptoms of dysequilibrium were more frequently reported in temporal lobe lesions than in other sites. Carmichael, Dix and Hallpike (1954) documented the modifying influence of the temporal lobes upon the vestibular nuclei, and hypothesized that disruption of these pathways may produce dysequilibrium. Alternatively, a mass lesion may form a focus for epileptic activity in the temporal lobe which may be associated with vertigo (see below).

Infection

Bacterial infection

Bacterial meningitis may involve the eighth cranial nerve, as mentioned previously. Tuberculous meningitis is now rare in western communities, but should be considered, particularly in third world countries, in immigrant populations and in any debilitated, deprived patient.

Intracranial complications of bacterial ear infection are considered in Volume 3, Chapter 12. An extradural abscess - a collection of purulent fluid between the dura mater and the bone of the middle or posterior fossa - is the most common intracranial complication of ear infections (Schuknecht, 1974). The patient presents with malaise, together with a fever, headache and vomiting, without firm neurological signs, making the diagnosis difficult. Computerized axial tomography or magnetic resonance imaging may reveal the diagnosis, but it may be necessary to proceed to surgical intervention to confirm the diagnosis. The spread of infection across the dura to the epidural space may result in thrombophlebitis of the lateral venous sinus, subdural abscess, meningitis and/or cerebral abscess, but fortunately, the use of antibiotics has made these conditions rare.

Venous thrombophlebitis is associated with fevers, rigors and profuse sweating. Involvement of the cerebral veins may produce recurrent focal epileptic attacks or status epilepticus (Pennybacker, 1961). Thrombosis of the superior sagittal sinus involves the arachnoid granules, impairing function and leading to otitic hydrocephalus.
A subtotal abscess may be confused with meningitis, as both may present with fever, severe generalized headache, vomiting and meningism. In the case of an abscess, focal signs may develop within 24-48 hours. Examination of the cerebrospinal fluid and the new scanning techniques which are available, have allowed the rapid differentiation of these pathologies.

Cerebral abscess, secondary to ear infection, is usually localized in the middle third of the temporal lobe or the anterior part of the lateral lobe of the cerebellum. An upper quadrant hemianopia will suggest a temporal lobe lesion with involvement of the optic radiation, and speech may be abnormal if the abscess is in the dominant hemisphere. These signs may be associated with a mild weakness of the face and arm. A cerebellar abscess is usually more obvious as the patient complains of severe neck stiffness, with marked gait ataxia and incoordination of the ipsilateral limbs. Speech may be slurred and an asymmetrical gaze paretic nystagmus may be observed. Scanning is again the diagnostic procedure of choice if an abscess is suspected.

The classic work on neurosyphilis (Merritt, Adams and Solomon, 1946) reported the occurrence of dizziness in 24% of cases of cerebrovascular syphilis, and in 14% of patients with meningeal syphilis. Early acquired syphilitic deafness usually reflects meningeal disease (Vercoe, 1976) and, unlike other forms of syphilitic deafness, usually recovers with treatment (Becker, 1979; McNulty and Fasselt, 1981). Unlike syphilitic labyrinthitis, in which normal cerebrospinal fluid findings are common, the diagnosis of acute meningo-vascular syphilis may be confirmed by elevated protein levels and cell count, as well as positive serological tests of the cerebrospinal fluid.

Viral infection

Viral encephalitis may involve the vestibular nuclei, as well as the nerve roots. The clinical history and findings on examination may suggest the diagnosis, which is confirmed by examination of the cerebrospinal fluid.

