Chapter 14: Vestibular disorders

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Embryology

The membranous labyrinth of the inner ear develops from the otic capsule between the fourth and twelfth weeks of intrauterine life. The cartilage around the capsule differentiates into the osseous labyrinth at 6-7 weeks' gestation and, at that time, is separated from the membranous portion by the perilymphatic space (Dayal, Farkushidy and Kokshanian, 1973). As the vestibular system is older phylogenetically, each stage in the development of this system is in advance of that of the auditory system, and is therefore less vulnerable to environmental insult.

The semicircular canals arise from the utricular portion of the otic vesicle and have attained gross morphology at the 30 mm stage. The cochlear duct extends from the saccular portion and has two and a half coils at the 500 mm stage (Anniko, 1983). The neuroepithelium is differentiated in the utricle at 7 weeks and the semicircular canals at 8 weeks, being complete in all cristae and maculae at 12-14 weeks. By comparison, the basal turn of the cochlea is not fully differentiated until mid-term (Dayal, Farkushidy and Kokshanian, 1973; Anniko, 1983). The vestibular nerve is among the first of the central nervous system tracts to myelinate at around 16 weeks, coinciding with the myelination of the intersegmental tract systems of the cervical spinal cord (Hamilton and Mossman, 1972). The auditory nerve is not myelinated until 20-24 weeks (Eisenberg, 1983). The vestibular system is functional well before birth with a feeble Moro reflex present as early as the ninth to tenth weeks (Holt, 1975) and the vestibulo-ocular reflex is present at 24 weeks (Hamilton and Mossman, 1972).

The different types of congenital dysplasia of the inner ear reflect this sequence of events. Vestibular dysplasia is usually confined to the less common and more severe anomalies. As these arise early in fetal life they may be associated with other morphological abnormalities (Chandra Sekhar and Sachs, 1975).

Four types of inner ear dysplasia are recognized:

1. Michel aplasia: total aplasia of the osseous labyrinth.

Scheibe dysplasia is the most common type. In children with this congenital anomaly the deafness is often not accompanied by vestibular dysfunction and there may be no associated abnormalities in other systems. X-ray studies of the petrous temporal bone will also be normal. Some environmental hazards will cause only microscopic abnormalities of the neuroepithelium. Hultcrantz and Anniko (1984) demonstrated changes in the crista ampullaris after gamma irradiation on the twelfth gestational day in mice. Wright et al (19820
demonstrated otoconial abnormalities after administration of prostaglandin inhibitors in pregnancy.

**Development of vestibular-based functions**

The function of the vestibular system can be divided into two categories: the maintenance of posture and stability of vision. In both capacities it integrates with other sensory and motor systems and cannot be considered in isolation. Assessment of vestibular function depends upon the integrity of systems such as the oculomotor tracts and close control of variables related to these systems.

**Maintenance of posture**

The primary archaic responses (disappearing by the age of 6 months) such as the Moro response, and the secondary inherent responses (persisting into childhood), for example the parachute reaction, are elicited as part of the neurological examination of the infant. They are not performed specifically to assess vestibular function. In the presence of otherwise normal development some of these may be used in this context and therefore deserve description.

**The primary archaic responses**

**The Moro response**

Sudden bilateral extension of the upper limbs followed by flexion is evoked by sudden jarring of the cot or by suddenly dropping the head backwards by 2 cm. To limit the stimulus to movement of the head in space, the infant is held on the examiner's forearms with hands supporting the infant's head. The examiner drops from the standing position to the crouching position by bending the knees. The baby is thus suddenly lowered and the reflex obtained (Eviatar and Eviatar, 1978). This response is always present at birth in normal children and disappears by the sixth month.

**Tonic labyrinthine response**

An increase in extensor tone when supine and flexor tone when prone is not always demonstrable in normal infants but is sometimes found in infants with cerebral palsy (MacKeith and Robson, 1970).

**The secondary inherent responses**

**Righting responses**

These reflexes maintain the head in the upright position and arise at the level of the red nucleus by integration of visual, proprioceptive and vestibular stimuli. To test for vestibular function these reflexes should be elicited with the baby blind-folded.

(a) The earliest righting reflex appears at 10 weeks and consists of extension of the head when the baby is held in ventral suspension.
(b) Later head righting reflexes can be obtained by changing the infant's position rapidly from upright to prone or supine. The head will be lifted to restore it to the vertical position.

(c) From 4 months the infant will tilt the head to maintain it vertical if the trunk is tilted through 30° (oblique suspension). At 5 months this manoeuvre is accompanied by the lower limbs moving away from the side to which the infant has been tilted (MacKeith and Robson, 1970).

(d) The propping reaction is elicited in the same manner with the baby in the sitting position. The head should remain vertical. The upper limb on the side to which the trunk has been tilted abducts as does the contralateral lower limb.

**Protective reactions**

**Parachute reaction**

(a) Downward parachute: the baby is held in vertical suspension and moved suddenly downwards. Up to the age of 5 months this elicits the Moro response, but above this age the lower limbs extend and abduct.

(b) Forward parachute: held in ventral suspension and moved forward and down the upper limbs abduct and extend and the fingers splay. This reflex can also be elicited by holding the baby in vertical suspension and moving rapidly to ventral suspension. For vestibular testing this should be performed with the infant blindfolded (Eviatar and Eviatar, 1978).

**Motor milestones**

The ages of sitting unsupported, crawling and walking unaided bear some relation to vestibular function (Kaga et al, 1981), but are also dependent upon the neurodevelopmental state of the infant. Those who bottom shuffle instead of crawling are known to walk later than other normal children (MacKeith and Robson, 1970). In the neurological examination of children the abnormalities found are more often due to failure to develop function than to its loss (MacKeith and Robson, 1970). The importance of vision for postural control, in particular low contrast vision in the peripheral field, cannot be overemphasized (Marron and Bailey, 1982). Attempts have been made to separate those aspects of motor control due to the vestibular system from other measurements of neurodevelopment, but the results were disappointing (DeGangi, Berk and Larsen, 1980; Bundy and Fisher, 1981). One measure of postural status that can be quantified is body sway using the vestibulospinal stability test (Black et al, 1977). Postural sway decreases during the first decade and then remains stable until it increases again in old age. However, it is not related to vestibular function alone. The development of motor and perceptual skills over a 3-year period in normal children and those with minimum brain dysfunction, perceptual and attention deficit, showed that many of the differences seen at the age of 7 years had disappeared 3 years later (Rasmussen et al, 1983; Gillberg, 1985). The development of motor milestones cannot be considered in isolation from the overall development of the child.
Stability of vision

The vestibulo-ocular reflex can be evoked by rotational or caloric stimuli, and enables clear vision to be maintained while the head is in motion. It consists either of nystagmus or of deviation of the eyes equal and opposite to the movement of the head. The fast phase of vestibular nystagmus is central in origin and is affected by maturational changes within the reticular formation. Visual-vestibular interaction has a complex effect upon the vestibulo-ocular reflex and is responsible for many of the changes that take place with increasing maturity.

Response to rotation

In the neonatal period, rotation is most conveniently achieved by holding the infant at arms length in vertical suspension while the examiner rotates about his own axis. As the infant is on the circumference of the circle, looking inwards, the eyes will open and deviate in the direction of rotation.

For the first few weeks of life there is deviation of the eyes alone. Between the fourth and sixth week nystagmus is superimposed upon this deviation. Both optokinetic and vestibular nystagmus will be in the same direction so that once nystagmus intervenes the test cannot be used to assess vestibular function. After this age the alert baby will usually fix his eyes on the examiner's face (Farmer, 1964; Eviatar and Eviatar, 1978).

In the laboratory, rotation testing is undertaken with the subject seated in a chair capable of impulse, ramp or sinusoidal acceleration. Infants are held supported on an adult's lap and tolerate sinusoidal acceleration particularly well. To maintain the lateral semicircular canal in the plane of rotation, the head should be flexed 30°. This position cannot be maintained for long periods in infants (Kaga et al, 1981).

The position of the head during recording of rotation or caloric responses affects the quality of induced nystagmus (Schrader, Koenig and Dichgans, 1985). Care must therefore be taken to ensure that the infant's head is maintained in the vertical plane throughout the test procedure.

Perrotatory nystagmus was recorded in 182 infants in the first year of life by Eviatar and Eviatar (1979). The percentage of babies with perrotatory nystagmus in the first 90 days of life was greatest in full-term infants. By 180 days of age all except the preterm infants exhibited nystagmus. These latter findings were thought to be due to immaturity of the central nervous system at birth.

