Latest Diagnostic and Management Aspects of Pediatric Vestibular Medicine

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Learner Outcomes

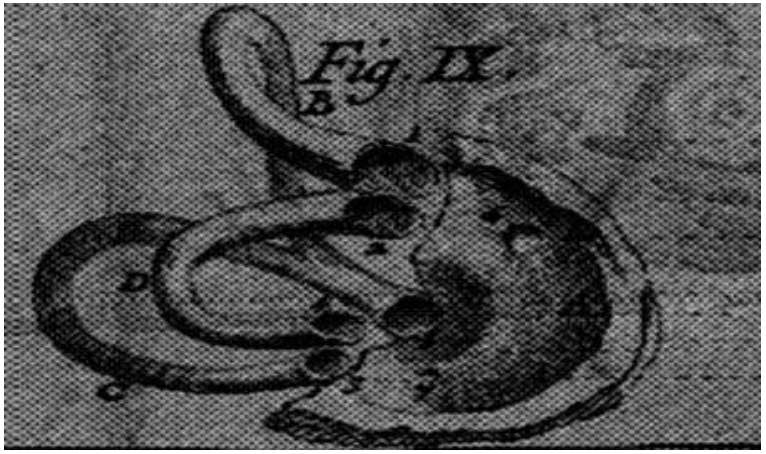
- After this course, participants will be able to list the latest advancements in vestibular diagnostics used to assess vestibular disorders in pediatric patients.

- After this course, participants will be able to discuss the differences between vestibular diagnostic procedures suitable for adults and those appropriate for pediatric populations.

- After this course, participants will be able to describe the utilization of cutting-edge diagnostic tools, such as the video head impulse test and the mastoid vibration test, in pediatric vestibular assessment.

Introduction

We are living in a golden era of vestibular medicine



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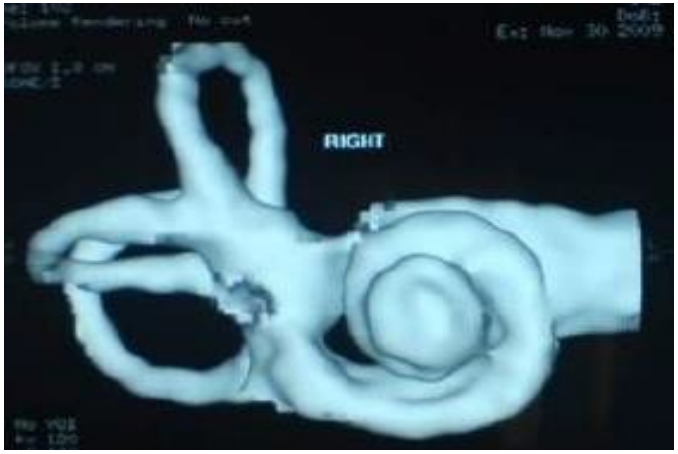


2016

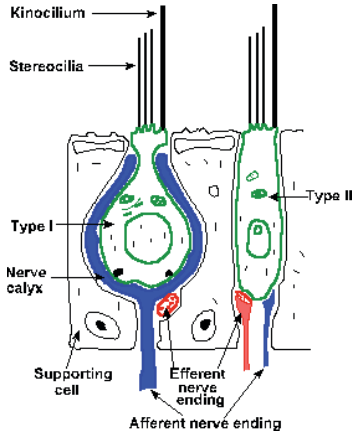
This session discusses advances in paediatric vestibular medicine

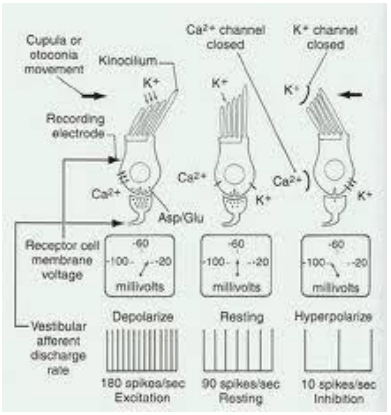
- Advances in paediatric vestibular medicine training
- Advances in vestibular anatomy, physiology and genetics
- Advances in paediatric vestibular history
- Advances in paediatric diagnostic assessment
- Advances in paediatric vestibular imaging
- Advances in paediatric vestibular management
- Advances in paediatric vestibular assessment of compensation

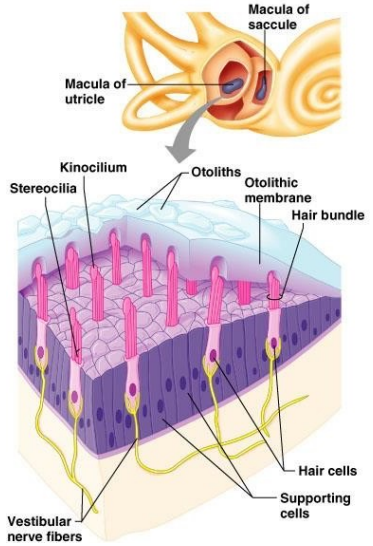
Advances in paediatric vestibular training

- A joint venture by the Barany Society and the International Vestibular Society
- In the UK, full residential programme in Audiovestibular Medicine includes tenures in all paediatric medical disciplines most importantly paediatric neurology, paediatric movement disorders, paediatric ophthalmology and eye movement disorders, paediatric cardiology and developmental paediatrics
- A paediatric vestibular specialist must have a broad knowledge of paediatrics in order to obtain robust insight into a child as a whole and not just the inner ear
- Including paediatric vestibular sessions in flagship otolaryngology/otology/neurotology/ paediatric/neurology/developmental paediatric conferences
- Offer fellowships in designated world centres for paediatric vestibular medicine
- Initiate and organise paediatric vestibular courses all over the world

Advances in knowledge in anatomy, physiology and pathology







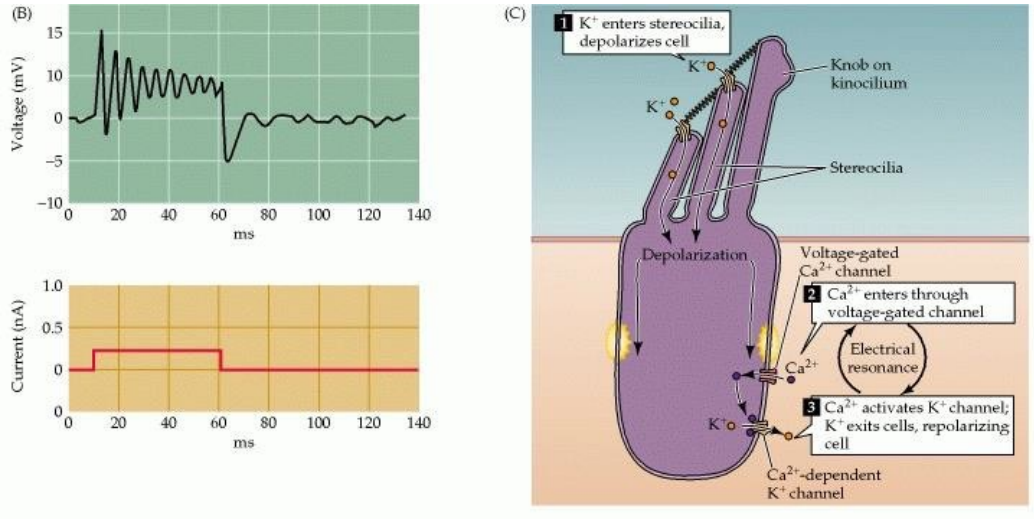
- Type 1 cells are fast acting and are VOR modifiers whilst type 2 are the transducers; genetically encoded ion transport mechanisms are identified (Hamid 2014)
- Otolith crystal integrity depends on Vitamin D metabolism (Dasgupta and Witana 2011)
- Vestibular sensory epithelium is highly tonotopic (van de Berg 2014)

Tonotopicity of the vestibular sensory epithelia

- Knowledge is emerging that the vestibular sensory epithelia is frequency specific or tonotopic and individual frequency zones may be involved in disease sparing other regions (de Berg et al 2014; Shubert and Minor 2004)

- Velocity of angular and translational motion is proportional to the frequency of movement and the vestibular sensory epithelia transduce a range of frequencies into an action potential

- The human physiological range is between 0 Hz to 10 Hz within velocities of 0 to 500 degrees per second; at very low frequencies (0.5 Hz, 30 degrees per second), the optokinetic system is responsible for maintaining VOR



- Vestibular sensory cells exhibit resonance at a characteristic frequency of head movement or active/passive linear acceleration which is voltage dependant (Eatock and Songer 2012)
- This leads to infer that the vestibular sensory hair cells are tuned to incoming frequencies where they generate maximum action potential and thus are frequency specific

Genetics of vestibular disorder

Genetic syndromic without otic structural deformities

- JLN (cardiological)
- Alport (renal)
- Alstorm (cardiological, eyes, DM)
- Usher (Eyes)
- Chromosomal aberrations (6q)
- The ataxia group (SCA, EA, ARASCS - cerebellum)
- The HSMN group (CMT)

Genetic nonsyndromic

- DFNA 9
- DFNA 11
- DFNA 15
- DFNA 28
- DFNAB102/103
- DFNB 2
- DFNB 4
- DFNB 12

Genetic Meniere's

- RERGL and PIK3C2G 12p
- FAM136A
- DTNA
- PRKCB
- DPT
- SEMA4D

Other genetic

- Familial MD (MICA)
- Motion sickness (PVRL3)
- Metabolic conditions (XLHR)
- Von Hippel Lindau syndrome
- DIDMOAD (Wolfram)
- ANSD (otoferlin, pejvakin)

Advances in taking a good history

Children might not able to describe their symptoms in which case the parents/carers need to assist. They usually and quite astutely observe ‘vestibular behaviour’ that are vital clues to the history

Remember, even if the history suggests that the problems is non vestibular in origin, one should still do at least a screening balance function assessment with a clinical vestibular examination

The history is the most important way to get an idea about aetiology

Vestibular behaviour must be sought from parents and carers

These depend on asking leading questions

- Change of facial expression following playground activities and movements
- Fright or pallor
- Clutching
- *Bumping into things*
- Clumsiness
- *Sudden pole axing falls*
- *Periodic episodes of nausea or vomiting*
- Delayed motor functions
- *Difficulties in playground activities*
- Oscillopsia
- Difficult to track challenging visual targets
- Poor head eye coordination
- Loss of postural control
- Difficulty with ambulation in the dark
- Abnormal movements/behaviour
- Difficulties in challenging movements (swimming)
- Autonomic symptoms

Other history

- Birth history
- Family history
- Educational performance
- Ear symptoms and otological history
- Accompanying neurological symptoms
- Development history especially motor development
- Other medical illness
- Head injury in the past
- Ototoxic history
- Musculoskeletal and other medical symptoms
- Gluten sensitivity and history of autoimmune disorders
- Past history of sepsis
- Psychological and cognitive history

Latest diagnostic tests

The aims of the diagnostic tests are to obtain frequency specific information of the vestibular sensory epithelium that will be used to devise customised vestibular rehabilitation, to aid in the aetiological process and to perform a functional assessment of balance

Vestibular screening in infants and neonates

- Vestibular screening in children successfully performed at 6 months of age mainly for children with sensorineural hearing loss utilising the cVEMP; the VIS-Flanders group has been actively working on this and observed that cVEMP is more sensitive than either the vHIT or the rotatory chair test to diagnose a vestibular deficit

- In the absence of the cVEMP, primitive neurological reflexes give you a good idea about vestibular function in children as they are primarily contributed by the vestibular system

- The remote camera system Synapses vHIT is the only system that can screen lateral semicircular canal in children under the age of 1 years

- Indirect ways of screening vestibular function include assessment of motor skills

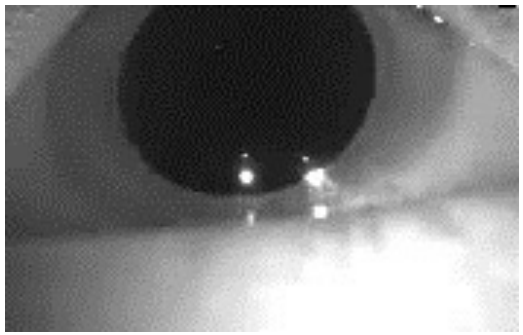
Selection of tests

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, chair
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

