Meniere's syndrome/endolymphatic hydrops in children

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This session

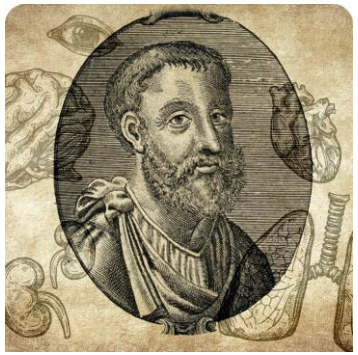
- Brief history of MD
- Pathophysiology of MD
- Diagnostic criteria of MD
- Epidemiology and aetiology of MS in children
- The clinical algorithm to diagnose MS in children
- Treatment of MS in children
- The Alder Hey series
- Conclusions

Brief history of MD



…..vertigo representing a primary affection of the head is preceded by ringing of the ears, pain and heaviness in the head, or a disturbance of the sense of smell and damage of the sensory parts originating there……..

Archigenes of Apamea, 1st century AD

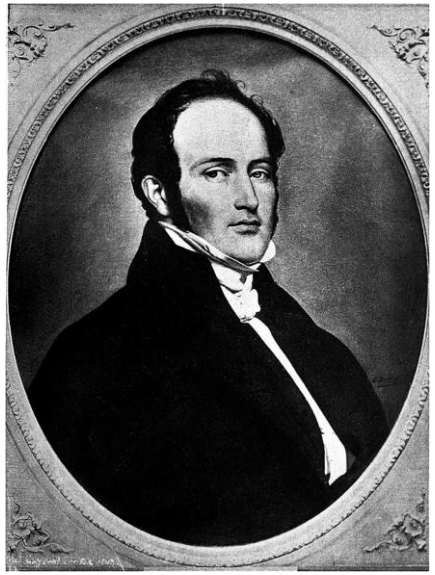






Picked up by Galen (2nd Century AD); Rhazes (9th century AD) and Avicenna (10th century AD)

And then a long wait until 19th century……



Prospere Meniere (1799-1862)
1861

II. *Sur une forme de surdité grave dépendant d'une lésion de l'oreille interne*, par M. Ménière. (*Commissaires :* MM. Cruveilhier, Baillarger et Barth.)

(Extrait par l'auteur.)

L'auteur résume ce travail en un certain nombre de propositions :

1° Un appareil auditif, jusque-là parfaitement sain, peut devenir tout à coup le siége de troubles fonctionnels, consistant en bruits de nature variable, continus ou intermittents, et ces bruits s'accompagnent bientôt d'une diminution plus ou moins grande de l'audition.

2° Ces troubles fonctionnels ayant leur siége dans l'appareil auditif interne, peuvent donner lieu à des accidents réputés cérébraux, tels que vertiges, étourdissements, marche incertaine, tournoiements et chute, et de plus ils sont accompagnés de nausées, de vomissements et d'un état syncopal.

3° Ces accidents, qui ont la forme intermittente, ne tardent pas à être suivis de surdité de plus en plus grave, et souvent l'ouïe est subitement et complétement abolie.

4° Tout porte à croire que la lésion matérielle qui est cause de ces troubles fonctionnels réside dans les canaux demi-circulaires

- The auditory apparatus may be suddenly affected causing tinnitus, and diminution of hearing;
- The inner ear is the site which can suddenly be affected causing attacks of vertigo, dizziness, uncertain gait, staggering and falling, and may be accompanied by nausea, vomiting and syncope;
- Attacks are intermittent followed by hearing loss of increasing severity;
- The lesion is likely to be in the semicircular canals.

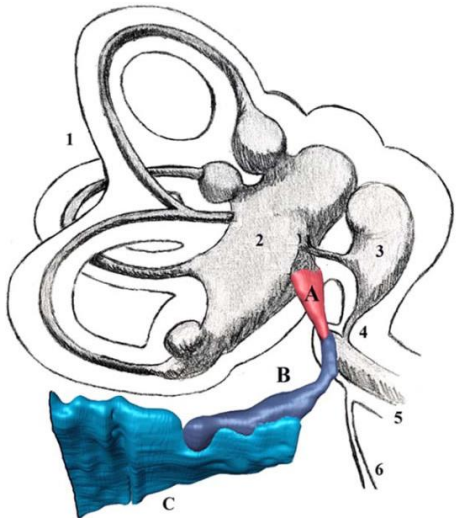
….and 165 years on we are yet to find what happens……

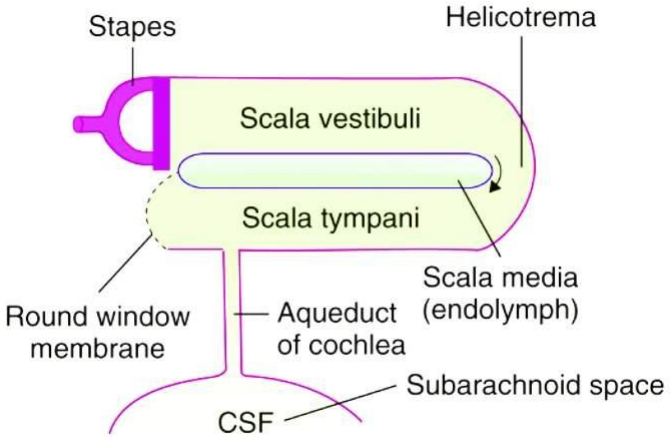
Pathophysiology of MD

- Menière’s disease is a clinical syndrome that consists of episodes of spontaneous vertigo usually associated with unilateral fluctuating sensorineural hearing loss (SNHL), tinnitus and aural fullness

- It is associated with the accumulation of endolymph in the cochlear duct and the vestibular organs in histopathological studies

