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Comprehensive Hearing Care

Hearing Your Best for Life

Anatomy & Physiology

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Basic Vestibular A&P

- Equilibrium is most basic sense
- Vestibular system evolves earlier
- Hearing arises from vestibular system
- Not just ear and peripheral system:
  - Cerebellum
  - Eyes
  - Muscles of postural stability
  - Descending motor tract

Basic Vestibular A&P

- Vestibular system consists of a number of complex anatomical structures and reflex pathways
- Structures code a change in acceleration of the head and/or body
- They also interpret the pull of gravity
- NOT sensitive to a consistent speed/motion

Basic Vestibular A&P

- Important definitions:
  - With hearing
    - Peripheral = cochlea, ME
    - Central = VIIIth nerve and up
  - With vestibular
    - Peripheral = end organ (ear), VIIIth nerve
    - Central = cerebellum, brainstem, brain
  - Class question:
    - Is a vestibular schwannoma a central or peripheral lesion?

- Basic rules that govern vestibular science:
  - Ewald's 1st Law → endolymph moves in opposite direction of head, causing eyes to move in same direction as endolymph
  - Ewald's 2nd Law → excitation stronger than inhibition

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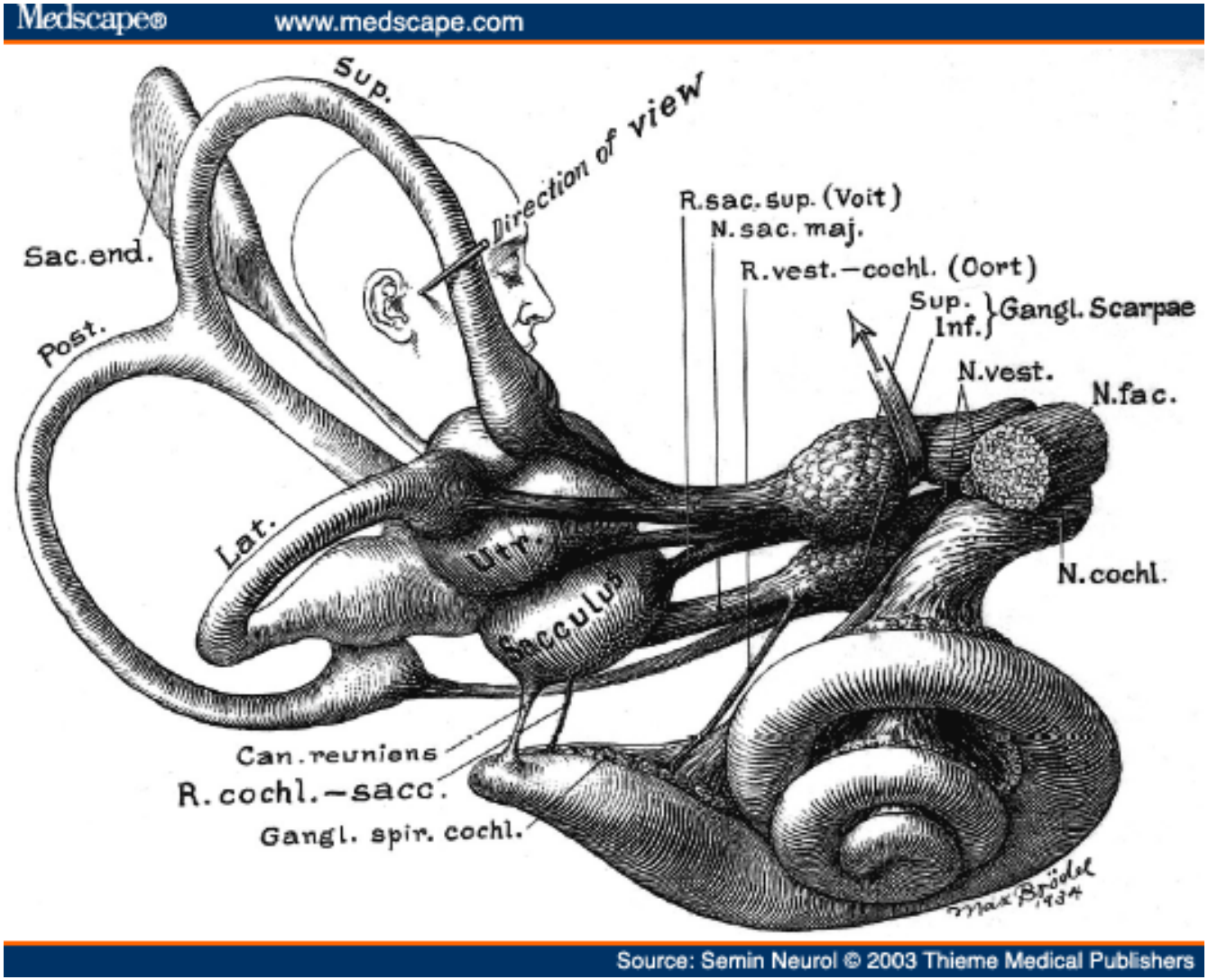
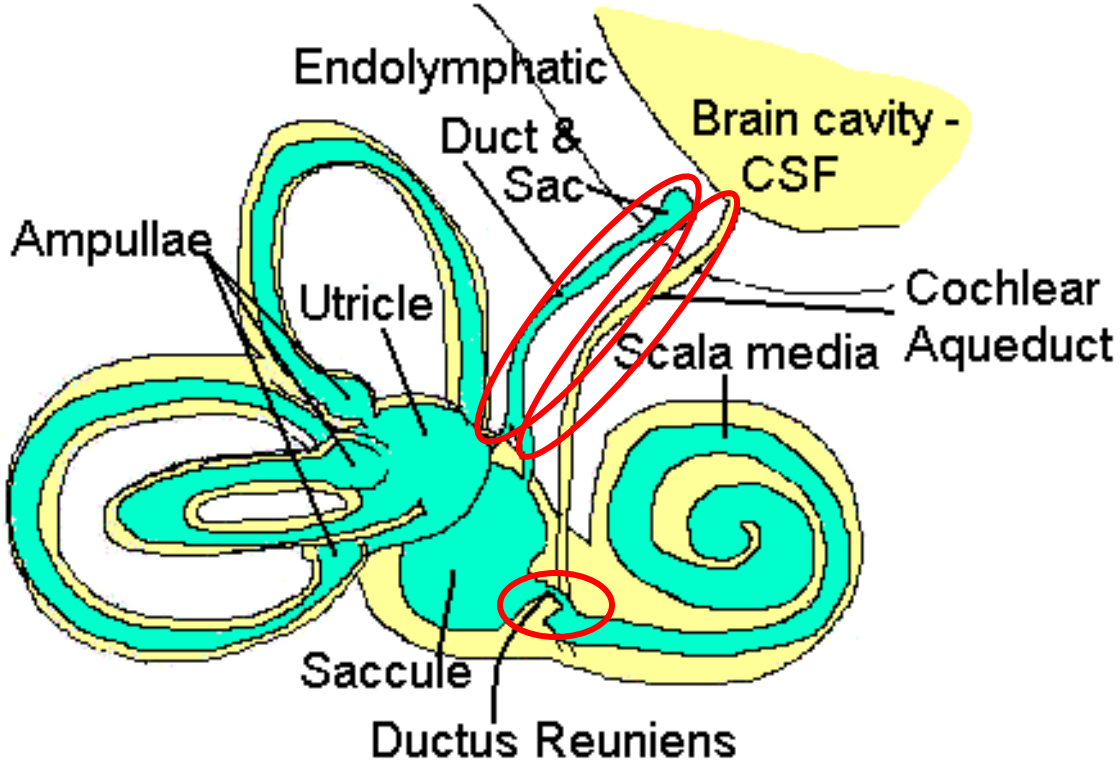
Peripheral A&P