Postrotatory nystagmus was found to be replaced by tonic deviation of the eyes in two earlier studies of rotation responses in neonates (Groen, 1963; Mitchell and Cambon, 1969). Testing in complete darkness gives better results than using a blindfold (Cyr, 1980; Ornitz, 1983). Absence of nystagmus may be due to lack of alertness in the young or preterm infant (Ornitz, 1983). This factor was illustrated by Reding and Fernandez (1968) who demonstrated absence of nystagmus and presence of tonic deviation to rotation during non-REM sleep in children aged 6-9 years.
The frequency of perrotatory nystagmus is low at birth and increases uniformly during the first 6 years of life, although it does not reach adult values until 10-15 years of age (Tibbling, 1969; Kaga et al, 1981). In parallel with frequency of beat, the amplitude of the nystagmus is large in infants, gradually decreasing as frequency increases. The duration of perrotatory nystagmus increases rapidly during the first year of life, and more slowly in the following 6 years (Tibbling, 1969; Kaga et al, 1981). The standard deviation in older children was greater than for adults. Theoretically the maximum velocity of the slow phase of vestibular nystagmus is a better indicator of vestibular function than is frequency or duration, but the validity of this measurement depends upon accurate calibration which may be difficult in the very young child. In spite of this, the maximum velocity of the slow phase of both primary (perrotatory) and secondary (postrotatory) nystagmus has been recorded. The maximum velocity of the slow phase of primary nystagmus can be considered as representative of vestibular responsiveness. Tibbling (1969) found that young infants exhibited a high slow component velocity which decreased with age. This was supported by Ornitz et al (1979) for responses measured in total darkness. Eviatar and Eviatar (1979), testing with the infant blindfolded, found that the slow phase velocity increased up to the sixth month and then remained stable. Herman, Maulucci and Stuyck (1982) reported a higher gain of vestibulo-ocular reflex in children. Secondary nystagmus which reflects vestibular adaptation was also found to be of higher velocity in infants (Ornitz et al, 1979) and the secondary nystagmus: primary nystagmus ratio was significantly greater in early infancy. It was postulated that strong adaptation was protective when the infant was subjected to unpredictable passive motion.

**Caloric responses**

Studies of ice water calorics in the neonatal period include those of Mitchell and Cambon (1969), Eviatar and Eviatar (1979), Pignataro et al (1979) and Donat, Donat and Lay (1980). Caloric nystagmus was found to be absent in some very young infants but Eviatar and Eviatar (1979) reported that it was present in 90% of infants at the age of 9 months. Frequency, velocity, amplitude and latency showed age-related changes similar to those for rotation. Donat, Donat and Lay (1980) described caloric-evoked tonic deviation of the abducting eye with disconjugate deviation of the adducting eye. This internuclear ophthalmoplegia was thought to be due to lack of myelination of the medial longitudinal fasciculus. Iced water is a painful, albeit alerting, caloric stimulus.

The bithermal caloric test was shown to give uniform and reproducible results in children aged 2-7 years (Koenigsberger et al, 1970), measuring duration with eyes fixed on a target. Values for duration and left/right difference are quoted. Michishita (1967) recorded frequency, duration and slow phase velocity of caloric nystagmus from birth to 15 years of age with very similar results to those for rotation reported by Tibbling (1969). Calculations for canal paresis and directional preponderance indicated that differences would have to exceed 25% and 35% respectively to be indicative of pathology.

The variance for these calculated values is similar to that found in rotation testing (Kaga et al, 1981) in normal children but, to some extent, arises from technique. It is difficult to maintain the child in a constant state of alertness. Also the external meatus is small so that inequalities of irrigation can easily occur. Ornitz (1983) has stressed the requirement for total
darkness when testing children, as even small sources of light can be sufficient for visual inhibition of nystagmus.

**Eye movement control**

*Adventitious eye movements*

Both drift and microsaccades (Carpenter, 1977) are common in the dark, and are particularly prevalent in children. The 'Bell's phenomenon' in which the eyes deviate upwards upon closure is universal and will inhibit nystagmus. Bell's phenomenon is reversed during a mental alerting task with the eyes closed (Goebel et al, 1983). When testing children care is needed to maintain eye opening and mental alertness. Recording electronystagmography by DC rather than AC helps to distinguish adventitious movements from nystagmus and makes possible the recording of tonic deviation. Eviatar and Eviatar (1981) were, however, able to document the absence of spontaneous and positional nystagmus in normal infants under the age of one year using AC recording.

**Smooth pursuit**

At birth smooth pursuit is only possible at low velocities. This is thought to reflect foveal immaturity (Kremenitzer et al, 1979). At greater velocities catching up saccades are required (Herman, Maulucci, and Stuyck, 1982). At 8 weeks an infant will follow a target with intermittent motion (Bower, Broughton and Moore, 1971). Atkinson and Braddick (1979) felt that tracking was probably fully developed by the age of 3 years, but Gilligan et al (1981) demonstrated improvement up to the age of 10 years. Conjugate eye movements were complete at the age of 3 years. Herman, Maulucci and Stuyck (1982) demonstrated maturation of visual motor skills up to the age of 18 years.

**Saccadic movements**

These are present at birth but are not accurate until much later. Infants require more than one saccade to reach a target or may overshoot (Bower, Broughton and Moore, 1971).

**Optokinetic nystagmus**

This is present at birth when it may be utilized to test vision (Gorman, Cogan and Gellis, 1957). It is, however, only present at low velocities and is accompanied by tonic deviation of the eyes in the direction of the slow phase, instead of the fast phase as in adults (Kremenitzer et al, 1979).

**Visual-vestibular interaction**

This is concerned with the effect of visual stimuli upon vestibular evoked eye movements. As visual motor and pursuit skills mature this effect will increase. Herman, Maulucci and Stuyck (1982) found that, in children, the vestibular stimulus dominates the retinal stimulus so that visual suppression of the rotation-induced vestibulo-ocular reflex was found to be less in children than adults. Visual suppression is due to low contrast peripheral field sensitivity rather than foveal vision so that immaturity of the retinal stimulus does not
account for this immaturity. The development of coupling between the visual and vestibular systems in childhood is an extremely complex area which has been largely neglected by physiological research (Ornitz, 1983).

**Physiological vertigo syndromes**

Physiological vertigo will occur when there is a mismatch of input from different sensory systems. The multisensory vertigo syndromes were comprehensively reviewed by Brandt and Daroff (1980). Height vertigo, visual vertigo, somatosensory vertigo, auditory vertigo, head extension vertigo, and bending over vertigo receive scant attention in children but differ very little from their presentation in adults. Motion sickness, however, presents a quite different problem as it is much more prevalent in childhood.

**Motion sickness**

The incidence of this complaint depends upon the parameters used (Money, 1970), but by any criterion the disorder is far more common in children than adults. Surprisingly, there have been very few studies of motion sickness in this age group. Sharma (1980) found 16.6% of male and 29.2% of female children to be affected. Motion sickness is believed to result from conflicting kinetic input from visual, vestibular and non-vestibular proprioceptive systems. It results in the same symptoms as those produced by excessive vestibular stimulation (Money, 1970).

An intact visual system, is not required and blind children frequently experience motion sickness. During childhood an individual progresses from field dependency (relying on visual clues for orientation in space) to one of field independence (relying on internal clues). Deich and Hodges (1973) related field dependence to motion sickness in fifth, seventh and college grade students (10-18 years). Both field dependence and motion sickness decreased with age but there was not relation to field dependence to motion sickness, nor sex to field dependence. Girls are more susceptible to motion sickness than boys, and this is thought to be cultural as their vestibular experience is often less than that of boys (Deich and Hodges, 1973; Lentz and Collins, 1977; Sharma, 1980).

Infants below the age of 2 years are usually resistant to motion sickness, either because they travel supine (Money, 1970) or because of lack of visual input (Brandt and Daroff, 1980). Children are most susceptible to motion sickness from 2 to 12 years of age with a subsequent gradual decrease in symptoms (Lentz and Collins, 1977; Brandt and Daroff, 1980; Sharma, 1980; Kuritzky, Ziegler and Hassanien, 1981). Decrease in motion sickness with age is attributed to experience moderating the vestibular response and to the development of central inhibition (Groen, 1963) with improvement in vestibulo-ocular reflex suppression (Hood, 1980; Fluur, 1983). Motion sickness is more prevalent in children who suffer from migraine (Toglia, Thomas and Kuritzky, 1981; Del Bene, 1982). In addition, visual vertigo and vestibular dysfunction contribute to motion sickness in this group.

**Vestibular stimulation**

Repetitive vestibular stimulation is pleasurable and calming to infants whether from self-stimulation or by those caring for the child. Self-stimulation in the form of body rocking
or head banging or rolling begins at about 6 months of age when the gain of the vestibulo-ocular reflex is at its highest (Ornitz, 1983). It does not usually last beyond the age of 2 years. Self-stimulation has been found to accelerate motor milestones (Sallustro and Atwell, 1978). Applied vestibular stimulation has been demonstrated to improve the development of ocular-motor skills and the integrative functions of the cerebellum (Clark, Kreutzberg and Chee, 1977; Weeks, 1979). In an older age group, vestibular stimulation affected the duration of postrotational nystagmus in three children with learning disability (Ottenbacher, 1982). It follows therefore that vestibular hypofunction is sometimes related to delay in motor milestones (Rapin, 1974; Eviatar and Eviatar, 1981), and is implicated in children suffering from cognitive, perceptual and attentional problems. This is discussed in more detail at the end of this chapter.

