IAPO-Interamerican Association of
Pediatric Otorhinolaryngology

Dizziness in Children

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THE VESTIBULAR SYSTEM - Embryology

The otic capsule develops early in gestation between the fourth and twelfth weeks of intra-uterine life. As the vestibular system is phylogenetically older than its auditory counterpart, each stage in development is in advance of the auditory system, and therefore less vulnerable to environmental insult The vestibular nerve myelinates by 16 weeks *in utero,* and by 24 weeks there is even a primitive vestibulo-ocular reflex (VOR) present. After birth, at four months of age, the baby can tilt its head to keep it vertical. Recorded bithermal caloric responses can be made in 9 month old babies if necessary, to measure the VOR. Vestibular nystagmus in children, however, tends to be of a lower frequency and greater amplitude. Maximum slow phase velocity readings are often similar to those in adults, but the normal range for the canal paresis and directional preponderance calculations are wider than that seen in adults. Congenital nystagmus resultant from macular visual loss differs however.

Infantile Reflex Responses and Motor Milestones

The Moro response comprises a sudden bilateral extension of the upper limbs evoked by sudden jarring of the cot or dropping the head backwards by a few centimetres. This response is present in normal children at birth, and disappears by the sixth month. The secondary inherent responses are righting responses and protective reactions. From four months, the infant will tilt the head to maintain it vertical if the trunk is tilted through 30 degrees. The ages of sitting unsupported, crawling, and walking, bear some relation to vestibular function, but also depend upon neurodevelopment. Vision also plays an important part in postural control.

ASSESSMENT OF THE DIZZY CHILD

Symptoms of vertigo in children

Childhood vertigo results from a mismatch of information from the three different sensory systems: vestibular, visual, and proprioceptive. Vertigo, however, is much more difficult to recognise in babies and children than in adults; children are not able to describe what they are experiencing, and may present with other somatic symptoms such as cowering in the corner of the cot, falling to the ground crying, burying their head in their hands, and vomiting. Interestingly, children born with a congenital lack of normal vestibular function often have no balance disturbance

at all, and in the paediatric age group, vision remains by far the most important sense for locomotor and balance acquisition.

It is helpful to direct the history taking with a number of principal and most likely diagnoses in mind. The principle conditions in the paediatric age group to consider are shown in **Table 1**, although a more complete list appears in **Table 2 A and 2 B.**

Table 1. Common vestibular conditions

Common Vestibular Conditions
● Benign Paroxysmal Vertigo of Children
● "Basilar" Migraine
● Epilepsy
● Central Causes of ataxia and loss of balance
● Vestibular Neuronitis
● BPPV
● Adult causes eg Ménière's Disease.

Table 2 A. Causes of Childhood Vestibular Symptoms

Conditions with Hearing Loss
● OME
● Suppurative Ear Disease
● Cholesteatoma with fistula
● Temporal bone trauma
● Barotraumatic Perilymph fistula
● Meniere's Disease
● Post Traumatic Vertigo
● Enlarged Vestibular Aqueduct Syndrome
● Other Congenital Temporal Bone Anomalies e.g. CHARGE association
● Dehiscent Superior semicircular canal Syndrome
● Drug Induced Ototoxicity
● Congenital Syphilis
● Herpes Zoster Oticus
● Congenital CMV infection
● Metabolic conditions – Hurler's Syndrome, - hypothyroidism
● Usher's syndrome

Table 2 B. Causes of Childhood Vestibular Symptoms

Conditions with Normal Hearing
● Motion sickness
● BPV - Benign Paroxysmal Vertigo of Childhood
● Basilar Migraine
● Seizure disorders
● BPPV
● Post Traumatic Vertigo
● Viral Labyrinthitis or neuronitis
● Posterior fossa Tumours
● Cardiac Causes: Syncope & Arrhythmias
● Acute Poisoning
● Multiple sclerosis & Lyme Disease
● CNS Infections - Coxsackie A & B or echovirus encephalitis or HIV infection
● Meningitis – viral or bacterial
● Chiari Malformations
● Hereditary Cerebellar Ataxias
● Acute Cerebellar Ataxia

The presentation of vertigo varies according to the age of the child quite dramatically. Whilst it is possible for two year-olds to experience acute vertigo, young children cannot describe this, and may even present with torticollis. If there is a delay in motor milestones children may present with poor balance or falling; this can also be associated with simple conditions like glue ear. By five years of age, short-lived dizzy episodes can be described, the common cause being benign paroxysmal vertigo of childhood. By the teenage years, migrainous vertigo, psychogenic vertigo, and the adult vertiginous conditions, are much more common.

It is helpful to establish the nature of the dizziness, whether true vertigo, loss of balance, or a light-headed faint feeling. How long do the symptoms last? The duration and periodicity can be useful guides as may precipitating factors, such as head or neck injury.

