Endolymphatic hydrops in children

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Introduction

Idiopathic Meniere's Disease (MD) is one of the commonest vestibular disorders in adults that presents with episodic vertigo, tinnitus and aural fullness/sensorineural hearing loss. In the early part of the disease, they may present with third window conductive hearing loss[1]. The Barany criteria defines probable or definite MD by a set of criteria[1]. MD is attributed to an accumulation of endolymphatic fluid in the membranous labyrinth (endolymphatic hydrops ELH) and can be due to primary or secondary causes. The idiopathic variety is primary whilst secondary endolymphatic hydrops can be due to various causes e.g. head injury, brain tumours, autoimmune ear conditions, vestibular migraine (VM) and metabolic conditions[2]. The secondary variety is called Meniere's syndrome (MS). MD in children is very rare due to endolymphatic fluid metabolism and MS is more common[3,4]. Diagnosis in children depends on a robust medical algorithm for best outcomes[3,4]. We present 4 such cases of MS in children who have had extensive investigations to detect the secondary causes of MS for the first time in literature.·

Barany Criteria for Meniere's Disease/Syndrome

Definite Meniere's Disease/Syndrome

A. Two or more spontaneous episodes of vertigo each lasting 20 minutes to 12 hours
B. Audiometrically documented low- to medium frequency sensorineural hearing loss in one ear, defining the affected ear on at least one occasion before, during or after one of the episodes of vertigo
C. Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear
D. Not better accounted for by another vestibular diagnosis

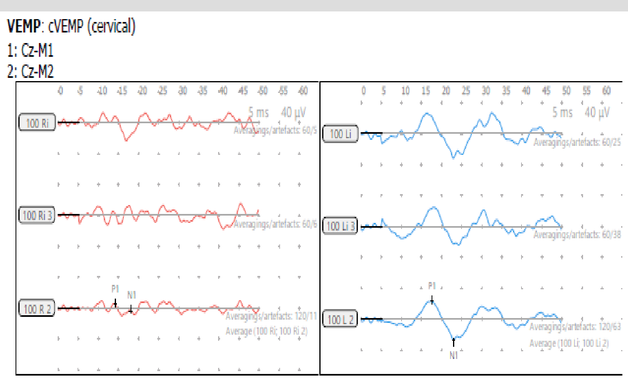
Probable Meniere's Disease/Syndrome

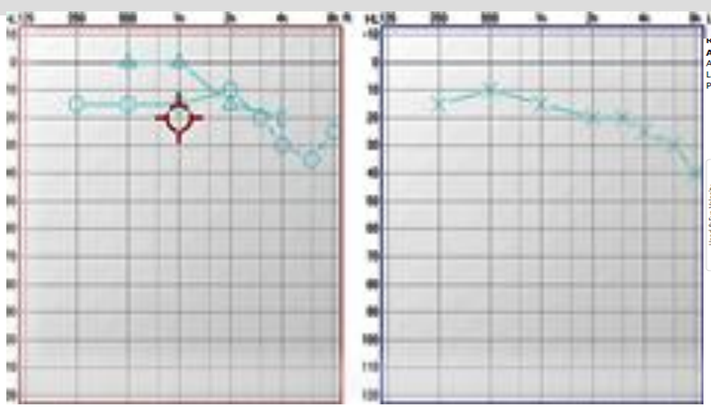
A. Two or more episodes of vertigo or dizziness, each lasting 20 minutes to 24 hours.
B. Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear
C. Not better accounted for by another vestibular diagnosis