Lopez-Escamez et al 2015

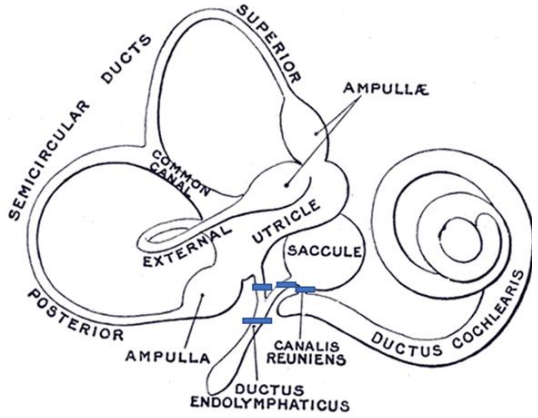


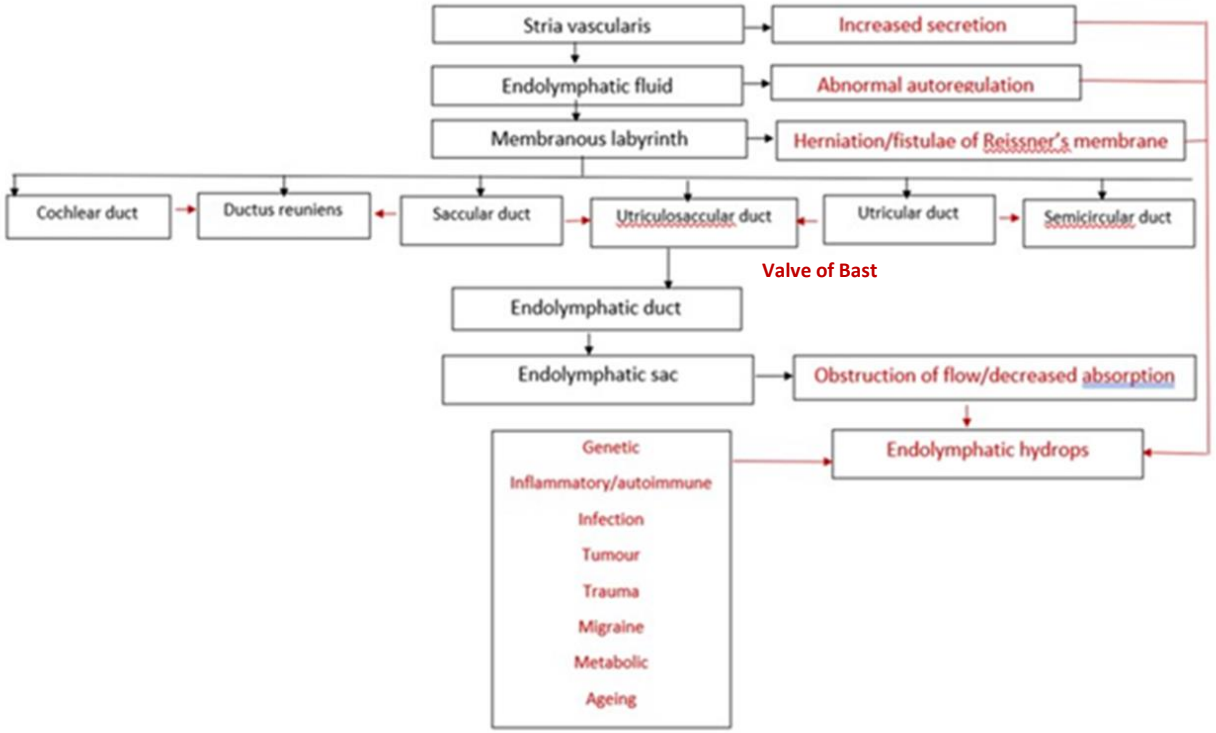


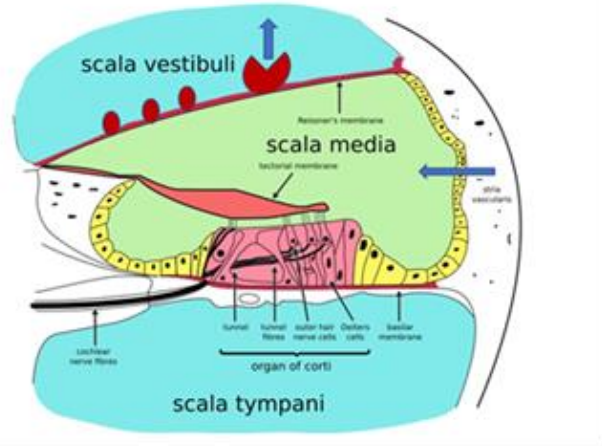
Normal endolymphatic flow

- Endolymphatic fluid is actively secreted by the stria vascularis
- Cochlear endolymphatic compartment larger than vestibular compartment
- The endolymphatic sinus/canalis reuniens is where the 2 flows converge between the ductus reuniens from the saccule, the utricular duct from the utricle and the cochlear duct
- This forms the endolymphatic duct
- The endolymphatic duct terminates in the endolymphatic sac
- The valve of Bast at the utricular – cochlear junction is critical to regulate endolymphatic pressure
- The helicotrema at the apex of the cochlea is the only place where there is mixture of perilymphatic and endolymphatic fluid

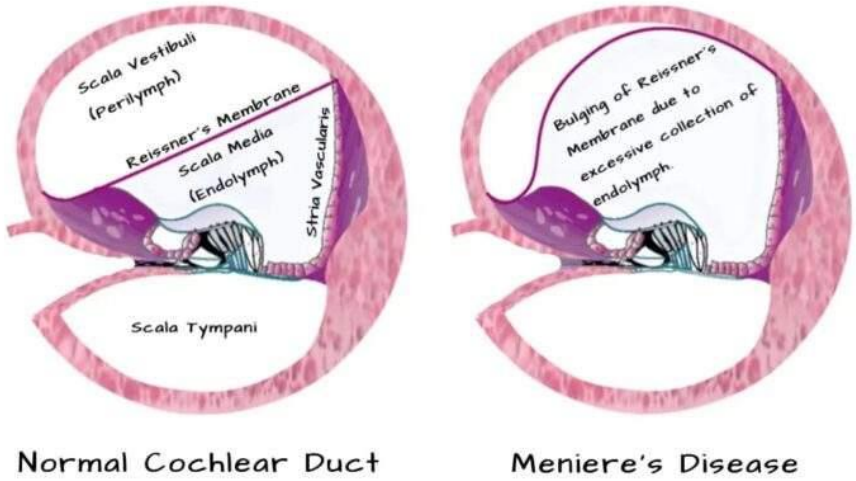
The hydrops can be due to a blockade in the endolymphatic flow or a deranged secretion/absorption ratio between stria vascularis activity and endolymphatic sac







The hallmark is endolymphatic hydrops or increase in the endolymphatic space



Normal Cochlear Duct Meniere's Disease

Other theories

- Hydrostatic
- Ischaemic and vascular
- Electrolyte imbalance
- Saccular otolith debris

Then follows a series of events

Raised endolymphatic pressure.

↓

Dilated cochlear duct may fills the scala vestibuli

↓

Bulged Reissner's membrane herniates through helicotrema into the apical part of scala tympani

↓

Rupture of the membrane that separates perilymph (low potassium extracellular fluid) from endolymph (potassium-rich intracellular fluid)

↓

Mixing of fluids

↓

Sudden change in the rate of vestibular nerve firing

↓

Acute vestibular imbalance producing sudden hearing loss & vertigo.

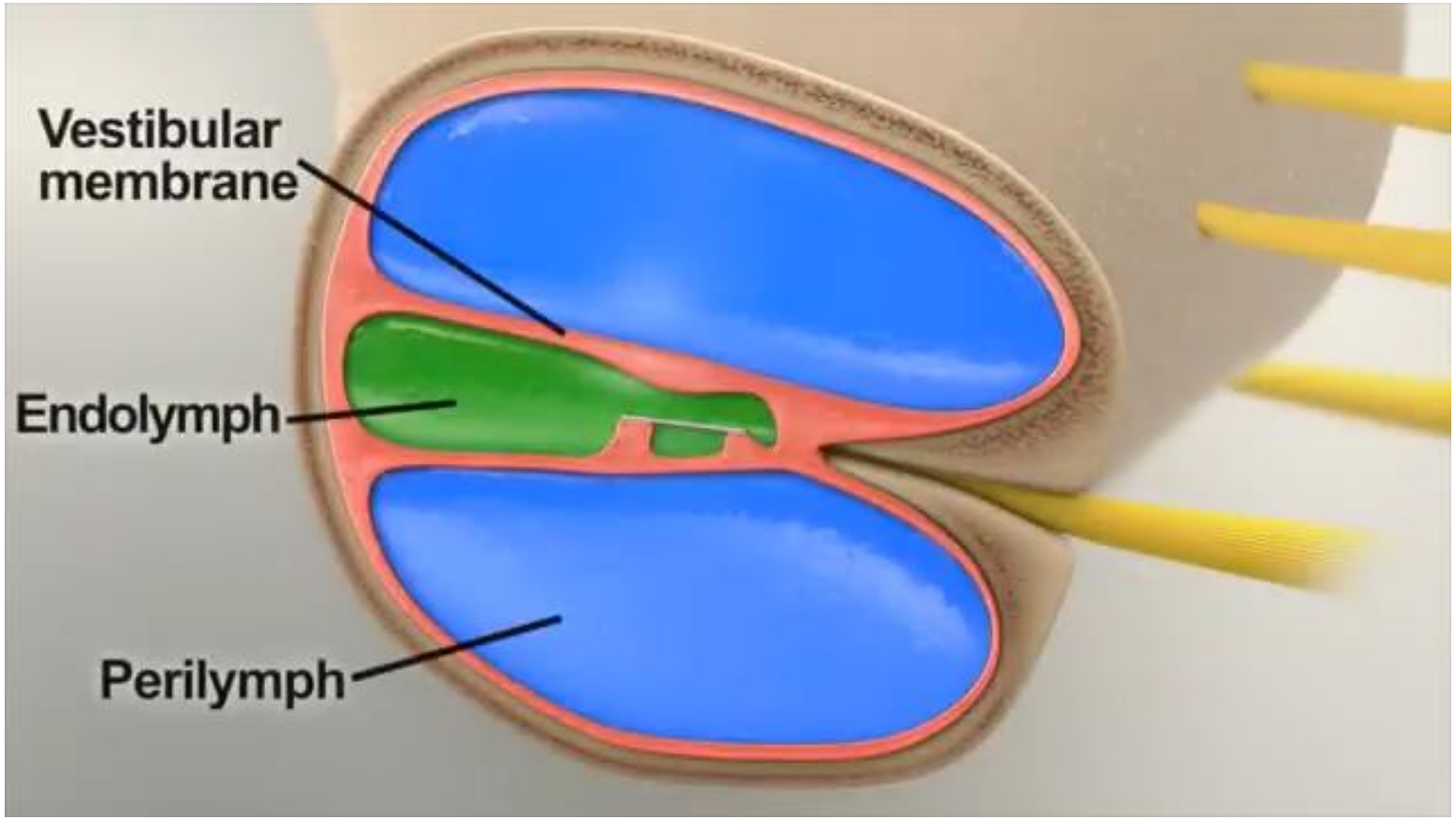
↓

When perilymphatic compartment is restored to normal, symptoms subside.

↓

Aided by the collapse, the rupture heals, and the process is repeated.

https://www.entlecture.com/menieres-disease/ - Bagla 2026



https://www.youtube.com/watch?v=qrk7OyAB_ss

Diagnostic criteria

Diagnostic criteria – definite MD, Barany Society 2015

- 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.

Diagnostic criteria – probable MD, Barany Society 2015

- 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.