Assessment in children under 1 year of age

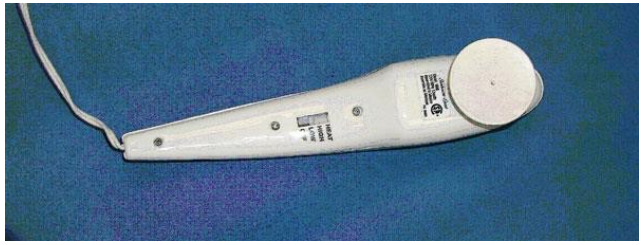
- Moro reflex
- Tonic neck reflex
- Head righting reflex
- Parachute reaction
- Doll's eye reaction
- Farmer's test

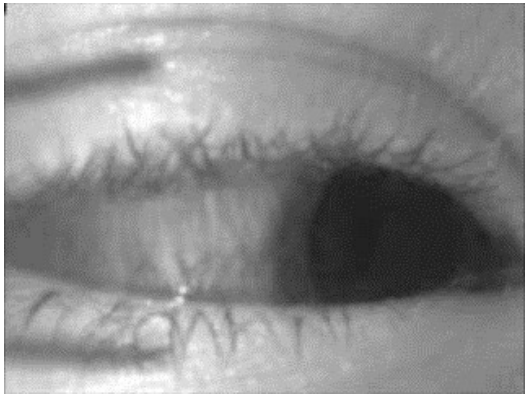
All these reflexes are influenced by the vestibular system and are crucial to assess vestibular function until 5 months of age



Spontaneous & gaze positional nystagmus 0^0 response LSCC

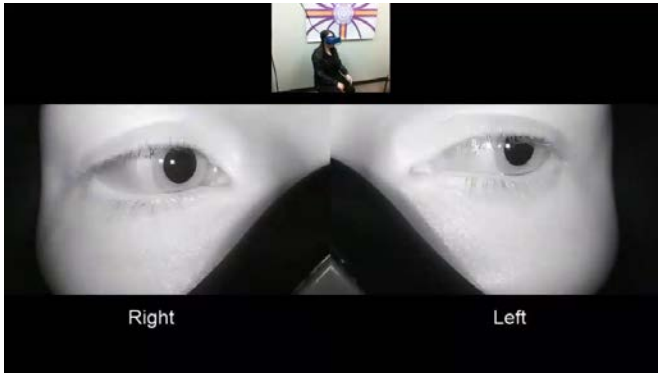
- Static response
- Indicates active lesion
- Amplitude indicator of prognosis
- High sensitivity and specificity
- Useful for prognosis

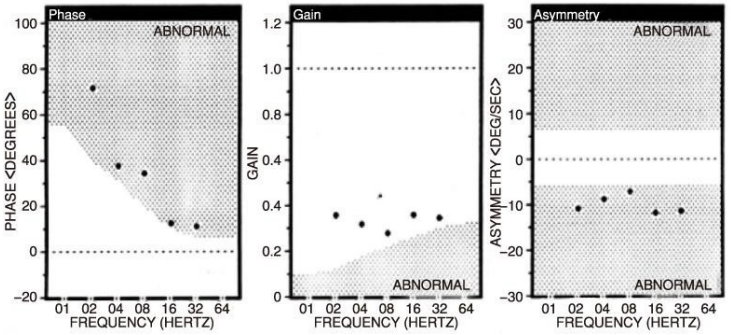




Mastoid vibration test – 0.2 Hz response LSCC

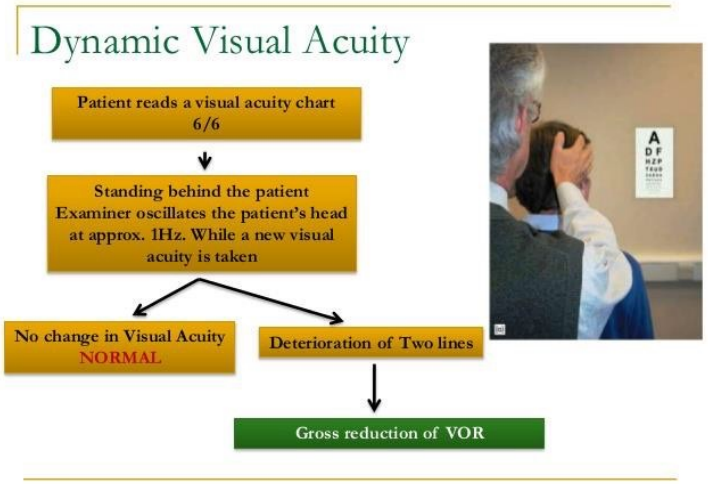
- Good correlation with caloric tests
- Good sensitivity and specificity (Nuti and Mandala 2005)
- Nystagmus beats towards the affected ear and can be stimulated contralaterally
- Useful for prognosis





Rotatory chair tests – 1-2 Hz response LSCC

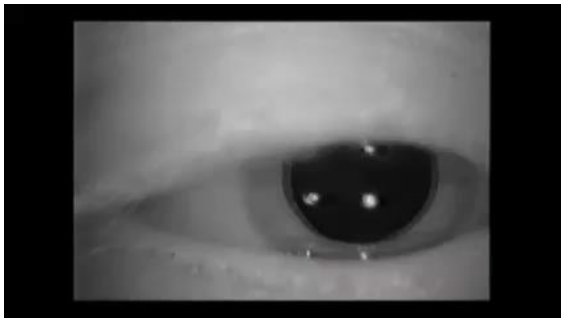
- Constant angular acceleration
- Impulse angular acceleration
- Step test angular acceleration
- Sinusoidal acceleration
- Optokinetic after nystagmus
- Off vertical axis rotation & unilateral centrifugation
- VOR cancel

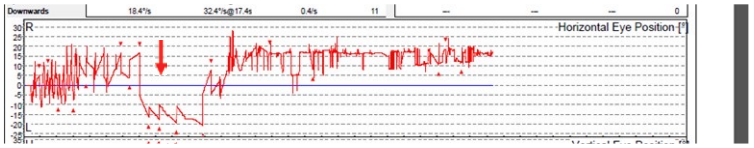


Dynamic visual acuity – 2-3 Hz response LSCC

- Based on the principles of retinal slip on head movement
- Found in unilateral but most useful indicator in BVH
- Computerised variety (cDVA) with high sensitivity and specificity
- Changes with age and vision
- Useful for prognosis

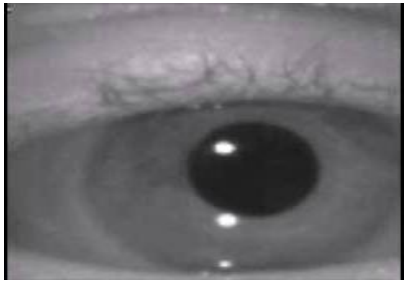
The head shake test – 3-4 Hz response LSCC

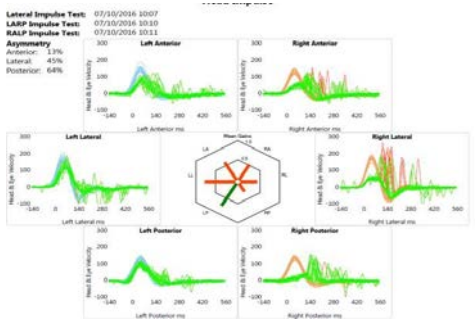




- 20 headshakes – horizontal and vertical
- Horizontal post headshake nystagmus for vestibular asymmetry
- Vertical post headshake and perverted headshake for central
- Useful for prognosis

The clinical and video head impulse test vHIT – 5-7 Hz response all canals

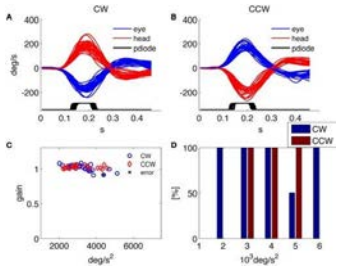


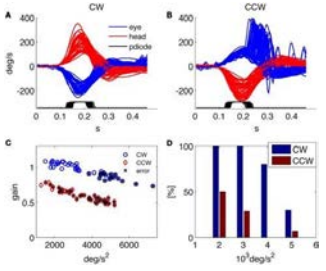


Courtesy: Dr Sylvette Vacher-Wiener

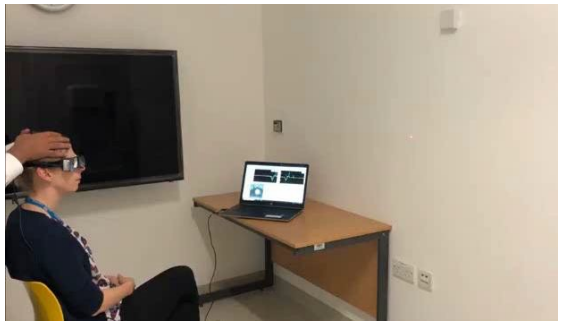
- In expert hands high sensitivity and specificity
- Thrust with fixed target
- Measures all 6 canals
- Revolutionised approach to vestibular diagnostics and can be performed in young children; norms available
- Useful for prognosis with evolution of saccades and VOR gain

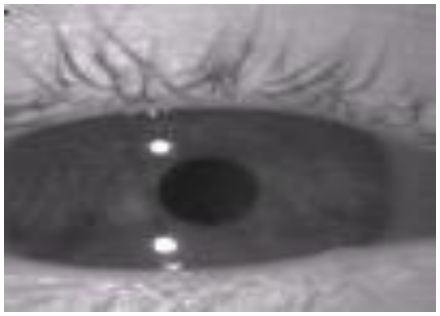
The functional head impulse test – 5-7 Hz all canals





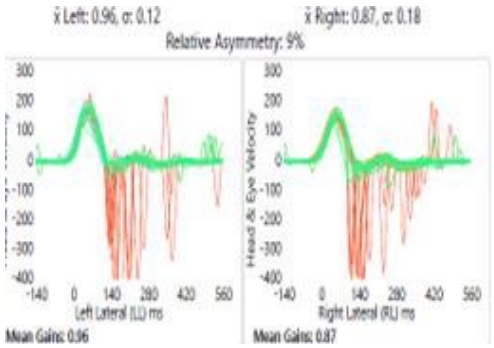
- Measures high frequency functional angular VOR in canals
- Good correlation with video head impulse test
- Advantage in measuring a whole range of head velocities not available in the standard vHIT software

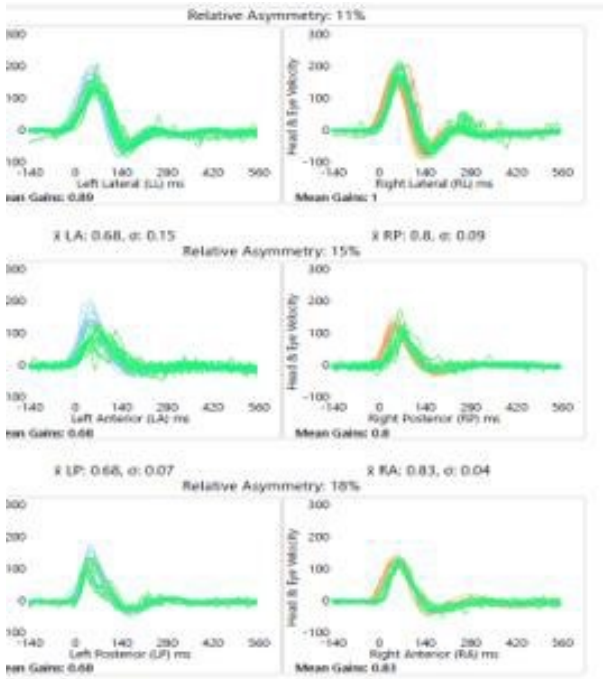


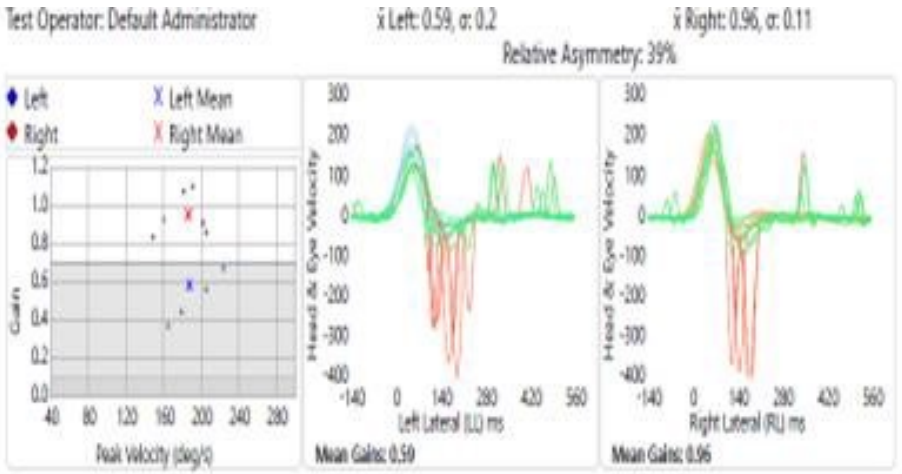


The suppression head impulse test SHIMP

- Measures high frequency lateral motion sensor compensation
- Thrust with moving target
- Useful for evolution of disease
- May be abnormal in presence of normal vHIT
- Useful in central lesions
- Useful for prognosis with VOR gain, evolution of peak saccadic velocities