- Ductus reunitis connects scala media to saccule
- Cochlear aqueduct runs between scala tympani and brain
  - Opening between C.A. and CSF not patent in adults
- Endolymphatic duct connects endo sac and saccule/utricle
- Cochlear aqueduct and endo duct might help with pressure regulation???

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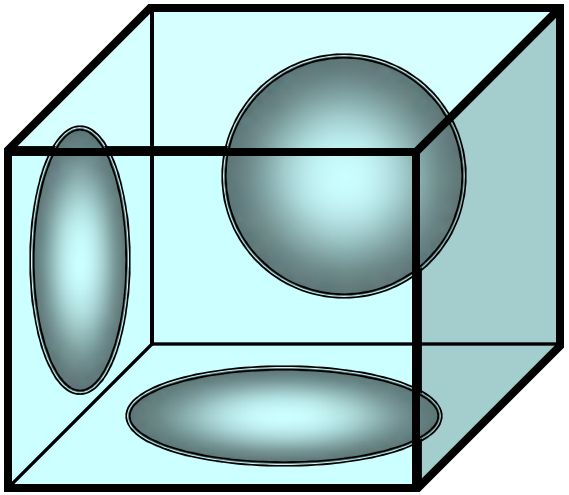
Semi-Circular Canals

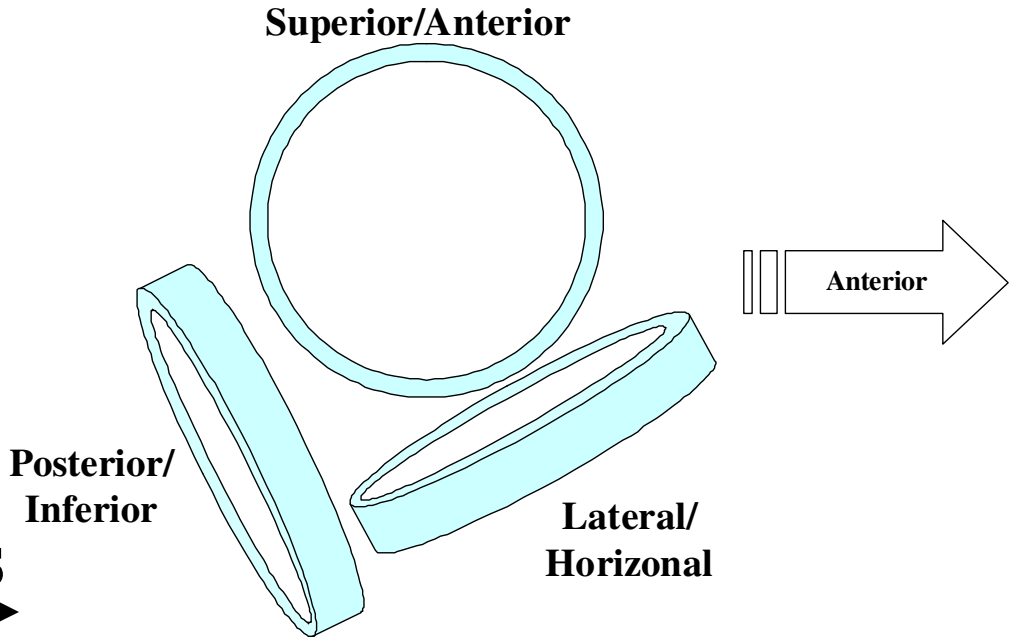
Semi-Circular Canals

- Sensitive to angular change in acceleration = yaw, pitch, and roll
- Oriented at right angles to one another
- Horizontal canal sits at 30-degrees
- Ampulated ends contain crista ampullaris



The semicircular canals lie perpendicular to each other

The canals are actually tilted at about 30 degrees



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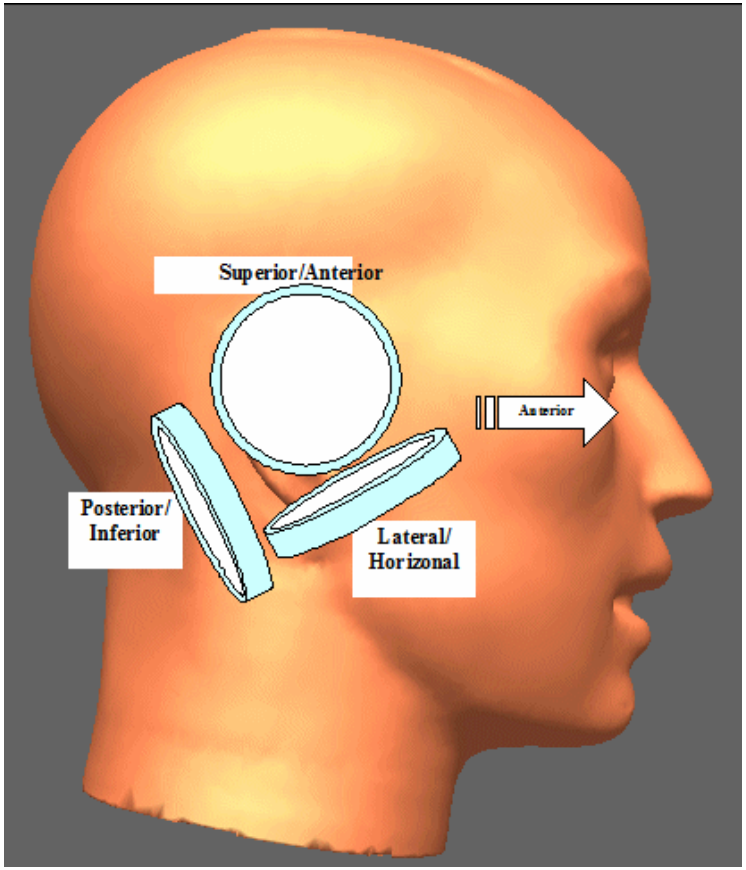
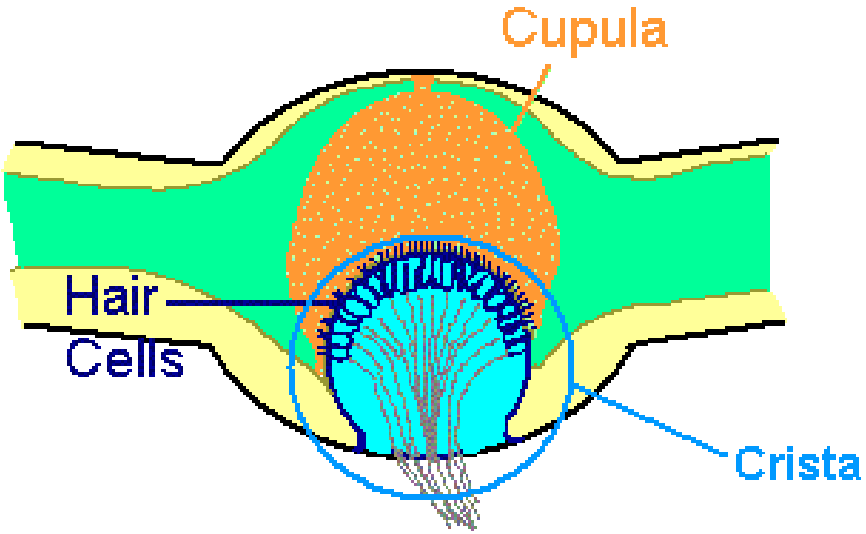
Semi-Circular Canals



Semi-Circular Canals



- Cupula has mass
  - Lags behind endolymph w/ initial movement
  - Will eventually catch up to endolymph
  - After movement stops, endo will stop and cupula will continue to move

