Balance and vestibular disorders in children

BY SOUMIT DASGUPTA, NIAMH MCKENNA AND SAFAA DAWABAH

Discover the world of paediatric vestibular assessment and management from the team at Alder Hey Children's Hospital, which is revolutionising services in this field.

izziness and balance problems generate significant morbidity in children of all age groups.

Vestibular disorders are the commonest causes of balance issues in children.

These disorders can lead to cognitive, psychological and somatic symptoms influencing the overall development of the child [1]. Evaluating dizziness in children requires experience, knowledge and extensive training in adult vestibular medicine, general and allied paediatric medical disciplines, especially developmental paediatrics and paediatric neurology.

Awareness regarding balance and dizziness in children in the global perspective is low, with only a handful of dedicated paediatric vestibular centres in the world. It is important to recognise that adult vestibular medicine is very different from paediatric vestibular medicine where norms and diagnostic processes cannot be extrapolated.

The vestibular system in children

The cochleovestibular system develops in utero from six weeks and is fully formed at birth. The uterine phase of development is guided by genetically controlled targeted development for synaptic and neural connections that attain function after birth. After birth, development is driven by sensory cues in the environment. Priming of vestibular function proceeds, following a critical temporal period dependent on neurotransmitter expression and regulation of synaptic connections.

The development of the vestibular system is shown in Figure 1. Key reflexes resolving space up to six months of age are the primitive developmental reflexes [2]. The vestibulo-ocular reflex becomes functional from two months and the vestibulo-spinal reflexes mature in adolescence. Complete central vestibular integration for the final balance percept incorporating the optical system, the proprioceptive system and the cognitive centres with an eventual output to the motor system is attained at

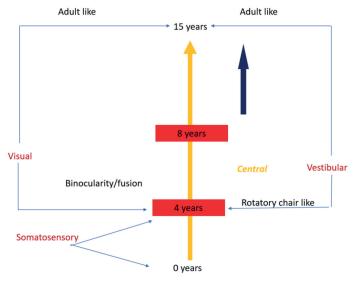


Figure 1: Development of vestibular system

15 years of age [3]. Knowledge about this development is essential to interpret meaningful age-specific vestibular function test battery.

The vestibular system in children is just not limited to a vestibular sensation; it also maintains posture and muscle tone, produces kinetic contractions to generate ocular stability and is involved in cognitive and spatial discrimination [4].

Epidemiology of balance disorders in children

The epidemiology of dizziness and balance problems in children in a large US study was 5.3% and increased with advancing age, possibly because of better reporting by older children, out of which at least 18% had a significant problem [5]. In children presenting with balance complaints, as many as 35.84% were diagnosed with a vestibular problem, often with accompanying sensorineural hearing loss [4]

Aetiology of balance disorders in children

The aetiology of balance disorders in children is different from those in adults

in terms of phenotype and occurrence. The causes are vast and overlap across paediatric medical disciplines. Medically inexplicable conditions, a third of development incoordination disorders and a third of sensorineural hearing losses can be accounted for or accompanied by a central or peripheral vestibular disorder. The causes are given in Table 1. The commonest cause of paediatric dizziness is vestibular migraine and, unlike in adults, viral acute vestibular neuritis, BPPV and Ménière's disease are rare in children and are usually secondary [4].

Assessment of the vestibular system in children

A comprehensive and detailed diagnostic algorithm is aimed at diagnosing peripheral and central vestibular disorders and a likely aetiology. It also addresses diagnosing a physical or a psychological cause for vestibular decompensation that generates symptoms. Therefore, a vestibular diagnosis is not just limited to the vestibular system alone. An accurate diagnosis leads to a very successful outcome from management and, thus, diagnosis is of paramount importance.