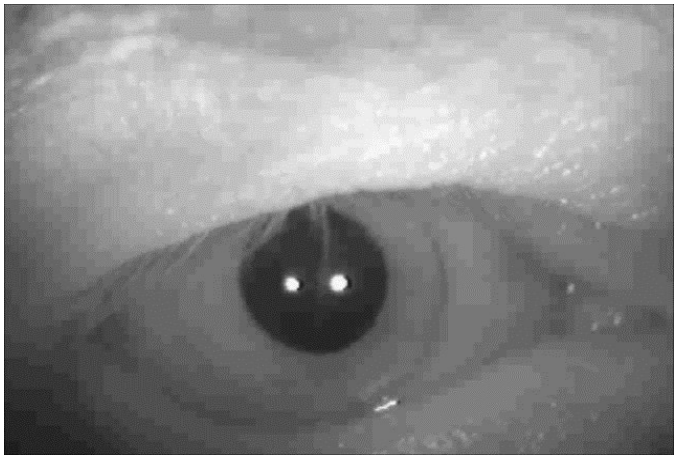






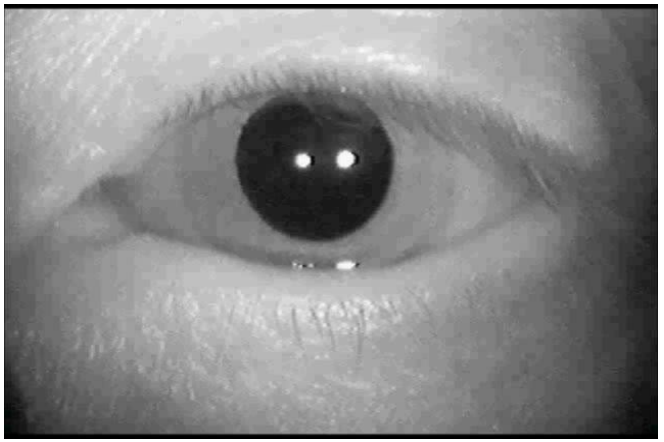
Normal vHIT but asymmetry R>L

Hyperventilation induced nystagmus HVIN

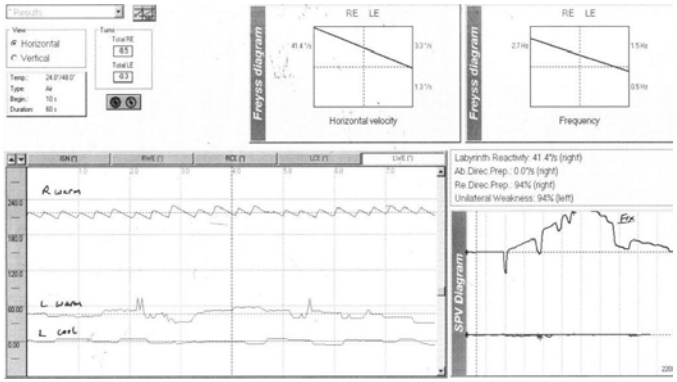


- Indicates lesion in the CPA; thus aetiological test
- Sensitivity is medium with high specificity
- Change of direction of nystagmus may indicate increased size of lesion

Hennebert's test



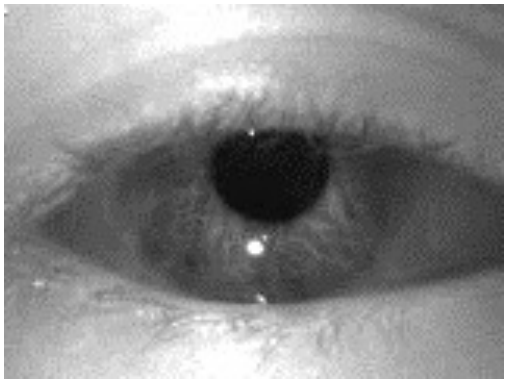
- Tragal pressure or forced expiration against closed glottis/Valsalva
- Characteristic of third window syndromes
- Indicates aetiology
- Unknown sensitivity and specificity



Caloric response – 0.04 Hz response LSCC

- Oldest devised test
- Irrefutable proof of vestibular weakness
- Measures non physiological 0.04 Hz zone
- High sensitivity and moderate specificity

The head heave test – translational high frequency utricular response

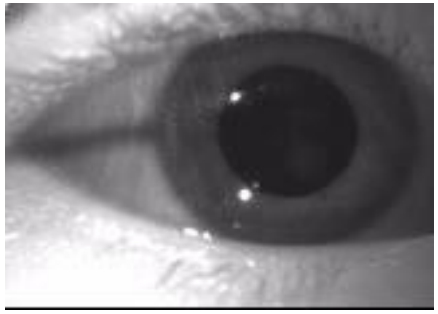


- Good correlation with oVEMP (personal series)
- Operator dependent
- May be found normally if not performed correctly
- Limited series to comment on sensitivity

Subjective visual vertical – static utricular response

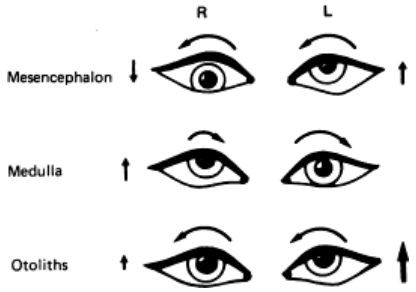
- Can be simply done with i phone app and with a bucket and protractor or can be computerised
- May be noisy due to other conditions – torticollis or strabismus
- Specificity is over 80% but sensitivity is low

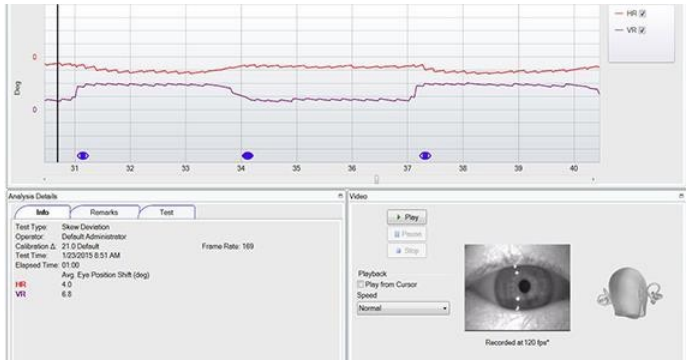
Ocular counter roll test – mid frequency utriculosaccular response



- Recording eye movement on passive and active head tilt
- Low sensitivity and specificity due to cervical
- Does not need expensive equipment to measure when used VNG videography and playback
- May be a useful adjunct to other tests

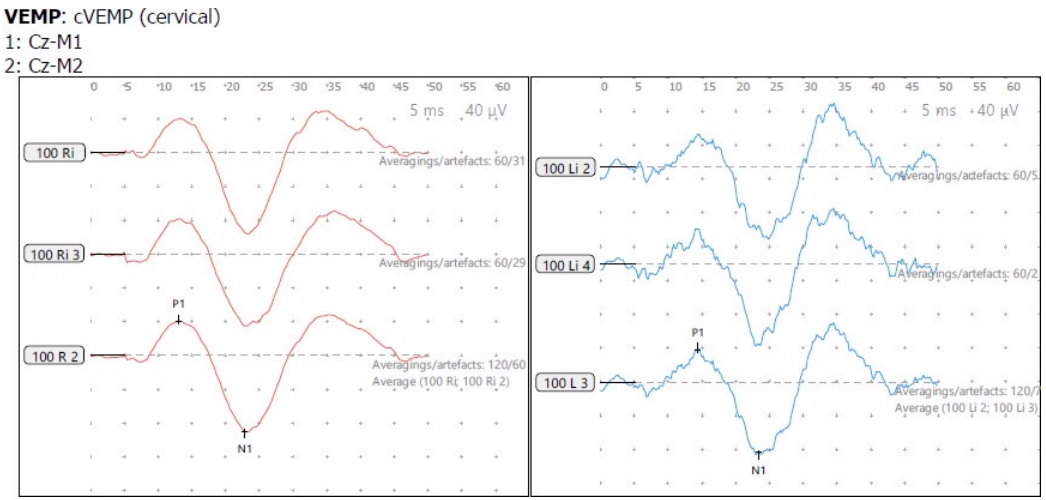
Skew deviation – otolith/central mismatch





- 30% of skews may be due to otolith dysfunction
- Can be measured with software
- In vestibular diagnostics should be backed up with video evidence
- Vestibular sensitivity and specificity undetermined but important for HINTS

Cervical vestibular evoked myogenic potential - low frequency saccular response



Asymmetry

N	P1	N1	P1–N1 %
100 R 2, 100 L 3	1.32	0.53	-3.5

Latencies && amplitudes (right ear)

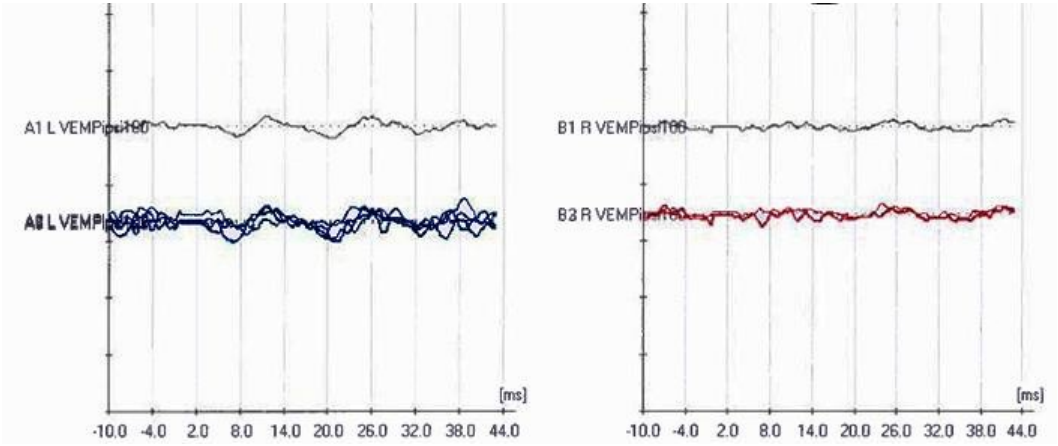
N	P1 (ms)	N1 (ms)	P1–N1 (µV)	Averaged EMG (µV)	Rectified amplitude (µV)	Samples
100 Ri				50.6		60
100 Ri 3				48.6		60
100 R 2	13.0	22.8	129.9	50.6	2.6	120

Latencies && amplitudes (left ear)

N	P1 (ms)	N1 (ms)	P1–N1 (µV)	Averaged EMG (µV)	Rectified amplitude (µV)	Samples
100 Li 2				58.7		60
100 Li 4				61.5		60
100 L 3	14.3	23.3	121.2	58.7	2.1	120

- Uses sternocleidomuscle cervico-colic response
- Always thresholds, rectified amplitudes and amplitude asymmetry considered
- Increased amplitude and decreased thresholds indicate third window

