

# Advances in vestibular medicine

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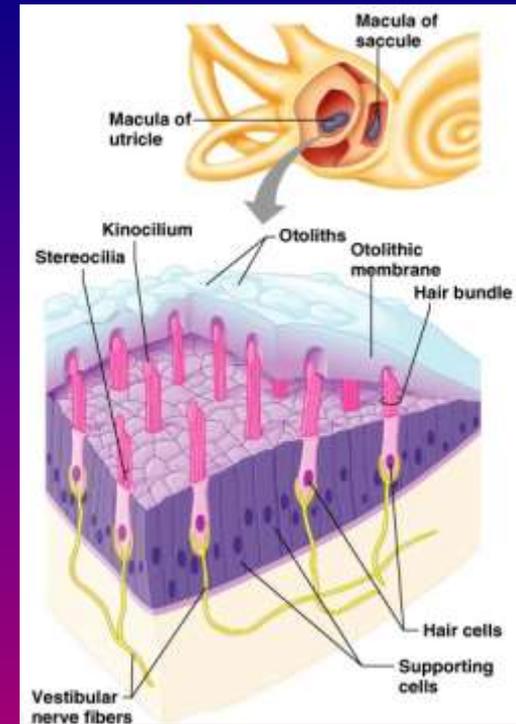
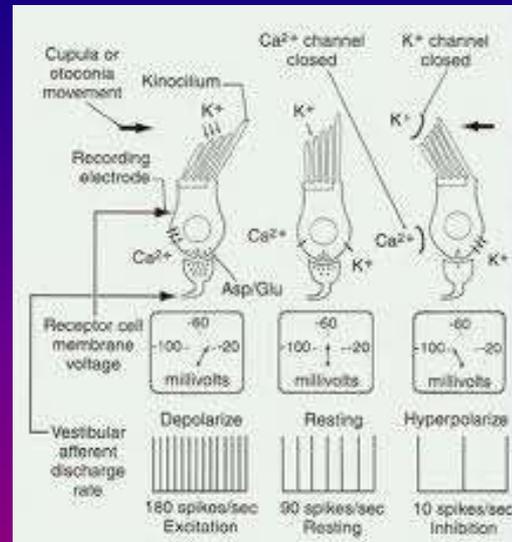
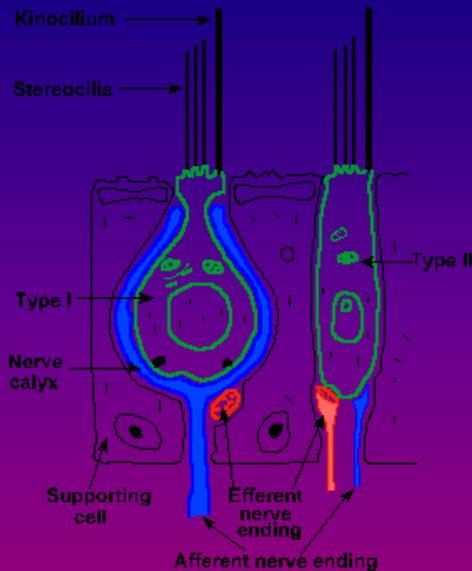
Manchester Centre for Audiology and Deafness

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## This lecture

- Recent anatomical and physiological advances
- Advances in vestibular diagnostics
- The vestibular gram
- Advances in management
- Conclusions

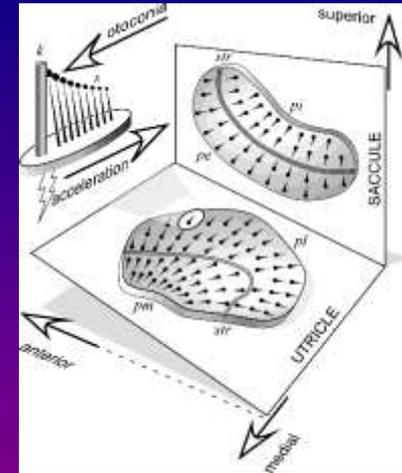
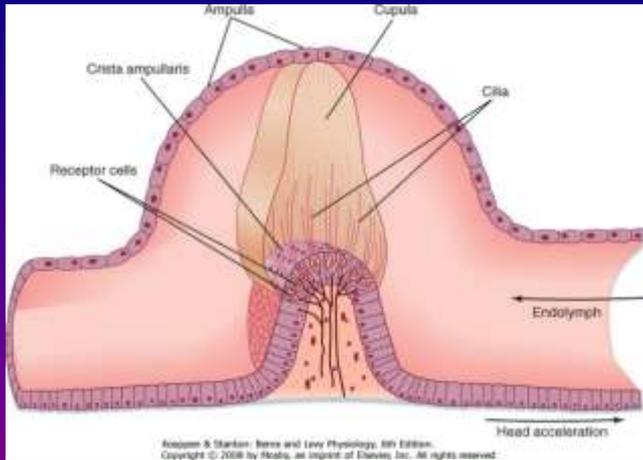
Advances in knowledge in anatomy and physiology