Symptomatology and history taking

Vestibular disorders may present with dysequilibrium or be asymptomatic (Brookhouser, Cyr and Beauchaine, 1982). In the young child vertigo frequently goes unrecognized. It should be suspected if the child lies face down wedged against the side of the cot with the eyes closed, not wanting to be moved (Farmer, 1964). Sudden falling or tipping over, with crying, pallor, and sweating are also indicative of vertigo (Basser, 1964). In older children vertigo should be suspected if they are unwilling to get out of bed after the acute phase of an illness has passed. Lack of vestibular function is usually symptomless as children have such good compensation for loss of vestibular input (Dix, 1948). It should, however, be suspected if there is inexplicable delay in the acquisition of motor milestones, learning to skate or cycle, or spatial disorientation in a darkened environment. Such children may be fearful of any situation in which the lights are turned out, as in a film show.

History

Acute episodic vertigo

Attacks of vertigo can begin in the second year of life before the child is able to describe the symptom, objective observations being all that is available. The infant may suddenly cry out in fear and drop to a crawling position, cling for support to any available object, or be observed to be ataxic. Nystagmus is sometimes reported, and the child is pale. If the attack lasts long enough, sweating or vomiting may occur. Once the child is a little older descriptions such as 'the skies are falling' or 'the world is going round' are given. Headache with the attack should be suspected if it is accompanied by screaming.

In children around the age of one year vestibular dysfunction can present as torticollis with abnormal tilting or twisting of the head. It is important to establish whether there is any loss of consciousness, however brief, during the attack or subsequent amnesia, or bizarre movements. The speed with which the attack comes on, its duration, and the manner in which the child returns to normal are often pathognomonic.

In addition to the description of the acute attack the following details must be established: accompanying symptoms, precedency, precipitating or aggravating events, frequency or periodicity of attacks, age of onset and sequelae. Accompanying symptoms to
be explored are hearing loss, tinnitus, headache, visual obscurcation, lassitude, photophobia, and neurological deficit such as ocular palsies, hemiparesis or paraesthesia.

Preceding and precipitating events to be elicited are as follows: exertion, barotrauma or head injury; changes in position; infections, especially otitis media, meningitis and viral illnesses. As pyrexial illness is so common in children, care must be taken to establish a causal relationship. For example, labyrinthitis occurs with, or immediately after, a viral illness whereas cerebellar ataxia is delayed by 7-10 days.

For recurrent attacks of vertigo the chronology of symptoms and their periodicity must be established as some vertiginous disorders of childhood are strikingly periodic. The vertigo syndromes of childhood tend to be different according to the age of the child. For example, paroxysmal torticollis is a disorder of the first 3 years of life; benign paroxysmal vertigo is usually confined to the second to tenth years, and basilar artery migraine begins in the first year of life and subsequently changes to common migraine. When confronted with one form of vertigo the presence of any pre-existing forms should be explored. The acute attack of vertigo must be distinguished from other recurrent paroxysmal epileptic and non-epileptic disorders (Collins, 1983/84) which are presented in detail in the section on differential diagnosis.

**Persistent vertigo, imbalance or motor delay**

Dysequilibrium due to primary vestibular deficit must be distinguished from ataxia due to central nervous system disease (Harrison, 1962a; Beddoe, 1977; Bussis, 1983). Whereas delay in the acquisition of motor milestones or balancing skills can result either from poor sensory input or central nervous system immaturity, deteriorating function is more likely to be due to posterior fossa disease (Curless, 1980). The distinction must therefore be made between delay, arrest or deterioration in motor and postural performance. The presence of any concomitant ocular-motor, visual or neurological deficit is ascertained. If there is no imbalance in daylight, specific enquiry should be made regarding the child’s performance in the dark. A history of fear of the dark or of being blindfolded can be helpful. Some children with vestibular disorders will be afraid of swings and roundabouts and of rough play.

Small children frequently fall, but children with vestibular disorders may be noticed to be more clumsy and fall more frequently than their peers. Distinction is made between unsteadiness on rapid movement and ataxia of gait. The presence or absence of motion sickness is relevant. As bilateral vestibular weakness is usually symptomless a high index of suspicion is required.

**General health**

The development, visual, and hearing status of the child is ascertained. The presence of any middle ear, renal or endocrine disease, serious medical disorder or any physical deformity is established. The child’s personality is relevant to migrainous disorders and also to hyperventilation. Problems with interpersonal relationships and school performance can lead to anxiety.
The frequency, location and duration of headache or vomiting is recorded in the same manner as for the attack of vertigo. Repeated physical trauma or exposure to toxic chemicals could cause dizziness. A detailed drug history, including commonly prescribed drugs for minor disorders, for example piperazine for infestation, is mandatory.

**Past history**

The health of the mother during pregnancy with regard to vial illness and also to systemic illness requiring the administration of ototoxic antibiotics (McCracken, 1976; Wright et al, 1982) is explored. In the perinatal period prolonged or repeated anoxia will result in brain damage rather than vestibular destruction. The administration of aminoglycoside antibiotics in association with loop-inhibiting diuretics is associated with vestibular and cochlear ototoxicity (Finitzo-Hieber et al, 1979; Camarda et al, 1981; Davis et al, 1982; Finitzo-Hieber, McCracken, and Brown, 1985). In childhood a history of trauma, surgery, infectious diseases, viral illness, meningitis or the administration of ototoxic antibiotics is noted.

**Development history**

Motor milestones, social and speech development are all relevant to vestibular function. School performance and learning ability are ascertained.

**Family history**

Enquiry should be made after the following familial disorders: migraine, seizures, deafness, endocrine disease, renal disease, motion sickness, and syndromes with aural manifestations or ataxia. Infections which cross the placental barrier, such as syphilis, may affect a number of siblings. The general state of health of the family is important as other members of the family will have been exposed to the same environment. The possibility of consanguinity is explored.

**Physical examination**

The examination of the child suspected of vestibular deficit or dysfunction must include complete otolaryngological, neurological and neurodevelopmental investigation. Particular attention is required in the following areas:

*The ear:* the tympanic membrane, which should preferably be examined under the operating microscope, noting any congenital abnormality of the external ear, however minor.

*The nose and throat:* infection, congenital abnormality, the state of the airway.

*Hearing and speech:* an objective assessment using distraction or toy tests of hearing and listening to the child's expressive language. For details the reader is referred to other chapters in this volume. The parents' subjective report is insufficient.

*Tuning fork tests.*
Eye movements: the cover test for latent strabismus, in addition to pursuit, convergence and nystagmus.

Postural control: righting reflexes, positional nystagmus, hopping, heel-toe walking, kicking a ball, gait and stance with eyes open and closed, head tilt, asymmetry of movement, removal of proprioceptive input.

Neurological: hand-eye coordination, cranial nerves, tendon reflexes, tone, developmental abnormalities, congenital defects and syndromes, abnormal movements.

Vision: visual acuity, ophthalmoscopy, heterochromia.

General examination: congenital defects, pigmentary disorders, musculoskeletal defects and disorders, heart sounds.

Investigation

In many instances it is possible to reach a diagnosis on the basis of careful history and detailed examination. Most of the causes of vertigo in childhood are not associated with any abnormality on investigation, which can be unproductive if not used selectively. Audiovestibular and electroencephalographic investigation frequently provide useful information and, in some instances, X-ray and laboratory tests are required. Referral for a paediatric opinion is often advisable.

Auditory tests

Pure-tone audiometry is essential in all children capable of giving reliable responses and is supported by speech discrimination tests. Impedance testing should also be routine.

Electrophysiological tests

An EEG should be performed whenever there is any possibility of altered consciousness associated with vertigo. It is also helpful in cases of persistent or progressive dysequilibrium. Evoked response audiometry may be required to assess hearing, but also provides information on neurological status.

Vestibular tests

Electronystagmography allows evaluation of vestibular function without the influence of visual and proprioceptive input. Providing the following precautions are taken to allow for the differences encountered when testing children, electronystagmography and the bithermal caloric test can be undertaken even in very young children.

(1) To overcome vestibulo-ocular reflex suppression by visual fixation, caloric or rotation responses should be measured in total darkness with mental alerting. An infra-red viewer is required to ensure that the eyes remain open and the child alert.
Compared with adults the caloric nystagmus of children is of lower frequency and greater amplitude. Although the mean maximum velocity of the slow phase is similar, the variance is higher and the response is often dysrhythmic. Calculations for canal paresis and directional preponderance should take this wide range of normal values into account. Published data for caloric responses in children are anomalous as illumination and recording conditions are not uniform. Each laboratory should therefore define its own normal data for the bithermal caloric test. Although convenient, the iced water caloric test is not recommended for children as it is distressing and is less accurate. As a small ear canal can cause inequality of irrigation, the volume should be measured after each caloric test.

(2) If electronystagmography is used to record the response, a DC machine will minimize the likelihood of adventitious eye movements masquerading as nystagmus (as may occur with AC equipment).