Ocular vestibular evoked myogenic potential – low frequency utricular response



- Uses contralateral inferior oblique response
- Always thresholds and amplitude asymmetry considered

Vestibulospinal tests – assess multiple input sensory integration

- Romberg and sharpened Romberg
- Unterberger
- Single leg stance
- Tandem gait
- Retropulsion
- CTSIM – M and dynamic posturography
- Quantification of gait rarely needed

Diagnostic biomarkers for Meniere's Disease

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

Neurological examination

- Cranial nerves
- Tone, power, reflex
- Gait and posture
- Cerebellum
- Sensation

Musculoskeletal examination

- Any obvious craniofacial dysmorphia
- Leg length discrepancy
- Lower limb varus and valgus deformities
- Joint deformities
- Elasticity of joints

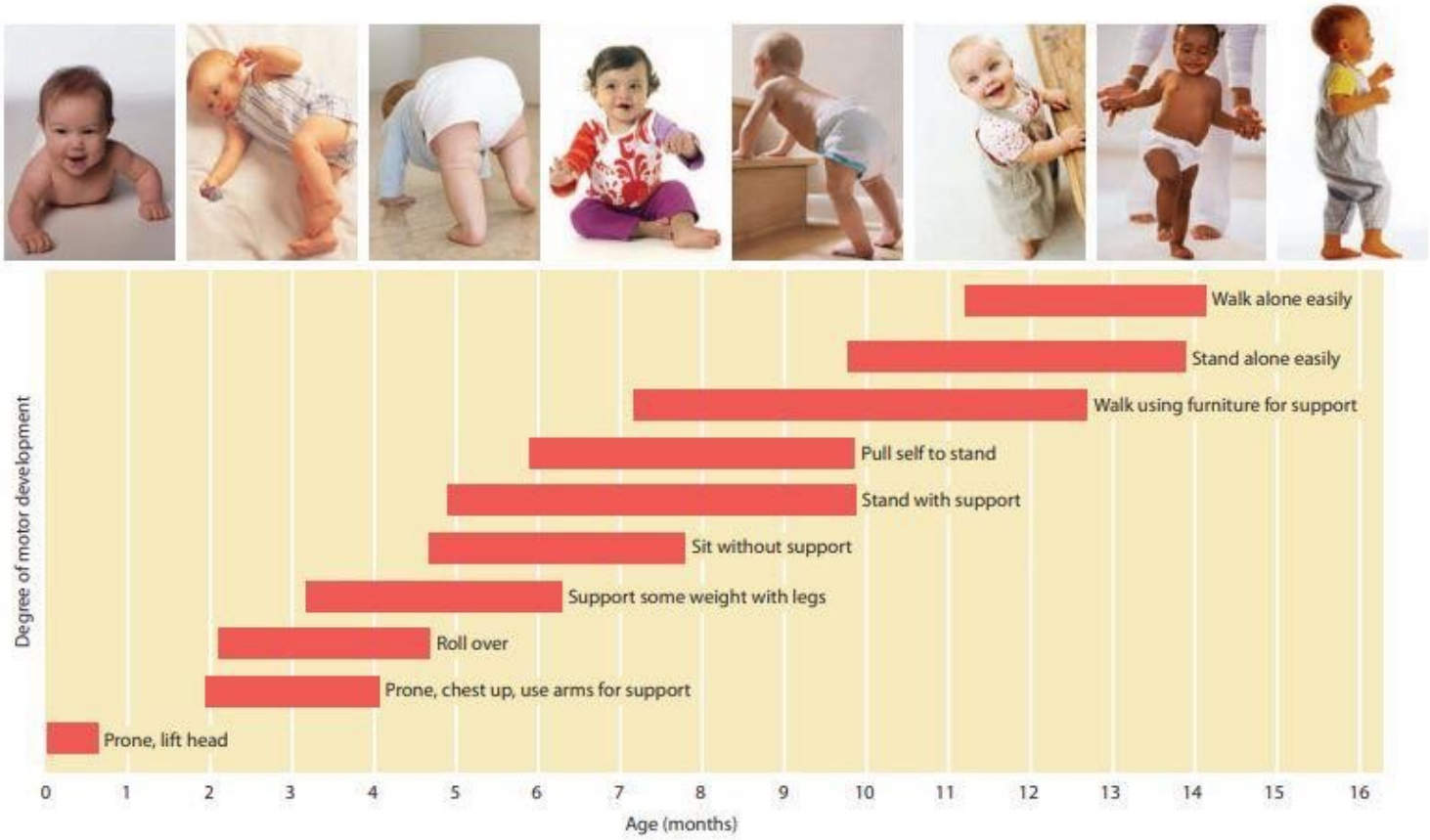
Cardiovascular examination

- Lying, sitting and standing BP
- Lying, sitting and standing pulse

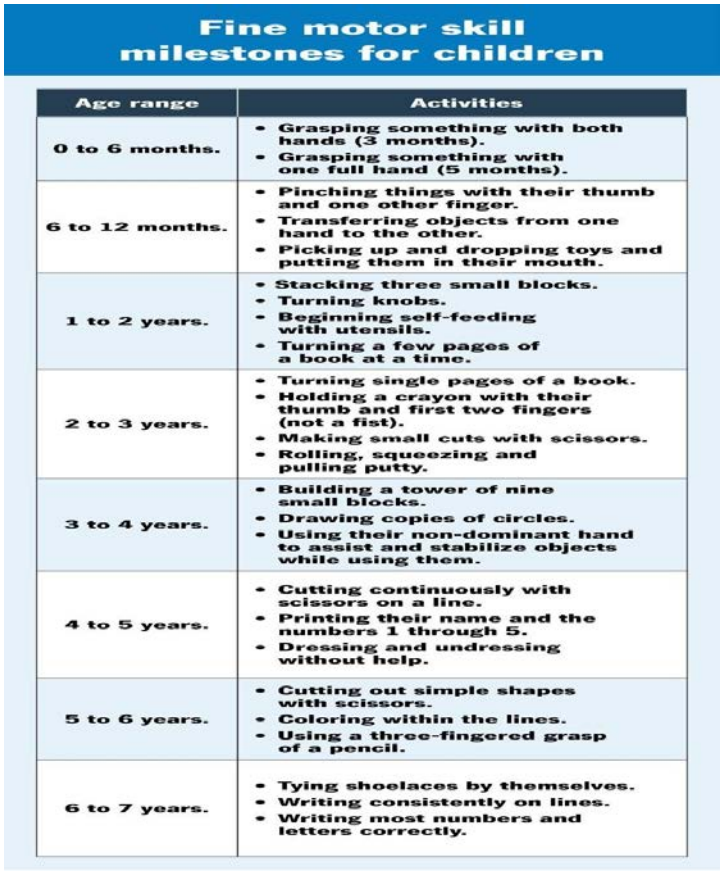
Ophthalmological examination

- Oculomotor eye movements
- Convergence

Gross Motor assessment



Fine Motor assessment



Fine motor skill milestones for children

Age range	Activities
0 to 6 months.	• Grasping something with both hands (3 months). • Grasping something with one full hand (5 months).
6 to 12 months.	• Pinching things with their thumb and one other finger. • Transferring objects from one hand to the other. • Picking up and dropping toys and putting them in their mouth.
1 to 2 years.	• Stacking three small blocks. • Turning knobs. • Beginning self-feeding with utensils. • Turning a few pages of a book at a time.
2 to 3 years.	• Turning single pages of a book. • Holding a crayon with their thumb and first two fingers (not a fist). • Making small cuts with scissors. • Rolling, squeezing and pulling putty.
3 to 4 years.	• Building a tower of nine small blocks. • Drawing copies of circles. • Using their non-dominant hand to assist and stabilize objects while using them.
4 to 5 years.	• Cutting continuously with scissors on a line. • Printing their name and the numbers 1 through 5. • Dressing and undressing without help.
5 to 6 years.	• Cutting out simple shapes with scissors. • Coloring within the lines. • Using a three-fingered grasp of a pencil.
6 to 7 years.	• Tying shoelaces by themselves. • Writing consistently on lines. • Writing most numbers and letters correctly.

Cleveland Clinic

Courtesy: Cleveland Clinic

Paediatric vestibular balance psychometric and observational assessment

- The Vanderbilt Dizziness Handicap Inventory
- The Berg Balance Scale
- Alberta Infant Motor Development Scale

Audiological assessment

- PTA/TYMP/SRT
- OAE/ABR

Cardiovascular assessment

- Blood pressure/pulse – lying, sitting, standing

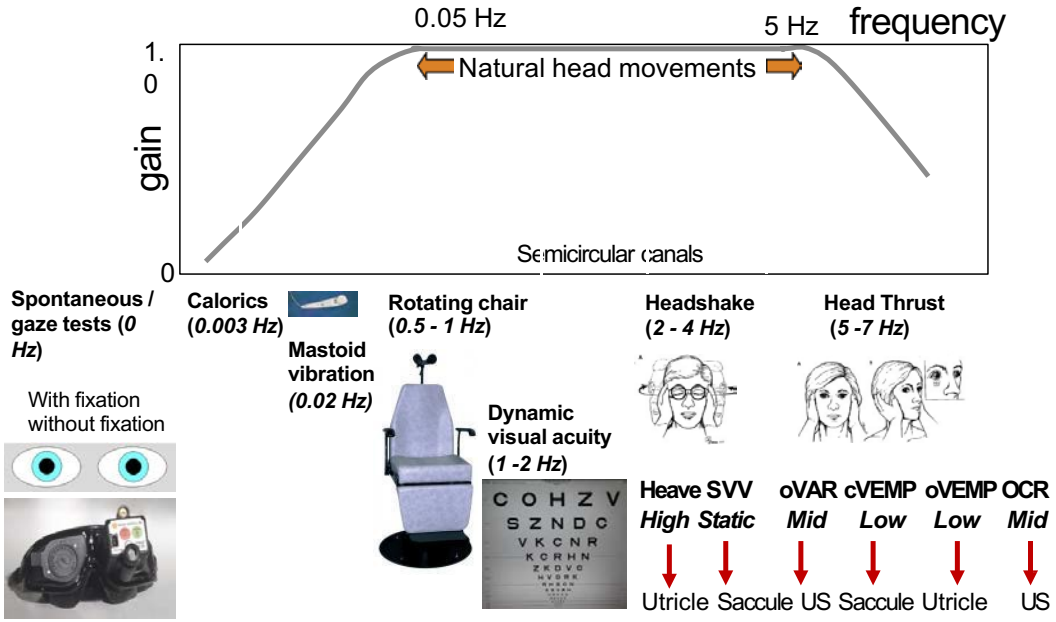
Ophthalmological assessment

- VA/VF
- Convergence
- Oculomotor
- Ophthalmoscopy

Musculoskeletal assessment

- Skeletal survey especially lower limb
- Hyperelasticity

Vestibular assessment and vestibular gram



Hyperventilation test
Hennebert's test
fHIT and SHIMP
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

Psychometric assessment

- DHI
- Berg/Alberta

Assessment of vestibular system in children is a unique artistic skill set that requires knowledge, wisdom and years of dedicated paediatric training and overall requires improvisation

Imaging advances

Imaging – to assess bony and membranous labyrinth

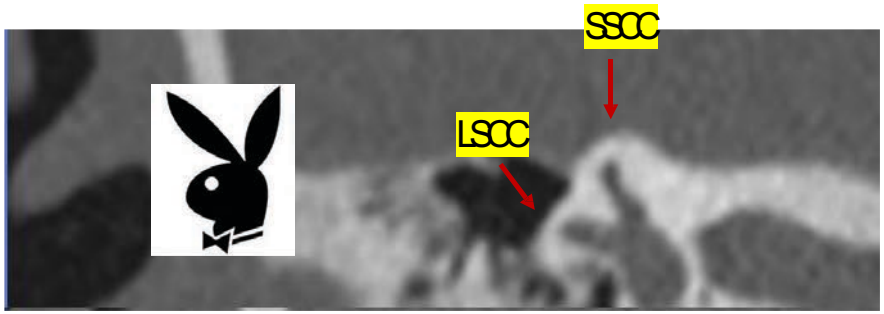
CT scans:

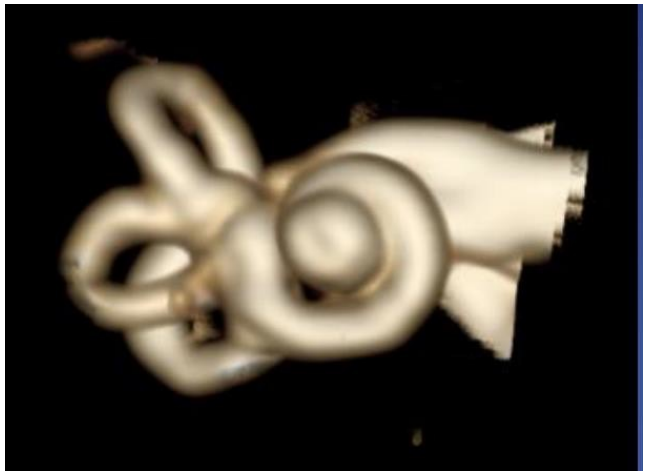
- Standard T2 weight without contrast
- Stenver and Poschl planes at minimum 0.6 slices with an average reformat at 0.3.mm for dehiscences
- Cone beam CT

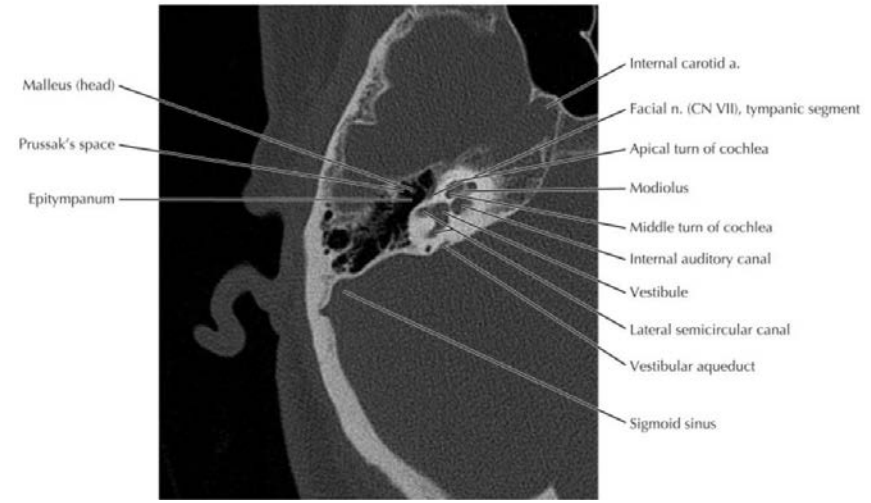
MRI scans

- Standard T2 weight with and without contrast when indicated
- HYDROPS technique for Meniere's disease

HRCT - Normal

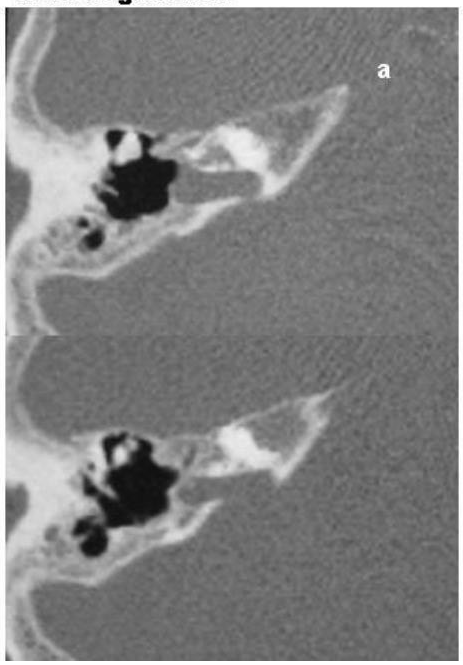

SSCC
LSCC



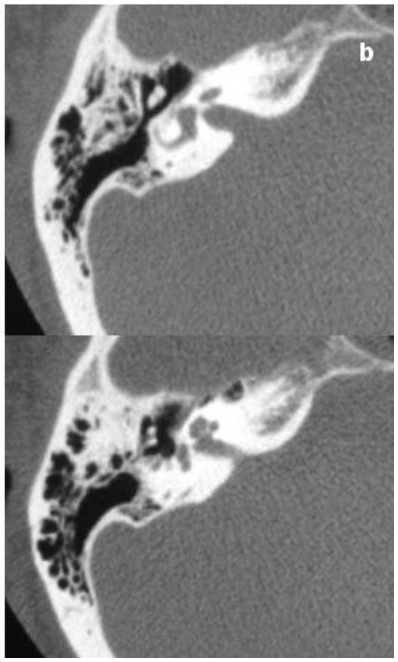

Malleus (head)
Prussak's space
Epitympanum
Internal carotid a.
Facial n. (CN VII), tympanic segment
Apical turn of cochlea
Modiolus
Middle turn of cochlea
Internal auditory canal
Vestibule
Lateral semicircular canal
Vestibular aqueduct
Sigmoid sinus

HRCT – Congenital anomalies

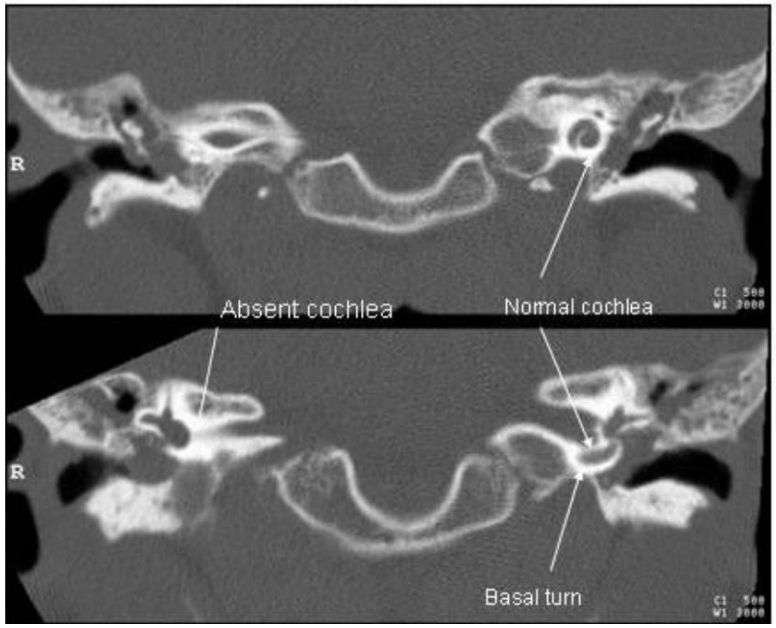
Michel aplasia – Arrest during 3rd week of gestation



