Balance Disorders in Children
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Balance Disorders in children
Epidemiology
Episodic vertigo / dizziness
The deaf child with imbalance
The hearing child with imbalance
Rare syndromes to know

Epidemiology
Balance disorders are common in children
5.6% prevalence age 3-17 years (Li, 2016)
40-90% deaf children have vestibular dysfunction (Verbecque 2017, Shinjo, Jin & Kaga 2007, Santos 2015)

Dizziness
Do you ever get dizzy? 5.7-14.5%
Dizziness sufficient to stop activity ~ 2%

Diagnoses seen depend on where you work
Leicester Royal Infirmary (tertiary referral ENT balance clinic)
Great Ormond Street Hospital for Children (tertiary referral children’s hospital)
Pooled data from publications
How symptom patterns help diagnosis

Leicester Balance Centre Paediatric Diagnoses (Turner, Rea 2019)

- Vestibular migraine**: 44%
- Imbalance: 19%
- Vertigo: 34%
- Vertigo + HL: 17%
- Imbalance + HL: 22%
- Hearing Loss (HL): 23%
- BPVC**: 8%
- Cyclical vomiting**: 2%
- Arnold Chiari: 2%
- Vestibulopathy: 4%
- Other, including psychogenic: 10%
- Cardio-pulmonary (POTS hyperventilation etc): 2%

(**migraine variants)
Developmental aspects

Maturation
The vestibular system is functional at birth but central connections are immature and continue to develop until 10-15 years age [smooth pursuit, VORS]

With increasing age, there is a progressive domination of the somatosensory and vestibular systems, with less reliance on visual orientation inputs [Zipori 2018]

A knowledge of normal development is essential to assess vestibular symptoms/signs in a child.

Red Flags
Does not crawl by 1 year
Does not walk by 18 months
Cannot climb stairs at 3 years

How are children like adults?
The vestibular system functions in the same way – basic physiology is unchanged

The same pathology can occur in both age groups – trauma, drugs, meningitis, vestibular neuritis

BUT.......!
Children are very different from adults

How are children different?

**History**
Vocabulary and language
Second hand presentation
Presentation of psychological problems hidden
Impact of problem affects behaviour, education, socialisation

**Examination**
compliance
development of locomotor skills,
impact of other developmental problems eg global developmental delay, dyspraxia, cerebral palsy, ADHD, autism
Emerging pathology eg epilepsy, neurological disorders
Diagnoses

In children:

- More common: Benign Paroxysmal vertigo of childhood (BPV)
  - Vestibular migraine
  - Otitis media
  - Psychogenic

- Uncommon: BPPV
  - Meniere’s

Episodic vertigo
  - “Unprovoked”
Migrainous vertigo or migraine equivalents
Visual vertigo
Psychological

Autoimmune disease
Meniere’s disease
Cyclical vomiting
Epilepsy
Cardiac arrhythmias
Anaemia and other metabolic conditions

Migrainous Vertigo and Migraine Equivalents
Account for 30-40% of presentations with vertigo
Travel sickness
Recurrent knee or abdominal pain
Benign torticollis of infancy (BTI)
Benign paroxysmal vertigo of childhood (BPVC)
Migrainous vertigo

Migraine and episodic syndromes:
ICHD-3 [2018]
1.6 Episodic syndromes that may be associated with migraine

1.6.1 Recurrent gastrointestinal disturbance
1.6.1.1 Cyclical vomiting syndrome
1.6.1.2 Abdominal migraine
1.6.2 Benign paroxysmal vertigo
1.6.3 Benign paroxysmal torticollis
1.2.2 Migraine with brainstem aura
A1.6 [Appendix] Episodic syndromes that may be associated with migraine
A1.6.4 Infantile colic
A1.6.5 Alternating hemiplegia of childhood
A1.6.6 Vestibular migraine

Benign Paroxysmal vertigo of Childhood (BPVC)
Very common – 2% children
A migraine precursor
Peak age 2-5, abates 10-12
Sudden onset severe frightening vertigo, nystagmus, N +/- V lasting 1-5 minutes, rarely hours
Light and motion sensitivity, nystagmus, vomiting, ataxia, pallor possible

Clinical features: BPVC
Episodes may occur in clusters
Clinical examination normal in between episodes
50-70% family history of migraine
Vestibular tests and hearing normal

Management: BPVC
Diagnosis is clinical. Posterior fossa tumours, seizures and vestibular disorders to be excluded. EEG, MRI

Treatment: Reassurance

Treatment with cyproheptadine, flunarizine and propranolol has been tried, if troublesome [Gelfand 2018, Teixeira 2014]

30-60% approx. develop migraine in adulthood. They may have other episodic syndromes called ‘migraine march’
Vestibular migraine (VM) in children
ICHD-3: ‘VM may start at any age’ Diagnostic criteria as in adults.
Commonest cause of spontaneous episodic vertigo in children!

Diagnostic criteria: VM [ICHD 3, 2018]
At least 5 episodes fulfilling criteria C and D
Current or previous history of migraine without aura or migraine with aura
Vestibular symptoms of moderate or severe intensity, lasting 5 min to 72 hours
At least 50% of the vestibular episodes are associated with at least one of the three migrainous features
1. Headache with at least two of the following characteristics: one-sided location, pulsating quality, moderate or severe pain intensity, aggravation by routine physical activity
2. Photophobia and phonophobia
3. Visual aura
E. Not better accounted for by another vestibular disorder or ICHD-3 diagnosis

Clinical features: VM in children
Headaches less often unilateral [Sonal Sekhar 2012]. Duration of headache in children shorter 2-72 hours [ICHD, 2018]. Pain is frontotemporal usually.
Often history of headaches may not be obtained. In various studies 30-70% had headaches [Langhagen 2016]
Photophobia and phonophobia inferred from behaviour.
Most migraine headaches are without aura. [Langhagen 2016]