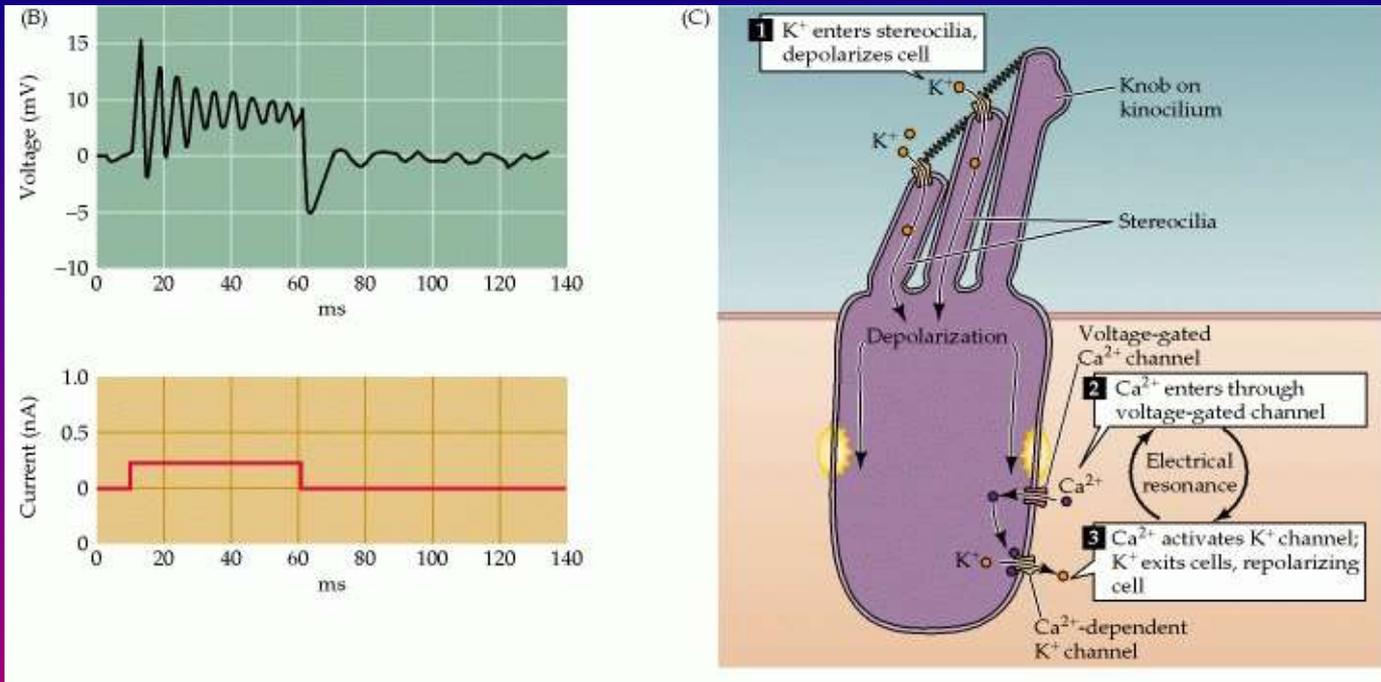
- Type 1 cells are fast acting and are VOR modifiers whilst type 2 are the transducers; genetically encoded ion transport mechanisms are identified (Hamid 2014)
- Otolith crystal integrity depends on Vitamin D metabolism (Dasgupta and Witana 2011)
- Vestibular sensory epithelium is highly tonotopic (van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



- Cupular endolymphatic flow deflect vestibular sensory cells in the canals to an ampullofugal or ampullopetal direction
- Striolar endolymphatic deflection deflect vestibular hair cells in the utricle/saccule to a striolo-petal or striolo-fugal direction

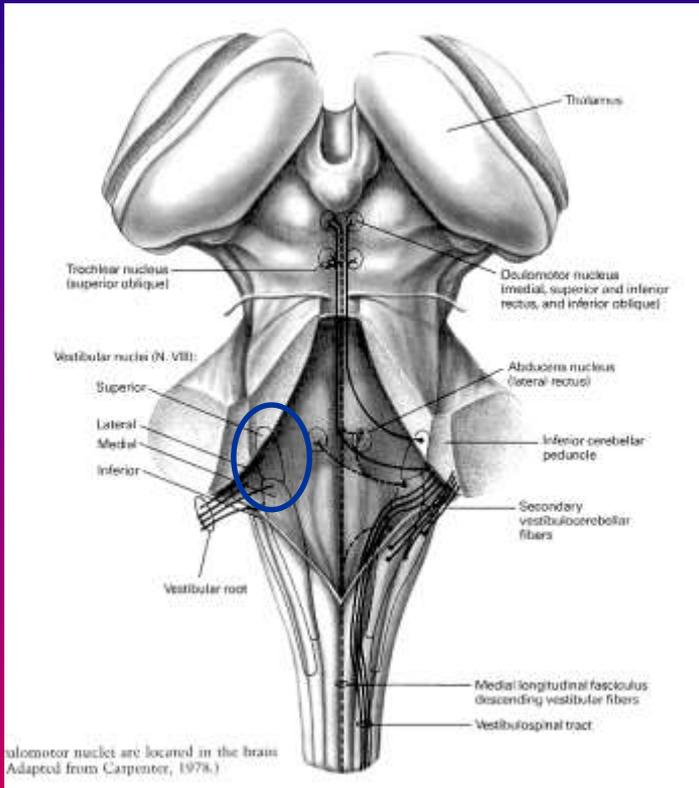


- Vestibular sensory cells exhibit resonance at a characteristic frequency of head movement or active/passive linear acceleration which is voltage dependant (Eatock and Songer 2012)
- This leads to infer that the vestibular sensory hair cells are tuned to incoming frequencies where they generate maximum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