Normal

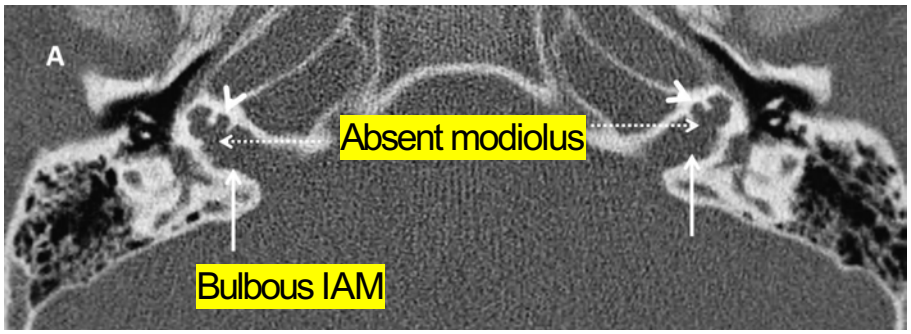


Michel's aplasia

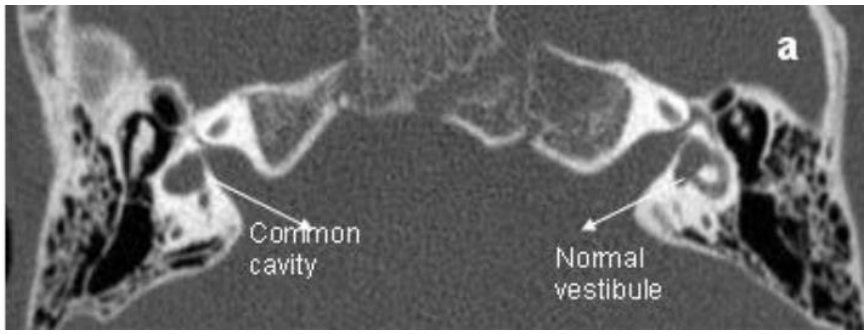


Hypoplastic SSCC/absent cochlea

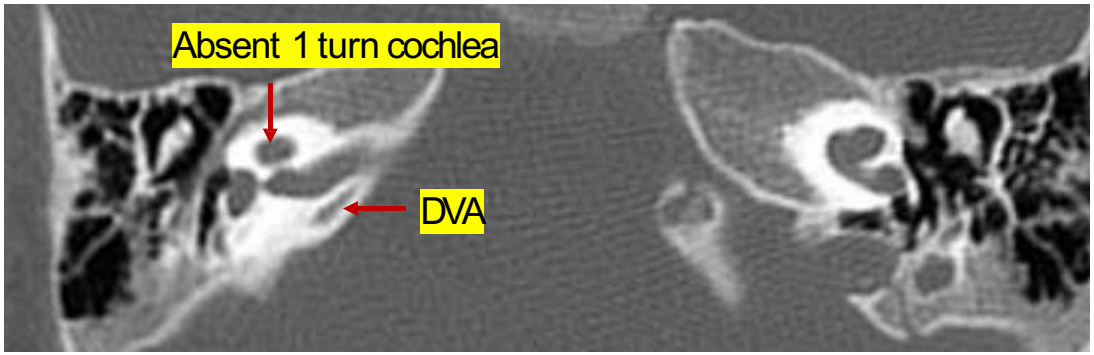
HRCT – Congenital anomalies



X linked gusher

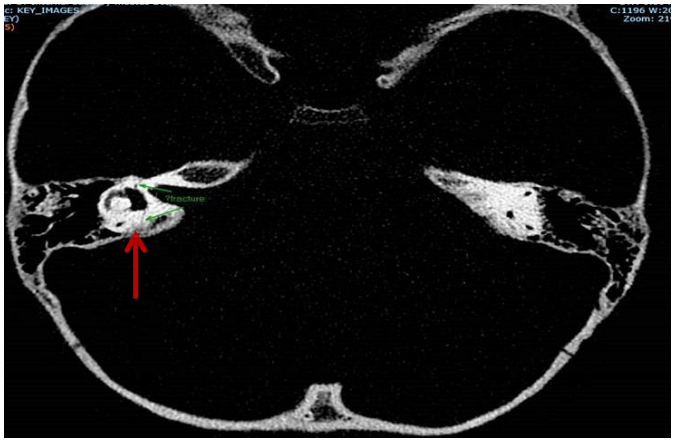


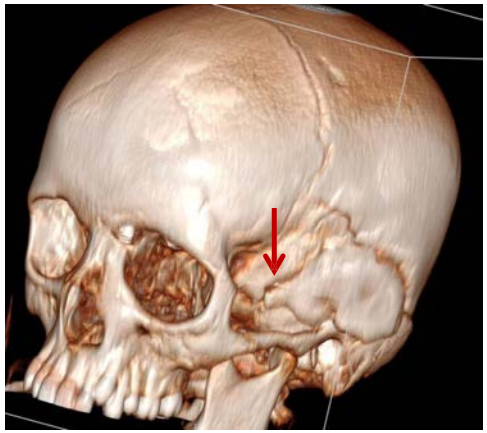
Common cavity

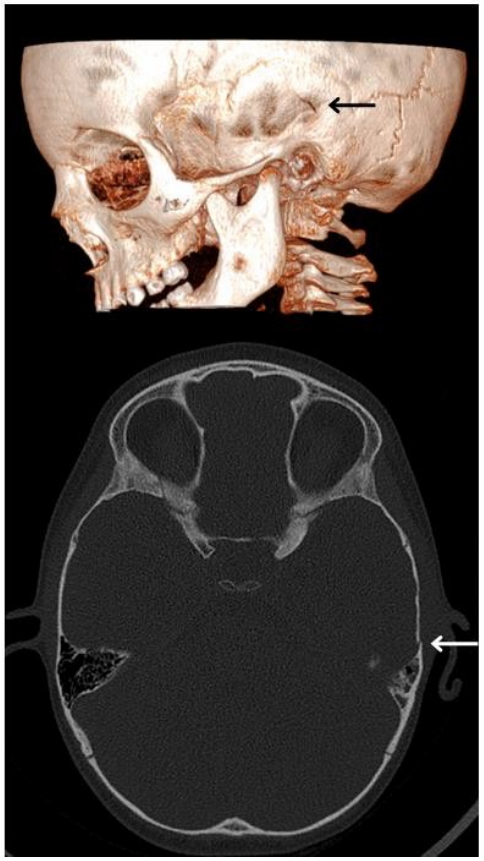


Unilateral Mondini

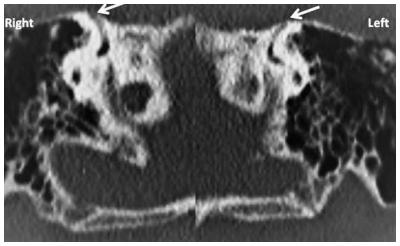
HRCT – Trauma



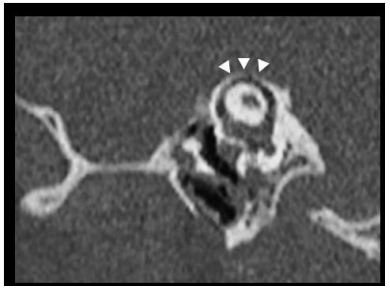




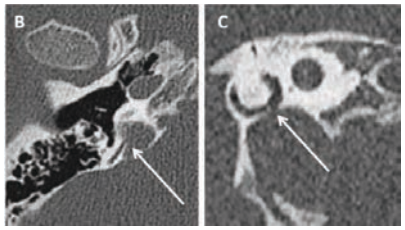
HRCT – Canal dehiscences



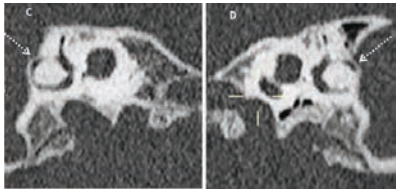
SSCDS



Near SSCC

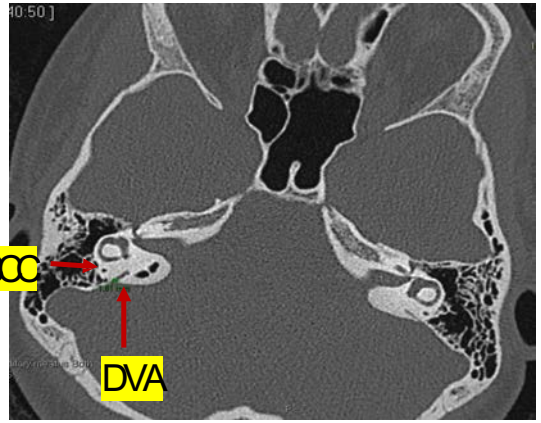


PSCCDS

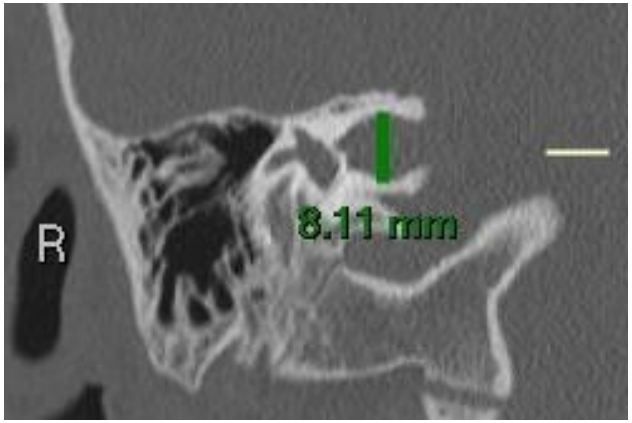


Near PSCC

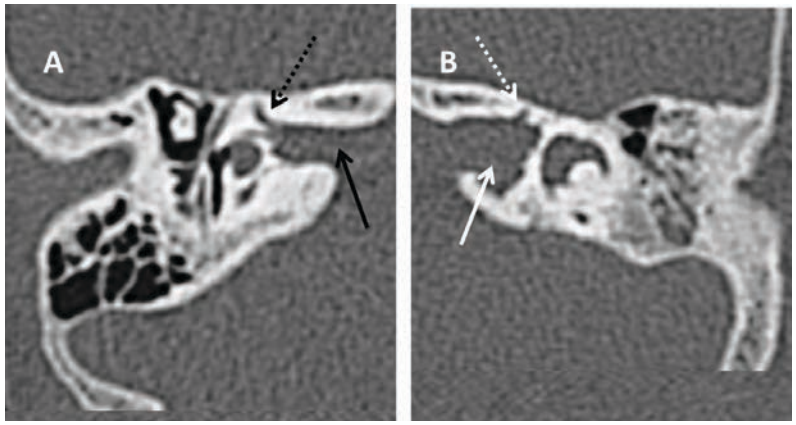
HRCT – Other third windows



DVA

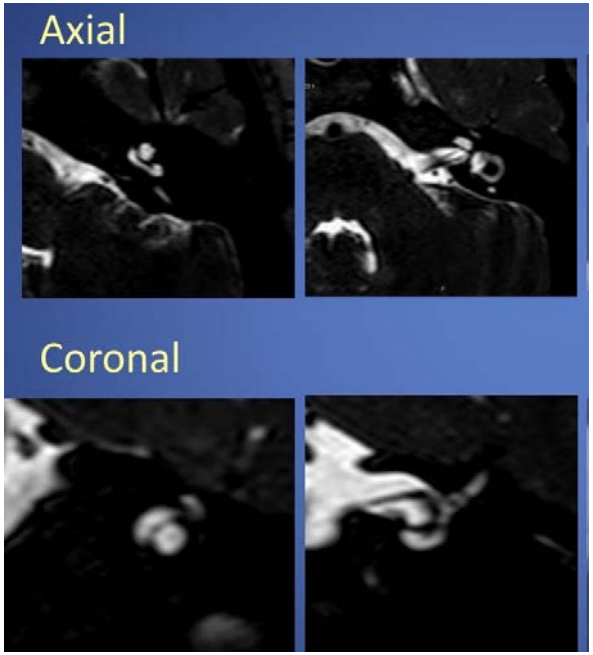


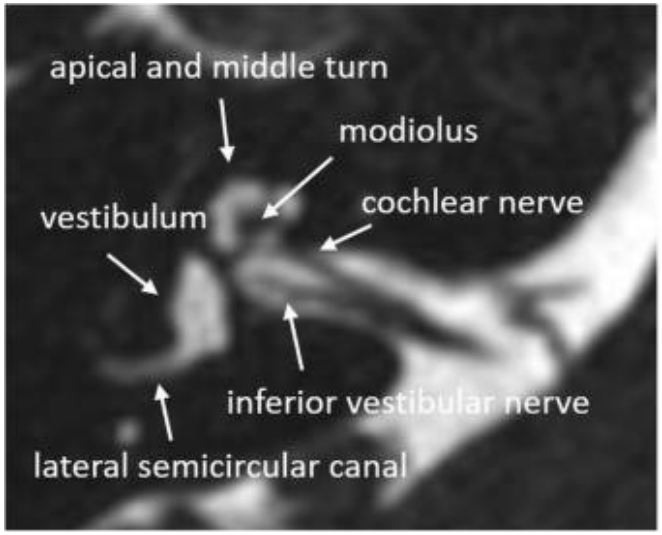
Dilated IAM

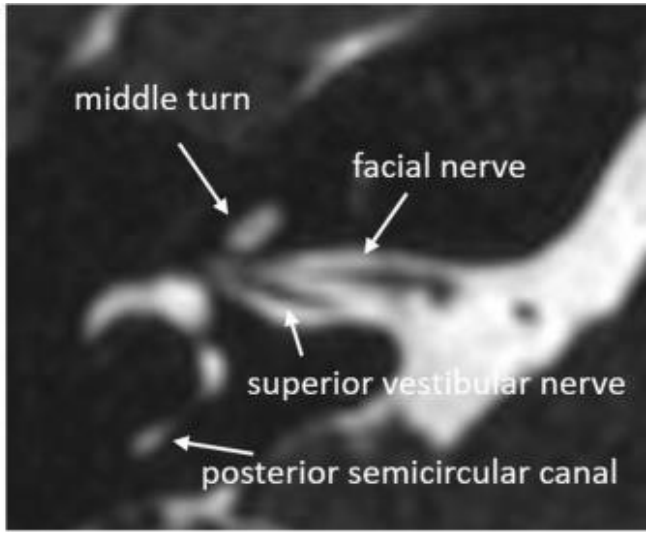


Facial canal dehiscence

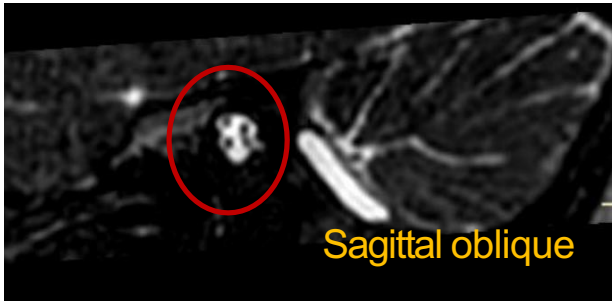
MRI - Normal



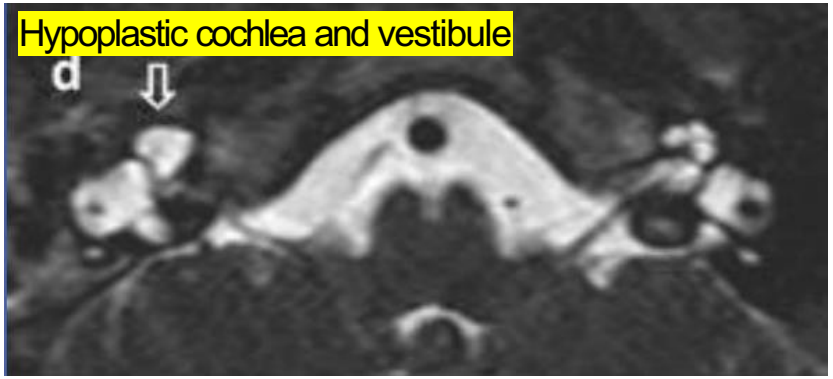


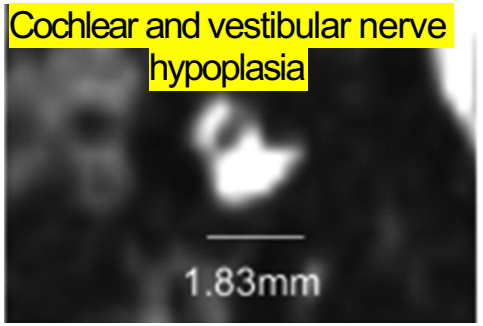


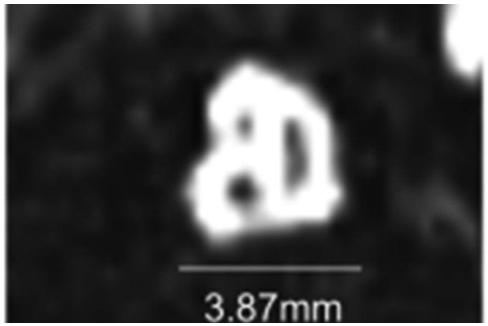
Widmann, Get al 2020

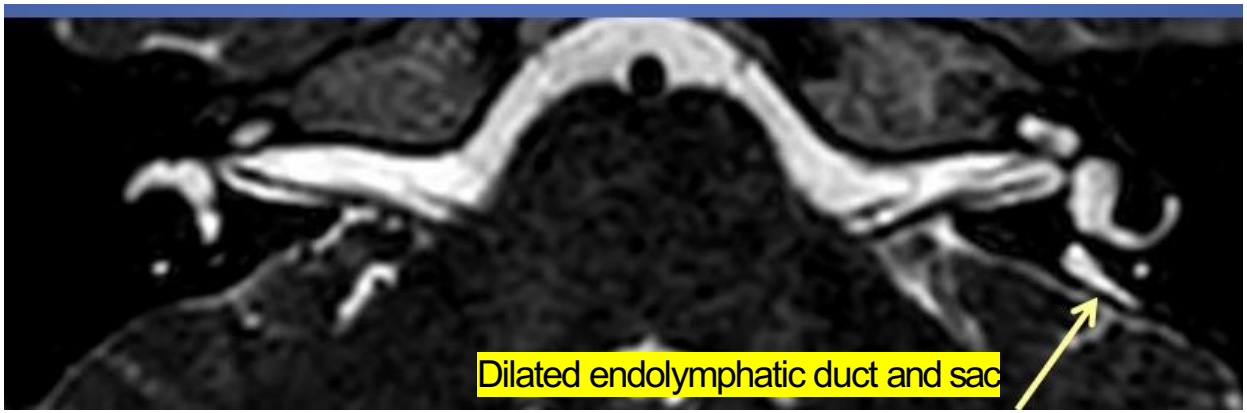


MRI – Congenital anomalies

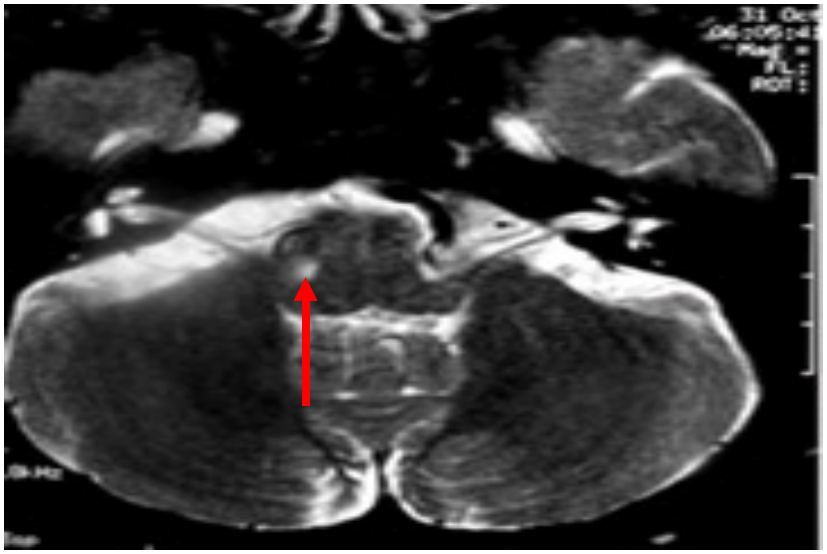


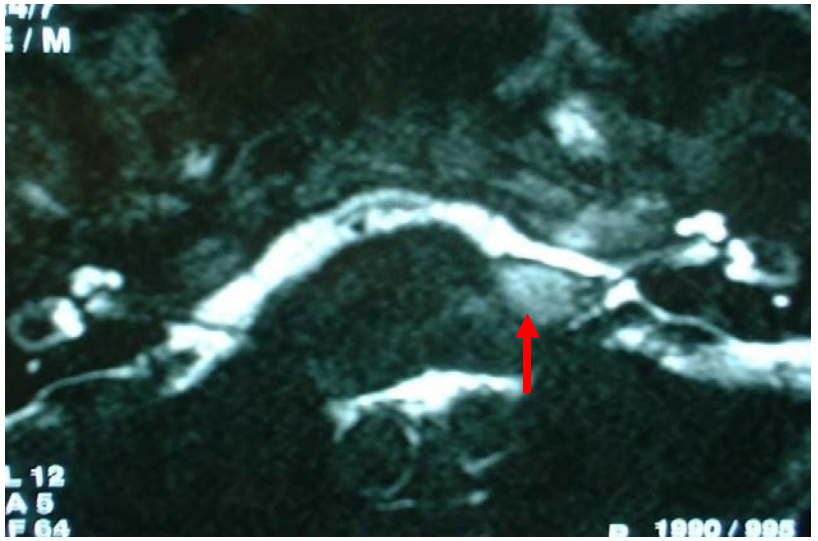






MRI – space occupying lesions

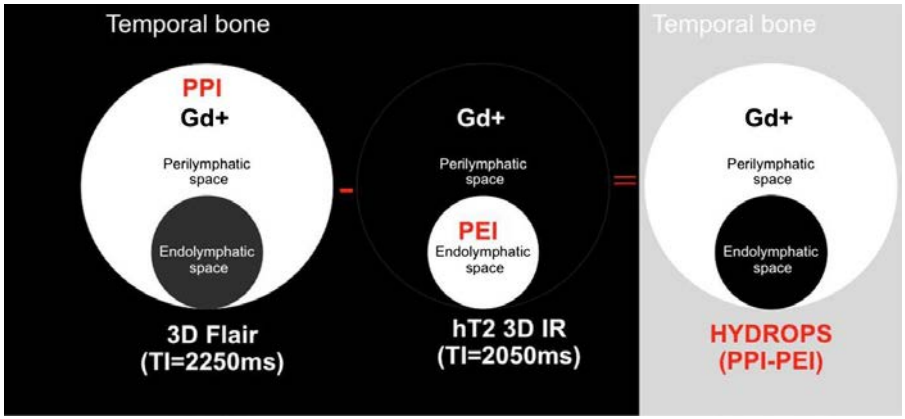


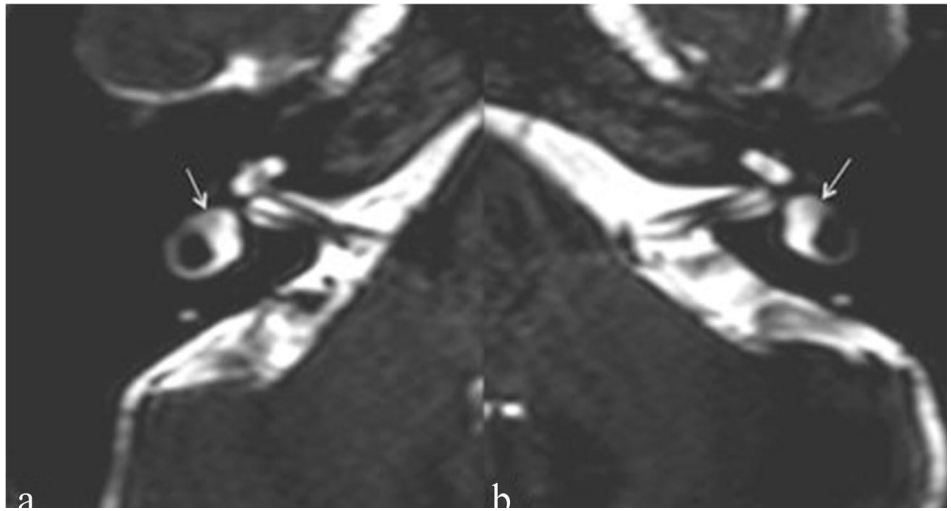


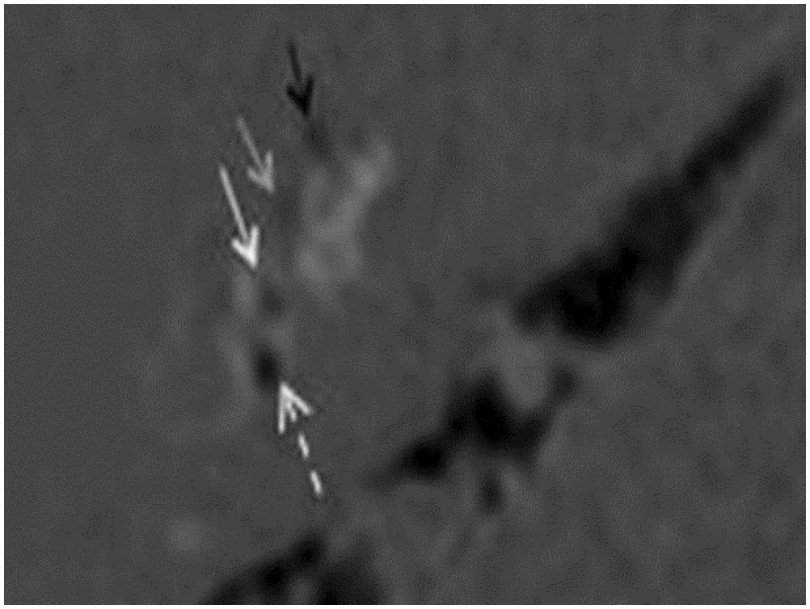
MRI for Meniere's Disease

- Initial T2 weight
- Injection of GBCM intra tympanic or intravenous and wait for 4 hours
- 3D FLAIR with conventional turbo spin echo or variable flip angle turbo spin
- Shortening inversion time
- Semiquantitative or qualitative interpretation
- Calculated volumetric analysis
- Not of much use in children as phenotype is not backed up by MRI (Chen 2023)