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Table 1: Aetiology of dizziness and balance disorders in children		
System	Pathologies	
Neurological system	Central migraine variants, epilepsy, demyelinating syndromes, neurovascular conditions (CVA, hypoxic-ischaemic encephalopathy), brain tumours and space-occupying lesions, neurodegenerative conditions, muscular dystrophies, hereditary sensorimotor neuropathies (HSMN), cerebellar ataxias	
Cardiovascular system	Vasovagal syncope, postural orthostatic tachycardia syndrome (POTS), cardiac structural abnormalities, conduction defects	
Ocular system	Refractive errors, convergence insufficiency, strabismus	
Psychological	Anxiety, panic attacks	
Rheumatological / musculoskeletal system	Connective tissue disorders, lower limb and skeletal spine abnormalities, hyperelasticity	
Haematological system	Blood dyscrasias, anaemia	
Metabolic system	Diabetes, thyroid and pituitary disorders, Vitamin (especially Vitamin D) deficiencies, electrolyte disturbances	
Neurodevelopmental system	Development incoordination disorder, sensory integration disorder, autistic spectrum disorder, attention deficit disorder	
Vestibular system	Vestibular migraine and migraine variants (BPVC) Peripheral vestibular lesions Infective—OME, CSOM, labyrinthitis, vestibular neuritis, meningitis and sepsis Inflammatory—autoimmune ear disorder, granulomatous lesions, multiple sclerosis of VIII nerve Space occupying lesions – posterior fossa and endolymphatic sac tumours and arteriovenous malformations Traumatic—head injury, perilymph fistula, iatrogenic, radiation Degenerative—endolymphatic hydrops, BPPV Ototoxic—gentamycin, cisplatin, vincristine, diuretics and antimalarials Third window disorders – enlarged vestibular aqueduct, semicircular canal dehiscence Genetic syndromic—cochleovestibular dysplasias, e.g. CHARGE, BOR, Pendred, Usher, Alport, Alstorm and Jervall Lange-Nielsen Genetic non-syndromic—DFNA9, DFNA12, DFNB2, DFNB4, DFNB12 Non-genetic structural abnormality—Arnold-Chiari malformation, space occupying lesions in the posterior fossa Auditory neuropathy spectrum disorder	

The assessment includes detailed anamnesis, multiple system examination followed by a neurotological examination and laboratory tests. Children up to the age of eight years may not be able to describe a perception of dizziness or imbalance. Eliciting vestibular behaviour from parents is thus pivotal to establish patterns for a diagnosis. In infants under the age of six years, the primitive reflexes are assessed. A clinical examination followed by objective quantification of vestibular function is required in all cases. Choice of tests to glean meaningful information is dependent on the developmental age of the child. Most tests are eminently feasible including in children with autism or attention disorders in a two-tester scenario. Removal of optic fixation and magnification of eye movements is recommended. A full audiological assessment, imaging studies and blood tests are required for determining pathology. An age-determined selection of tests is given in Table 2 and the full battery in Table 3.

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Age	Test
Birth to 6 months	Developmental reflexes, VNG
6 months to 12 months	Clinical, cVEMP, VNG, remote vHIT, chair
12 months to 3 years	Clinical, cVEMP, VNG, remote vHIT, VST, chair
3 years to 6 years	Clinical, oVEMP, cVEMP, mounted vHIT, SHIMP, VNG, VST, chai
5 years to 10 years	Clinical, oVEMP, cVEMP, mounted vHIT, SHIMP, VNG, VST, chair, fHIT, Vanderbilt DHI, balance scores
10 years to 18 years	Clinical, oVEMP, cVEMP, mounted vHIT, SHIMP, VNG, VST, chair, fHIT, psychometry DHI, balance scores