The vestibular epithelia is therefore a cochlear analogue

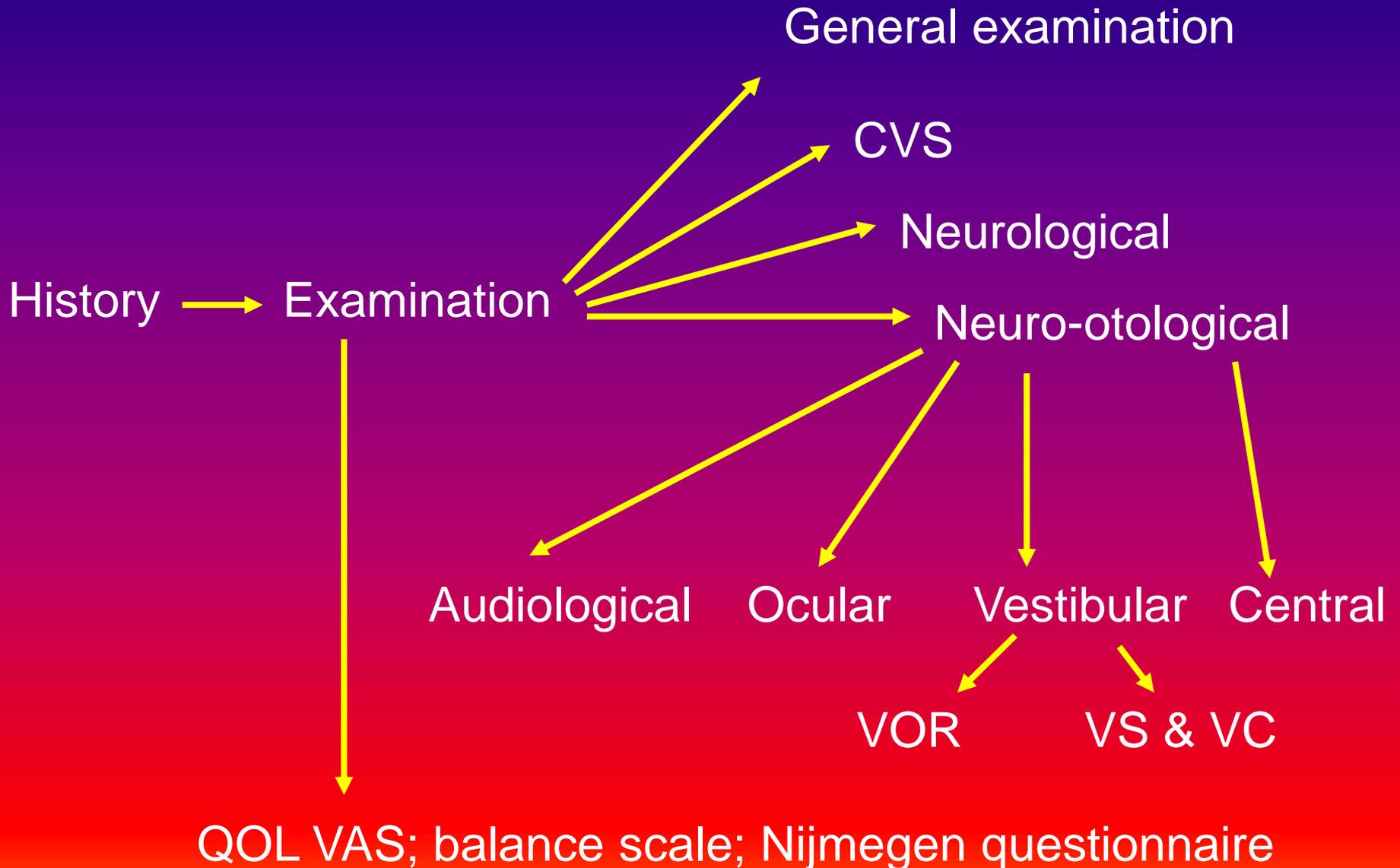
# Central vestibular connections (Patterson et al 2007)



- The medial vestibular nucleus is involved significantly in compensation through the GABA glycine mechanism
- Stress plays a significant role in modifying vestibular damage response through the hypothalamic pituitary axis

# Advances in vestibular diagnostics

# The clinical algorithm



# History

- Lloyd Minor in 1998 identified SCDS for the first time which suggested that in all cases of vestibular presentations, third window symptoms should be asked :
  - Gaze evoked tinnitus
  - Conductive dysacusis
  - Hennebert's phenomenon
  - Autophony
  - Tullio's phenomenon
- Over reliance on visual sensors occurs after vestibular assault generating the condition visual vertigo (Bronstein 2004)
- Mal de Debarquement syndrome (MDS) identified by Brown (1987) now incorporates persistent disorientation following passive motion or non motion stimulation (van Ombergen 2016)