The presence of frequent headaches and whether they occur with vertigo or at other times is valuable. Associated vomiting may be an indication of an acute true vertigo, of a migraine phenomenon or of the presence of raised intracranial pressure. Associated hearing loss, otalgia or otorrhoea are important. It can be helpful to categorize childhood dizziness into conditions with normal hearing, and those with associated deafness (**Table 2 A and 2 B**).

A neurological history is helpful, specifically if there is anything to suggest tem-

poral lobe seizures, visual or olfactory hallucinations. The developmental history, in terms of the motor milestones, or any regression should be ascertained. The presence of a recent pyrexial illness, the drug history, both current, past and indeed *in utero*, can be important, and various sorts of poisoning should be borne in mind in the child who becomes acutely ill with vertigo and may have ingested something while playing. In the ill febrile child a range of serious infectious diseases should be considered. In a slightly older child, it may be more apparent that the problem is one of fainting or hyperventilation, or that there are cyanotic attacks or palpitations in association with the dizziness.

The family history is relevant, especially one of maternal migraine, or familial sensorineural deafness, or of NF2.

Examination of the dizzy child

The routine paediatric examination will include the tympanic membranes. Facial nerve function, tongue movements, and the gag reflex should be checked. It is important to look at eye movements and particularly to search for nystagmus, which can be unmasked with the use of Frenzel glasses. The standard clinical balance assessments can be undertaken such as Romberg's test, Untenberger's stepping test and the tandem heel-toe gait: it is helpful to make this fun for the child, by introducing games, like hopping and kicking a football, to better assess balance function. Optokinetic nystagmus may be viewed by watching a rotating drum and a marked directional preponderance may be diagnosed. Dix Hallpike positional testing should be undertaken also.

Cerebellar ataxia is seen on heel-toe tandem gait with dysmetria, but with normal ranges of lower limb motion and unchanged gait velocity and stride length. Characteristically, gait is wide based with dyssynergia and dysrhythmia, and balance is poor. [2]

Investigations

Audiometry is mandatory. This should comprise a pure tone audiogram or alternative threshold assessment such as visual reinforcement audiometry (VRA) if the child's age or development, demand it. Objective testing with brainstem auditory evoked responses may be indicated. Tympanometry should also be undertaken.

Routine blood tests to exclude anaemia or other blood dyscrasias are worthwhile. The white cell count, inflammatory markers (ESR or CRP) may give a clue to an infective condition which could have led, for example, to cerebellar encephalitis. Serology should exclude congenital syphilis and HIV disease might be considered.

Depending on the history and the level of concern, other investigations might include formal bithermal caloric testing with video-nystagmography (VNG) or electronystagmography (ENG) recordings.

Imaging the head and inner ears with MRI scanning and/or a high resolution CT scan for the bony labyrinth and temporal bones will be indicated in selected children. For example, reassurance that there is no space occupying lesion in a child with headaches and vertebrobasilar migraine, or defining an enlarged vestibular aqueduct in association with sensorineural hearing loss, could be important. If the

diagnosis is clinically obvious however, then it is unnecessary to undertake brain scanning.

Where the history indicates it, referral for an electroencephalogram (EEG) and neurological opinion or for an electrocardiogram (ECG) and cardiac review may have to be considered.

CAUSES OF CHILDHOOD VESTIBULAR SYMPTOMS

The diagnostic flow chart (**Figure 1**) summarises the diagnostic process in managing the child with vestibular symptoms. The conditions are discussed in more detail below.