Therefore, since there are no specific biomarkers for MD, the diagnosis is based on anamnesis and fulfilment of criteria proposed as well as excluding other vestibular conditions

Important points to consider for diagnosing MS in children

- Remember that in children, you might not be able to glean a history and will have to depend on carers/parents to describe audiovestibular behaviour that demands skill and patience and a thorough training in paediatrics
- In a developing ear, stria vascularis activity changes over time, therefore, spontaneous remission of MS in children is a possibility
- In the early phases, the RM herniations act as micro-fistulae leading to a third window with CHL and other features especially autophony
- This leads to a different phenotype of MS in children both in terms of clinical features and in laboratory diagnosis and as a result often not diagnosed
- Classical features become apparent in mid to late teens
- May exist as a part of a syndrome

Epidemiology and aetiology of MS in children

Prevalence

- Unilateral in 66%
- Spontaneous remission in a third (anecdotal) due to resolution of primary cause, cerebral plasticity and growth of the stria vascularis
- Variable vestibular function test outcomes

MD percentage	Author
1.68% (n = 119)	Riina 2005
1.8% (n = 54)	Balatsouras 2006
Rare (review)	Vacher-Wiener 2008
0.4% (n = 159)	O'Reilly 2010
2.5% (n = 37)	Shupak 2011
Nil (n = 206)	Sommerfleck 2016
8.3% (n = 411)	Lee 2016
6% (n = 374)	Wang 2017
0.7% (n = 580)	Dasgupta 2021

Publications

- A systematic review found only 12 studies with 57 children studied in case series between 1958 and 2024 in terms of paediatric MS (Saniasiaya and Prepageran 2025)
- No study has investigated the aetiology of MD in detail
- All studies performed basic CT/MRI that were normal
- In the cohort studies some familial patterns were observed
- One study (Hausler 1987) observed in their cohort of 14; 35% with a cause accounting for a secondary MD ranging from vestibulocochlear dysplasia, head trauma to infection with mumps and H. influenzae
- Many studies observed an overlap with features of vestibular migraine

Proposed aetiologies

- Genetic - RERGL, PIK3C2G, FAM136A, DTNA, PRKCB, DPT, SEMA3D, USH1A, TECTA, OTOG, COCH, STRC and MYO7A
- Autoimmune – AIED or systemic
- Allergy
- Infection
- Structural defects
- Trauma
- Vascular
- Endocrine/autonomic
- Dietary
- Migraine
- *Idiopathic (commonest variety in adults the classical MD)*

Von Hippel Lindau disease

- AD with tumour suppressor gene at 3p25-26
- Endolymphatic sac tumour in teens and later presenting with MS phenotype
- Variable VFT

X linked hypophosphatemic rickets (XLHR)

- PHEX gene (Xp22.1) mutation FGF23 excess with phosphate metabolic error
- Excess production of EL at stria vascularis level
- Presents with MS phenotype with variable VFT
- Burosomab with good outcome

Comparison between adult and paediatric MD

Parameter	Children	Adults
Pathophysiology	More stria vascularis integrity; evolving SV immune priming and hormonal regulation; more metabolic activity in SV making it more resistant to change unless secondary ELH	Less stria vascularis integrity; immunity primed; less metabolic activity and well-established hormonal regulation making it more vulnerable to change for primary MD
Anamnesis	Difficult to elicit to fit criteria	Easier to elicit to fit criteria
Hearing loss	Variable, may be normal	Usually, low frequency to start with
Vestibular function	Variable	Up to 80% deranged
Aetiology	Almost always secondary	Majority idiopathic
Progression	Slow over several years; spontaneous recovery	May be fast over months; no spontaneous recovery but ‘burn out’

Differential diagnosis of MS in children

- Vestibular migraine – coexists in 50% and usually does not present with aural fullness or hearing loss; the duration of dizziness is prolonged and more intense than MS - MS does not present with photophobia or phonophobia

- *Third window syndromes – may mimic MS but also presents with third window symptoms; remember MS itself can present as third window as it can be classed as a third window – on the other hand the first stage of an SSCDS is a Meniere's phenotype*

- Peripheral cochleo-vestibulopathy (trauma, tumour, infection, inflammation, dysmorphia) – does not present with fluctuating symptoms but remember that this can lead to MS later

- Central vestibular syndromes – usually audiological symptoms are missing and characterised by typical oculomotor abnormalities

Diagnosis in children

Symptoms

- Classical triad might not be described – episodic hearing loss, dizziness, tinnitus
- Aural fullness
- Third window symptoms especially autophony
- Deranged vestibular behaviour
- Cognitive and psychological reactions

In children often surrogate history from carers

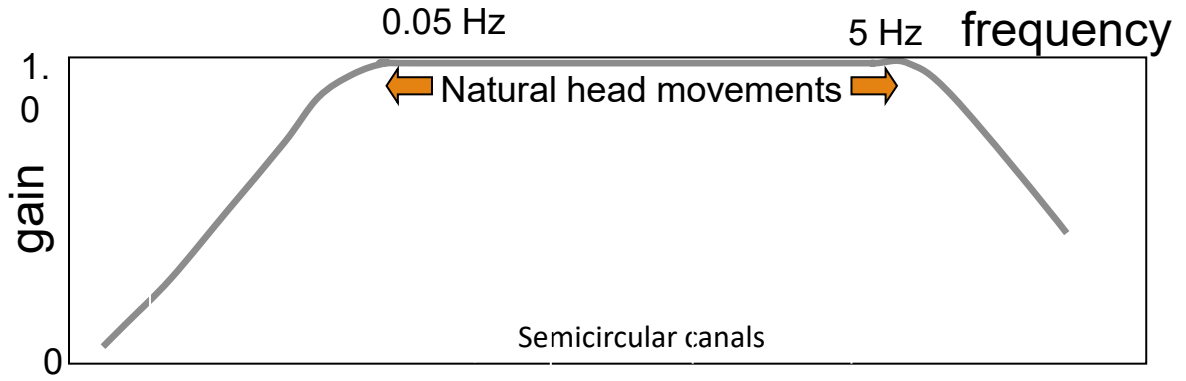
Signs

- Hennebert's sign
- Normal or abnormal PTA with low frequency involvement to start with normal middle ear
- Variable vestibular function tests
- Deranged vestibular behaviour
- Cognitive and psychological reactions

Assessment

Audiometric evaluation

- Middle ear studies
- Pure tone audiometry
- Speech audiometry
- Otoacoustic emissions
- E Coch G (not used)



Spontaneous / gaze tests (*0 Hz*)

With fixation
without fixation





Calorics (*0.003 Hz*)



Mastoid vibration *(0.02 Hz)*

Rotating chair (*0.5 - 1 Hz*)



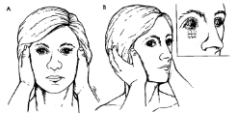
Dynamic visual acuity (*1 -2 Hz*)



Headshake (*2 - 4 Hz*)



Head Thrust (*5 -7 Hz*)



Heave	SVV	oVAR	cVEMP	oVEMP	OCR
High	*Static*	*Mid*	*Low*	*Low*	*Mid*
Utricle	Saccule	US	Saccule	Utricle	US

Musculoskeletal and cardiological assessment

Imaging and blood tests

Hyperventilation test
Hennebert's test
fHIT and SHIMP
Vestibulospinal tests
Static and dynamic posturography

Neurological assessment

- Examination
- Smooth pursuits
- Saccades
- Vertical headshake and perversion
- VOR cancel
- Optokinetic assessment
- Skew deviation
- Caloric disassociation and fixation index
- Oculomotor movements for central signs

Calorics in MD

- Calorics is the oldest objective test used in MD
- Calorics show canal paresis in at least 50% cases
- The postulated reason is that calorics being indicative of an ultra low frequency response close to the static response (nystagmus) denotes ELH rather than vestibular weakness and indicative of active disease (Choi 2017)
- ***More important and promising is the dissociation of calorics and vHIT , i.e. the latter is normal whilst calorics are not (Limviriyakul 2019)***
- Overall, calorics on its own is not a biomarker for MD

Electrocochleography in MD

- Identified in 1977, ECocgG saw significant research until early 2000s as a reliable biomarker where SP/AP ration <0.5 was proposed to be diagnostic of MD with click stimuli
- Later studies identified the relative lack of sensitivity and specificity of the test and as result had fallen out of repute (Hornibrook 2017)
- However, recent research shows that tone burst stimuli (100 dBNHL with SP threshold and AP/SP ratio) have a high sensitivity and specificity (>80%) and is promising (Hornibrook 2017)
- ***This is a resource intense and an invasive procedure and it is not needed***