Peripheral vestibular examination –  
Semicircular canals

## Spontaneous & gaze positional nystagmus (Kavanagh 1986)

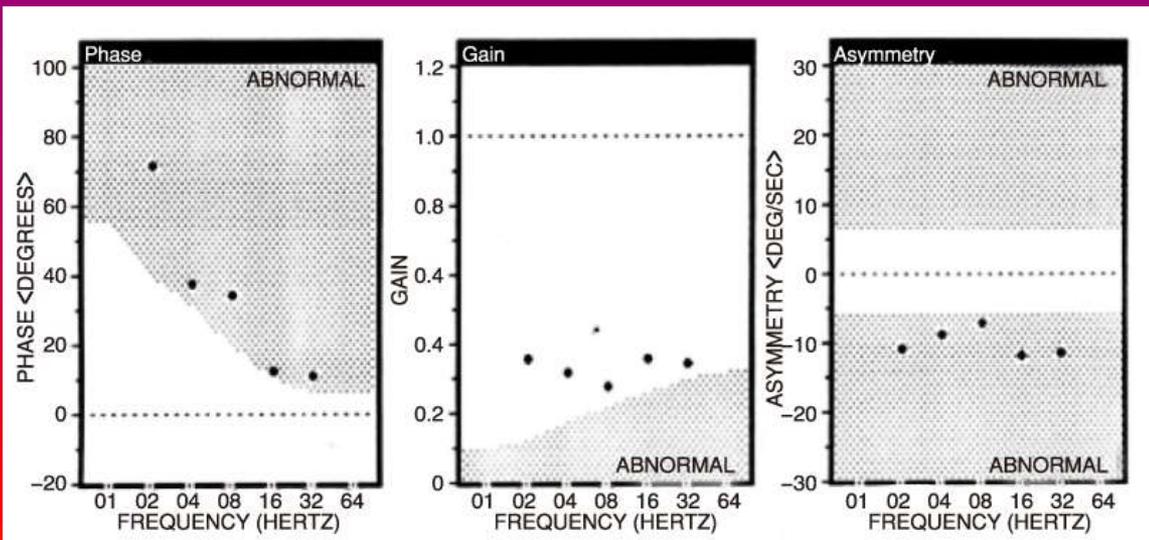
- Static response
- Indicates active lesion
- Amplitude indicator of prognosis
- High sensitivity and specificity

## Caloric response (Goncalves 2008)

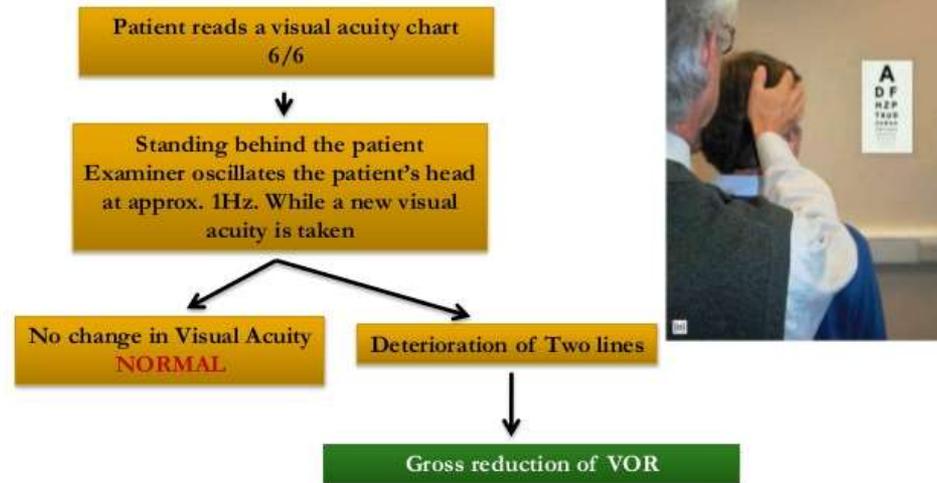
- Oldest devised test
- Irrefutable proof of vestibular weakness
- Measures non physiological 0.04 Hz zone
- High sensitivity and moderate specificity
- Inverted calorics are central

## Rotatory chair tests (Mach 1885; Barany 1907)

- Constant angular acceleration
  - Impulse angular acceleration
  - Step test angular acceleration
  - Sinusoidal acceleration
  - Optokinetic after nystagmus
  - Off vertical axis rotation & unilateral centrifugation
  - VOR cancel
- 
- The first 4 tests measure lateral semicircular canals at frequencies between 0.5 and 2 Hz are used only as adjuncts and have a very low specificity but a high sensitivity. Results obtained by gain, phase and symmetry (Hain 2015)
  - Optokinetic with smooth pursuits are important information for low velocity angular motion (Baloh and Honrubia 2011)
  - OVAR measures translational function
  - VOR cancel measures cerebellar function



## Dynamic Visual Acuity



(Cohen 1984)

- Measures low to mid frequency vestibular function
- Useful for prognosis and effects of rehabilitation
- Based on the principles of retinal slip on head movement
- Found in unilateral and active unilateral lesions
- Computerised variety (cDVA) with high sensitivity and specificity (Herdman 2010)
- Changes with age and vision

# The head shake test (Bonvier 2008)



- 20 headshakes – horizontal and vertical
- Horizontal post headshake nystagmus for vestibular asymmetry
- Non vestibular post headshake for central
- Checks mid frequency response LSCC (2-3 Hz)
- Useful for prognosis
- Not very sensitive or specific and does not localise side

## The head thrust (Halmyagi and Courthoys 1988)



- The bedside HT measures high frequency canal function between 4 to 7 Hz at velocities between 150 degrees to 500 degrees per second
- Operator dependant but in expert hands sensitivity and specificity in the 70% range (Jorns Haderli 2007)
- All 6 canals can be measured
- Only overt saccades seen
- Useful for prognosis