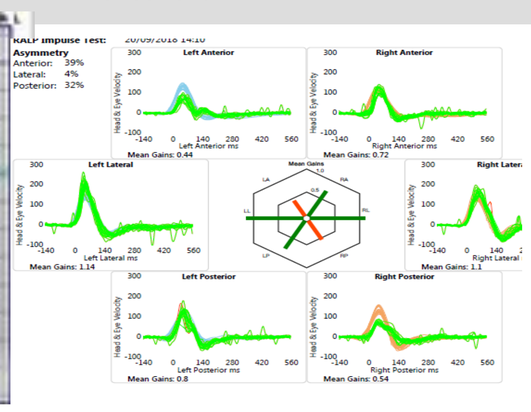
The Children

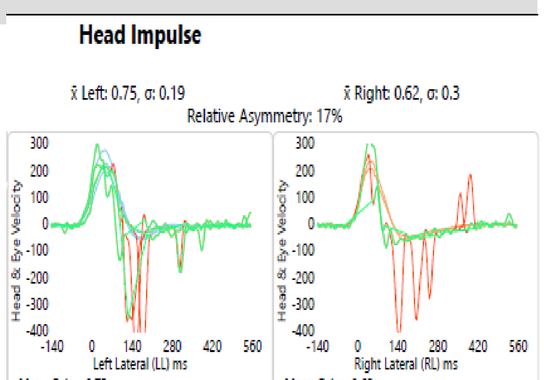
Out of 580 children seen in Alder Hey paediatric vestibular clinic between June 2018 and December 2019, only 0.7% fulfilled Barany criteria for definite or probable ELH. They all underwent audiovestibular investigations with peripheral hearing test battery, objective vestibular quantification with videonystagmography (VNG), video head impulse test (vHIT), suppression head impulse test (SHIMP), cervical vestibular evoked myogenic potential test (cVEMP) and static posturography in addition to a genetic, infective, metabolic and autoimmune profile; average age being 14 years.

Feature	Child 1	Child 2	Child 3	Child 4	Feature	Child 1	Child 2	Child 3	Child 4
History	Classical definite MS +Tullio	Classical definite MS with headache	Classical definite MS with VM and recent onset travel sickness	Probable MS with VM	**Imaging**	Normal	Normal	Normal	Orbit, parietal and pituitary involvement of tumour
Family history	Absent	Present	Absent	Absent	**Inflammatory markers**	Raised ESR and IgA	Raised ESR and Hep 2 positive ANA	Raised ESR	Normal
Aetiology	Coeliac disease	Hep 2 positive ANA	One episode of acute vestibular event	Cranial Langerhans histiocytosis treated with chemotherapy	**Other medical comorbidities**	Coeliac disease	Nil	Nil	Pan hypopituitarism diabetis insipidus
Hearing	Flat 30dBHL mild SNHL right	Bilateral mixed loss in high frequencies	Low frequency 30 dBHL CHL left	Normal	**Other investigations**	Normal	Normal	Normal	Normal
Balance	Functional deficit; positive headshake; absent cVEMP right	Functional deficit and positive HT	Functional deficit and SHIMP asymmetry	Functional deficit; abnormal SHIMP and OCR	**Treatment and outcome**	Betahistine, diuretics and intratympanic steroids; cognitive support; excellent	Betahistine and prochloperazine; excellent	Betahistine and topiramate; excellent	Betahistine and propranolol; cognitive support; excellent









. **cVEMP Child 1** **PTA Child 2** **vHIT Child 3** **SHIMP Child 4**

Discussion

ELH is rare in children. In our cohort, the observed systemic/cranial autoimmune disease as well as vestibular migraine are recognised associations of ELH. ELH can present with a conductive element or a mixed hearing loss due to a third window effect. MS in children follow a different trajectory to those in adults probably as a result of the difference in stria vascularis integrity. In a vast majority, the cause is secondary, so active effort is recommended to detect an aetiology. Children are symptomatic, therefore, the diagnosis should be reliable as this will dictate management. Holistic management with pharmacological/vestibular rehabilitation/cognitive intervention leads to excellent prognosis. This is the first study investigating secondary causes of paediatric ELH in detail.

Conclusions

The condition ELH in children can be reliably identified with a rigorous paediatric diagnostic algorithm that must include the vestibular system and systemic investigations to detect a cause. Management is rewarding with a favourable outcome.

References

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