Video head impulse in MD

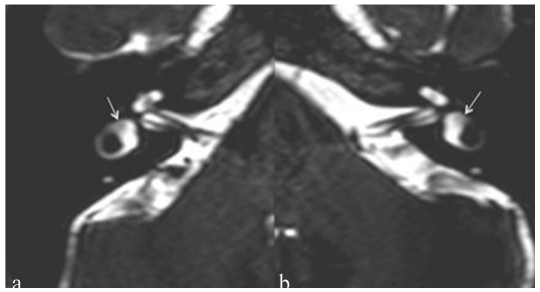
- vHIT usually normal in MD unless there is an obvious hypofunction due to the disease
- VOR gain is not a good indicator of MD but may show fluctuation between phases of the disease, i.e. ictal and post ictal or quiescent (Yacovino 2016)
- Recently, saccades rather than VOR gain appear to be better indicators when the VOR gain is normal (Kaci 2020)
- Overall, vHIT on its own is not a biomarker for MD

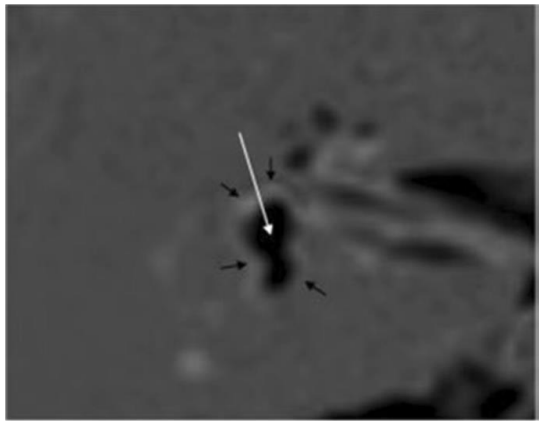
Vestibular evoked myogenic potential in MD

- VEMP results in MD are not confirmatory with conflicting results and study methodology
- ***VEMPS will show lowering of threshold in early stages due to the third window phenomenon*** and can be deranged in advanced disease with vestibular weakness
- Amplitude asymmetry evolution and change may indicate different phases of the disease (Maheu 2017)
- ***Recently frequency tuning of VEMP at the 1000Hz/500Hz response is showing promise*** (Singh 2023)
- Overall, VEMP on its own is not a biomarker for MD

MRI in MD

- Up to 80% sensitivity and 80% specificity (Xie 2021)
- More the volume of the hydrops better is the MRI correlation and correlates with AV function (Gurkov 2011)
- Still in infancy and lack of rigorous scientific evidence for routine use
- Does not pick up probable MD in most cases
- Time consuming and resource intense, ***may be very difficult for children; toxicity of contrast***
- Consensus yet to be achieved regarding qualitative and quantitative volumetric analysis; protocol







Multi frequency/wideband tympanometry

- Limited studies but non invasive and easy to perform requiring dedicated software
- Likely due to ELH pressure on stapes footplate leading to alteration of middle ear elasticity and stiffness
- Probe at different frequencies from 226 Hz to 8000 Hz
- Measured parameters are resonant frequency and absorbence both of which are abnormal in MD (Meng 2022)
- Too early to infer

A useful protocol for diagnosis

Detailed history/anamnesis

Suggested definite/probable MD

Full physical clinical examination

- Audiovestibular
- Cardiology
- Neurology
- Ophthalmology
- Musculoskeletal
- General

Detailed audiometry

- PTA
- Tympanometry (including WBT)
- SRT
- OAE

Vestibular quantification

- Caloric disassociation vHIT
- VNG nystagmus/Hennebert
- vHIT normal VOR gain but saccades
- ***VEMP low threshold/AR/increased frequency tuning ratio***
- Tone burst ECochG

Other tests

- Autoimmune marker
- MRI
- HRCT
- Blood tests
- Genetic testing

High index of suspicion when:

- Any 2 of episodic dizziness with ***aural fullness/hearing loss and tinnitus*** that takes minutes to hours to recover
- No photophobia or phonophobia in pure MS but present when VM
- Variable hearing loss – normal to CHL to mixed to SNHL
- Negative BC threshold in early stages
- Variable OAE normal to absent
- Balance symptoms and abnormal vestibular function tests especially cVEMP tuning
- Stigmata of syndromic medical condition

Treatment of MS in children

Diagnosis of allergy and avoidance of allergen - Often overlooked, any allergy should be treated rigorously although the link between allergy and MD is not clearly established, experiments in the animal kingdom are suggestive (Weinreich and Agarwal 2014)

Auditory rehabilitation : Auditory rehabilitation with hearing aids is difficult due to the nature of the hearing loss (Valente 2006); mapping of cochlear dead regions and DPOAE growth studies may be beneficial

Tinnitus retraining is important as pharmacological intervention may not affect tinnitus

Vestibular rehabilitation

May play a role in the chronic phase of the disease for those receiving medications or intra tympanic intervention (Gottshall et al 2005)

Stress management and cognitive therapies:

MD generates distress and negative behavioural responses, the management of MD is incomplete without addressing these issues; includes applied relaxation, breathing exercises, stress management and CBT (Yardley 2006)

Lifestyle changes

Hydration, salt intake, diet, low grade exercises

Pharmacological management

Class of drug	Agents in use	Evidence level	Hearing outcome	Disorientation outcome
Histamine analogue	Betahistine	Empirical; Cohrane not validated;	Nil	Improvement
Antihistamines	Cinnarizine (Ca channel blocker)	Empirical	Nil	Improvement
Diuretics	Thiazides, loop and carbonic anhydarse inhibitor	Empirical, Cohrane not validated	Nil	Improvement
Phenothiazines	Prometazine, prochloperazine	Some evidence	Nil	Alleviation

Class of drug	Agents in use	Evidence level	Hearing outcome	Disorientation outcome
Steroids – oral or intratympanic	Dexamethasone, methyl/prednisolone	Cochrane statistical validation	40% improved	Significant improvement 80-90%
Intratympanic aminoglycoside	Gentamycin	Cochrane statistical validation	Hearing loss risk	80% improved
Antimigraine therapy	B blockers; TCAD; gabapentin; pizotifen, topiramate	Correlation MD/ migraine established	Nil	Significant improvement
Antiviral	Acyclovir	One paper establishes connection	40% improved	65% improved
Vasodilators	Ca blockers	Empirical	Nil	Some improvement

Surgical management (seldom needed in children)

- Ventilation tube application
- Tenotomy of the middle ear muscles
- Endolymphatic duct blockage at the level of the valve of Bast
- Endolymphatic sac surgery
- Vestibular neurectomy
- Labyrinthectomy

The Alder Hey experience




Audio Vestibular Medicine
Vestibular Laboratory



- 0.7% of dizziness in children, all fulfilling EAONO/Barany criteria for diagnosis of MD (2018 – 2019) – average age 14 years
- Full audiological quantification along with PTA, OAE and SRT
- Full vestibular quantification with VNG especially Hennebert's sign, vHIT and VEMP
- MRI, CT scan, autoimmune profile, metabolic screen in all instances
- Secondary ELH in 90%, only in one case, no cause found
- Aetiologies – intracranial pathologies, VM, AIED, systemic AIED, pan allergic spectrum, trauma, genetic, metabolic

Other symptoms

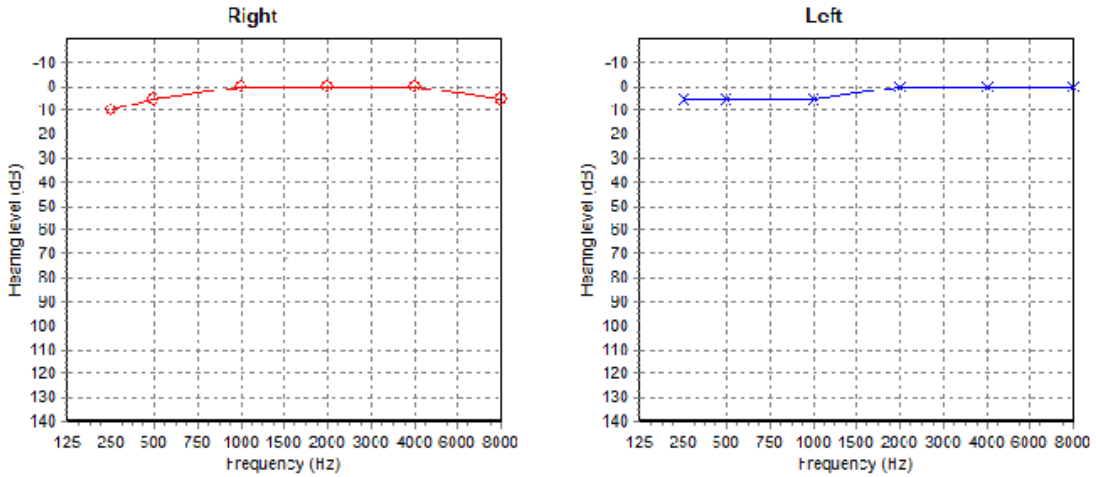
- Tullio and autophony
- Imbalance
- VM symptoms
- Psychological symptoms
- Cognitive symptoms
- Missing school
- Family history positive