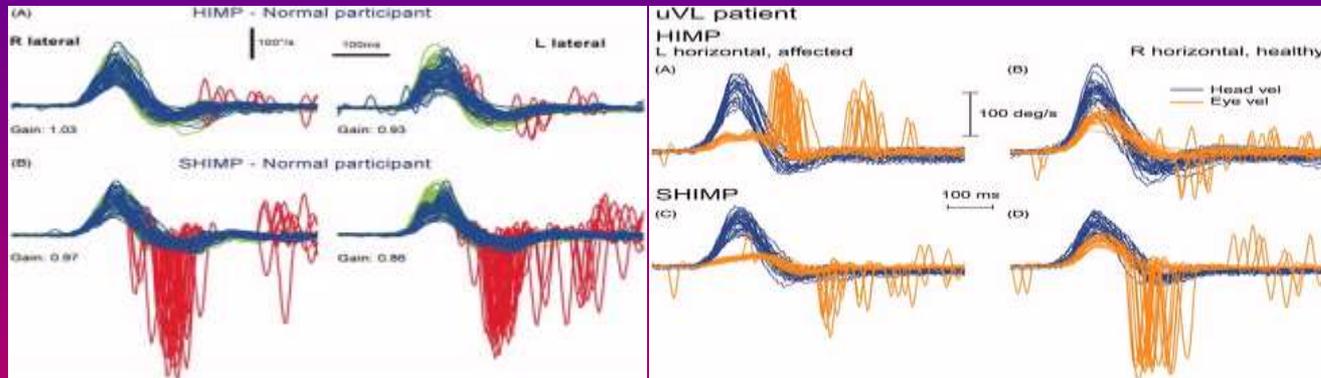
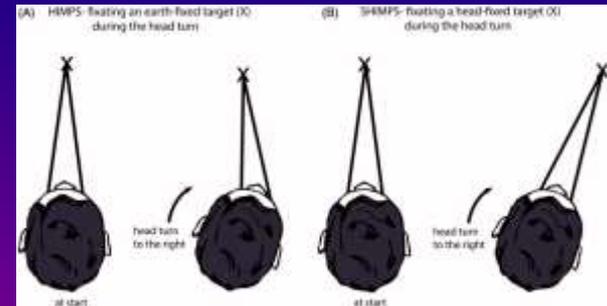
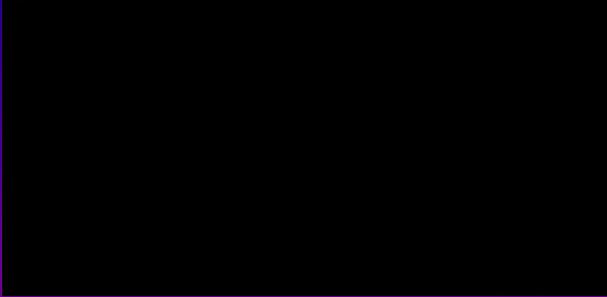
# The video head impulse test (McDougal 2009)

- Detects covert and overt saccades and useful for prognosis with gradual return of VOR gain and conversion of saccades
- Both covert and overt saccades seen
- Still evolving
- Operator dependant
- Cannot measure velocities in excess of 350 degrees per second
- Sensitivity and specificity from 60% to 100% (Bartolomew 2014)

## Caveats in video head impulse test

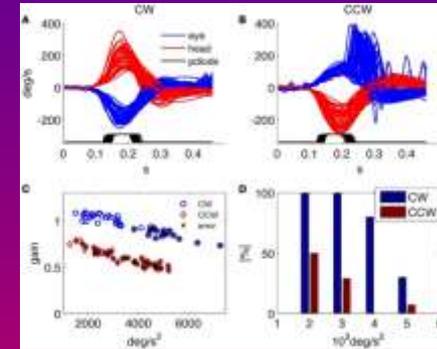
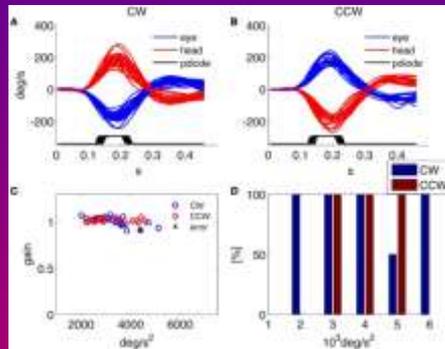
- Saccades and not VOR gain which indicate weakness
- Saccades beyond 400 milliseconds are physiological
- Saccades need to be repetitive and complete
- Normal VOR gain with saccades indicate compensation
- Low VOR gain without saccades could be artefactual or found in central lesions and migraine
- Anterior canal sparing weakness found in ototoxicity and head injuries
- Saccade clustering indicates compensation
- Oculomotor palsies might contaminate traces

# The suppression video head impulse test SHIMP (McDougal 2016)



- Head fixed target
- Normal subjects make anti compensatory saccades; hypofunction reduces these saccades
- Indicates residual vestibular function
- In my opinion does not yield any extra information and does not distinguish between compensated and decompensated lesions
- Is heavily influenced by the cerebellum as requires VOR suppression; depends on amplitude which is affected by many factors

# The functional head impulse test (Ramat 2012)



- Recently introduced measures high frequency functional angular VOR in canals
- Good correlation with video head impulse test
- Yet to be tried in children
- Advantage in measuring a whole range of head velocities not available in the standard vHIT software