Clinical features: VM in children
Auditory symptoms in 20%-hearing loss, tinnitus, abnormal ABR (cf MD)
Nausea, vomiting, prostration common
Triggers: stress, lack of sleep, diet, menstrual cycle
[Langhagen 2016]
Clinical features: VM in children
Motion sickness in 50%, Family history in 60%
Clinical exam: Often normal, mild central ocular motor signs 10-60%
Vestibular tests may be abnormal 0-50%, e.g. caloric/VHIT/VEMP, inconsistent reports

[Langhagen 2016]

Vestibular migraine (VM)

Management: VM
No specific guidance for vestibular symptoms in children, but management along lines of migraine headaches
Keep a diary of symptoms for 8 weeks (diet, menstrual cycle)
Assess impact on school, anxiety caused due to symptoms

Symptom Diary – preferably for a minimum of 8 weeks

Dizziness and the deaf child

Why test balance in deaf children?
Aetiology
Part of syndrome eg Usher, Prognosis
developmental delay? Future ability Cochlear implant decision/preparation Treatment Habilitation/rehabilitation Safety Swimming and diving Mountaineering etc

The Deaf Child with Imbalance
Frequently delay in motor milestones walking age of 18/12 or greater is indicator (Möller 2002)

More common with inner ear deformity More common with severe and profound losses Very common after bacterial meningitis (about 80% or more)

Consider Congenital eg Usher’s, JLN, congenital CMV, congenital rubella Acquired eg trauma, acoustic neuroma / NF 2, Cogan’s syndrome

Usher’s Syndrome Incidence: 1:20,000 children 3-6% deaf children Autosomal recessive Multiple genes Some defects affect steriocilia Retinitis pigmentosa is a typical finding
Usher’s Syndrome
Usher Syndrome I
Born profoundly deaf
Slow development motor skills
Walk very late (18/12+)
Retinitis pigmentosa in 1st decade
Ashkenazi Jews

Usher Syndrome III
Progressive hearing loss
Half have balance disorders
Common in Finnish
Rare in USA

Jervell and Lange-Nielson Syndrome (1957)
Autosomal recessive
Incidence 1 in 1,000,000
Scandanavia: 1 in 200,000
Affects K+ channel function
Causes prolonged QT interval on ECG
Profoundly deaf at birth
Present with syncopal episodes on exertion
Untreated 50% die before age 15

The hearing child
Imbalanced but hearing

**Sensory**
- Vestibular hypofunction
- Blind/visually impaired
- Glue ear

**Central**
- Developmental delay
- Cerebral palsy
- Dyspraxia
- Vestibular processing disorder

**Neuromuscular**
- Still's disease
- Muscular dystrophy
- Lower limb deformities

Persistent, vague or fluctuating
- Otitis media with effusion
- Post – AVN / labyrinthitis
- Psychological
- Menarche
- Intracranial tumours e.g. acoustic neuroma
- Anaemia, Vitamin D deficiency and other metabolic conditions
- Heredodegenerative diseases e.g. Refsum (ataxia), DIDMOAD (DI,DM, optic atrophy, deafness)
- Demyelination

Single episode
- Vestibular neuritis
- Trauma
- Infection – meningitis, encephalitis
- Autoimmune e.g. Cogan’s
- Perilymph fistula
- CVA

Or first in a series = episodic
Episodic vertigo - provoked
Positional vertigo  \textit{RARE}
Any cause of vertigo – feels better sitting up

Chiari malformation
Posterior cranial fossa tumour
Perilymph fistula
Benign paroxysmal positional vertigo (BPPV) (?)

\textbf{The history is key}

Episodic vertigo - provoked
Migraine – stress, dietary factors
Drugs – caffeine, alcohol etc
Psychological - episodes of abuse
Visual vertigo
WVA (90% have Pendred’s and often vestibular hypofunction)
Vasovagal
Perilymph fistula
Dehiscent semicircular canals
EA2 – episodic ataxia 2 - CACNA1A - Ch 19p

WVA: Epidemiology
5-15\% of children with SNHL
Female to male 1:1 - 3:2
Bilateral in 80-90\% (but hearing loss may be unilateral)
Most but not all will develop HL

\textbf{Anatomy I}

\textbf{Anatomy II}
Mid-point diameter normally: 0.5-1.4mm (mean 0.8mm)
Length: 10mm
EVA> 1.5mm diameter
It is not the EVA that is important but the associated enlarged vestibular duct and endolymphatic sac. The EVA is only a marker of potential disease.
Pathophysiology of WVA: Pendred’s Syndrome

? In 40-90% of cases
Autosomal recessive
Mutation of PDS gene on Chromosome 7
The SLC26A4 gene occurs in the cochlea, kidney, and thyroid and is an iodide/chloride transporter
About 60% develop a goitre in their teens or later
May account for 5% of cases of paediatric SNHL

About 40% develop vestibular symptoms

WVA Symptoms: Balance
About 50% of patients experience vestibular symptoms. In some this worsens after CI.
In children may be chronic poor balance
Paroxysmal dizziness triggered by exertion

WVA Symptoms: Hearing loss
Often normal neo-natal hearing
HL usually develops in first few years
HL can develop in teens
Very rarely in adulthood

WVA Conservative Treatment
Avoidance of head trauma is the key. Children cannot be excluded from all sports.
Let them do what has caused them no harm.
Avoid contact sports
Avoid diving, weightlifting, and wind instruments
Very rare after commercial flights
Hearing aids are the main stay

In Conclusion:
Balance is important
Children are different from adults – you need specific paediatric competences to work with children
Think about the vestibular system when managing deaf children
Investigation and management is multidisciplinary