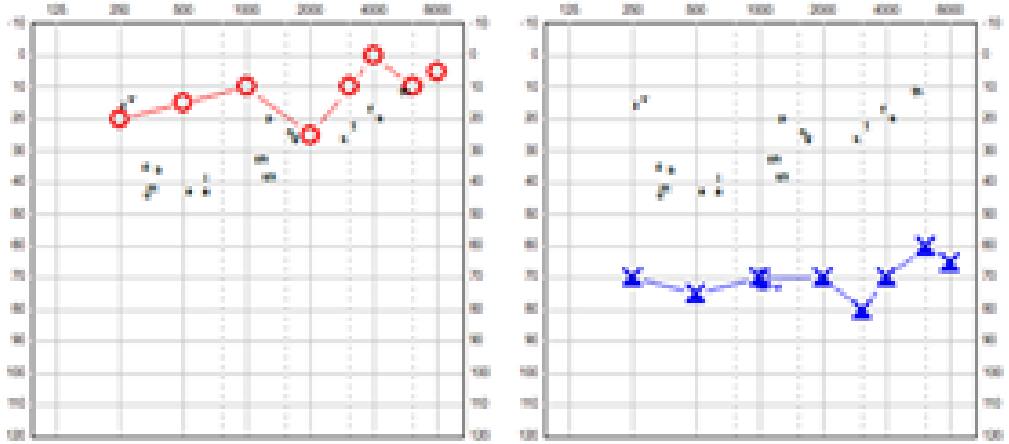
Signs

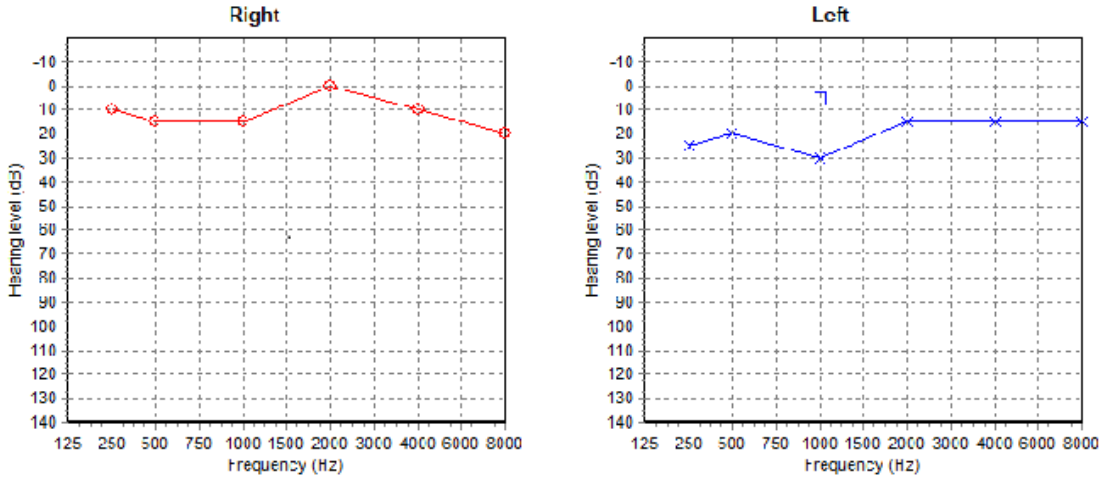
- Unilateral in about 75%
- CHL/SNHL and mixed hearing loss
- Multifrequency VNG vestibular weakness
- Occasional vHIT and SHIMP abnormalities
- VEMP abnormalities
- Missing school
- Family history positive

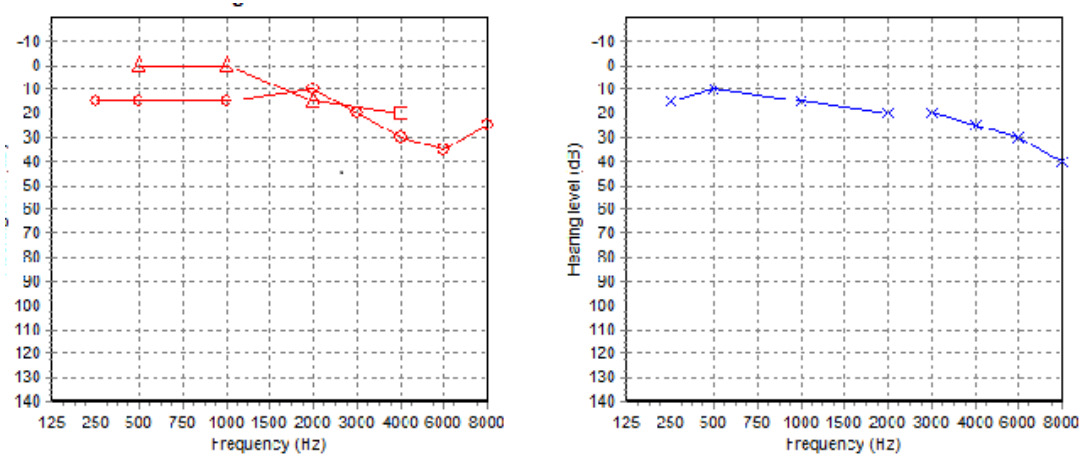
Investigations

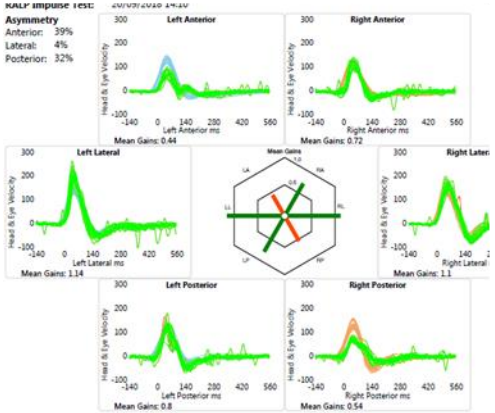
- Imaging normal to intracranial pathologies (tumour and demyelination)
- Raised inflammatory markers in majority
- Systemic AIED (coeliac)

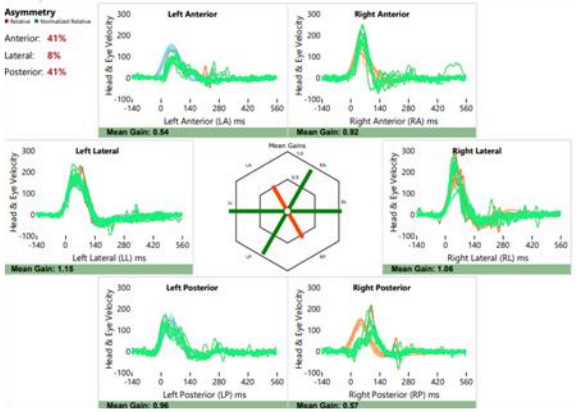

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Hearing level (dB)
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125 250 500 750 1000 1500 2000 3000 4000 6000 8000
Frequency (Hz)

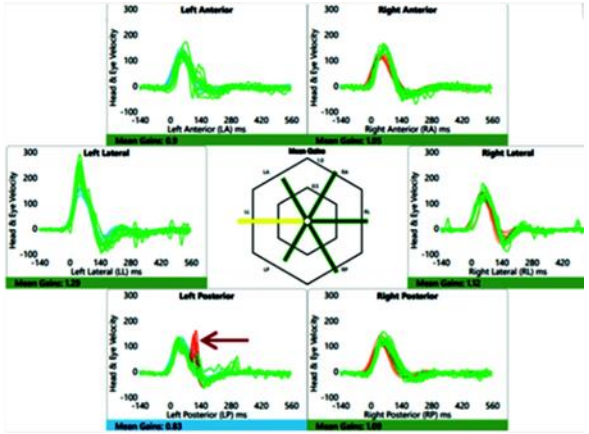


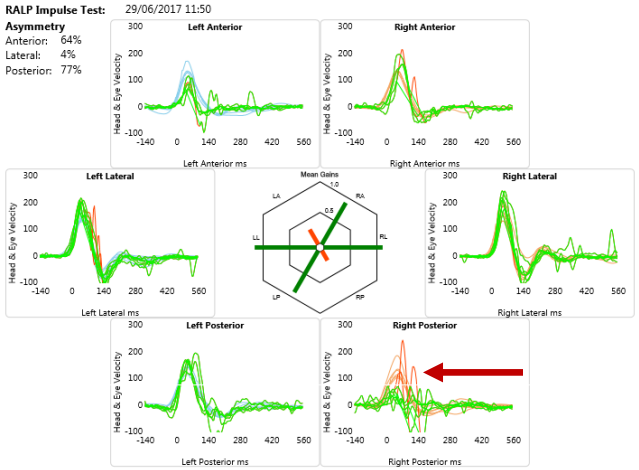

Right
Left
Hearing level (dB)
-10 0 10 20 30 40 50 60 70 80 90 100 110 120 130 140
125 250 500 750 1000 1500 2000 3000 4000 6000 8000
Frequency (Hz)