# Hyperventilation induced nystagmus HVIN (Mandala 2013)

- Hyperventilation x 30 seconds
- Indicates lesion in the CPA; thus aetiological test
- Sensitivity is medium (65%) with high specificity (98%)
- Change of direction of nystagmus may indicate increased size of lesion
- Uncomfortable and cannot be used in epilepsy

## Hennebert's test (Shuman 2012)

- Tragal pressure or forced expiration against closed glottis/Valsalva
- Characteristic of third window syndromes
- Indicates aetiology
- Unknown sensitivity and specificity but personal series indicated more common than Tullio's phenomenon and slightly less common than mixed hearing losses

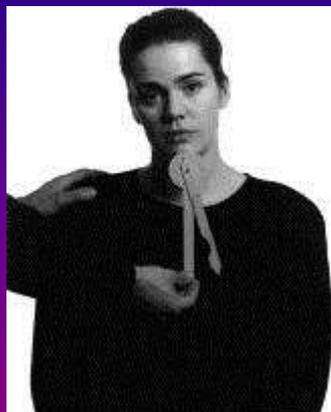
## The mastoid vibration test (Nutti and Mandala 2005)



- Mastoid stimulation on one side at 60-100Hz elicits horizontal nystagmus in vestibular weakness
- Non localising and directional preponderance may be utilised to identify unilateral weakness
- Strong correlation to calorics, thus very low non physiological stimulation
- Sensitivity and specificity reported very high in caloric weakness
- May be found in central lesions (Bonvier 2008)

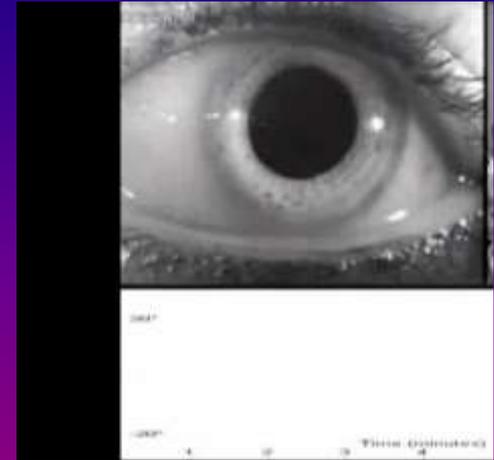
# Translational motion sensor tests

## Subjective visual vertical (Bohmer 1995)



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

## Ocular counter roll test (Otero-Milan 2017)



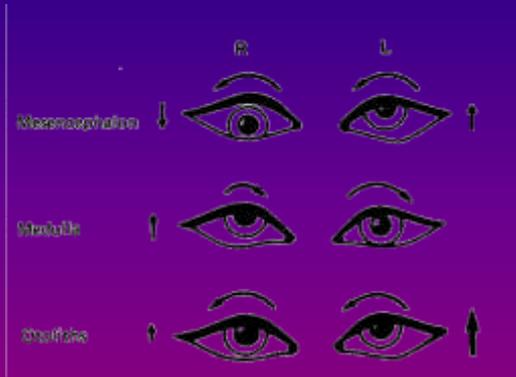
- Recording eye movement on passive and active head tilt
- Static tilt measures utricle whilst dynamic roll measures saccule
- ? Mid frequency response
- Low sensitivity and specificity due to cervical and truncal proprioception cues
- Does not need expensive equipment to measure when used VNG videography and playback
- May be a useful adjunct to other tests

## The head heave test (Ramat 2001)



- Translational analogue of head thrust
- Checks utricle high frequency responses
- Good correlation with oVEMP (anecdotal)
- Operator dependent
- May be found normally if not performed correctly
- Limited series to comment on sensitivity and specificity

# Skew measurements (Brandt and Dietrich 1991)



- With a new video head impulse software, can be objectively measured
- Usually central but can be due to otolith response in third windows with upward asymmetrical deviation of both eyes in the vertical direction

## The positional tests – Dix Hallpike, roll test and the deep head hanging test for BPPV

- Positional tests diagnose BPPV and do not indicate vestibular weakness but rather malfunctioning
- High sensitivity and specificity
- 10 different BPPVs recognised each treatable with particle repositioning exercises (Buki 2012)

# Vestibular evoked myogenic potentials

- Cervical VEMP measures saccule and neural connections (Colebatch 1992) and ocular VEMP measures utricle and neural connections (Courthoys 2012)
- Evolving field, so sensitivity and specificity difficult to compute but reported to be 60 – 70% sensitive and 80-90% specific
- May be useful for cochlea vestibular nerve lesions
- Very useful in third windows especially canal dehiscences
- Operator dependant and covariables in neck generated noise
- Checks one frequency of otolith response



## Off axis vertical rotation (Douglas 2012)

- Limited clinical value as can be nauseating and variations within subjects
- Measures otolith function at continuously changing orientation of the head and the body in respect to gravity

## The vestibulospinal tests

- Useful adjuncts to other tests
- Tests vestibulospinal reflexes
- Poorly localising
- Can be involved in central lesions
- Low sensitivity and specificity on their own  
(Babic 2003)