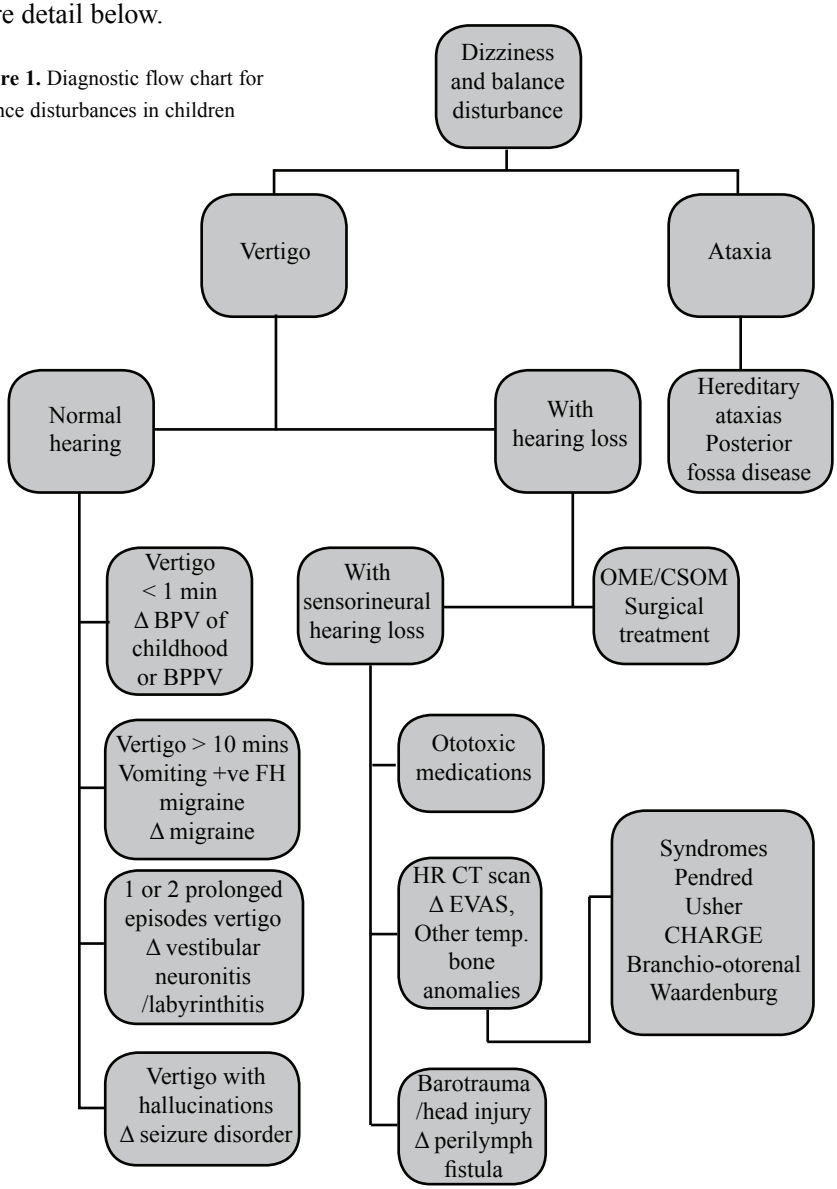

Figure 1. Diagnostic flow chart for balance disturbances in children

VESTIBULAR CONDITIONS WITH NORMAL HEARING

Table 1 and **Table 2 A and B** list most of the conditions which can present with childhood vertigo, dizziness or balance problems. Although the differential diagnosis is extensive, in over half the children who present to the paediatric otolaryngologist with dizziness or disequilibrium, the cause will either be glue ear, benign paroxysmal vertigo of childhood, or dizziness as a migraine phenomenon.[3] A further study indicates the most common causes for vertigo in children to be migraine in 31%, and benign paroxysmal vertigo of childhood (BPVC) in 25%. Other less frequent causes include trauma with deafness, delayed endolymphatic hydrops, benign positional vertigo, and more rarely, cerebellopontine angle tumour, seizures, acute vestibular neuritis or juvenile rheumatoid arthritis. In this study, abnormalities sere found in hearing in 24%, in positional testing in 5%, in 11% of bithermal caloric tests, and in 65% on rotational chair testing.[4]

Motion Sickness

Motion sickness is caused by a conflict in the kinetic input, often with an excessive vestibular stimulation. Girls are more commonly affected than boys, and it tends to settle at puberty. Interestingly, motion sickness can occur in people with blindness, but can also be caused by purely visual stimulation. There is a mild association with migraine and vestibular dysfunction and significantly greater motion sickness for female as compared with male subjects.[5]

Benign paroxysmal vertigo of childhood

Benign paroxysmal vertigo of childhood is not a positional vertigo and is quite different from BPPV. It occurs in children who are four or over, with no obvious precipitating factors. It is a very frequent cause of paediatric dizziness, being found in 35% of children with dizziness in one series.[6] The child experiences short-lived acute vertigo for 30 to 60 seconds. He or she may fall or hold onto something suddenly and cry, becoming pale and sweaty. There is then a rapid return to complete normality a few minutes later. Nystagmus is present during attacks which are recurrent and variable in frequency. They can continue in older children, but usually subside. Half of these children, however, go on to develop migraine in adolescence. Most have a family history of migraine. Caloric abnormalities are quite likely to be present, if recordings are made. A very similar condition presents in a slightly younger age group as benign paroxysmal torticollis. All torticollis is not related to vertigo however. There is a suggestion that creatinine kinase levels are likely to be elevated in BPV of childhood, and measurement may be helpful in diagnosis.[7] BPV has a very favourable long term prognosis. In one study the condition had resolved by about 8 years and on long term follow up 21 % had developed migraine but none had any vertigo or balance disorder.[8] The differential diagnosis of a child or baby presenting with a marked torticollis, incidentally, is large and varied and should include: congenital torticollis, paroxysmal torticollis with vertigo, mastoiditis or neck abscess, skull base tumour and neurological extrapyramidal spasmodic torticollis or psychogenic spasmodic torticollis.