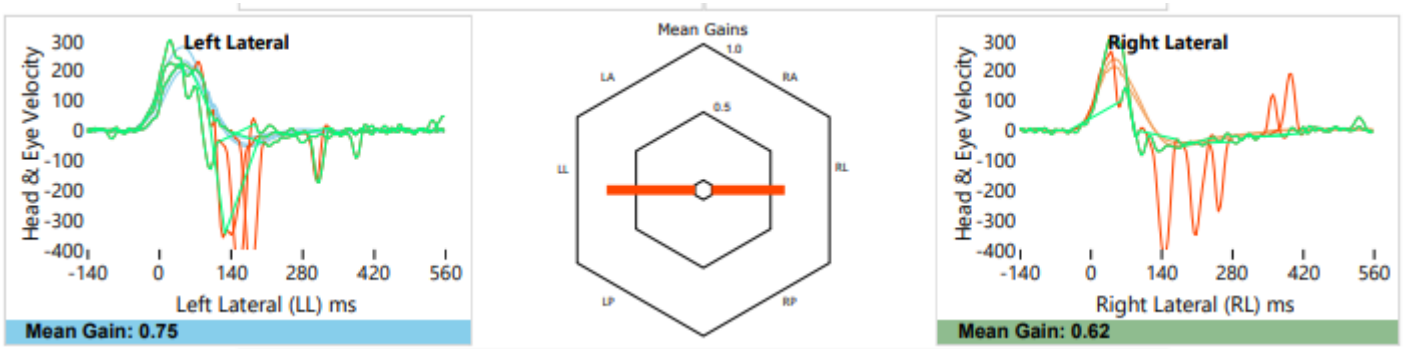

Hearing level (dB)
-10 0 10 20 30 40 50 60 70 80 90 100 110 120 130 140
125 250 500 750 1000 1500 2000 3000 4000 6000 8000
Frequency (Hz)

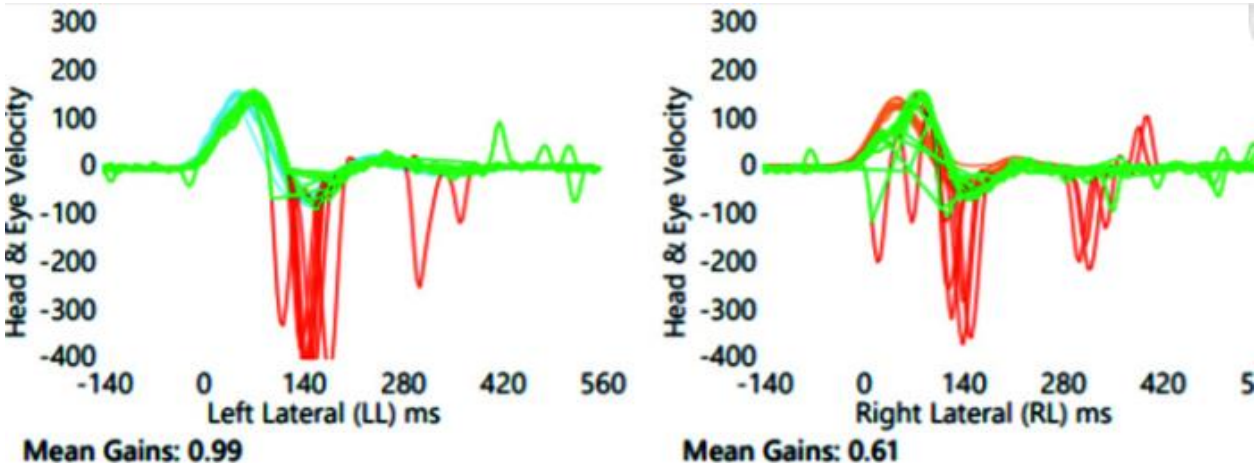

Asymmetry
Anterior: 39%
Lateral: 4%
Posterior: 32%
Left Anterior
Right Anterior
Left Lateral
Right Lateral
Left Posterior
Right Posterior

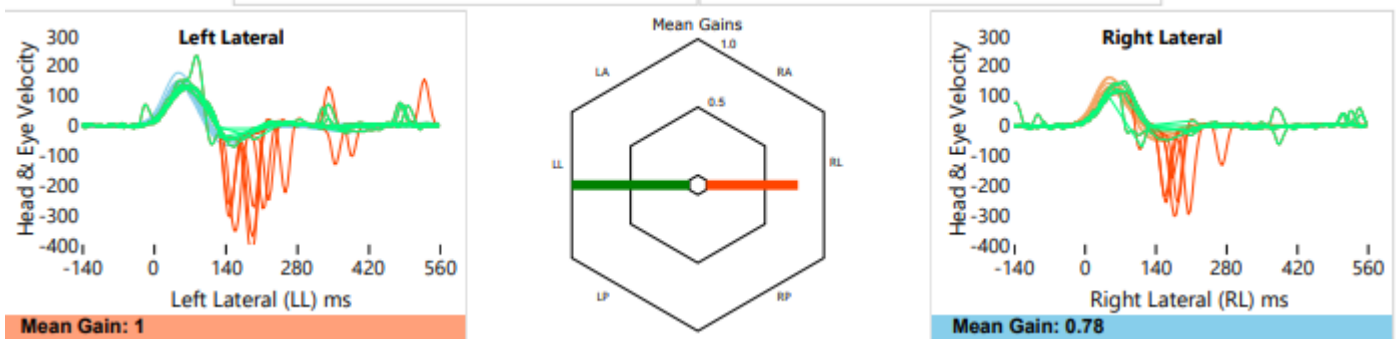

Asymmetry
Anterior: 41%
Lateral: 8%
Posterior: 41%
Left Anterior
Right Anterior
Left Lateral
Right Lateral
Left Posterior
Right Posterior

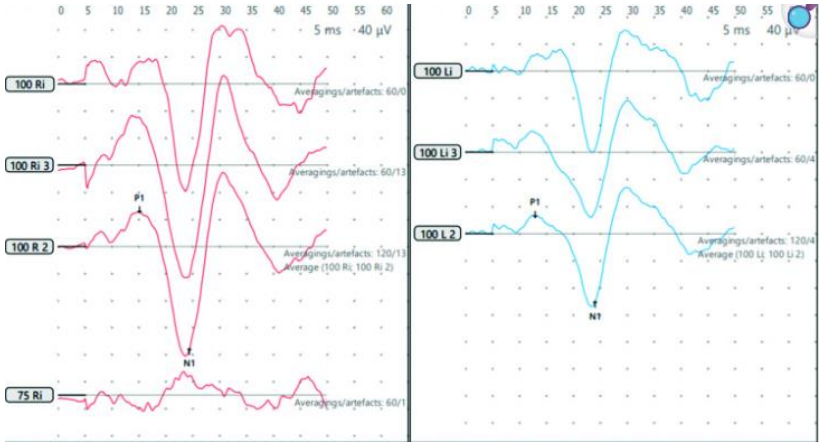

Left Anterior
Right Anterior
Left Lateral
Right Lateral
Left Posterior
Right Posterior


RALP Impulse Test: 29/06/2017 11:50
Asymmetry
Anterior: 64%
Lateral: 4%
Posterior: 77%
Left Anterior
Right Anterior
Left Lateral
Right Lateral
Left Posterior
Right Posterior

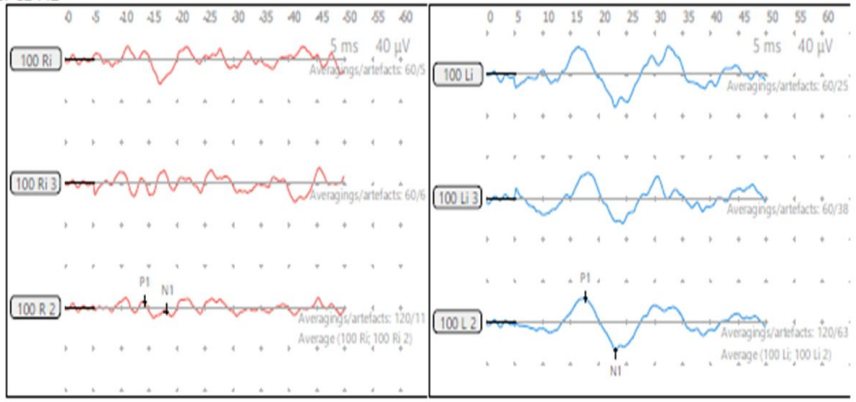

Left Lateral
Head & Eye Velocity
300
200
100
0
-100
-200
-300
-400
-140
0
140
280
420
560
Left Lateral (LL) ms
Mean Gain: 0.75
Mean Gains
1.0
0.5
LA
RA
LL
RL
LP
RP
Right Lateral
Right Lateral (RL) ms
Mean Gain: 0.62


