Endolymphatic hydrops in children

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Introduction

Idiopathic Ménière’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. The Barany criteria defines probable or definite MD by a set of criteria. 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. 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. Diagnosis in children depends on a robust medical algorithm for best outcomes. 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

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<tr>
<th>Definite Meniere’s Disease/Syndrome</th>
<th>Probable Meniere’s Disease/Syndrome</th>
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<tr>
<td>A. Two or more spontaneous episodes of vertigo each lasting 20 minutes to 12 hours</td>
<td>A. Two or more episodes of vertigo or dizziness, each lasting 20 minutes to 24 hours.</td>
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<td>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</td>
<td>B. Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear</td>
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<td>C. Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear</td>
<td>C. Not better accounted for by another vestibular diagnosis</td>
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<td>D. Not better accounted for by another vestibular diagnosis</td>
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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.

The children presented to Alder Hey Children’s Hospital with several features that are typical of Meniere’s Disease (MD) in adults. The children all had a variable infective, metabolic, autoimmune and/or genetic aetiopathologies including coeliac disease, hypothyroidism, diabetes and Langerhans cell tumour. They all underwent extensive investigations to detect the secondary causes of MS for the first time in literature.

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