Migraine Related Vertigo

The child with migraine related vertigo should have a history of migraine, namely nausea or vomiting or phonophobia or photophobia with severe headaches and at times vertigo attacks. The migraine history can be either with or without visual aura.

Migraine Equivalent Vertigo

This term is applied to the patient in which the vertigo attacks are the sole symptom of migraine and the clue to the diagnosis is usually a very strong family history of migraines.

Basilar Migraine

Basilar migraine is usually diagnosed when there is a clear history of migraine together with at lease two of the following episodic brainstem features: dysarthria, vertigo, tinnitus, hyperacusis, diplopia, ataxia, or bilateral paraesthesia.

Migraine is a common multifactorial neurovascular disorder. Several genes have been discovered for migraine, the CACNA1A gene on chromosome 19p13, encoding for part of a neuronal calcium channel is one. Other loci and genes associated with migraine have also been described. Familial hemiplegic migraine (FHM) has been similarly linked.

Migraine in general affects 5% of schoolchildren, and can occur at any age. Half the children with basilar artery migraine do not have headaches, but when present, the headaches are synchronous with the dizzy attacks in only one third. Vomiting with attacks is common and the child is likely to be unwell for a few days at a time. If there is no headache, the history of some transient neurological disturbance such as hemiparesis, ataxia, or facial paresis will indicate the diagnosis. An important clue to this diagnosis is the family history; 85% have a history of first degree relative suffering from migraine. In one study into migraine related vestibulopathy common vestibular test abnormalities included unilateral reduced caloric responsiveness, and treatment was usually directed at the underlying migraine condition by identifying and avoiding dietary triggers and prescribing prophylactic anti-migraine medications. Symptomatic relief was also provided using anti-motion sickness medications, vestibular rehabilitation, and pharmacotherapy directed at any associated anxiety.[9] Review of the symptoms of aura revealed the following: migraine with typical aura in 63.1%; migraine with prolonged aura in 4%; FHM in 6.5%, basilar migraine in 15.9%; and ophthalmoplegic migraine in 4%. The average age at the onset of the migraine was 8.8 years and of the appearance of aura 10.9 years. The most frequent aura symptoms were visual. Hemiparaesthesia was seen in 34%, vertigo in 14.5%, hemiparesis in 13%, and ataxia in 6.6%. Vaproate and Flunarizine were effective for migraine prophylaxis.[10]

Videonystagmography studies in children with migraine, undertaking spontaneous nystagmus, gaze nystagmus, eye tracking test, optokinetic and positional nystagmus, and caloric testing, showed that all patients with migraine had abnormalities in vestibular testing. Analysis of the results suggested a mainly central localisation of vestibular dysfunction.[11]

There is 30 % overlap between children with chronic tension type headache (CTTH) and migraine headache, usually without aura. Nausea, noise and light intolerance is common in these patients but vertigo is unusual.

Vestibular neuronitis

This presents as it does in adults, with acute severe vertigo, nausea and often vomiting but normal hearing. Children recover more quickly from this disorder than do adults. Half the patients can have repeated episodes, although within a few years attacks become progressively less severe and are likely to cease.

Benign paroxysmal positional vertigo (BPPV)

Whilst in adults BPPV most commonly occurs spontaneously or follows vestibular neuronitis some time previously, in children it is rare, and BPPV is more likely to occur following a head injury or marked whiplash injury. It has a good prognosis. In one study on children's temporal bones in Boston, 12.7% of paediatric temporal bones examined had basophilic deposits, many of them with otoconial crystals in the semi-circular canals. That is much higher than the incidence of children who had any vertigo symptoms, a temporal bone finding mirrored for this condition in adults.[12] The author believes that the pathophysiology of BPPV depends upon an interaction between canalith particles exerting a gravitational effect and a loss of normal reflex suppression of semicircular function for otolith or superior vestibular nerve dysfunction.