(3) Acceptance of the test is helped by the display of a photograph of a child wearing electrodes. Throughout the procedure the child should be included in the conversation as any sense of isolation produces anxiety.

(4) Stability of gaze for recording spontaneous nystagmus is improved by proprioceptive clues.

(5) Attention for smooth pursuit tracking is maintained by the use of an intermittent light source.

(6) Optokinetic tracking can be recorded from birth provided a full field stimulus is used. Attention for the small optokinetic drum is so difficult that results are poor even in children 5-15 years old.

(7) Rotation responses either to sinusoidal harmonic rotation or to ramp acceleration and deceleration are of greater gain in children than adults, with rather less fixation suppression. The test is well tolerated and gives invaluable information on the overall vestibular status of the child.

Vestibulospinal stability test: this new technique of quantifying body sway gives a rapid objective recording of stability and has a place in the screening of children where more demanding investigations are not tolerated.

Radiology

Plain skull films and mastoid X-rays are performed if indicated. Inner ear abnormalities which are detectable on petrous bone tomography are likely to be associated with vestibular abnormality, if not total lack of function. Tomography is therefore more rewarding than in the investigation of straightforward genetic cochlear deafness. Computerized tomographic scanning is obligatory where there is any suspicion of central nervous system disorder causing unsteadiness, particularly if it is progressive.
Urinalysis

This is indicated for estimation of protein and red blood cells in children with sensorineural deafness and vestibular disorder. It should be repeated at yearly intervals as renal involvement can be progressive.

Blood tests

Serology: screening for congenital syphilis is mandatory in all children with progressive cochlear and vestibular dysfunction in the first or second decade. In young children a viral screen is advisable if congenital deafness is present.

Blood chemistry: sugar, creatinine, calcium phosphate, electrolytes, T3 and T4 are advocated.

Haematology: full blood count, erythrocyte sedimentation rate (ESR), and autoimmune profile are indicated when vertigo is accompanied by fluctuating deafness.

Electrocardiography is indicated in congenital deafness and vestibular deficit of unknown aetiology to detect those syndromes with cardiac and labyrinthine dysgenesis.

Pathological vertigo syndromes

Vestibular disorders with auditory symptoms

External meatus

Impacted cerumen may cause dysequilibrium.

Middle ear cleft

Otitis media with effusion

This condition usually gives rise to no more than mild positional vertigo or occasional slight dizziness. There are, however, reports of balance disturbance and episodic rotational vertigo which respond to the insertion of tympanostomy tubes (Clinical Otolaryngology, 1978; Fried, 1980; Blaney, 1983).

Blaney reported serous otitis as the cause for five out of 25 children presenting to the clinic with rotational vertigo. Busis (1983) regarded eustachian tube dysfunction as the commonest cause of vestibular disturbance in children. Markedly negative middle ear pressure in association with thick mucus in the middle ear cleft may cause dramatic attacks of transient rotational vertigo accompanied by pallor and nausea, which are not precipitated by movement and with little change in hearing.

Alternatively, parents of preschool children with serous otitis not infrequently notice that the child 'walks clumsily', 'falls all over the place' or 'walks into things'. They may query whether he or she has adequate vision to account for this. These symptoms of imbalance
respond immediately to correction of middle ear pressure. In the author's experience 50% of children with serous otitis may have some balance disturbance. This responds to correction of the underlying condition in one-half of those affected. The presence of balance disturbance should weight the scales in favour of surgical intervention for serous otitis even if the hearing is relatively unaffected.

**Suppurative otitis media**

Either acute or chronic suppuration may serve as an irritative or invasive focus involving the labyrinth. Some of the topical antibiotics used for this condition are also vestibulotoxic. In spite of this, suppuration relatively rarely causes vertigo.

**Cholesteatoma**

Both congenital (Schwartz et al, 1984; Wang et al, 1984) and acquired cholesteatoma may invade the labyrinth and give rise to vertigo. In any child with a discharging ear or attic disease in association with vertigo the fistula test should be performed.

**Labyrinthine**

**Congenital vestibular deficit**

Of all the congenital sensory deficits this is the most silent. It is usually associated with congenital deafness, but vestibular hypofunction may also contribute to the handicap of other disorders such as Down's syndrome (Zarnoch, 1980). When congenital deafness is accompanied by episodic vertigo or abnormal vestibular responses, polytomography may reveal an enlarged vestibule with dilatation of the ampullated portions of the horizontal and superior semicircular canals, in addition to an abnormal cochlea (Hill, Freint and Mafee, 1984; Brama, 1985). Congenital vestibular deficit with deafness is more often symptomless and only detected by examination.

There have been a number of studies of vestibular function in deaf school children (Arnvig, 1955; Sandberg and Terkildsen, 1965; Swisher and Gannon, 1968; Brookhouser, Cyr and Beauchaine, 1982). In an unselected population in schools for the deaf the incidence of reduced or absent caloric or rotation responses varies from 10 to 36% depending on the technique used. Brookhouser, Cyr and Beauchaine (1982) demonstrated that caloric responses are sometimes suppressed in children. They recommend that results be recorded with eyes open in the dark and with a mental alerting task, using sign language where required. In this manner they found 22% to have caloric abnormalities. These were correlated with results of the tandem Romberg test in the Jendrassik position.

Although not correlated with the severity of the deafness the presence of caloric weakness is useful for the determination of aetiology and therefore prognosis. Arnvig (1955) found caloric reduction most prevalent in those with deafness due to meningitis (91%) and those with retinitis pigmentosa, who accounted for one-half of the children with reduced vestibular function. Kumar, Fishman and Torok (1984) reported that vestibular deficit in Usher's syndrome is found in that variety of the disease that has the worst prognosis for visual
deficit. Karjalainen et al (1985) postulated that vestibular tests may facilitate the detection of heterozygote carriers of Usher's syndrome with central vestibular abnormalities.

Waardenburg's syndrome accounts for 2% of all congenital deafness and 30% of these patients will also have congenital vestibular failure (Schweitzer and Clack, 1984). Patients with Waardenburg's syndrome and vestibular disorder experience dizziness or episodic vertigo rather than delayed milestones or imbalance. In Hurler's syndrome there is disruption of the statoacoustic nerve by deposits of Hurler cells between the nerve fibres (Friedman et al, 1985). This disruption may cause a retrocochlear auditory and vestibular deficit in addition to the degeneration of the organ of Corti.

A congenital vestibular disorder may therefore be anticipated in those cases of congenital deafness which are associated with other defects particularly of the head and neck, vision, or petrous temporal bone.

Apart from prognosis and diagnosis vestibular assessment allows better rehabilitation. Harris et al (1984) linked vestibular weakness in congenital deafness due to cytomegalovirus with delayed motor milestones and imbalance. Brookhouser, Cyr and Beauchaine (1982) recommended that all deaf children have vestibular assessment for recreational and occupational counselling. They suggested that this be performed early in those with deafness greater than 110 dB as vestibular weakness is more likely in this group.

There is no advantage in caloric assessment for the purposes of aural rehabilitation using 'vestibular hearing' (Swisher and Gannon, 1968) as the caloric test stimulates the lateral semicircular canal. It is the saccule that is responsible for vestibular hearing (Lenhardt, 1985). This is one explanation of the better hearing in those with normal vestibular function.

**Trauma**

Head injury produces vertigo by perilymph fistula (see below), or by labyrinthine concussion with or without fracture of the temporal bone. Vertigo may be short-lived even if destruction of the labyrinth is complete as children adapt quickly to altered vestibular function. Recording nystagmus in the absence of visual fixation soon after the injury is important for medico-legal reasons as vestibular responses may remain abnormal permanently.

It is possible to lose vestibular function without deafness in a longitudinal fracture. Vartiainen, Karjalainen and Karja (1985) reported one such case of vertigo in 61 children following acute blunt head injury. This child had vertigo due to a traumatic perilymph fistula. The incidence of sensorineural deafness was 7%. Perilymph fistula may follow even minor trauma to the head.

**Infection**

Bacterial infection is transmitted to the labyrinth from the middle ear (see above) or via the cochlear aqueduct from meningeal infection. Deafness due to meningitis is accompanied by loss of vestibular function in 66% (Arnvig, 1955). Infection with congenital syphilis presents with deafness, tinnitus and vertigo in a pattern not dissimilar from endolymphatic hydrops (Wilson and Zoller, 1981). Viral labyrinthitis is more usual, the
pathogenesis being the same as in adults via the modiolus with the difference that it is easily overlooked (Tieri et al, 1984). Cochlear function may be affected. The child is often too young to describe the symptoms and the lack of mobility is ascribed to the causative illness. The vertigo subsides in a few days without recognition of labyrinthine involvement (Hyden, Odkvist and Kyle, 1979). Subsequent caloric weakness may be demonstrated even in the absence of vertigo. The viruses most often associated with labyrinthitis in adults and children are mumps, measles, varicella zoster and cytomegalovirus (Davis and Straus, 1973; Straus and Davis, 1973; Ronson and Hinchcliffe, 1976; Karmody, 1983). Epidemic labyrinthitis does not involve the cochlea.