Contrast medium diffuses to the perilymph but not the endolymph in MD and generates a perilymph positive image (PPI - hyperintense) whilst the endolymph remains dark - by altering the time of inversion recovery, the endolymphatic space becomes darker generating a positive endolymph image (PEI – hypointense) – the subtraction of PPI - PEI generates the HYDROPS (hybrid of the reversed image of the positive endolymph signal and native image of the positive perilymph signal) to visualise the endolymphatic space







Normal

HYDROPS

Sousa et al 2021

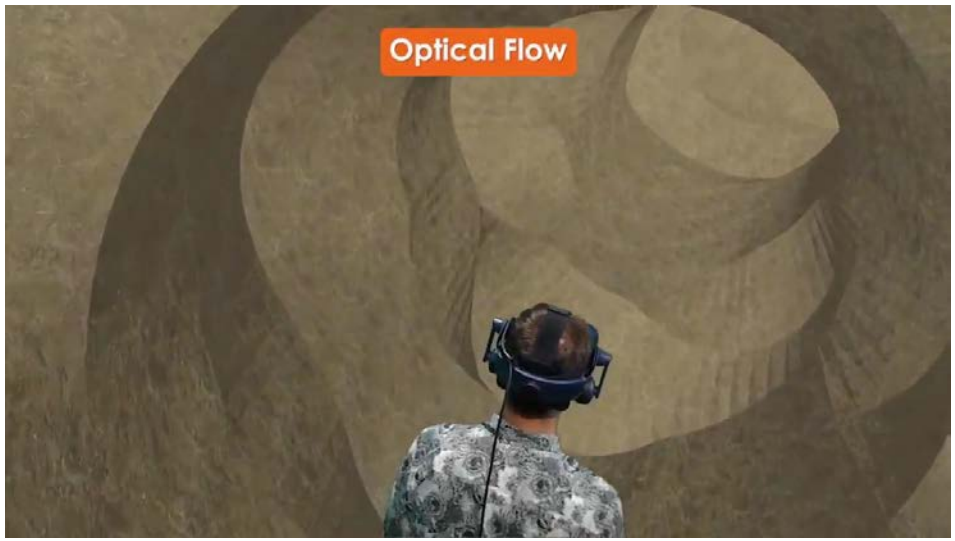
Advances in management

- Vestibular management is now ***multidisciplinary***
- ***A cognitive approach*** is essential
- ***Pharmacological management*** is recommended to be used ***as a short term measure when absolutely indicated*** for MD, VM and first 3 weeks of an acute vestibular event
- ***Vestibular rehabilitation is now customised*** to frequency specific responses with sensory integration strategies and outcome measured by different tests and psychometric questionnaires – ***it should not be used as a blanket approach***
- ***Balance belt*** is a non invasive device showing good outcome
- ***Vestibular implant*** research is gathering momentum
- ***Virtual reality*** is used to habituate, substitute and adapt to individual needs
- ***Life style changes and situational counselling*** is essential
- ***Music therapy*** has shown promise

The multidisciplinary approach

- Management of hearing loss → Audiology
- Management of paediatric and surgical comorbidities → Medical/surgical specialities
- Sensory integration strategies → Occupational therapy
- Neuro and musculoskeletal physiotherapy if required → Specialised physiotherapy
- Psychological intervention when appropriate → Psychology
- Correction of visual problems → Ophthalmology and optometry
- Music therapy and hearing therapy → Psychology and audiology
- Engagement significant others → Education, family, social services