Post Traumatic Vertigo

The complaint of dizziness and headaches quite commonly follows head injury in children. The relatively high incidence of these persistent post-traumatic symptoms in children and adolescents presents a diagnostic challenge. It is often difficult to differentiate between functional complaints generated by psychological trauma or compensation-seeking, and an organic aetiology. [13]

Seizure Disorders

Recurrent unprovoked seizures due to epilepsy are either generalised or localised (focal). Seizure disorders can give rise to vertigo in two ways: firstly, in the aura of a generalised (Grand Mal) fit; secondly, as vertiginous epilepsy or vestibulogenic epilepsy. In temporal lobe or occipital lobe focal epilepsy, there may be transient loss of consciousness or amnesia; the child may describe the sensation of movement and may have visual or auditory hallucinations. There may be motor or emotional components. Convulsive epilepsies are generally unmistakable. Absence epilepsies may be recognized by the provocation of an episode during hyperventilation. Complex partial seizures in children can be difficult to distinguish from behavioural problems, shuddering attacks, paroxysmal vertigo, breath-holding spells, cardiogenic syncope, night terrors, and movement disorders, such as paroxysmal kinesigenic choreoathetosis.[14]

A comparison of the elementary visual hallucinations of 50 patients with migraine and 20 patients with occipital epileptic seizures showed that epileptic seizures are predominantly multicoloured with circular or spherical patterns as opposed to the predominantly black and white linear patterns of migraine. This simple clinical symptom of the elementary visual hallucinations may be helpful in distinguishing between classic or basilar migraine and visual partial epileptic seizures.[15] Referral

to paediatric neurology is required if vertiginous epilepsy is suspected.

Psychogenic (conversion reaction) vertigo

Psychogenic dizziness should be diagnosed after excluding organic pathology. Sometimes seen in adolescents, more commonly girls, it is said to occur in children who are put under parental pressure to achieve. Recurrent fainting episodes can be seen in adolescence, when the possibility of a cardiac cause should be considered. In the Surdo-cardiac syndrome, for example, there is a prolonged QT interval, fainting, and a risk of sudden death.

Psychogenic vertigo as a conversion reaction can be seen alone or in association with psychogenic hearing loss. The discrepancy between symptoms and findings in audiometric or vestibular tests is the essential clue for reaching a diagnosis of a conversion disorder. Referral to a psychiatrist may be necessary, because many patients have problems in school or at home, and recovery may take a long time.[16]

Miscellaneous conditions with normal hearing

There are a number of other conditions which can be worth considering in the infant or child with unusual symptoms which could mimic vertigo. These include: toddler breath-holding attacks of cyanosis.

Poisoning

Acute poisoning from plants, chemicals or drugs should be considered in the ill child with dizziness nausea and vertigo amongst other symptoms.

Ataxias

The ataxias and other primarily neurological, hereditary or degenerative conditions are rare. They are discussed below.

VESTIBULAR CONDITIONS WITH ASSOCIATED HEARING LOSS

Otitis Media with effusion (OME) and Chronic Suppurative Ear Disease (CSOM)

Glue ear (OME) may be detected in the clumsy child with poor balance who is more prone to falls than his siblings or peers. CSOM with perforation and infections can influence general balance and CSOM with cholesteatoma carries the possibility of a fistula to the lateral semi-circular canal or oval window accounting for dizziness or leading to suppurative labyrinthitis.

Ménière's disease (MD)

Childhood or adolescent onset of MD, though uncommon, is well documented. In one series sporadic MD started at this early age in less than 3%, though in the less common familial MD in more than 9%, no doubt due to the phenomenon of anticipation. The clinical features are indistinguishable from those in adults, though early onset tends to be associated with more aggressive disease and a likelihood of relatively early bilateral involvement.[17]

Associated temporal bone abnormalities and hearing loss

In numerous conditions and syndromes, there is sensorineural hearing loss with temporal bone anomalies. In only a small number of these conditions are children likely to present with vestibular symptoms. Vision remains the most important special sense in acquiring balance and children with bilateral vestibular impairment, may be delayed somewhat in motor development but rarely present

with vertigo. Not surprisingly, conditions in which there is abnormal or absent vestibular development and visual loss are more likely to present with balance disturbances. Usher's syndrome children have vestibular hypofunction and may therefore have balance difficulties when vision is also impaired.

In CHARGE syndrome (**C**oloboma, **H**eart Abnormalities, **A**tresia of the Choanae, **R**etardation of Growth / Mental, **G**enitourinary defects, **E**ar Abnormalities) there are frequently abnormalities such as a primitive otocyst and these children may have absent semicircular canals and an aberrant facial nerve. A study from Ann Arbor studied patients with severe sensorineural hearing loss and agenesis of the semicircular canals. Most were CHARGE syndrome, some were non-syndromic, one had Noonan's syndrome. They did not present with vertigo.[18] X-linked hereditary deafness is another example in which vestibular symptoms are uncommon despite vestibular hypofunction. Vestibular hypofunction may be present in Down syndrome.