Semi-Circular Canals

- Hair cells:
  - Tall central kinocilium
  - Gradually shorter cilia around kinocilium
  - Adjacent cilia connected via tip links
  - Movement of cilia either opens or closes ion channels via tip links
  - Opening causes excitation, while closing causes inhibition (in reference to spontaneous firing)

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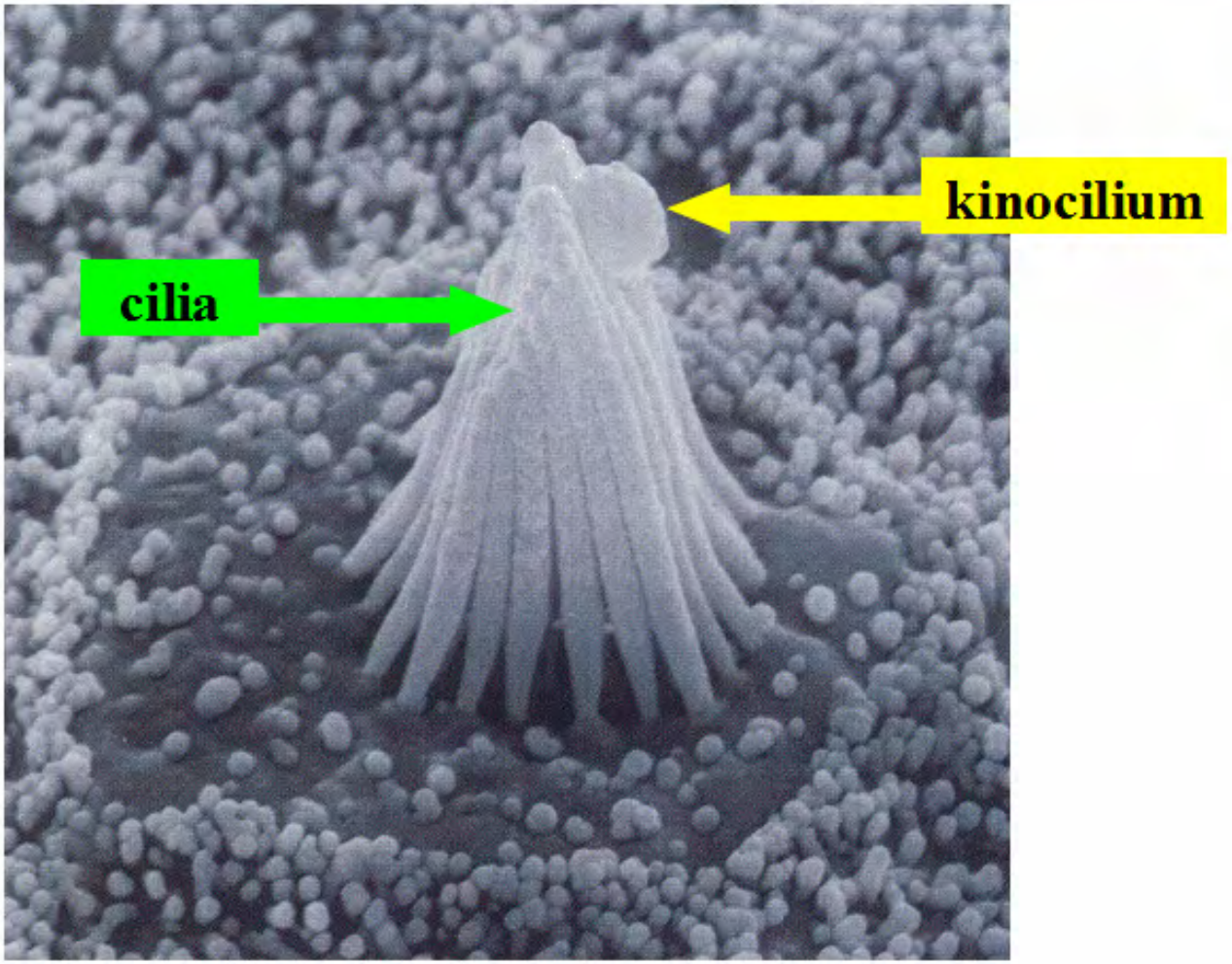
Semi-Circular Canals