Virtual reality



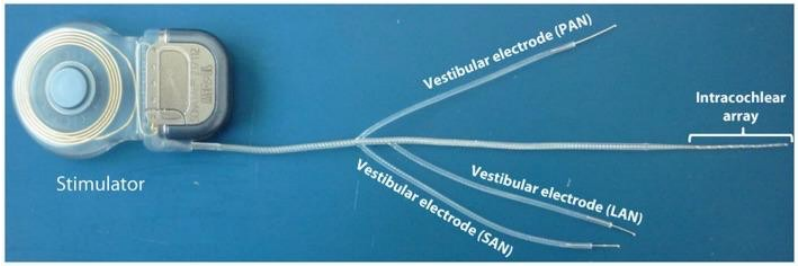
Courtesy – Virtualis and Frank Assaban

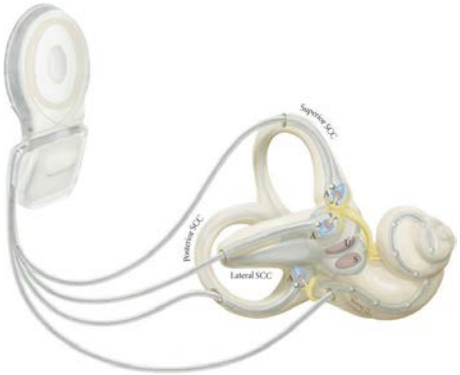
- Part of rehabilitation including visual vertigo
- Good desensitising techniques
- Good outcome reported

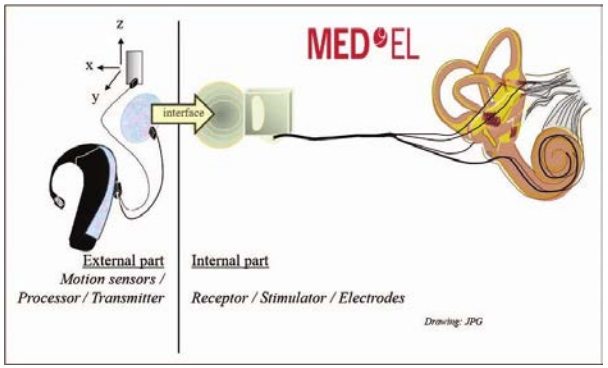
The Balance Belt

- Indicated in refractory vestibular/balance disorder with symptoms
- Works by haptic/vibrotactile stimulation delivered at a calibrated rate to improve proprioception
- Efficacy reported so far good but more studies needed

The Vestibular Implant







- Engineered with modifications of a cochlear implant
- Electrodes inserted into canals or the otoliths
- Indicated in refractory and resistant cases of bilateral vestibular hypofunction
- Only 4 groups working in the world at the moment and the outcome is varaibale

Summary of the management algorithm

Phenotype determination

↓

Vestibular and central quantification

↓

ACCURATE DIAGNOSIS

Idiopathic

Symptomatic treatment

- **Cognitive approach**
- Life style changes
- **Involvement of significant others and MDT**
- Pharmacological intervention
- **Vestibular rehabilitation**
- Device management when indicated

Investigate decompensation

Cause found

Symptomatic treatment

- **Cognitive approach**
- Treatment of cause, i.e. surgery, BPPV etc
- Life style changes
- **Involvement of significant others and MDT**
- Pharmacological intervention
- **Vestibular rehabilitation**
- Device management when indicated

Advances in assessment of dynamic and static vestibular compensation

VNG

- Alexander's nystagmus changes in grade
- Headshake horizontal nystagmus amplitude change or absence
- Disappearance of mastoid vibration nystagmus
- Changes in HVIN and Hennebert
- Head heave saccade changes or absence
- Ocular counterolling changes or absence

Video head impulse test

- Improvement in VOR gain
- Changes in saccade morphology – clustering and PR score

Suppression head impulse test

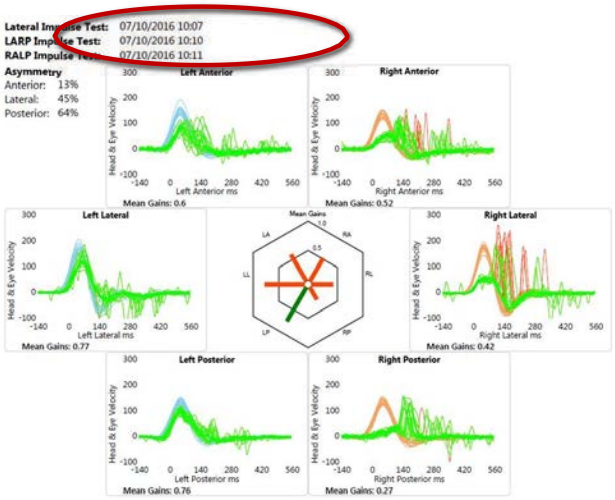
- Improvement in VOR gain
- Changes in saccade morphology – clustering and PR score
- Changes in asymmetry and peak saccadic velocity

Others

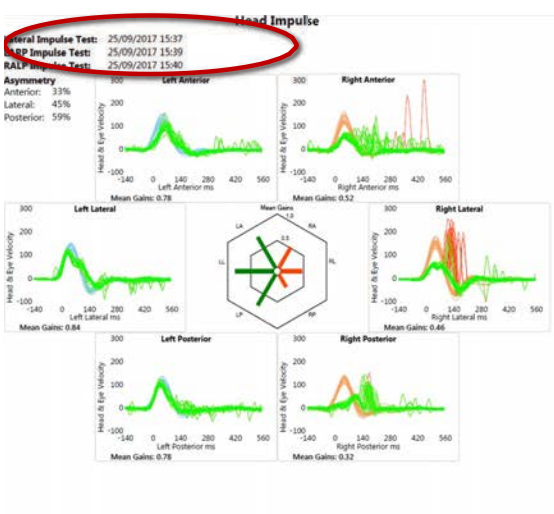
- Improvement in chair VOR gain and amplitude in VEMP
- Static and dynamic posturography changes
- Improvement in balance scale scores

Anamnesis

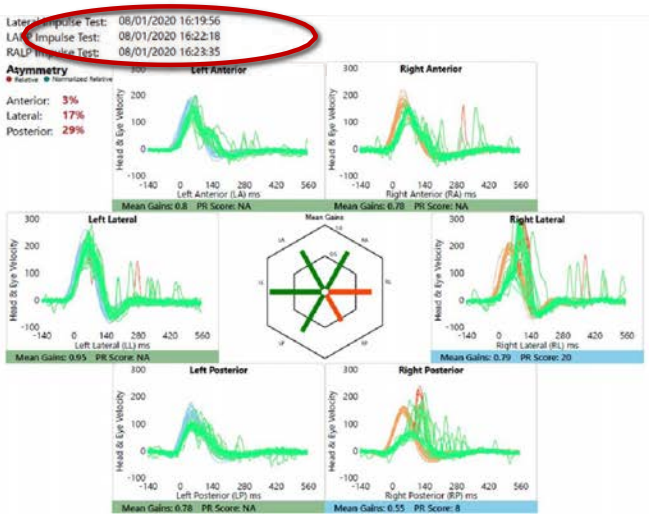
- Improvement in functional performances
- Improvement in cognition
- Improvement in motor skills
- Improvement in school performance and social interactions
- Improvement in duration/frequency/ intensity of VM/ELH episodes



October 2017; 0.52, 0.42, 0.27
Overts dispersed

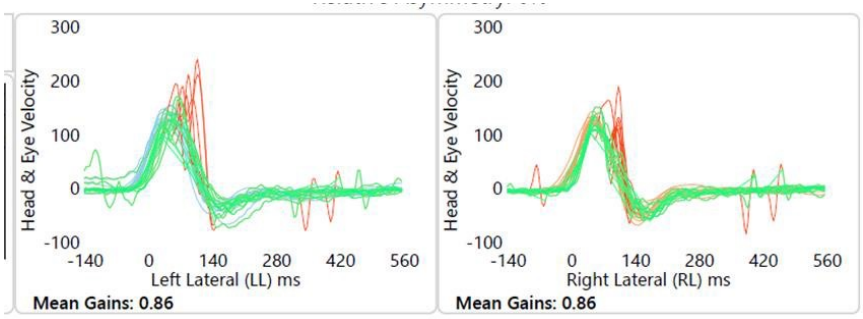


September 2017; 0.57,0.46, 0.32
Coverts/overts clustered

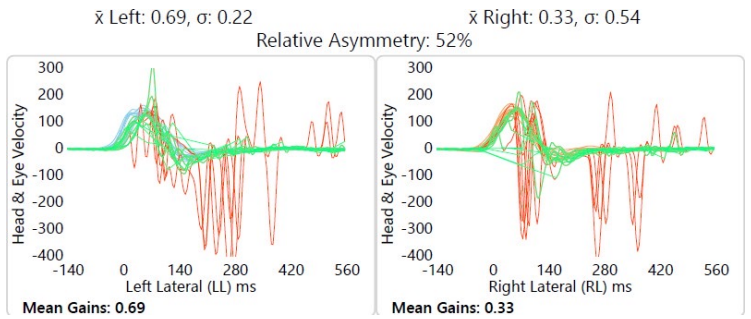


January 2020; 0.78, 0.79. 0.55
Mainly coverts, clustered

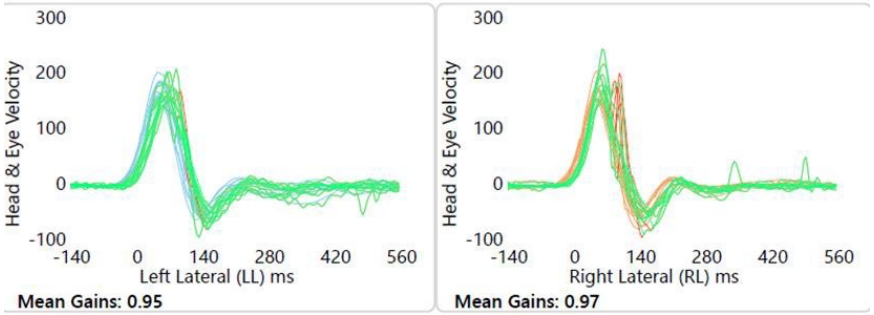
Acute vestibular neuritis (R) - asymptomatic after rehabilitation



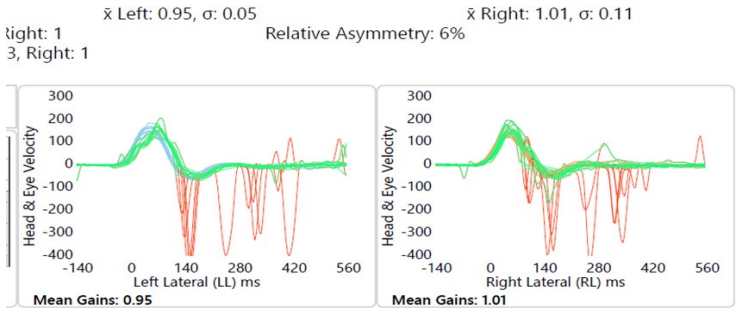
August 2022 → Coverts, normal VOR gain 0.86



August 2022 → PSV 319/269; asymmetry 52%; VOR 0.69/0.33



January 2023 → Coverts (L), normal VOR gain 0.97



January 2023 → PSV 224/228; asymmetry 6%; VOR 0.95/1.01

Acute bilateral AIED – well compensated with mild residual problems after rehabilitation

Conclusions

- Vestibular knowledge is still evolving at a rapid rate
- Vestibular function can now be extensively quantified
- Latest imaging techniques and vestibular processing can be mapped
- We are now at an age of vestibular implant
- Genetics of vestibular disorder being identified

Alder Hey Academy

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Alder Hey Children's Hospital Paediatric Audiovestibular Department presents

UNIVERSITY OF LIVERPOOL

Alder Hey Paediatric Vestibular Course
October every year

We have come a very long way but still we are collecting pebbles on the seashore

Thank you