Basal ganglion disorders

Imbalance, alterations in gait, and postural abnormalities are commonplace in Parkinson's disease, which is one of the most common neurological disorders, occurring in about 1 in every 200 of the population over the age of 50 years. The disease is characterized by rigidity, akinesia and 4 Hz 'pill-rolling' tremor, which is present at rest. In idiopathic Parkinson's disease, postural instability is a late feature, whereas Purdon-Martin (1967) demonstrated that, in postencephalitic parkinsonism, there is a failure of the normal protective righting reflexes, following a sudden tilt. Reichert, Doolittle and McDowell (1982) have reported that patients with Parkinson's disease have reduced, or absent, vestibular responses, compared with a controlled population. In addition, the literature abounds with reports of abnormal eye movements in Parkinson's disease (De Jong and Melvill-Jones, 1971), although much of the work is contradictory. Nonetheless, certain generalizations may be made. Corin, Elizon and Bender (1972) have confirmed the common observation that convergence and vertical gaze palsies occur in a substantial proportion of patients with this disorder. Upgaze is more commonly involved than downgaze and the saccadic system is usually more severely affected than the pursuit system. Hence, it would appear that in Parkinson's disease, not only an impaired vestibular input, but also derangement of visual control and alterations in the
modulation of the multisensory input required for balance, may all contribute to dysequilibrium.

The introduction of L-dopa has done much to alleviate the idiopathic form of this disorder, but may exacerbate other extrapyramidal disorders. Accurate diagnosis is therefore essential and idiopathic Parkinson's disease must be differentiated from drug-induced (phenothiazine and butyrophenones) Parkinson's syndrome, postencephalitic Parkinson's disease, benign essential tremor and other basal ganglion disorders. The incidence of drug-induced parkinsonism increases with age (Ayd, 1961), as does idiopathic Parkinson's disease. Following withdrawal of a causative drug, it may take as long as 18 months for recovery to occur.

Parkinson's disease must be differentiated from other akinetic rigid syndromes, for example that of cerebrovascular disease, which is often accompanied by pyramidal signs and dementia (Parkes et al, 1974), while tremor is almost never present, in contrast to Parkinson's disease. Demented patients with Alzheimer's disease may develop frank signs of basal ganglia dysfunction with akinesia, rigidity and instability late in the natural course of the disorder (Pearce, 1974). The Steele-Richardson-Olszewski (1964) syndrome may superficially resemble Parkinson's disease. It is characterized by marked axial rigidity, expressionless facies, mild dementia and a supranuclear palsy, while the combination of absence of tremor and relatively normal tone in the limbs may help in the differentiation of the two conditions. Vertical eye movements are affected before horizontal gaze, and voluntary eye movements before pursuit. Frequently, downgaze is more involved than upgaze and saccadic velocities are reduced (Dix, Harrison and Lewis, 1971). This disorder usually presents in the sixth decade of life, or later, and is progressive, with death occurring within 7 years of the first symptom. There is little response to conventional antiparkinsonian therapy.

Bannister and Oppenheimer (1972) have described a group of conditions which are now referred to as the multisystem atrophies. Individual patients may exhibit a combination of symptoms and signs of autonomic neuropathy, cerebellar degeneration and strionigral degeneration. These disorders are all associated with balance impairment, although the exact pathophysiology of the derangement remains unclear.

**Cerebellar disease**

Cerebellar disease may present with imbalance, nystagmus and vertigo, but episodic symptoms are unusual in comparison with persistent unsteadiness. In 1922, Holmes described a cerebellar syndrome comprising gait and limb ataxia, hypotonia, disordered eye movements and slurred speech. Midline cerebellar lesions may produce truncal ataxia as the only sign and, to the untrained eye, this may be misinterpreted as an hysterical gait disorder. Frequently, routine neurological examination on the bed will fail to reveal any abnormality. In the adult, secondary metastases in the cerebellum are the commonest cause of a clear-cut cerebellar syndrome, but ischaemia in the vertebrobasilar territory should also be considered. Cerebellar ectopia (Arnold-Chiari malformation) may also present with ataxia. Cerebellar degeneration may be associated with malignancy, phenytoin intoxication, alcoholism and myxoedema. Early diagnosis may lead to effective treatment in these groups.
Spinocerebellar degenerations have been considered above. Vestibular and oculomotor abnormalities are well documented, and there are numerous reports in the literature of hereditary syndromes with various combinations of spinocerebellar degeneration, neuropathy, muscular wasting and sensorineural deafness (Matthews, 1950; Sylvester, 1958; May and White, 1968; Musiek, Weider and Mueller, 1982).