Table 3: Assessment of vestibular system in children		
Assessment	Methods	
Vestibular behaviour/history	Dizziness / vertigo / lightheadedness over the age of eight years, imbalance and unsteadiness, visual vertigo, difficulties in playground activities, fright or pallor, clutching, bumping into things, clumsiness, sudden pole axing falls, periodic episodes of nausea or vomiting +/- migrainous features, delayed motor functions, loss of postural control, difficulty with ambulating in the dark, abnormal movements or behaviour, difficulties in challenging movements (swimming), oscillopsia, difficult to track challenging visual targets, poor head eye coordination, poor reading skills	
Other history	Birth history, family history, ear symptoms especially hearing loss, development history especially motor development, other medical illness, head injury in the past, ototoxic history, musculoskeletal pain, gluten sensitivity and history of autoimmune disorders, past history of sepsis, history relating to social behaviour and peer interaction, academic history and behaviour in school, psychological and cognitive history with quality-of-life history	
Developmental assessment	Fine and gross motor skills and cognitive / social development	
Primitive reflexes – until six months of age	Moro reflex, tonic neck reflex, head righting reflex, parachute reaction, Doll's eye reaction	
System examination	General, cardiological, musculoskeletal, ophthalmological	
Audiological examination	Otoscopy, tympanometry, acoustic reflexes, pure tone audiometry, otoacoustic emissions and auditory brainstem if indicated	
Neurological examination	Cranial nerves, tone, power, reflex, cerebellum, oculomotor eye movements for nystagmus and smooth pursuits and saccades, VOR suppression, skew deviation, optokinetic assessment	
Neurotological examination	 Angular motion semicircular canals with and without optic fixation – nystagmus, calorics, mastoid vibration, rotatory chair, headshake, dynamic visual acuity, video head impulse test, suppression head impulse test, hyperventilation test, Hennebert's test Gravitational otolith system with and without optic fixation – subjective visual vertical, ocular counter rolling, head heave, cervical and ocular vestibular evoked myogenic potential Vestibulospinal tests with and without foam cushion – Romberg, Unterberger, tandem gait, retropulsion, sharpened Romberg Static and dynamic posturography and gait analysis in selected cases 	
Validated scales	Paediatric Berg Balance Score, Vanderbilt paediatric DHI	
Other tests	Blood, imaging – CT for bony labyrinth and MRI for membranous labyrinth	

Vestibular infant screening

Ghent in Flanders, Belgium, is the first centre in the world to successfully implement vestibular infant screening with cervical evoked myogenic potential test from six months onwards in children with sensorineural hearing loss. Alder Hey in the UK is starting a similar programme with the cVEMP, the remote camera video head impulse test and assessment of primitive reflexes. The objective is to intervene early in vestibular disorder. This is a new field and evidence is awaited.

Management of paediatric vestibular disorders

Following diagnosis, treatment is dictated by the aetiology, the level of intrusion in the child's life and is holistic and multidisciplinary involving, in addition to clinicians, parents / carers and the school. These include sensory integration strategies, psychological intervention when appropriate, correction of visual problems and music therapy, and involvement of multidisciplinary specialists as dictated by the diagnosis.

A key element to managing the paediatric population with vestibular disorders is to understand illness cognition models, which are applicable to both children and their carers. A concrete diagnosis achieves cognitive closure and leads to a sense of wellbeing and, more importantly, reconciliation and self-directed coping strategies built around lifestyle. This leads to a significant improvement in quality of life. Lifestyle changes for specific conditions like vestibular migraine and endolymphatic hydrops with diet and hydration are recommended. Situational counselling regarding activities that involve significant vestibular engagement is important, and adequate physical activity should be encouraged. Treatment of the primary cause, whether through surgery, pharmacological management, or particle repositioning manoeuvres, should be pursued when appropriate. Conditions such as space-occupying lesions, semicircular canal dehiscence, vestibular migraine, paroxysmia, endolymphatic hydrops, and BPPV may require these

treatments. Similarly, any cause of decompensation must be managed properly.

Vestibular rehabilitation should not be a blanket treatment option for all children presenting with dizziness and balance problems. The decision depends on the quality of lives of children and the proper quantification of a vestibular weakness and evidence of decompensation. If no weakness can be quantified, then it depends on the presence of a vestibular history (central or peripheral) and vestibular behaviour affecting QOL. When selected correctly, it remains the best option to treat a vestibular syndrome in a child as it promotes natural central compensation [4]. It should be instituted early after diagnosis and needs to be customised according to individual needs. The outcome that is measured by vestibular function tests and pre- and post-rehabilitation validated psychometric questionnaires is very favourable and rewarding. It has limited effectiveness in active and irritant vestibular lesions like vestibular migraine, Ménière's syndrome and vestibular paroxysmia.

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Conclusions

Diagnosing balance disorders in children is challenging as they are so complex. These are often interpreted as neurological problems and journey times can be long. Intervention makes a significant difference in many children's and parents' lives, so recognition / diagnosis is crucial. It is holistic, multidisciplinary and very rewarding, with favourable outcomes from management.

AUTHORS

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