The Enlarged vestibular aqueduct syndrome

The enlarged vestibular aqueduct syndrome (EVAS) is a rare congenital anomaly. Vestibular disturbance uncommon with EVAS, but is seen in 4 % of children. Fluctuant and progressive sensorineural hearing loss is the norm and is bilateral in 87 % of cases. A vestibular aqueduct radiologically wider than 1.5 mm at its midpoint or wider than 2mm at the operculum is defined as enlarged.[19] Most patients remain with stable hearing in at least one ear over a four-year period. It can occur in non-syndromic conditions, but is also found in 50 % of patients with Waardenburg's syndrome (types 1 and 2), in which there may be significant widening of the vestibular aqueduct at its mid-point together with other temporal bone anomalies.[20] These children tend to have profound or severe hearing loss. Up to 30 % of children with Waardenberg's syndrome have vestibular impairment and some experience episodic vertigo. EVAS is also seen in Pendred and the Branchiootorenal syndromes, often with an associated Mondini deformity in the former. Patients with EVAS may show an autosomal recessive inheritance.[21] Avoidance of head injuries is recommended, but may not influence the progression of deafness, the exact mode of its pathogenesis being as yet, ill understood. Surgery to occlude the Vestibular aqueduct remains controversial, currently conservative management is advised.

The patent cochlear aqueduct

The cochlear aqueduct at its narrowest portion is 0.14 mm wide. It widens as it opens into the posterior fossa with a very variable size at this point. The late Peter Phelps stated that after histological study of 1,400 normal temporal bones, and 29 with dysmorphic labyrinths, he had failed to show a dilated cochlear aqueduct, and believed that patients with sensorineural deafness attributed to this in fact related to defects at the fundus of the internal auditory canal.

The dehiscent superior semi-circular canal syndrome

The dehiscent superior semi-circular canal syndrome has been described but is quite rare. It can be demonstrated on high resolution CT scanning. Typically vertigo or oscillopsia is evoked by loud noises or by stimuli that result in changes in middle ear or intracranial pressure. The Tullio phenomenon and Hennebert sign

may therefore be found. Three quarters of patients also experience chronic disequilibrium which is often the most debilitating symptom. [22] The condition may also present with an apparent conductive hearing loss. Evoked eye movements, by Valsalva against pinched nostrils, tragal compression, or sounds over 100db at 500-2000Hz produce vertical and torsional components. Surgical repair via the Middle Fossa approach is successful. There is little suggestion that this condition is seen in children.

Perilymph Fistulas

Perilymph fistulas in children are usually seen in association with temporal bone anomalies and pre-existing severe or total hearing loss in the affected ear. They may present with recurrent meningitis or with CSF behind the tympanic membrane. Perilymph fistula can arise directly from blunt trauma to the middle ear or from temporal bone fractures and indeed iatrogenically, after ear surgery for CSOM or post-stapedotomy. More rarely, marked barotrauma may lead to a fistula from the round or oval windows. In all these situations surgical exploration to seal the fistula is indicated. Spontaneous perilymph fistula in the normal temporal bone however is probably almost never seen.

In the late 1980s there was a vogue for clinically diagnosing a spontaneous perilymph fistula in children and adults who presented with symptoms of hearing loss, vertigo and sometimes tinnitus. These patients were subjected to surgical exploration of the middle ear with sealing of the apparent fistulae. In general the hearing outcomes from surgery did not seem to correlate with the finding of a fistula and indeed it can be very difficult at surgery to be sure if there is any real perilymph leak.

To conclude, perilymph fistulas can be a cause of hearing loss, vertigo or tinnitus and these symptoms may be fluctuant and possibly progressive. There is currently no good diagnostic test for a small fistula. In the paediatric population the most frequent cause is a congenital fistula. Severe or profound hearing loss is in this instance always associated with temporal bone anomalies when a peripymph/CSF leak may be present with fluid behind the tympanic membrane. A defect in the stapes and continuity with the fundus of the internal auditory meatus is one such example. This can be found with a true Mondini deformity in which case some hearing from the basal turn of the cochlea is possible. These cases will require surgical exploration and closure of the leak, not to improve or restore hearing but in an attempt to prevent subsequent meningitis.

Traumatic perilymph fistulas with a normal temporal bone anatomy are rare. They are described following head injury, penetrating injury to the middle ear, with or without temporal bone fracture but diagnosis is difficult. [23] A persistent perilymph fistula following ear surgery requires re-exploration. Severe barotrauma can also produce a fistula from the round window or oval window. Clinical suspicion will lead to the decision to explore the ear surgically. More obvious bony erosion with fistula is not infrequently encountered in the presence of cholesteatoma. Exploration and closure of the fistula is indicated. Spontaneous perilymph fistula, in the absence of head injury direct injury or barotrauma can be virtually discounted.