Pure cerebellar ataxia may be differentiated from these various syndromes of spinocerebellar and olivopontocerebellar degeneration, although they are rare, as the majority of patients have additional features such as neuropathy, retinitis pigmentosa or other central nervous system disorders. Zee et al (1976) have documented the vestibulo-ocular manifestations in a group of patients with late-onset dominantly inherited cerebellar ataxia - deranged pursuit, nystagmus, oculomotor dysmetria and abnormal vestibulo-ocular reflex suppression.

The hereditary neuropathies include a large number of disorders which are usually classified on the basis of the clinical presentation and pattern of inheritance. Certain of these conditions have been biochemically identified, for example Refsum's disease, but the majority have not. The importance of diagnosing Refsum's disease, by measuring the serum level of phytanic acid, is that progression of the disease can be halted by exclusion of phytanic acid from the diet. The occurrence of neuro-otological abnormalities in this group of patients is rare, but the diagnosis should be considered in a patient with dysequilibrium and a family history of neuropathy (Denny-Brown, 1951; Hallpike, 1967b; Allen et al, 1978).

**Frontal lobe lesions**

Frontal lobe lesions may lead to a disordered gait and balance disturbances (Denny-Brown, 1958; Meyer and Barron, 1960). True ataxia, produced by a frontal lobe lesion, has been attributed to compressions of corticocerebellar connections (Frazier, 1936), but an apraxia of gait is the more usual result of frontal lobe dysfunction. Apraxia is characterized by the loss of ability to use the lower limbs appropriately, which cannot be accounted for by any demonstrable sensory or motor impairment. Associated frontal lobe signs include grasp reflexes, gegenhalten and difficulty in copying movements, such as kicking a ball.

**Hydrocephalus**

Both communicating and non-communicating hydrocephalus may give rise to a gait disturbance, associated with loss of sphincter control and cognitive impairment, leading to dementia. Scanning may provide the diagnosis and, in appropriate cases, benefit is derived from ventricular shunting.

**Epilepsy**

The association of vertigo and epilepsy has been considered rare, but a recent report has suggested that this is not necessarily correct (Kogeorgeos, Scott and Swash, 1981). Vertigo may be a manifestation of an aura, or part of a temporal lobe seizure, but has also been identified with other forms of epilepsy (Lennox, 1960; Schneider, Calhoun and Crosby, 1968). In 1907, Gowers reported dysequilibrium in 90 of a series of 505 cases of epilepsy.
Smith and Docherty (1982) have recently reported a patient with temporal lobe epilepsy who complained of oscillopsia and demonstrated nystagmus.

**Syringobulbia/syringomyelia**

Syringomyelia is characterized by a ballooning of the central cavity of the spinal cord and is commonly associated with the Arnold-Chiari malformation. The condition is diagnosed by the presence of a dissociated sensory loss (loss of pain and temperature sensation), together with muscular weakness and wasting, secondary to anterior horn cell involvement, and trophic changes in the affected limbs. Extension of the lesion upwards into the medulla and pons is known as syringobulbia. Clinically, this may be associated with trigeminal pain, vertigo and a bulbar palsy. Disruption of the vestibular afferent nerves from the neck muscles commonly results in the development of nystagmus before the sensory loss extends on to the face.

**Peripheral neuropathies**

Sensory neuropathy, as a result of degeneration of the dorsal root ganglia, is a rare, but striking, syndrome characterized by the subacute onset of a severe sensory ataxia with gross sensory loss, particularly of posterior column sensibility, but often of all forms, with loss of reflexes. The commonest causes include carcinoma, syphilis and diabetes (McAlpine and Page, 1951; Bosanquet and Henson, 1957). Historically, tabes dorsalis is the best recognized of these conditions. Hereditary sensory neuropathy may also present with marked dysequilibrium (see above).

Having discussed the general medical, otological and neurological causes of vertigo, there remain two conditions of importance which fall outside this classification, namely cervical and ocular vertigo.

**Cervical vertigo**

Cervical vertigo is defined as 'vertigo induced by changes of position of the neck, in relation to the body'. Since the early work of Barany (1918), it has been known that movements of the neck may provoke attacks of dizziness and induced nystagmus (Voss, 1925; de Kleijn and Nieuwenhuyse, 1927). There is much controversy as to the underlying pathophysiology of cervical vertigo, but it is well documented that the labyrinths are not essential for the development of cervical vertigo (Bos, 1972).