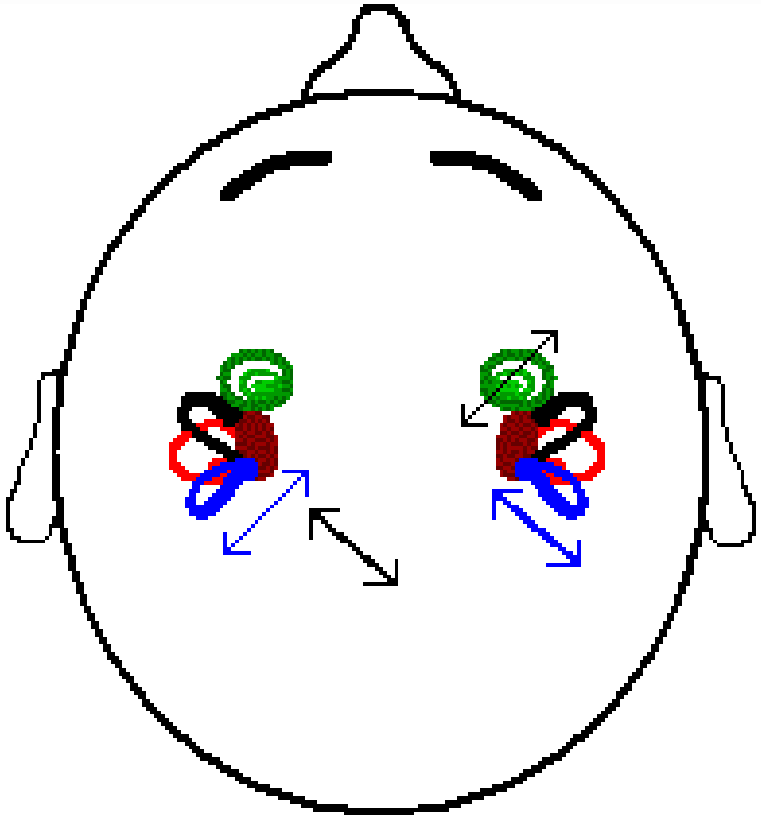


Semi-Circular Canals

- Oriented in matched pairs
  - Ears work in tandem = push-pull system
  - One stimulatory response, other inhibitory
  - Neural homeostasis achieved (when not moving) if equal response from both ears (clinical correlate: vn)

Semi-Circular Canals

- Thoughts
  - Ears have a spontaneous firing rate
  - Vestibular nucleus (central) is comparing/contrasting firing rate from both sides
  - Difference between sides = movement



The semicircular canals work in matched pairs, but the pairs are not actually the same canal on each side (depends on movement)

Semi-Circular Canals

- Movement of cupula
  - Utriculopetal
    - Towards from the utricle
    - Stimulatory in the horizontal canals
  - Utriculofugal
    - Away from the utricle
    - Stimulatory response in the posterior and anterior canals
  - Movement towards the kinocilium is always excitatory
    - HC = Kinocilium on side of utricle
    - PC & AC = Kinocilium on side of haircell away from utricle

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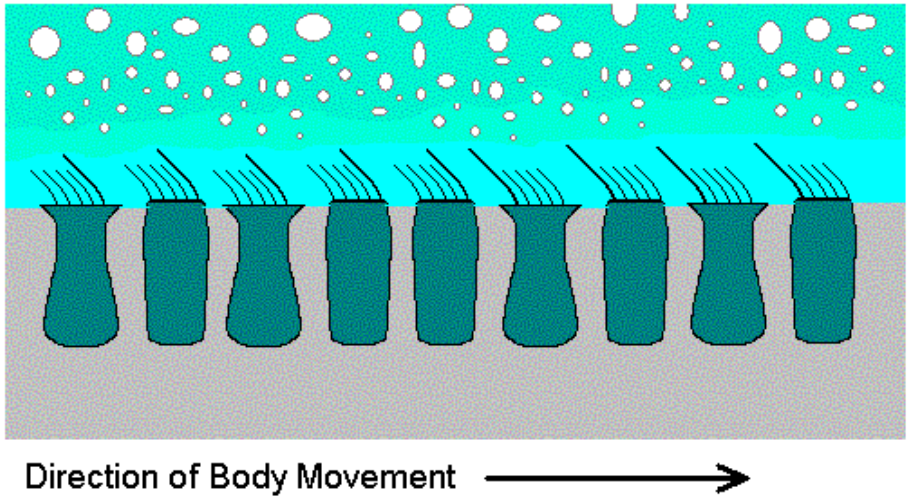
Utricle and Saccule

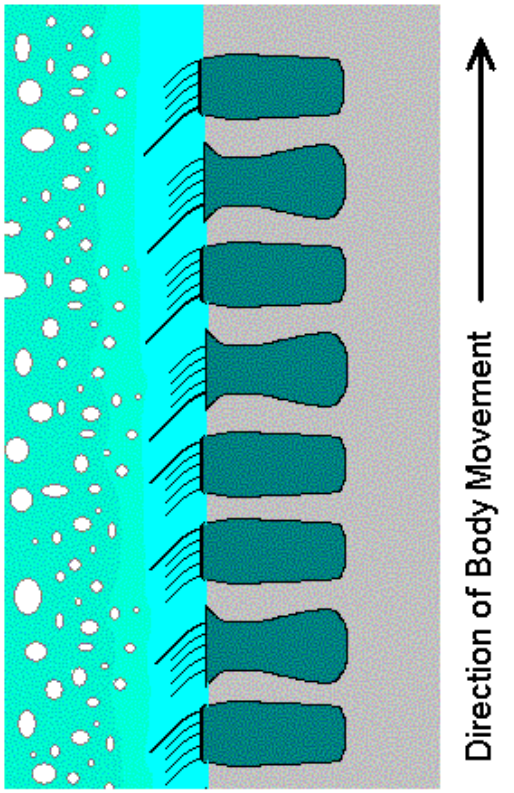
Utricle and Saccule

- Sensitive to linear acceleration
- Gravity detector
- Utricle oriented in horizontal plane
- Saccule oriented in vertical plane
- Each structure has slight curvature, which allows different sections to be stimulated by slightly different motions