Ménière's disease

Ménière's disease is uncommon in children, accounting for only 2% of Harrison and Naftalin's (1968) series of 423 cases with this condition. Earlier reports of childhood Ménière's disease include those of Brain (1938), Crowe (1938), Simonton (1940), Ombredanne and Aubry (1941), Ford (1944), Cawthorne (1954), and Golding-Wood (1960). Some of these were traced by Meyerhoff, Paparella and Shea (1978) when they presented a series of eight cases. Parving (1976), Beddoe (1977) and Sade and Yaniv (1984) documented an additional seven cases. Filipo and Barabara (1985) reported that 7% of referrals for Ménière's disease are under the age of 20 years.

From the published data, Ménière's disease would appear to be almost twice as common after the age of 10 years than it is in the younger children as mentioned by Gates (1980). Children presenting with deafness, tinnitus and vertigo merit a comprehensive search for aetiological factors. Adrenal, pituitary and thyroid gland insufficiency, allergy, autoimmune disease, syphilis, trauma, perilymph fistula, acoustic neuroma and congenital abnormalities of the inner ear, petrous temporal bone and base of skull should be considered (Meyerhoff, Paparella and Shea, 1978; Filipo and Barabara, 1985).

A large vestibular aqueduct has been associated with congenital sensorineural deafness and vertigo (Valvassori and Clemis, 1978; Hill, Freint and Mafee, 1984) but Rizvi and Elliott Smith (1981) found no significant correlation between a large vestibular aqueduct and endolymphatic hydrops when comparing this population with a control group.

The criterion for diagnosing Ménière's disease in childhood is no different from that in adults; namely a predominantly low tone cochlear deafness with tinnitus and episodic rotational vertigo. Between attacks vestibular function may return to normal (Busis, 1983). Some children with Ménière's disease in infancy have progressive sensorineural deafness (Sade and Yaniv, 1984), but others go into spontaneous remission with no residual cochlear or vestibular deficit. Harrison and Naftalin (1968) regarded this group as being 'pseudo-Ménière's', but it is possible that this is remission rather than recovery. Although Meyerhoff, Paparella and Shea (1973) regarded saccus decompression more successful than medical therapy in children, Filipo and Barabara (1984) reported a good response to diuretics in three out of five children. Other forms of medical therapy have not proved beneficial, with the exception of steroids for autoimmune and inflammatory endolymphatic hydrops (Bachynski and Wise, 1984; Hughes et al, 1984).
Of the 15 children with Ménière's disease under the age of 10 years reported in the literature five are known to have bilateral disease; a much higher proportion than in the older children (2/21). This reported high proportion of bilateral disease in the very young is in agreement with the author's own experience.

**Perilymph fistula**

Perilymph fistula is an important cause of sudden, fluctuating or progressive hearing loss associated with tinnitus, vertigo or imbalance. Unfortunately, it is extremely difficult to diagnose without surgical exploration of the ear (Halvey and Sade, 1983). A high index of suspicion for this disorder is required if surgical closure of the leak is to stabilize the hearing, abolish vertigo and prevent recurrent meningitis (Grundfast and Bluestone, 1978; Healy, Friedman and Ditroia, 1978; Althaus, 1981).

In children fistula is often associated with congenital abnormalities of the head and neck, for example craniosynostosis, Pendred's syndrome, Klippel-Feil syndrome, a large cochlear aqueduct, Mondini defect, or lateral dilatation of the internal acoustic meatus. It may be precipitated by such events as barotrauma, exertion or a Valsalva manoeuvre (Althaus, 1981; Milner et al, 1983; Supance and Bluestone, 1983; Weider and Musiek, 1984; Cremers et al, 1985). Malformation of the round window niche associated with perilymph leak can be present without any radiological evidence of labyrinthine deformity (Pashley, 1982) and on tympanotomy the round window is often obscured by mucosal folds (Flood et al, 1985).

In the absence of a clear history of trauma, exertion or surgery, this condition may be confused with other causes of progressive or sudden deafness. Halvey and Sade (1983) were unable to identify any significant preoperative differences between those with and without fistula at operation. A number of tests have been advocated to aid diagnosis. A unilateral sensorineural hearing loss with poor speech discrimination and a change in hearing thresholds in the recumbent position should alert the clinician (Busis, 1983; Flood et al, 1985). Very small changes in threshold are, however, difficult to establish in children and transtympanic electrocochleography may be required for diagnosis.

Vestibular function is often deranged in perilymph fistula. This can be documented by recording spontaneous or positional nystagmus, posturography, and the fistula test. The latter was thought to be more specific for perilymph leak than more general evidence of peripheral vestibular disorder when recorded with the eyes closed (Daspit, Churchill and Linthicum, 1980). Results have, however, been disappointing as the fistula test recorded by electronystagmography can activate a latent spontaneous nystagmus that is not necessarily due to a perilymph leak. Supance and Bluestone (1983), in a large study of perilymph fistulae in children, confirmed by tympanotomy, found that the fistula test did not correlate with the operative findings. Exploratory tympanotomy was carried out in 33 infants and children (44 ears) identifying a perilymph leak in 29 ears. Middle ear anomalies were found in 20 ears. Preoperative factors determined to be highly suggestive of perilymph fistula were sudden onset of sensorineural hearing loss, congenital deformities of the head, and abnormal findings on tomography of the temporal bones, especially Mondini-like inner ear dysplasias.

The vestibular abnormalities most frequently associated with perilymph fistula were positional nystagmus and abnormal platform posturography. Vertigo was present in 10 of the
33 children explored, of whom were found to have a fistula. Perilymph gushers during surgery for congenital stapes fixation may be anticipated in X-linked progressive mixed deafness; abnormal vestibular symptoms and signs and abnormalities on polytomography of the temporal bones may be found (Cremers et al, 1985).

In summary the features suggestive of a perilymph fistula are:

**History**

- Head injury, barotrauma, sneezing, Valsalva manoeuvre, laughing, blowing a wind instrument.
- Sudden or fluctuating sensorineural deafness.
- A sensation of ‘pop’ in the ear followed by deafness or vertigo.
- Tullio phenomenon.
- Continuous dizziness increased by postural change.
- A past history of recurrent meningitis.

**Findings**

- Spontaneous or positional nystagmus.
- Positive Romberg test, including posturography.
- Unilateral sensorineural hearing loss, change of hearing with position.
- Congenital abnormalities of the head and neck including minor abnormalities of the pinna.
- Abnormal petrous bone tomography, especially Mondini dysplasia, dilatation of the distal end of the internal auditory canal or an abnormally large vestibule.

**Ototoxic drugs**

Whether or not the aminoglycoside antibiotics used in the neonatal period cause significant ototoxicity is a debate which remains to be fully resolved. Animal studies suggest that young animals are more susceptible to vestibular and cochlear toxicity than are adults of the same species (Prieve and Yanz, 1984). Prospective, long-term studies of infants who received aminoglycoside antibiotics in the neonatal period have failed to provide convincing evidence that, in those children subsequently proved to be deaf or partially hearing, ototoxicity was the sole factor (Finitzo-Hieber et al, 1979; Finitzo-Hieber, McCracken and Brown, 1985). McCracken (1976) and Elfving, Pettay and Raivio (1973) similarly found it difficult to evaluate a cause-effect relationship with regard to vestibular deficit in two out of 13 infants treated with gentamicin.

As the incidence of vestibular ototoxicity is only of the order of 1% (Noone, 1982) large numbers of subjects are required if statistically significant differences are to be shown between groups. In spite of this difficulty Eviatar and Eviatar (1981, 1982), in a prospective study of 43 infants treated with aminoglycosides compared with 250 control preterm infants, found abnormal vestibular test results only in the treated infants. Head control was delayed in two of 26 treated with kanamycin, eight of 17 treated with gentamicin and only one untreated control. All except the one control baby and one of the gentamicin group had positional nystagmus with additional abnormalities of rotation of caloric responses. The
caloric abnormality was unilateral in six of the infants. Only two of the infants with vestibular dysfunction had an associated hearing loss. Delayed head control usually arises from cerebral palsy and it is therefore important to know whether vestibular function is normal so that a realistic prognosis can be given.

These recommendations were in agreement with those of Camarda et al (1981), demonstrating vestibular deficit in 52 subjects aged 2-26 years with ototoxic hearing loss and delayed motor control. The new antibiotic, netilmicin, would appear to have a far lower incidence of ototoxicity as only one case was found out of 804 neonates, infants and children treated (Chiu et al, 1983). The 572 neonates in this study were evaluated for ototoxicity by brainstem evoked responses in the neonatal period only and it is therefore possible that vestibular toxicity was missed in this age group. The only case of toxicity reported was a 14-year-old girl with vertigo, tinnitus and deafness.