Many pathological processes have been implicated in the aetiology of cervical vertigo: cervical spondylosis (Davis, 1953); trauma to the vertebrae (Rebattu and Lesne, 1962); avulsion of nerve roots (Decher, Rohr and Unterhanscheidt, 1958); trauma to the joints (Stoddard, 1952); lesions of the neck muscles (Gray, 1956); cervical adenopathy (Mayoux, 1953); involvement of the vertebral artery (Sandstrom, 1962) and brachial radiculitis or neuritis (Biemond, 1940). Despite the assertion by Ryan and Cope (1955) that 'we are convinced that the neck plays a larger part in the mechanism of vertigo, than is generally thought', knowledge has advanced little over the last 30 years.
Three mechanisms have been postulated to explain vertigo of cervical origin:

1. Sympathetic irritation resulting in vertebrobasilar ischaemia (Barré, 1926)
2. Intermittent vertebral artery compression by osteophytes, caused by cervical spondylosis (Sheehan, Bauer and Meyer, 1960)
3. Deranged somatosensory input from the cervical kinaesthetic receptors (De Jong, 1967; Wyke, 1979).

Pfaltz and Richter (1958) have postulated that cervical vertigo is probably not the result of a single pathophysiological mechanism, but of multifactorial aetiology.

Cervical articular mechanoreceptors have been documented, by both clinical and experimental observations, to be essential for both the sense of balance and cervical kinaesthesia (that is awareness of head and neck position) (Cohen, 1961; De Jong, 1967; Igarashi et al, 1969; Jongkees, 1969). An age-dependent degenerative loss of mechanoreceptors has been clearly documented (Arnold and Harriman, 1970) and, in the elderly, coincidental disease, for example cervical spondylosis, may exacerbate any resultant dysfunction.

A widely held belief, particularly about the elderly, is that vertigo and nystagmus are the result of vertebrobasilar ischaemia, secondary to compression of the blood vessels of the posterior circulation, by arthritic changes in the neck. It must, however, be noted that both Arslan (1952) and Aschan and Hugosson (1966) have emphasized that unilateral, or indeed, bilateral compression of the vertebral arteries, in the presence of a normal circle of Willis and internal carotid arteries, produces only minimal brainstem ischaemia.

The clinical differentiation of vertebrobasilar insufficiency from cervical vertigo is difficult. As noted above, it must be emphasized that vertigo, or dizziness, alone is unusual in vertebrobasilar disease (Troost, 1980). Kuilman (1959), Jongkees (1969) have outlined the symptomatology associated with cervical vertigo. Classically, neck and/or occipital pain occurring particularly in the morning, is associated with signs of cervical root compression: paraesthesiae in the arms, muscle weakness and wasting, corresponding depression of reflexes and reduction of posterior column sensation. Cochleovestibular symptoms are explained on the basis of chronic irritation of the sympathetic plexus, giving rise to circulatory dysfunction, while vestibular symptoms may also be the result of derangement of mechanoreceptors, giving rise to spurious information regarding the position of the head, relative to the neck. In Kuilman’s series (1959), approximately one-third of patients complained of tinnitus and approximately one-fifth of balance disorders. Jongkees (1969) described a number of patients with classical Ménière’s syndrome on the basis of cervical vertigo, but vestibular symptomatology was more prominent than cochlear symptoms.

Thus, a careful history is essential in making the diagnosis of cervical vertigo, while normal optokinetic responses, together with a positive passive neck torsion test, on examination, may support the diagnosis (Jongkees, 1969; Toglia, 1975). It must be emphasized that radiographic findings may prove misleading as 75% of people over the age of 50 years show osteoarthritic spurs, or other degenerative changes, in the cervical vertebrae which are not directly related to symptomatology (Pallis, Jones and Spillane, 1954). Treatment in the form of local heat, massage, correction of posture, immobilization of the neck in a
cervical collar, cervical contour pillow, neck traction, neck exercises, relaxation and psychotherapy all have a place in the treatment of this disorder (Jackson, 1958). If such simple measures fail, local anaesthetic infiltration may be of value in an acutely painful situation. The need for judicious gentle traction must be underlined, in view of the experience of Ryan and Cope (1955) of severe cervical vertigo, as a result of more vigorous traction.