Head & Eye Velocity
300
200
100
0
-100
-200
-300
-400
-140
0
140
280
420
560
Left Lateral (LL) ms
Mean Gains: 0.99
Right Lateral (RL) ms
Mean Gains: 0.61

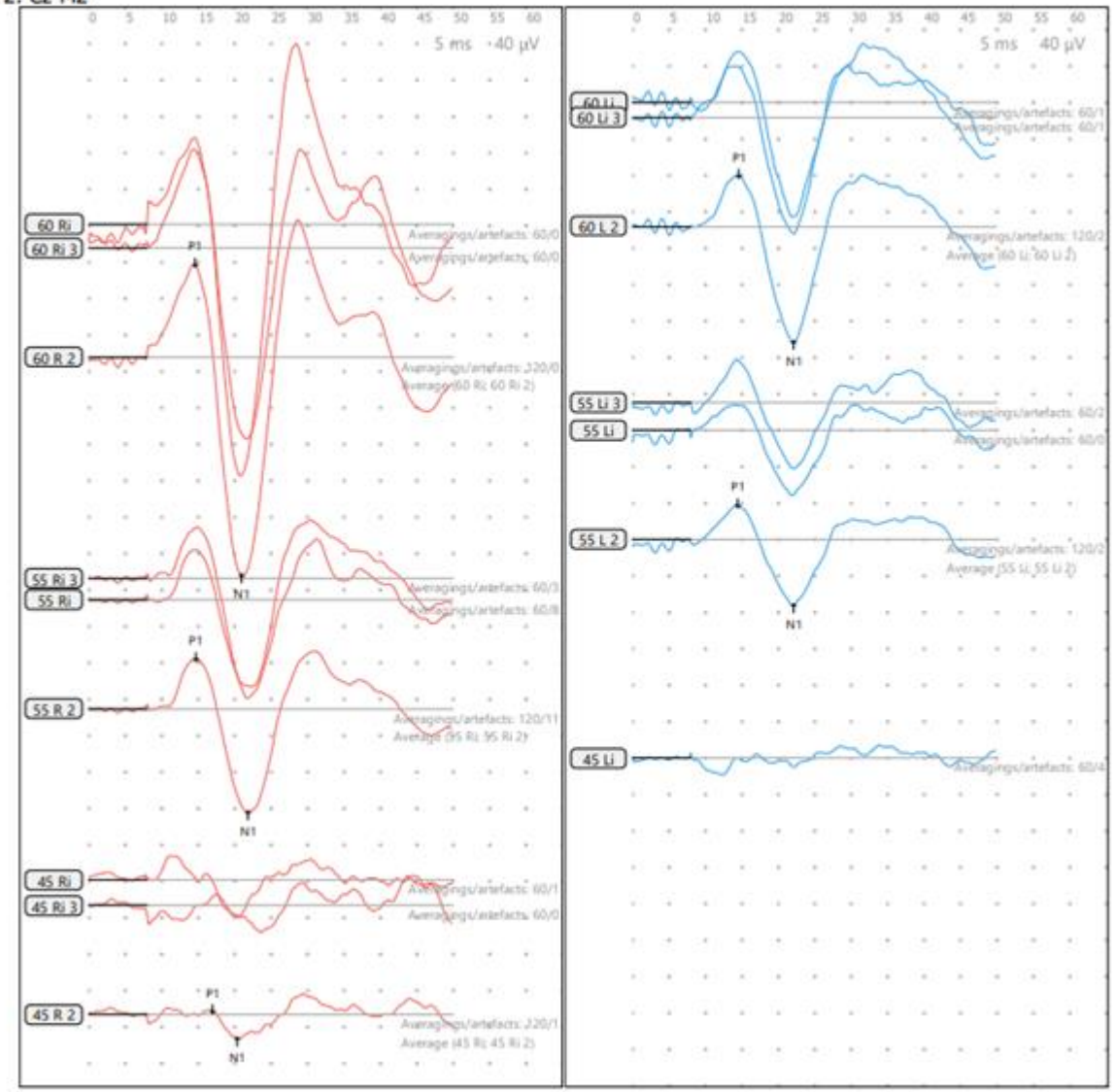

Left Lateral
Head & Eye Velocity
300
200
100
0
-100
-200
-300
-400
-140
0
140
280
420
560
Left Lateral (LL) ms
Mean Gain: 1
Mean Gains
1.0
0.5
LA
RA
LL
RL
LP
RP
Right Lateral
Right Lateral (RL) ms
Mean Gain: 0.78



VEMP: cVEMP (cervical)
1: Cz-M1
2: Cz-M2



VEMP: cVEMP (cervical) BC
500Hz VEMP
1: Cz-M1
2: Cz-M2



cVEMP 500 Hz

cVEMP	Right					Left					
Sound intensity	Latency ms		Amplitude P1-N1 µV	EMG µV	Rectified Amplitude µV	Latency ms		Amplitude P1-N1 µV	EMG µV	Rectified Amplitude µV	% Rectified Asymmetry
	P1	N1				P1	N1				
BC 60dBnHL	12.3	20.5	247.0	124.3	2.0	11.8	20.1	226.6	103.8	2.2	- 4.76%
BC 55dBnHL	12.3	19.6	174.6	102.1	1.7	11.9	20.0	258.8	160.8	1.6	3.03%
BC 45dBnHL	13.1	22.1	86.5	91.8	0.9	12.8	21.7	98.3	163.9	0.6	20.00%
BC 35dBnHL	13.1	22.9	31.3	75.6	0.4	15.3	21.2	33.4	176.9	0.2	3.03%
BC 25dBnHL	NCR	NCR	NCR	75.2	NCR	NCR	NCR	NCR	170.5	NCR	NA

cVEMP 1000 Hz

cVEMP	Right					Left					
Sound intensity	Latency ms		Amplitude P1-N1 µV	EMG µV	Rectified Amplitude µV	Latency ms		Amplitude P1-N1 µV	EMG µV	Rectified Amplitude µV	% Rectified Asymmetry
	P1	N1				P1	N1				
BC 60dBnHL	10.5	16.7	67.0	55.8	1.2	10.7	16.1	118.3	119.8	1.0	9.09%
BC 55dBnHL	10.2	17.3	37.0	75.3	0.5	10.8	16.9	40.7	90.9	0.4	11.11%
BC 45dBnHL	11.1	19.3	70.7	91.0	0.8	NCR	NCR	NCR	100.0	NCR	100.00%

High amplitude, low threshold

Tuning 1000Hz/500Hz = 1/2.2 = 0.45 at 60 dBnHL

oVEMP 500 Hz

oVEMP	Right				Left				
Sound intensity	Latency ms		Amplitude N1-P1 µV	EMG µV	Latency ms		Amplitude N1-P1 µV	EMG µV	% Asymmetry
	N1	P1			N1	P1			
BC 60dBnHL	10.7	15.5	28.2	8.9	10.2	14.0	26.1	9.1	3.9
BC 55dBnHL	10.7	16.1	21.3	9.9	10.6	14.3	15.7	12.3	15.1
BC 45dBnHL	10.5	16.1	8.7	9.7	12.6	17.7	2.9	12.0	49.3
BC 35dBnHL	10.3	16.4	4.1	10.1	10.3	18.4	11.1	16.2	46.3

oVEMP 1000 Hz

oVEMP	Right				Left				
Sound intensity	Latency ms		Amplitude N1-P1 µV	EMG µV	Latency ms		Amplitude N1-P1 µV	EMG µV	% Asymmetry
	N1	P1			N1	P1			
BC 60dBnHL	8.8	12.7	5.5	12.9	10.2	14.8	3.5	17.4	23
BC 55dBnHL	9.1	15.1	4.0	9.1	10.9	16.3	3.0	13.7	14.4
BC 45dBnHL	11.0	13.0	1.1	10.6	INC	INC	INC	16.4	INC