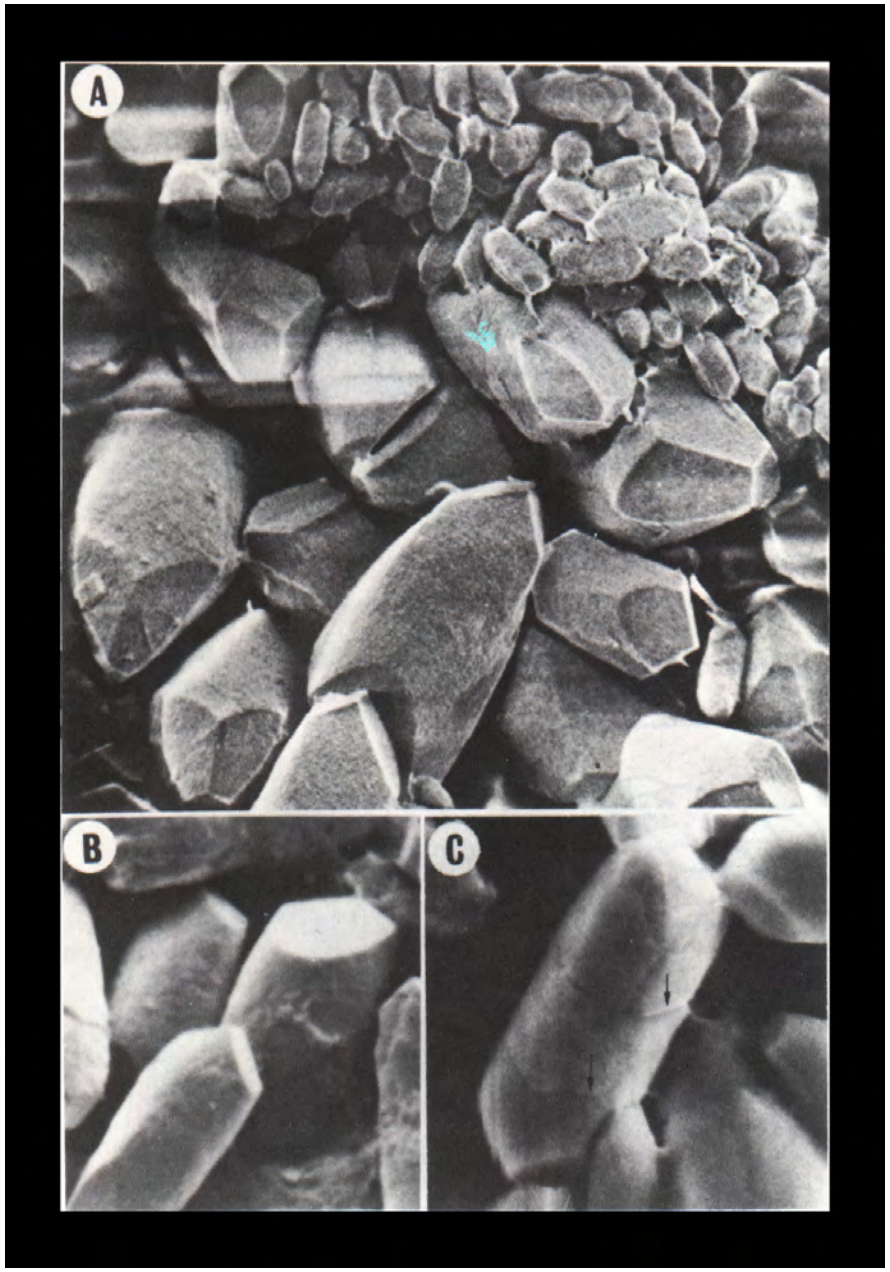
Utricle and Saccule

- Each contain a sensory structure- macula
  - Hair cells imbedded in a gelatinous layer
  - This layer contains otoconia, which add weight and create drag
  - The cause of BPPV and the Crisis of Tamarkin (more about these later)





- **Macula of the utricle is oriented in a horizontal plane**
- **Macula of the saccule is oriented in a vertical plane**



Electron Micrograph

(Lim, 1969)

Stones from Utricle show both small and large crystals

Close-up of crystals showing cylindrical forms

Utricle and Saccule

- Hair cells are similar to those in ampula
- Hair cells oriented in many orientations
  - Sensitive to motion in multiple directions
  - Kinocilium oriented in different directions/plans, so may stimulate or inhibit

Utricle & Saccule Biomechanics

- Otoliths are made from calcium carbonate (Latin for "ear stones")
- The endolymph in the utricle and saccule are calcium deficient
- The otoliths have a life span and are being constantly recycled – they dislodge from macula and float in endolymph

Utricle & Saccule Biomechanics

- As we age:
  - Our calcium absorption slows (vitamin D)
  - Protein matrix around otoliths break down and become less "sticky"
- This may cause a higher concentration of free-floating otoliths