**Perinatal risk factors**


Six factors were identified:

1. hereditary tendency to deafness
2. rubella or other virus infection in pregnancy
3. malformations of the ear, face, syndrome-like appearance, chromosome defects and alcohol fetopathia
4. asphyxia requiring more than 10 minutes resuscitation and/or intensive care treatment
5. very low birthweight infants
6. neonatal sepsis/meningitis.

All these factors will predispose the infant to ototoxicity, both vestibular and cochlear, making cause-effect difficult to disentangle. Ototoxicity is related more to total dose, and slow clearing from perilymph than it is to peak levels of drug concentration (Ohtani et al, 1982).

In older children there are a number of conditions in which treatment with an ototoxic antibiotic with or without loop inhibitor diuretics is likely (Davis et al, 1982), for example cystic fibrosis, severe burns, renal failure. Monitoring of patients receiving aminoglycoside antibiotics is usually achieved by pure-tone audiometry and measurement of blood levels. Although, ideally, caloric testing should be performed, the condition of the patient may make this impractical. There is the additional problem that patients become adapted to the caloric stimulus if it is performed frequently. Longridge and Mallinson (1984) have described a method by which the vestibulo-ocular reflex can be used as a screening test which correlates with caloric responses in those patients capable of discriminating the letter 'E'.

Neurotoxic chemicals such as solvents should also be considered in the aetiology of vestibulo-oculomotor dysfunction (Odkvist et al, 1982).
Retrolabyrinthine

Lesions of the eighth cranial nerve give rise to progressive deafness, tinnitus and imbalance. In childhood and adolescence these symptoms are more likely to be due to progressive, genetic, infective, or hydropic disorders of the labyrinth. Meningitis can give rise to audiovestibular deficit by infection around the statoacoustic nerve within the internal auditory meatus (Keane et al, 1983). Benign cerebellopontine angle tumours do occasionally occur in children, particularly those with a family history of von Recklinghausen's disease (see below). Each case of asymmetrical progressive sensorineural deafness or imbalance should therefore be investigated on its merits if these rare conditions are not to be overlooked.

Vestibular disorders without other ear symptoms

Referral of children with vertigo to an otologist is relatively rare. In a recent survey of 175 children aged 10-20 years seen in an otolaryngology clinic over a 33-month period, Ruben (1985) reported that none presented with vestibular symptoms. Blaney (1983), also reporting referrals to an otolaryngology clinic, listed 27 children presenting with dizziness in a period of 18 months. Children may not complain of vertigo or imbalance, either because they are too young or because they do not realize that it is an abnormal symptom (Gates, 1980). The recognition that the child's symptoms are due to vertigo often depends upon correct interpretation of the parent's description of the child's behaviour. Children with vertigo are therefore usually referred to paediatric clinics rather than an otolaryngology clinic. The disorders described in this section are not thought to arise primarily in the labyrinth but, nevertheless, cause rotational, episodic vertigo. Both benign paroxysmal vertigo of childhood and benign paroxysmal torticollis are regarded as migraine equivalents by many authors (Sanner and Bergstrom, 1979; Eeg-Olofsson et al, 1980; Koehler, 1980).

Benign paroxysmal vertigo of childhood

First described by Basser in 1964, this disorder is characterized by brief, sudden and severe episodes of spontaneous vertigo in otherwise healthy children. There are concomitant autonomic symptoms such as pallor, sweating, and occasionally vomiting. The onset is abrupt, causing confusion with seizures, but there is no loss or alteration of consciousness. Typically the child suddenly cries out with fear, drops to all fours or clings for support, and remains acutely unsteady for 30-60 seconds. He/she rapidly returns to complete normality within at most a few minutes and resumes whatever he/she was previously doing. Nystagmus may be observed during the attack, and the children, who are frequently articulate for their age, describe 'the world going round' or 'the walls falling down'. The attacks are recurrent, of variable frequency, and there are no precipitating factors or sequelae. Attacks typically commence before the age of 4 years (Fried, 1980), and disappear spontaneously in a matter of months or years. The paroxysms can occasionally occur in older children or the young teenager (Busis, 1983).

Almost one-half of the affected children go on to develop migraine in adolescence (Koenigsberger et al, 1970; Chutorian, 1972; Koehler, 1980). Most children have a family history of migraine (Koenigsberger et al, 1970; Eeg-Olofsson et al, 1980; Koehler, 1980; Mira et al, 1984). Sometimes the attacks are associated with headache and photophobia (Mira et al, 1984). Early studies of caloric tests in children with benign paroxysmal vertigo were
performed with the eyes open and fixed on a target. Caloric abnormalities were common compared to a control group (Koenigsberger et al, 1970; Koehler, 1980). Studies of caloric responses in the absence of fixation using electrondystagmographic (ENG) recording gave normal results between attacks (Eeg-Olofsson et al, 1980; Mira et al, 1984).

The paroxysm are distinguished from other causes of episodic vertigo by the absence of auditory symptoms, and the complete absence of abnormal findings between attacks, especially the EEG and X-ray studies. Although Beddoe (1977) and Busis (1983) recommend that the diagnosis of benign paroxysmal vertigo of childhood should not be made unless there is evidence of residual vestibular disturbance on the caloric test, this is not necessarily the case. As some reports found normal caloric responses in patients with this condition the presence of persistent vestibular disorder should alert the clinician to the possibility of recurrent perilymphatic fistula, Ménière's disorder, or posterior fossa pathology.

**Benign paroxysmal torticollis**

First described by Snyder in 1969 this condition commences before the first birthday and recovers spontaneously by the age of 5 years. The paroxysms are characterized by head tilting to one side which persists for hours or days. The symptom is worse in the upright position and attempts to right the head are met with resistance. Usually the child is not distressed but the attacks may be accompanied by pallor, agitation or vomiting. Ataxia may accompany the attack which may be preceded by rolling of the eyes. There is spontaneous and sudden recovery. Of 10 children described by Dunn and Snyder (1976), one progressed to develop migraine. Food allergies to milk or chocolate are also reported in this group. Both Dunn and Snyder (1976) and Sanner and Bergstrom (1979) believed this condition to be a variant of benign paroxysmal vertigo of childhood and to be related to migraine. Once again the children tend to be articulate and, once they are old enough, will describe 'the house turning'.

**Migraine: classical and basilar artery**

Migraine is a hereditary disorder affecting 5% of schoolchildren (Fried, 1980). In a study of 386 children with this condition, Watson and Steel (1974) found 43 to have vertigo of whom 23 did not have an associated headache. A diagnosis of migraine without headache is tenable if the patient has transient neurological disturbances as well (Busis, 1983). In childhood, the manifestations of migraine are more protean than in adults, and it is only as they mature that the symptoms change to include paroxysmal headache (Watson and Steel, 1974).

Classical migraine typically begins in the 5-15 year range (Watson and Steele, 1974), but in basilar artery migraine the onset of symptoms can be in the first 3 years of life (Golden and French, 1975; Eviatar, 1981). At this age the child is too young to complain of either vertigo or headache and the diagnosis must be made on the parents’ observations. Typically the attacks last 1-3 days during which time the infant becomes progressively lethargic, anorexic and photophobic. Initially unsteady, the infant eventually prefers to lie undisturbed, wedged against the side of the cot with the eyes tightly closed. Picking up the child may induce vomiting. Having had disturbed nights the child finally awakes screaming with pain, clutching at the head or pulling his hair. The attack then subsides over the next 24 hours and
the child returns to normal. Symptoms may last only a matter of hours, but in some cases neurological sequelae can last for weeks. The attacks are characterized by transient neurological disturbance and visual obscuration. Ataxia, ocular palsies, hemiparesis, facial palsy, drop attacks and blindness are likely to be present.

In basilar artery migraine there may be bilateral weakness or paraesthesia of the limbs, tinnitus and occasionally transient deafness. The attacks are separated by symptom-free intervals and may be remarkably periodic (Ouvrier and Hopkins, 1970; Watson and Steel, 1974; Golden and French, 1975; Eviatar, 1981).

Children at risk for migraine have a strong family history of the condition (Watson and Steel, 1974; Eviatar, 1981; Busis, 1983). Children with a previous history of motion sickness, vertigo, dizziness, visual vertigo, vomiting attacks, and periodic abdominal pain are at risk of developing classical migraine with headache (Papatheophilou, Jeavons and Disney, 1972; Kuritzky, Zeigler and Hassanein, 1981; Del Bene, 1982). Caloric abnormalities are present in a proportion of children with basilar artery migraine and adults with classical migraine (Eviatar, 1981; Toglia, Thomas and Kuritzky, 1981). Watson and Steel (1974) found no EEG abnormality in children with migraine, but Eviatar reported a significant number of children with vertigo and EEG abnormalities.

The abrupt onset of both headaches and vertigo in migraine are similar to the onset of these symptoms in epileptic disorders. Seizure headaches may be distinguished on the EEG evidence but also on the family history. Children with this condition have a family history of seizures rather than migraine (Swaiman and Frank, 1978).