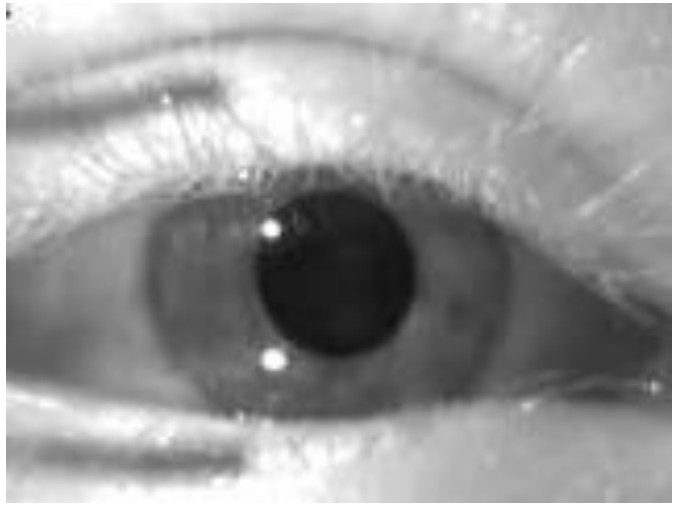
High amplitude, low threshold

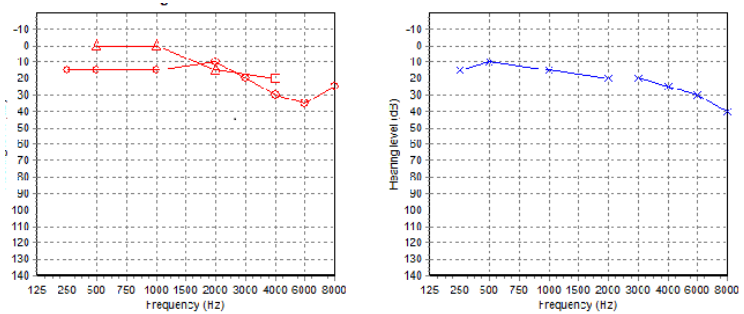
Tuning 1000Hz/500Hz = 17.4/9.1 = 1.9 at 60 dBnHL

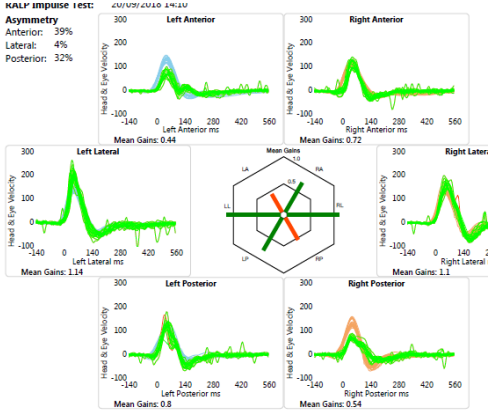
Treatment

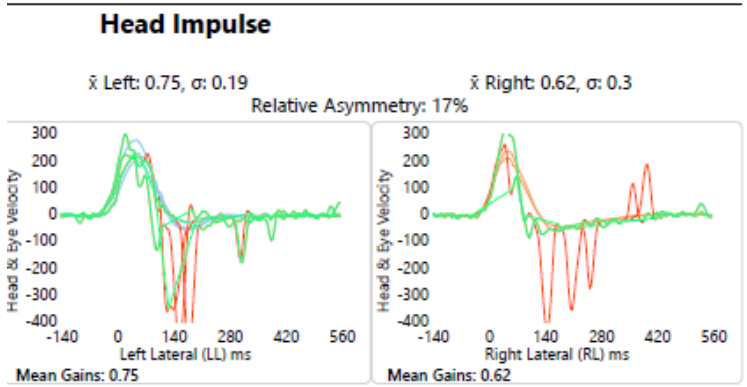
- Betahistine
- Diuretics
- Cinnarizine or prochlorperazine for breakthrough
- Propranolol when VM
- Cognitive and psychological support
- Lifestyle changes
- Intra tympanic steroids required in only 2 cases so far out of circa 30 cases 2018-2025

Excellent symptom control in all cases, vestibular rehabilitation hardly required

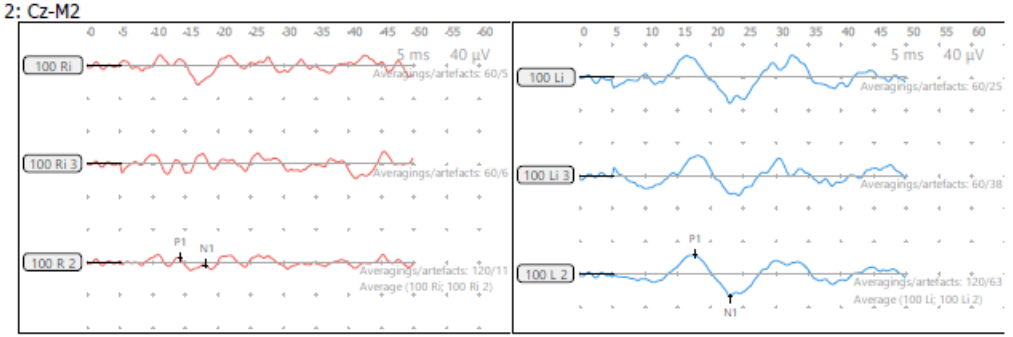


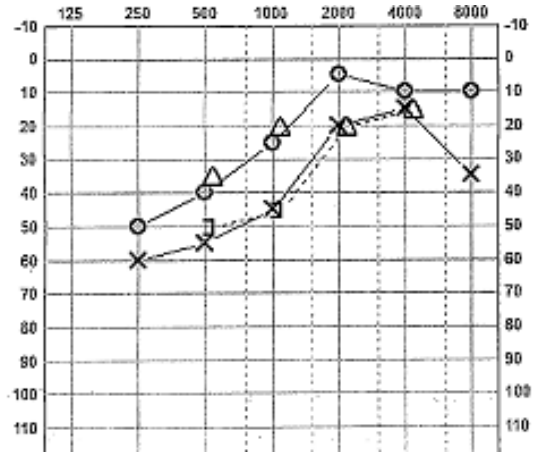


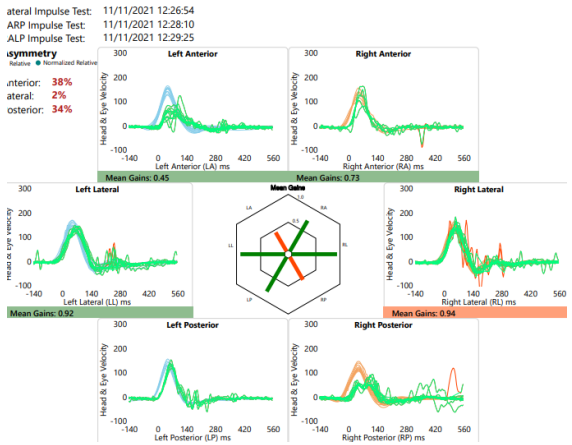


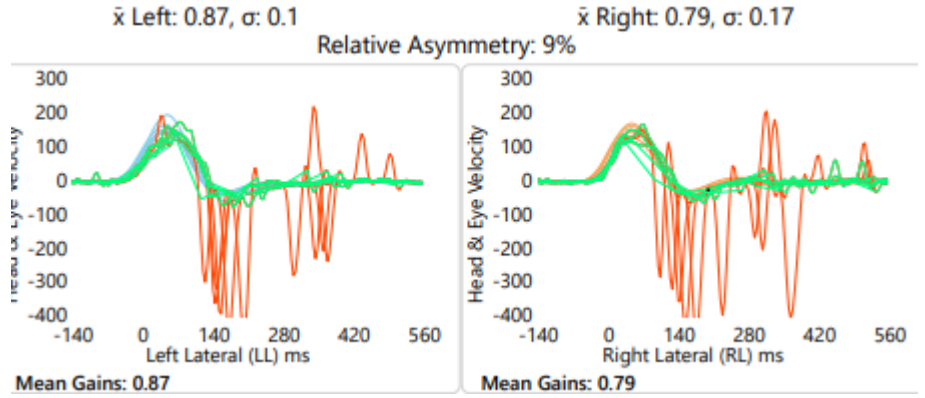


Teenager with coeliac disease and MS – required intratympanic steroids

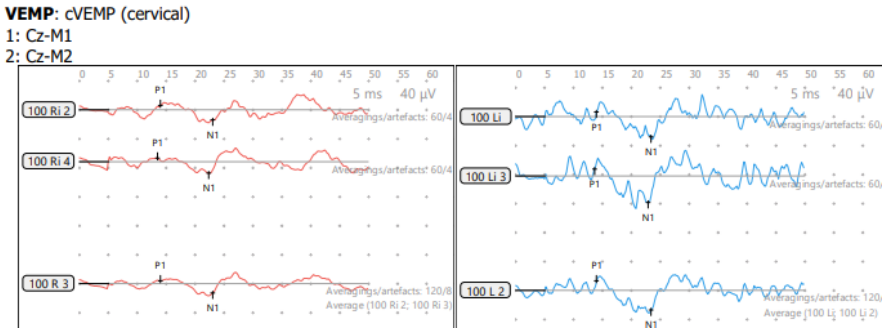












Teenager with XLHR – asymptomatic on burosumab

Conclusions

- Although rare, MD in children is a diagnosis that can be reliably made with a rigorous clinical algorithm
- MD 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
- Children respond to pharmacological management very well and monitoring or surveillance is essential for progression of disease

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- Promoting research and education to enhance understanding and care
- Developing evidence-based guidelines for assessment and management
- Facilitating collaboration through an online community and networking tools
- Raising awareness across pediatric disciplines



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