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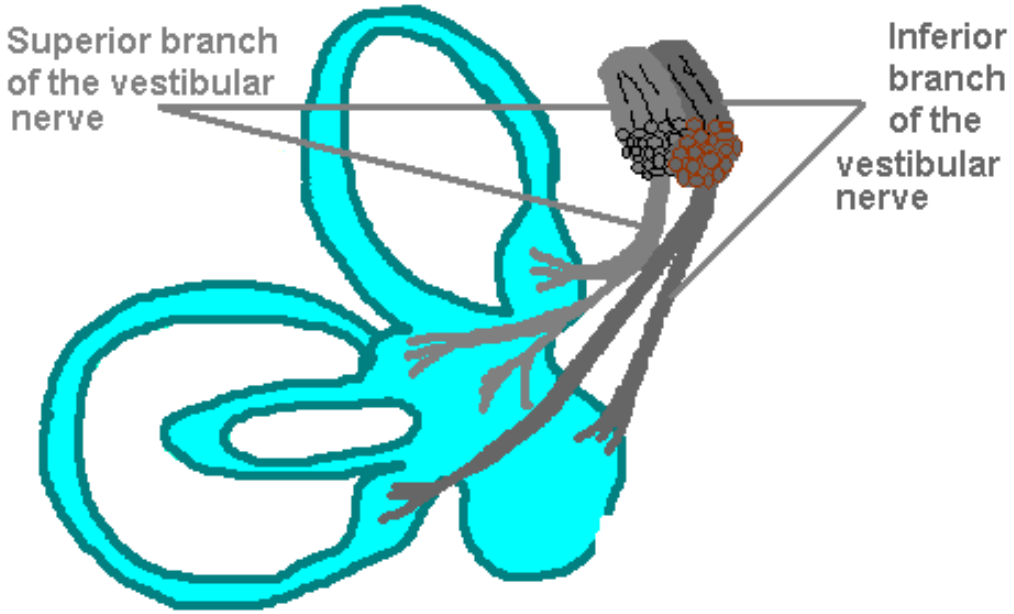
Cranial Nerve VIII and Blood Supply

Vestibular Branch of VIII

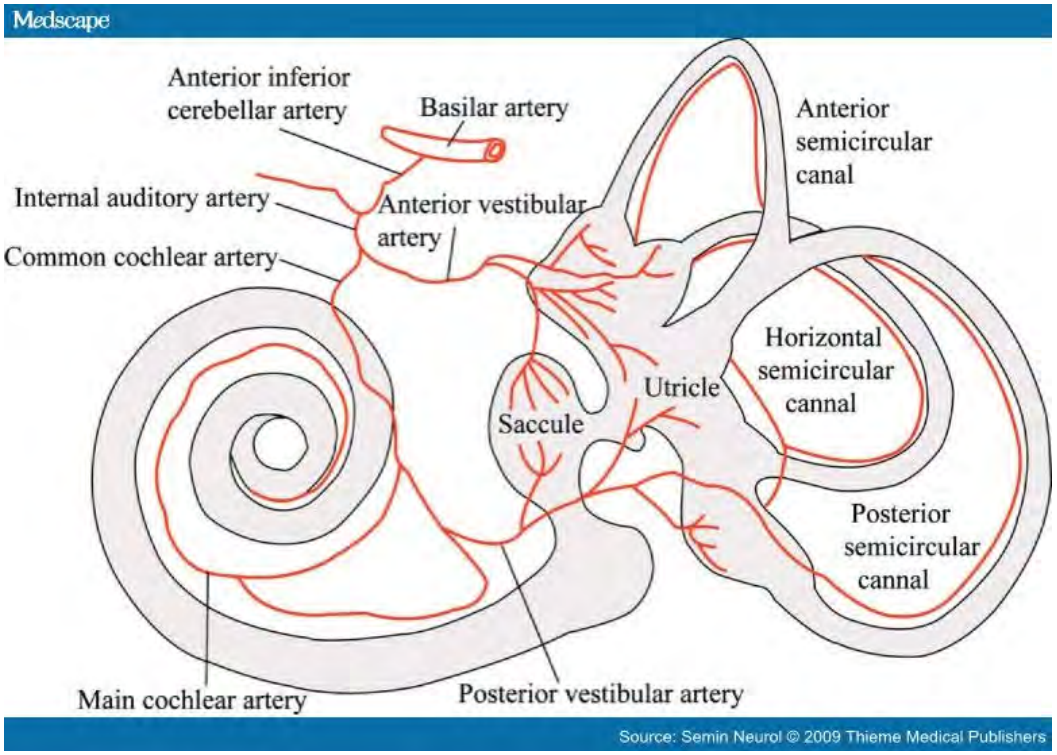


- Superior branch innervates HC, AC, and utricle
- Inferior branch innervates PC and saccule

Cranial Nerve VIII

- Differentiating which branches are involved helps with diagnosis
- We have tests that look at each branch individually
  - Calorics = superior branch
  - cVEMPs = inferior branch
  - ABR, hearing, etc. = cochlear branch
- If you see deficits in all 3 branches unilaterally, a mass lesion should be r/o

Blood Supply



- Inner ear fed by posterior circulation
- Common blood supply between cochlea and vestibular system at level of basilar artery (and outward)
- Differentiate after AICA

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Central A&P

Central Vestibular System

- Vestibular system multisensory
  - Vestibular input
  - Visual input
  - Somatosensory input
  - Motor input
- Information from each system is integrated in the vestibular nuclei
- Equilibrium and postural control information sent to other brain structures

Central Vestibular System

- Balance information sent from midbrain to multiple regions of the cortex
- Connections to a multitude of brain areas
- Diffuse pathways are difficult to study
- However, it is generally agreed upon that there are 4 major brain regions for balance