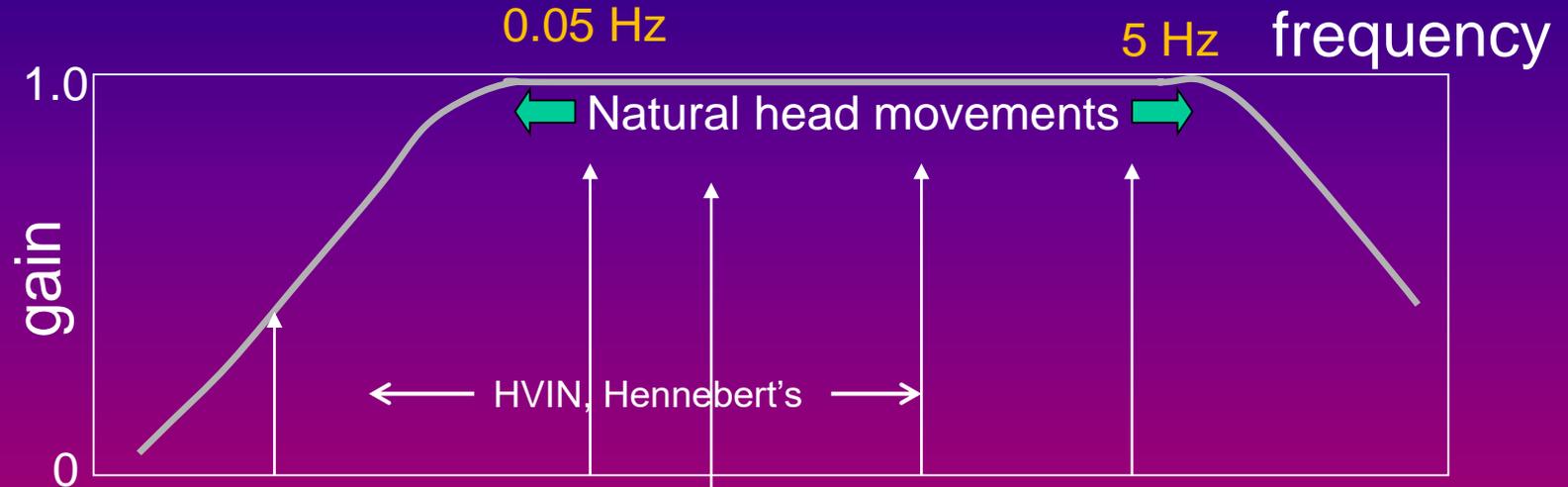
## Other assessments

- Dizziness Handicap Inventory
- HAD
- Quality of Life Visual Analog Score
- Nijmegen score for hyperventilation syndrome
- Anxiety scores if indicated
- Situational Characteristic Questionnaire SCQ
- Posturography only for prognosis and not for diagnosis
- Imaging studies (CT, MRI, Doppler) : <0.6mm HRCT for third windows and T1 and T2 weighted MRI for CPA tumours
- Blood tests when indicated

# The vestibular gram (Dasgupta and Bacon 2013)

- A vestibular gram not only indicates zonal weakness but also aids in aetiology as some pathologies affect specialised frequency zones for example high frequencies in vestibulotoxicity or all frequencies for example vestibular neuritis
- Frequency mapping of vestibular function leads to devising customised frequency specific rehabilitation for maximum possible outcome
- Research needed in this field

# The vestibular gram



Spontaneous / gaze tests (0 Hz)

Calorics (0.003 Hz)

Rotating chair (0.5 Hz)

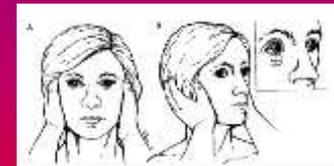
Headshake (2 - 4 Hz)

Head Thrust (5 Hz)

With fixation  
without fixation

Mastoid vibration (0.02 Hz)

Dynamic visual acuity (1 - 2 Hz)



Heave SVV oVAR cVEMP oVEMP OCR  
*High Static Mid Low Low Mid*

Utricle Saccule US Saccule Utricle US

## Case scenario

- 10 year old girl with sudden severe cochlear failure on the right and severe balance problems

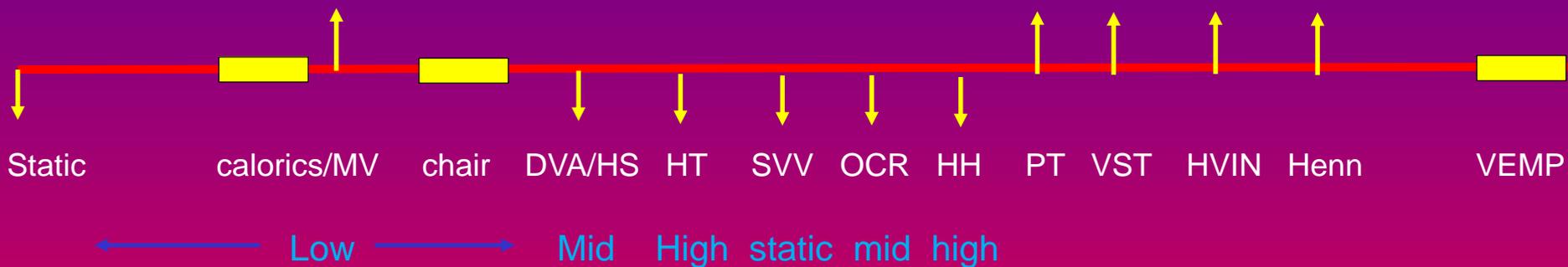
# The vestibular gram

## Legend

↑ Normal function

↓ Hypofunction

■ Not done



- Static, mid and high frequency angular sensor and low and high frequency gravitational sensor weakness; ongoing compensation; no TWS and CPA lesion; likely AIED/infective – high ESR and CRP
- Excellent response to steroids, static, mid and high frequency canal and low and high frequency otolith rehabilitation