**Vestibular neuronitis**

First described by Dix and Hallpike in 1952, vestibular neuronitis consists of an episode of vestibular failure characterized by rotational vertigo, vomiting and autonomic disturbance in association with an upper respiratory infection. The acute symptoms last days to weeks and then gradually subside. Approximately 50% of those affected have only one attack. Over the ensuing months to years the remaining 50% experience gradually decreasing episodes of vertigo precipitated by movement or lightheadedness. Children recover more quickly from this disorder than do adults (Gates, 1980).

Vestibular neuronitis is a disease of young adults, but may affect any age group. Harrison (1962b) reported three cases under the age of 20 years out of a total of 67. In a series of 50 children referred with vertigo Eviatar and Eviatar (1977) found vestibular neuronitis in five children. In a similar series of 27 children Blaney (1983) was unable to make this diagnosis in any child. Out of a total of 28 children presenting with vertigo to the present author over a period of 2 years, one case was thought to be due to this condition. Beddoe (1977) regarded vestibular neuronitis as common over the age of 10 years.

**Cervical vertigo and odontogenic vertigo**

Cervical vertigo is thought to arise from disordered input from the cervical nerves to the vestibular nuclei and is extremely uncommon in childhood. The mechanism of odontogenic vertigo arising from irritative foci in the maxilla, mandible or temporomandibular
joint is less clear, but impacted wisdom teeth should be considered as a cause of dizziness in young people (Eidelman, 1980).

**Benign paroxysmal positional vertigo**

In contrast to benign paroxysmal vertigo of childhood, benign positional vertigo is a purely peripheral labyrinthine phenomenon. It may be caused by head or whiplash injury, or arise spontaneously. In children it is usually caused by trauma. If positional vertigo arises spontaneously in a child it is often of the central type rather than the benign paroxysmal peripheral type (Curless, 1980). Eadie (1967) found that out of 115 cases with benign positional vertigo 9% were in the age group 11-12 years.

**Metabolic causes of imbalance**

**Hyperlipidosis**

Cochlear vessels involute from fetal life onwards (Johnsson and Hawkins, 1972). Hyperlipoproteinaemia is though to accelerate presbyacusis, but this is not a significant factor in children except in familial forms of this condition. Type II hyperlipoproteinaemia develops in infancy and is heralded by xanthomata in the tendons of the hands and feet. Hyperlipoproteinaemia can cause inner ear dysfunction characterized by the symptoms of Ménière's disease which improve on appropriate management (Pillsbury, 1981).

**Hypothyroidism**

Although this is an important cause of imbalance in the elderly, it is unlikely to give rise to symptomatic unsteadiness in children. Vestibular function may be found to be abnormal in cases of congenital deafness and myxoedema (Pendred's syndrome).

**Central vestibular disorders**

**Congenital anomalies of the skull base**

Structural anomalies of the upper cervical vertebrae and foramen magnum may present with ataxia or vertigo resulting from pressure on, and stretching of, the cerebellum, brainstem and lower cranial nerves. Platybasia is a familial disorder that produces upward displacement of the floor of the posterior fossa and narrowing of the foramen magnum. Neurological symptoms do not appear until the second or third decade and include progressive spasticity, incoordination, nystagmus and weakness of the lower cranial nerves. It may be associated with other malformations of the central nervous system including the Chiari malformations.

The Chiari malformations are characterized by cerebellar elongation and protrusion through the foramen magnum into the cervical spinal cord. In type I malformation the cerebellar herniation exists alone or with malformations of the base of the skull such as platybasia, basilar impression, or Klippel-Feil syndrome. Type I is often asymptomatic in childhood but may become clinically apparent in adolescence with hydrocephalus, signs of cervical cord compression, suboccipital headache, vertigo, laryngeal paralysis and progressive cerebellar signs. Downbeating vertical nystagmus has been reported to be associated with this
condition. Other central vestibular signs found on electronystagmographic recording such as saccadic smooth pursuit, optokinetic disruption, ocular dysmetria and failure of fixation suppression are present, but are not as diagnostic of Chiari malformation (Chait and Barber, 1979).

Type II Chiari malformation is the most common form of this condition, comprising type I malformation together with non-communicating hydrocephalus and lumbosacral spina bifida.

Type III may have any of the features of types I and II with occipital cranial bifidum or cervical spina bifida.

Types II and III present early in childhood with widespread neurological abnormalities, whereas type I may present with vertigo or ataxia in adolescence, or simply a failure to learn to cycle or skate. Diagnosis is by computerized tomographic (CT) scanning of the skull base, or preferably by magnetic resonance scanning (Longridge and Mallison, 1984).

Klippel-Feil syndrome, characterized by fusion and reduction in number of the cervical vertebrae, causes variable neurological symptoms. It is commonly associated with other malformations including congenital deafness.

Hereditary cerebellar ataxias

Heredodegenerative diseases that involve the cerebellum and begin in childhood are uncommon. They present with slowly progressive ataxia, and posterior fossa tumours must be excluded before making this diagnosis. The more common familial cerebellar degenerations are ataxia telangiectasia, Friedreich's ataxia and Ramsay Hunt syndrome, of which Friedreich's ataxia is associated with congenital deafness. Refsum's disease is of more relevance to the otologist as it presents with cerebellar ataxia, deafness, retinitis pigmentosa and polyneuritis. The combination of night blindness, cerebellar ataxia and vestibular deficit causes progressive difficulty in walking. The onset of symptoms is usually between the ages of 4 and 7 years. It is due to an underlying disorder of lipid metabolism and is detected by the presence of lipiduria and raised serum phytanic acid. A phytol-free diet lowers the serum phytanic acid with improvement in neurological signs (Menkes, 1985). Inheritance is autosomal recessive.

In contrast to the heredodegenerative diseases, acute intermittent familial cerebellar ataxia is a self-limiting disease inherited by a dominant trait with variable penetrance. This disorder gives rise to acute episodes of ataxia beginning in the first 2 years of life, and disappearing around the age of 15 years. The attacks are characterized by sudden onset of gait and truncal ataxia with upper limb ataxia, intention tremor and dysarthria. Headache, vomiting, nystagmus and sometimes seizures may occur. In childhood the attacks last about 4 weeks, decreasing in duration with increasing maturity. Eventually the attacks are mild and last only days at a time. Afflicted members of the family are symptom-free between attacks. There are no skin or biochemical disturbances during the attack. It is postulated that the attacks result from toxic effects upon the immature cerebellum from infections including ascariasis (Hill and Sherman, 1968).
Hereditary disease of the peripheral and cranial nerves

The most common heredodegenerative condition is Charcot-Marie-Tooth disease. Peroneal muscular atrophy is the usual presentation of this disease (Menkes, 1985) but congenital sensorineural deafness is present in a proportion of cases (Cornell, Sellars and Beighton, 1984). As the deafness is retrocochlear it is likely to be accompanied by vestibular weakness. Neurofibromatosis affects both peripheral and cranial nerves. The optic nerve is the most common and earliest site of involvement; bilateral acoustic neuromata are less common. The presence of seizures in early childhood associated with café-au-lait spots and with or without subcutaneous neurofibromata, should alert the clinician to the possibility of intracranial tumours (Menkes, 1985). Congenital nystagmus is a recessive trait in which there is fine and rapid nystagmus, usually pendular, which is asymptomatic but may be associated with vertigo and ataxia (White, 1969).

Infection

Bacterial meningitis may present with ataxia or this symptom may develop during the course of the illness. Ataxia at the time of meningitis is not necessarily associated with deafness. In a study of six such children only one was left with sensorineural deafness (Schwarz, 1972). Infection from the meninges to the labyrinth takes place via the internal auditory canal causing a purulent labyrinthitis with residual deafness and dysequilibrium (Eavey et al, 1985). The imbalance resolves although the vestibular weakness will persist together with the hearing loss. In these patients, the labyrinth usually becomes ossified (Becker et al, 1984). Symptomatic dizziness can persist in the absence of deafness and be associated with central manifestations. Eviatar and Eviatar (1977) identified post-meningitis dizziness in three of a series of 50 children presenting with vertigo.

Brainstem encephalitis gives rise to either vertigo or ataxia. The symptom is persistent and is accompanied by fever and neurological signs such as supranuclear or internuclear ophthalmoplegia, vertical nystagmus and directional preponderance of induced nystagmus (Ellison and Hanson, 1977; Curless, 1980; Fried, 1980).

Cerebellar encephalitis may present a similar picture and is distinguished from acute cerebellar ataxia by the identification of a causative organism (Menkes, 1985). Acute cerebellar ataxia is prevalent in the first 3 years of life and follows 7-21 days after a non-specific infectious illness. There is sudden truncal ataxia with other neurological symptoms and signs. In the very young child it may be difficult to distinguish from acute labyrinthitis. Symptoms may persist for several months and, although it is a self-limiting condition, there are sometimes permanent sequelae.