**Ocular vertigo**

In 1794, Erasmus Darwin wrote: 'many people, when they arrive at 50 or 60 years of age, are affected by slight vertigo; which is generally ascribed to indigestion, but in reality arises from the beginning defect of their sight ... These people do not see objects so distinctly as formerly and by exerting their eyes more than usual, they perceive the apparent motion of objects, and confound them with the real motion of them; and, therefore, cannot accurately balance themselves so as to easily preserve their perpendicularity by them'. Ocular vertigo is usually described as a sensation of unsteadiness or disorientation, and is caused by a mismatch of information arising from the visual system, with information from the labyrinths and/or the somatosensory system. When the visual input is defective, there may be a contradiction between the true visual input and the expected visual input, producing pathological visual vertigo (Brandt, 1984). Vision plays a major role in postural stabilization and attenuates body sway by 50-100% (Travis, 1945; Edwards, 1946). The effect of removing vision in normal and a sensory impaired patient is clearly documented in the figure.

Physiological visual vertigo may be induced by optokinetic stimulation, or by a critical distance between the subject and the closest stationary visible landmark, as for example in height vertigo. Pathological visual vertigo, however, may be secondary to involuntary eye movements, for example oscillopsia, or a mismatch between the true and the expected eye position in the head, as may occur with an acute extraocular muscle paresis (Brandt, 1984).

A common visual mismatch giving rise to symptoms of dysequilibrium is experienced by subjects wearing refractive lenses for the first time. Not only may this result in 'expected' versus 'real' image mismatch, but may also disturb an individual's meridian magnification and aniseikonia (inequality in size of retinal images) (Belmont, 1967).

**Conclusions**

This account of balance disorders provides the clinician faced with a dizzy patient with a working knowledge of the differential diagnosis. Priority must be given to determining whether the balance disorder is primarily of vestibular, neurological or general medical origin. A full and accurate history is invaluable, as always in clinical practice, and certain aspects of the clinical picture, which have been outlined at the beginning of this chapter, may narrow the differential diagnosis considerably. A complete physical examination must be performed if the single, or multiple, pathologies giving rise to disordered balance are to be identified. It should be re-emphasized that Drachman and Hart (1972) identified the multisensory deficit syndrome as the third major cause of unsteadiness, in their study of 125 unselected dizzy patients. The main abnormalities reported in these patients were peripheral neuropathy (85%), cervical spondylosis (71%), vestibular abnormalities, as shown by inadequate labyrinthine responses (64%) and visual loss, secondary to cataracts (35%). All subjects presented a combination of the above causes and most of them were said to be 'old and diabetic', but it
is clear that only by a thorough general medical examination are the multiple components of dizziness likely to be identified. On the basis of a detailed history and examination, specific investigative procedures may be required. Identifiable vertiginous syndromes, for example syphilitic labyrinthitis, acoustic neuromata and temporal lobe epilepsy, may require precise therapeutic regimens, but, in general, the pathophysiology of most vestibular disorders is poorly understood, and it is therefore impossible to consider specific therapy. Symptomatic treatment may be surgical or medical. In general terms, surgical intervention is indicated only for one of three main reasons:

(1) to secure the safety of the patient, for example in complications of otitis media
(2) to improve the quality of life in a patient suffering from severe rotational vertigo in whom all medical measures have failed
(3) to exclude a perilymph fistula.