Drug Induced Vertigo or Imbalance

Ototoxic medications, particularly amino-glycosides can cause marked vestibular dysfunction with, acute vertigo at the time of administration or poor balance, ataxia and motor delay. These drugs might have been administered systemically prenatally to the mother or in post-natal life, but occasionally topically in the presence of a chronic perforation or grommets. Fortunately however, hearing loss from systemic aminoglycosides given to the infant is unusual. Some degree of vestibular loss may be more common and under-diagnosed. Streptomycin and Gentamicin are more selectively vestibulotoxic. In one study, children who had previously been treated with streptomycin commonly showed delay in walking. [24] The antimalarials such as mefloquine, which is only slowly cleared from the body, can cause dizziness or hearing loss. The platinum based cytotoxic agents can cause ototoxicity, usually high tone hearing loss and tinnitus rather than dizziness.

Miscellaneous conditions with hearing loss

Infectious aetiologies such as congenital CMV infection can include sensorineural hearing loss with vestibular symptoms and metabolic diseases such as Hurler's syndrome can be seen with a retrocochlear type of hearing loss and vestibular impairment. Herpes zoster oticus can occur in children.

PERSISTENT IMBALANCE & ATAXIA – Central Disorders

Toddlers may present with imbalance and a delay in motor development, or with a subsequent deterioration in vestibular function. They can have falls, fear of the dark, abnormal gait, and vomiting. Primary developmental delay with motor delay and poor balance suggests a congenital or early acquired neurodevelopmental disorder, while regression of balance and locomotor function that was previously acquired indicates the need to exclude a space occupying lesion such as meningioma or medulloblastoma. A family history of NF2 would raise suspicion. Any severe illness or even major surgery in the baby or smaller child, will not infrequently lead to temporary loss of previously acquired skills such as ability to walk.

The Ataxias

Ataxia is a common mode of presentation of cerebellar, posterior column, and vestibular disease in children. The aetiology of ataxia covers a broad range, from infections to rare hereditary metabolic diseases. The importance of recognizing potentially reversible conditions such as vitamin E deficiency and Refsum's disease has been stressed. [25]

Hereditary cerebellar ataxias

The hereditary cerebellar ataxias present with a slowly progressive ataxia, though a posterior fossa tumour must be excluded by imaging. Cerebellar disorders have a variety of inherited and sporadic causes. Advances in genetics have led to the successful classification of over 20 forms of autosomal dominant and recessive cerebellar ataxias with variable phenotypes and have shed light on the underlying pathophysiology of many of these disorders. Successful disease-modifying or symptomatic treatments for these conditions, thus far, have remained limited. [26]

Refsum's Disease

Refsum's disease is a disorder of lipid metabolism with pigmentary retinopathy, demyelinating neuropathy, ataxia, and hearing loss. There is progressive difficulty in walking, and this comes on between the ages of 4 and 7 years. The site of the hearing abnormality in Refsum's disease may be 'post-outer hair cells' in some cases. [27]

Charcot Marie Tooth Disease

The most common hereditary degenerative condition is Charcot Marie Tooth Disease. Inheritance is autosomal dominant. Perineal muscle atrophy is usual, congenital sensorineural deafness is present in some cases, and there can be vestibular weakness. These children develop spinal scolioses and *pes cavus*.[28]

Acute Cerebellar Ataxia

Acute Cerebellar Ataxia occurs, usually in the first three years of life, in a child who was previously normal. It follows a viral febrile illness a few weeks beforehand. There is sudden ataxia, and the condition may take a number of months to slowly resolve, or leave some permanent sequelae.

The Chiari malformations

The Chiari Type 1 malformation is characterised by cerebellar tonsil herniation through the foramen magnum. These children most commonly present with bilateral vocal cord paralysis and associated upper airway obstruction, but they can also present with this sort of cerebellar ectasia causing a positional vertigo and a central type of nystagmus. The condition can be more severe and associated with syringomyelia, in which case there can be neurological improvement after foramen magnum surgical decompression. Type 1 may present to otolaryngologists.

The Type 2 Chiari malformation is the same as Type 1, except that there is a non-communicating hydrocephalus and lumbosacral spina bifida in addition. Type 3 can have any of these features, but with cervical or occipital bifida. The Types 2 and 3 have widespread neurological abnormalities.

Miscellaneous conditions

Demyelination can present in post-pubertal children, in which case vertigo is quite commonly seen. NF2 with posterior fossa meningioma or vestibular schwannoma are occasionally seen in children, and other intracranial posterior fossa lesions such as medulloblastoma may present with ataxia and vomiting.

Infectious causes

Infectious causes include Lyme disease. Viral infections include meningitis, Coxsackie A and B and echovirus; they can involve the central nervous system with vertigo, nystagmus and cerebellar signs. HIV infection is another possible cause. Bacterial infections include primary meningitis, labyrinthitis as a complication of meningitis or CSOM and tertiary or congenital syphilis.