Trauma

Dizziness and vertigo in the post-concussion syndrome are less common in children than in adults as the vestibular system is more plastic, but Eviatar and Eviatar (1977) reported four cases and Toglia, Rosenberg and Ronis (1970) 43 individuals under the age of 25 years in a series of 235 whiplash patients with closed head injuries. The numbers are small considering the frequency of closed head injury in the paediatric population. The possibility of a contrecoup injury to the temporal lobe vestibular cortex as it strikes the sphenoid ridge
should be considered if vertigo develops days or weeks after a closed head injury (Busis, 1983).

**Neoplasia**

Persistent, progressive ataxia or vertigo in the absence of pyrexial illness is likely to be due to posterior fossa neoplasia. The presence of other neurological symptoms and signs, particularly disorders of eye movement control point to a central rather than peripheral aetiology (Curless, 1980; Hood, 1980).

**Demyelination**

The clinical picture of multiple sclerosis as it occurs in children differs little from that seen in adults. The diagnosis is difficult to make in this age group as it depends upon long observation. The first attack is more likely to occur after, rather than before, puberty. Molteni (1977) reported vertigo as the presenting symptom in four out of 14 cases of childhood multiple sclerosis. Menkes (1985) found six with vertigo and 10 with dizziness and vomiting as presenting symptoms in 36 children with this condition. Both authors felt that this incidence was similar to that in adults.

**Seizure disorders**

Seizures give rise to vertigo in three ways: as an aura of a grand mal fit (de Jesus, 1980), as vertiginous epilepsy (Alpers, 1960), or as vestibulogenic epilepsy (Behrman and Wyke, 1958). Vertigo as an aura of a grand mal fit is the most common, and is easy to distinguish from other causes.

Vertiginous seizures are a variety of temporal lobe epilepsy which can be confused with other causes of acute episodic vertigo. Vertiginous seizures may be distinguished from benign paroxysmal vertigo as the attacks last seconds rather than minutes, there is a transient ‘absence’ or loss of consciousness, and they are followed by lethargy, postictal depression, or brief amnesia. The child does not describe the sensation of movement with the clarity of a child with paroxysmal vertigo. Concomitant nystagmus does not occur.

There is a high incidence of associated visceral complaints and sensory symptoms such as visual and auditory hallucinations. There may also be motor or emotional components. Vertiginous seizures should be distinguished from minor motor seizures in which balance may be lost as a result of muscle contraction and from myoclonic-astatic epilepsy of early childhood (Bower, 1981). A family history of seizures is common and attacks of vertiginous epilepsy usually change to focal fits in the second decade. The EEG is diffusely abnormal and the attacks respond to anticonvulsants. The incidence of vertiginous epilepsy is much higher in some series (Eviatar and Eviatar, 1977) than in others (Blaney, 1983). This is largely due to differing patient populations in different clinics, but caution should be exercised against the assumption that vertigo with an abnormal EEG is vertiginous epilepsy.

Vertibulogenic epilepsy originates in the reticular formation of the brainstem and is precipitated by vestibular stimulation. Unlike vertiginous epilepsy which arises in the
vestibular cortex there is concomitant nystagmus. Vestibulogenic epilepsy is a rare disorder that is not specifically a phenomenon of childhood.

**Differential diagnosis**

**Paroxysmal disorders**

Peripheral and central disorders of vestibular function must be differentiated from other recurrent paroxysmal disorders. In the young child unable to describe the symptom of vertigo, great care is required in history taking. Breath holding spells and seizures are most likely to be confused with vertigo. Breath holding spells have a peak incidence between 2 and 3 years. The attack consists of a precipitating event, for example sudden fear, frustration or trauma, followed by crying, exhalation, cyanosis and limpeness. The child regains consciousness, is transiently confused and then fully recovers. The mechanism is the Valsalva manoeuvre. The attack lasts 2-20 seconds.

In the second, less common, form of breath holding an unexpected painful stimulus precipitates sudden limpeness, pallor, loss of consciousness and apnoea. The child may quickly regain consciousness or progress to opisthotonos and seizures. Diagnosis is by the ocular compression test which induces cardiac asystole for more than 2 seconds in 35% of cases. There is a positive family history in 30% and the EEG between attacks is normal (Rabe, 1974).

Recurrent syncope is seen in infants and adolescents. Usually it is a reaction to emotional stress, mild hypoglycaemia and environmental factors, but the possibility of cardiac syncope should be considered. This is particularly the case with children with congenital deafness who may be expected to have episodic vertigo. The surdo-cardiac syndrome is familial and produces congenital deafness, a prolonged Q-T interval, fainting and sudden death. The attacks begin in late infancy (Rabe, 1974).

Infantile spasms occurring in mentally handicapped infants are associated with an abnormal EEG which is diagnostic. Myoclonic-astatic epilepsy of early childhood is similar to infantile spasms but occurs in children over the age of 2 years. In such a ‘drop attack’ the child is suddenly flung forwards or backwards as if pushed violently (the myoclonic attack) or suddenly collapses due to loss of muscle tone (the astatic attack). Consciousness may be lost momentarily (Bower, 1981).

Night terrors may be confused with positional vertigo or basilar artery migraine occurring at night. Night terrors occur in the first 3 hours of deep, non-REM sleep. The child suddenly sits bolt upright, screaming with terror, and remains unaware of the surroundings for 10-20 minutes before dropping back into quiet sleep (Rabe, 1974).

In slightly older children hypoglycaemia may give rise to faintness or dizziness which usually arises in relation to a fixed interval after a meal, typically during the morning.
Psychosomatic dizziness and hyperventilation

These disturbances may present as vestibular disorders and must be distinguished from organic disease. The older the child at the onset of symptoms the more likely is to be psychogenic (Beddoe, 1977). Two groups are most at risk: those under excessive social pressure to achieve (Fried, 1980), and adolescent females. Hyperventilation may be accompanied by nausea, vertigo, headache, palpitations, faintness and visual disturbance. Diagnosis is facilitated by reproducing the symptoms with hyperventilation (Pincus, 1978). Functional dizziness may be distinguished from vertigo by turning the child on an office chair to see whether rotation induced vertigo mimics the complaint (Drachman and Hart, 1972; Kerr, 1983).

Vestibular disorders associated with abnormal development, behaviour and learning ability

Vestibular stimulation has already been noted to enhance the development of eye movement control and coordination. Poor vestibular function should therefore be regarded as an additional handicap where it exists in a multihandicapped child (Zarnoch, 1980). Studies have been reported of vestibular disorders in motor delay (Rapin, 1974), learning disorders (Ayres, 1978) and autism (Tjernstrom, 1973). Studies in learning-disabled children in particular have largely failed to isolate vestibular function from other influences upon induced nystagmus. The abnormalities found can be ascribed to differences in visual-vestibular interaction in both the learning-disabled (Ornitz, 1983) and the autistic (Tjernstrom, 1973) groups. Learning disability is multifactorial and is associated with minor, widespread, pathology in the central nervous system (Fuller, Guthrie and Alvord, 1983; Gillberg, 1985). Scattered lesions within the brainstem and higher centres may give rise to central signs on electronystagmography which may be confused with immaturity of eye movement control seen in some normal children.

Disorders of eye movement control and vestibular sensitivity do occur in children with developmental, learning and behaviour problems but, in any study of such children, comparison with age-matched controls and careful control of variables such as illumination and mental alerting, are advisable (Ornitz, 1983; Snashall, 1983). Assessment of vestibular function and eye movements will provide information that is helpful in planning realistic management goals for these children.

Management of vertigo

Medical

The treatment of vertigo in children is the same in principle as that in adults with some minor differences. The management of motion sickness has received little attention. Many children require vestibular sedatives such as dimenhydrinate, but most can be managed by positioning the child in the vehicle so that there is not a mismatch of visual and vestibular information (Jay, Jay and Hoyt, 1980).

Benign paroxysmal vertigo usually requires no more than reassurance of the parents, but dimenhydrinate may be helpful. The migrainous disorders respond to reassurance,
antihistamines or beta blockers and dietary measures. Exclusion diets of known precipitants of migraine such as chocolate, cheese, oranges and tomato derivatives along with the 'E' factors often produce dramatic results.

The various seizure disorders respond to anticonvulsants to a greater or lesser degree.

Ménière's disorder is more difficult to manage in children than in adults as routine medication has poor compliance. The fluctuation in hearing is such that assessment of response to therapy is difficult. Low salt diet, diuretics and betahistine may be used in conjunction with antihistamines, but many children with this condition do just as well without treatment.

**Psychological**

Although most children have very little difficulty coping with vertigo once the initial fear has passed they can occasionally become psychologically disabled by this symptom. This is particularly likely in adolescent girls who experience recurrent positional or movement-induced vertigo. Strong reassurance may be required to persuade these patients to resume normal mobility.

**Surgical**

If secretory otitis is present it should be corrected surgically as this will provide the fastest and most consistent response. If there is suspicion of a perilymphatic fistula, surgical exploration should be considered especially if there is any bony abnormality of the temporal bone. If there is unremitting vertigo due to endolymphatic hydrops with or without distension of the vestibule, endolymphatic sac surgery may be appropriate.