The medical management of vertigo is discussed fully in Chapters 11 and 15 and may be summarized as comprising counselling, which is of the utmost importance, together with vestibular rehabilitation and appropriate vestibular sedatives.
**Table 5.5. Neurological, auditory, vestibular and oculomotor abnormalities at various levels in the central nervous system***

**Site**

**Neurological**

**Auditory**

**Oculomotor**

**Caloric + OKN**

### Nerve VIII

- **Veering to side**
  - Unilateral SN hearing loss
  - Ipsilateral SR elevation
  - Ipsilateral SR decay
  - Poor speech discrimination
  - Prolonged 1-V latency or absent ipsilateral ABR

- **EOMs: Normal**

- **Nystagmus: Vestibular to contralateral side**

- **Caloric: Ipsilateral CP ± contralateral DP**

- **OKN: Normal**

### Brainstem

- Constellation of cranial nerve signs + motor + sensory long tract signs
- ± Cerebellar signs (see below)
  - ± Symmetrical SN hearing loss
  - Elevation / absence SRs (bilateral ipsi + contralateral)
  - Derangement of sensitized speech tests
  - Bilateral ABR abnormalities

- **EOMs: Gaze paresis / INO**

- **Oculomotor nerve palsies**

- **Increased latency saccades**

- **Decreased maximum velocity saccades**

- **Ipsilateral / bilateral impairment of pursuit**

- **Nystagmus: All varieties**
  - **Caloric: Ipsilateral CP ± ipsi / contralateral DP**
  - ± Impairment of suppression
  - ± Perverted nystagmus
  - **OKN: Ipsi / bilateral decrease in SCV**
  - ± DP ipsi / contralateral

### Cerebellopontine angle

- **Early: V + VII nerves**

- **Late: VI nerve**

- **Ipsilateral cerebellar ataxia**

- **Bulbar palsy**

- **Papilloedema**

  - As for VIII nerve lesions
EOMs: Ipsilateral saccade dysmetria
Progressive ipsi / bilateral impairment of pursuit
Nystagmus: Vestibular (contralateral side)
Gaze paretic ± rebound ± vertical
  Caloric: CP (ipsilateral)
  ± DP (ipsi / contralateral)
OKN: Progressive ipsi / bilateral decrease SCV
  DP to side of lesion

Cerebellum
  Hypotonia
  Limb ataxia: past pointing, diadochokinesis
  Truncal ataxia
  Dysarthria
  N/A

  EOMs: Impaired pursuit (ipsi / bilateral)
  Normal saccade velocity
  Dysmetria
  Drifting eye movements
  Flutter ± micro / macro saccadic oscillations
  Nystagmus: Gaze paretic
  Rebound
  Periodic alternating
  Vertical
    Caloric: Hyperactive VOR
    Impaired VOR suppression
    Ipsilateral DP
    Dysrhythmic response
    OKN: Decrease SCV
    Ipsilateral
    DP ipsilateral
    Dysrhythmic response

Basal ganglia†
  Rigidity
  Bradykinesia
  Tremor
  N/A

  EOMs: Hypometric saccades
  Increased latency saccades
  Impaired pursuit (ipsi / bilateral)
  Supranuclear eye movement disorder
  Nystagmus: nil
  Caloric + OKN
  Ips / bilateral decrease SCV
  ± Deviation eyes in direction slow component
Cortex
Personality change
Impairment higher functions
Apraxia
Aphasia
Epilepsy
Hemiparesis
Sensory deficits
Visual field defects
Gaze abnormalities
Papilloedema

PTA - normal
Derangement of dichotic tests
+ directionalization tests (unilateral temporal lobe)
Auditory agnosia ± pure word deafness (bilateral temporal lobe)
ABR - normal

EOMs: Contralateral saccade inaccuracy (FP cortex)
Ipsilateral pursuit
Derangement (PO cortex)
Nystagmus: ± Fine gaze paretic (PO cortex)
Caloric: DP reverses with + without optic fixation (PO cortex)
OKN: FO cortex - normal
PO cortex - ipsilateral decrease SCV
ipsilateral DP.

OKN = optokinetic nystagmus
EOMs = extraocular movements
VOR = vestibulo-ocular reflex
SCV = slow component velocity
DP = directional preponderance
CP = canal paresis
FP = frontoparietal
PO = parieto-occipital
INO = internuclear ophthalmoplegia
SN = sensorineural
SR = stapedial reflex
N/A = not applicable
PTA = pure tone audiogram
ABR = auditory brainstem response