TREATMENTS FOR VERTIGO

Medical Treatments

The causative condition should be treated directly if possible. The mainstay of treatment however is usually explanation to the parents and the child, and reassurance.

Medical treatments for vertigo symptoms are summarised in **Table 3** and for migraine related vertigo in **Table 4**.

Table 3. Medical Treatments for Acute Vertigo Symptoms

● **Acute Otologic Vertigo attacks** ● Cinnarizine (15 mg tds if over 5 yr) / Cyclizine ● Hyoscine patches (1/4 patch per 72 hrs, if <3yr) ● Metoclopromide (maxolon) 1-2 mg/kg tds ● Domperidone 15-30 mg b.d rectally
● **Motion Sickness** ● Cinnarizine, Promethazine (phenergan), domperidone,_
● **Meniere's Disease** ● As for Acute Otologic vertigo ● Betahistine – weak central H1 agonist, moderate H3 antagonist & vasodilator ! ● Diuretics (bendroflumethaiazide) ● Ca channel blockers ?

Table 4. Medical Treatments for Migraine Related Vertigo in Children

● Exclusion Diet (chocolate & tyramine-free) ● Hyoscine Patches ● Cinnarizine ● Metoclopramide ● Rizatriptan (5HT1 antagonist)/ Sumotriptan/ zolmitriptan ● (35 % response only)
● Prophylaxi**s** ● Propanalol ● Amitriptyline ● Verapamil slow release ● Pizotifen ● Anticonvulsants - valproate, clonazepam

Symptomatically, the vestibular sedatives can be helpful: the antihistamines such as Cyclizine or Cinnarizine can be taken for more prolonged attacks; Hyoscine patches have been advocated and Domperidone is helpful for associated sickness. The dopamine antagonists, phenothiazines such as prochlorperazine are effective vestibular suppressants. However there is a greater risk of extrapyramidal side effects when using phenothiazines especially in children. They should be avoided in the under 10 kg baby. Should these medications have led to extrapyramidal effects such as olygogyric crisis, it can be treated acutely with the antagonist, Procyclidine, by injection.

HT3 antagonists such as Ondansetron are powerful antiemetics which block serotonin binding at vagal afferents in the gut and in the regions of the CNS involved in emesis, including the chemoreceptor trigger zone and the nucleus tractus solitarii. Although principally used in post-operative nausea and vomiting or with cyto-

toxic drug therapy, they may have a role in the vertiginous child if vomiting.
Migrainous vertigo can be treated with Metaclopramide, or serotonin 5-HT1 receptor agonists such as Sumatriptan. Rizatriptan is reported to be more effective than other drugs of this class, and other simple analgesics.[29] Dietary exclusions can be helpful. Preventative measures, if necessary would be the currently recognised ones; Pizotifen, Amitriptyline or Propanolol, and if those fail, anticonvulsants such as Valproate might be prescribed by a neurologist.
Ménière's Disease may be treated with Betahistine, low salt diet and possibly diuretics. Surgery may occasionally be indicated in severe variants of the disease.
The seizure disorders are usually well controlled with anticonvulsants under the paediatric neurologist's guidance.

Physical treatments for vertigo

If there is benign positional vertigo, the Epley manoeuvre, the Semont manoeuvre or the Brandt-Daroff exercises can be employed successfully. Other vestibular rehabilitation exercises for children who have suffered unilateral labyrinthine damage might be helpful in achieving full central compensation and speeding recovery.

Surgery for vertigo

Surgery relates to that indicated for specific underlying conditions. Unilateral glue ear with poor balance can be corrected by insertion of grommets (preferably bilaterally, the contralateral ear as a prophylactic measure).
If there is a perilymph fistula from barotrauma or from middle ear or mastoid disease, or following surgery, that should be explored and closed. Likewise suppurative ear disease and congenital or acquired cholesteatoma will require tympanomastoid surgery.
Perilymph/CSF fistula from congenital temporal bone anomalies should be closed surgically in an attempt to prevent future meningitis.
Childhood onset Ménière's disease tends to run an aggressive course with debilitating bilateral disease later in life. Destructive surgery is not advised at an early stage although endolymphatic sac decompression and drainage may have a place.

CONCLUSION

Children do not usually complain of vertigo; history and diagnosis can be elusive. The pattern of symptoms in the very young has a very wide differential diagnosis because of this.Once middle ear disease and congenital or hereditary sensorineural conditionshave been excluded, a large percentage will have dizziness associated with migraines. The family history is a helpful clue to this diagnosis. Posterior fossa neurological disease should be considered and in older children adult causes of vertigo may be seen. Reassurance about a condition with a favourable prognosis, and antihistamine such as Cinnarizine or if present anti-migraine treatments, are usually effective.

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