# Central balance assessment

- Assessing balance function also must include central nervous system examination especially eye movements of central lesions
- Any eye movement which does not follow the peripheral vestibular pattern is either central or ophthalmological
- There may be mixed lesions
- There may be at least 20 different eye movements known which are indicative of central balance processing problems some of which are vestibular processing disorders; therefore, a thorough knowledge is essential regarding neuro anatomy and physiology
- Outcome of treatment is different for peripheral and central lesions

# Advances in management

- Treatment of active or acute condition with anti labyrinthine medication or anti migraine therapy
- Carbamazepine for vestibular neuralgia
- Customised vestibular and frequency and site specific rehabilitation (for example gaze stabilisation for static deficits and otolith recalibration for otolith dysfunction – Basta 2009)
- VR when condition inactive with best outcome
- Visual vertigo desensitisation with optokinetic stimulation – real and virtual (Pavlou 2004)

- Particle repositioning for canal specific BPPV
- Treatment for cause for decompensation
- Surgery for ELH or SOL or VP or SCDS
- Active rotation under optokinetic stimulation for MDS (Dai 2014)
- A CBT specialist is essential in any balance team

## Habituation

- A reduction in symptoms caused by repetitive exposure to the movement which evokes the dizziness
- Occurs due to the central process of neural plasticity but is not well understood
- Exercises: anything that evokes dizziness
- Head turns, nods tilts
- Body movements

## Vestibular Adaptation

- Ability of vestibular system to make long term changes in the neuronal response to asymmetrical input: neural plasticity gain
- Change in VOR gain (and VSR)
- Needs retinal slip (movement of the visual image across the retina). Brain tries to ↓ error enhancing gaze stabilization

## Substitution

- Substitution of, or reweighting of importance of other visual and proprioceptive cues/ strategies to replace the lost or impaired function : central pre- programmed saccades
- Development of alternative strategies to help maintain balance
- May be most important in functional recovery and especially for bilateral hypofunction
- Visual cues may substitute at low frequencies for gaze stabilization

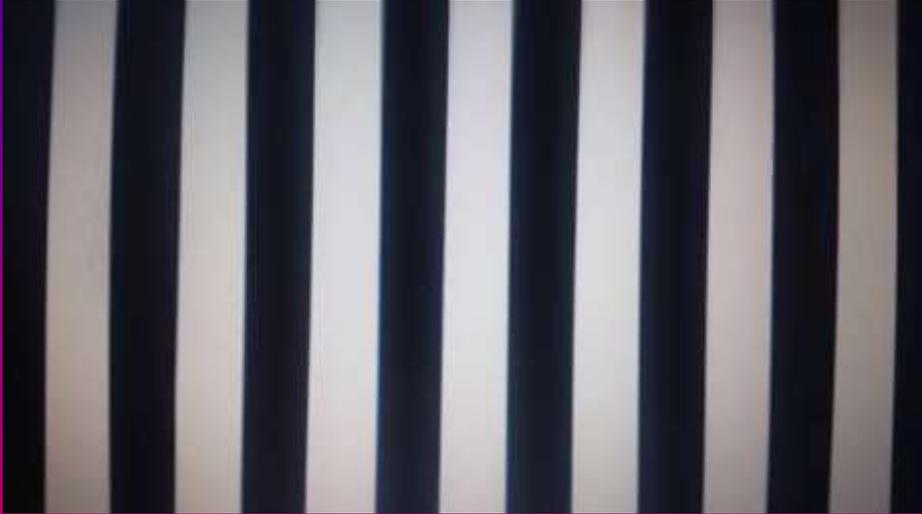
## Compensatory saccades

- Central pre-programming of saccades
- During or after head movements to stabilise gaze or large amplitude gaze shifts

## Neck mobilisation

- To promote VCR

# Optokinetic rehabilitation for visual vertigo and MdDS (Dai 2012)



# Otolith recalibration

- Standing on 2 legs with eyes open/closed
- standing on 1 leg with eyes open and closed
- 8 tandem steps (1 foot in front of the other) with eyes open
- standing with 2 legs on a foam support surface with eyes open/closed
- standing on 1 leg on a foam support surface (if you don't have a foam support surface, use a soft mattress)
- 8 tandem steps on a foam support surface,
- walking 3 m while rotating the head
- walking 3 m while vertically pitching the head in rhythm
- walking 3 m forward with eyes open/closed
- Follow your finger with your head in the opposite direction – for example if you move your finger to the right; your head moves to the left but your eyes move to the right

# Conclusions

- Vestibular Medicine is a hugely complex subject. This is because localising the exact site of the problem is crucial in the presence of multiple factors and overlapping. However, it can be done.
- With an aetiological diagnosis, localising the site of the lesion, decompensation syndromes, episodic irritation syndromes, psychological overlay and frequency response at which decompensation occurs; vestibular rehabilitation can be tailored adequately for best outcome.
- *Not everyone with a balance disorder will respond to a physiotherapy programme or medications.*



Labyrinthine vestibule – the final frontier, boldly going where no man has ventured before.....

Thank you