Central Vestibular System

- Four major areas of the brain for balance:
  - Voluntary motor movement = frontal lobe, specifically precentral cortex and connections to pyramidal sytem
  - Visual information = occipital lobe w/ connections to frontal cortex
  - Deep brain basal ganglia = help coordinate muscle movement
  - Cerebellum = control of posture, coordinate sensory and motor information

Central Vestibular System

- Efferent and afferent connections between systems
  - Information from each sensory system is received and integrated in the vestibular nuclei
  - Corrective postural and visual control information is sent to the muscles of the neck, trunk, legs, arms, and eyes
- Also connections with reticular formation (autonomic nervous system)

Reflex Pathways

Three primary pathways measured clinically:

- Vestibulo-Ocular Reflex (VOR)
  - VNG, VAT, CD-VAT
- Vestibulospinal Reflex (VSR)
  - SOT
- Vestibulo-Collic Reflex (VCR)
  - VEMP

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Vestibulo-Ocular Reflex

Vestibulo-Ocular Reflex (VOR)

- VOR
  - Helps with gaze stabilization during head/body movement
  - Allows desired object to stay on fovea, even when walking/running or moving head
  - Deficit = oscillopsia

Semi-Circular Canals

- Each SCC is connected to extra-ocular muscle
- Excitation:
  - Horizontal canal:
    - Ipsilateral medial rectus
    - Contralateral lateral rectus
  - Anterior canal:
    - Ipsilateral superior rectus
    - Contralateral inferior oblique
  - Posterior Canal:
    - Ipsilateral superior oblique
    - Contralateral inferior rectus
- Inhibition in corresponding antagonist muscles

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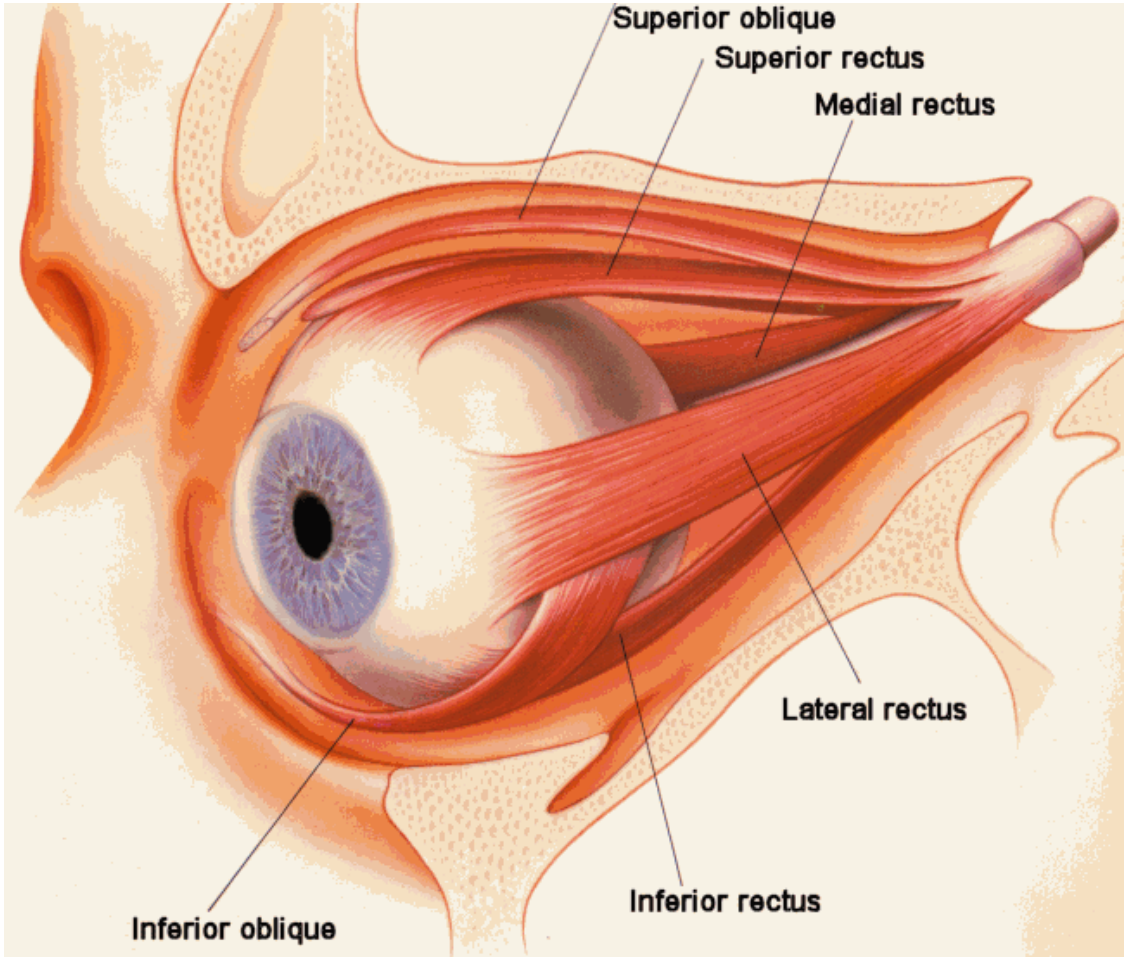
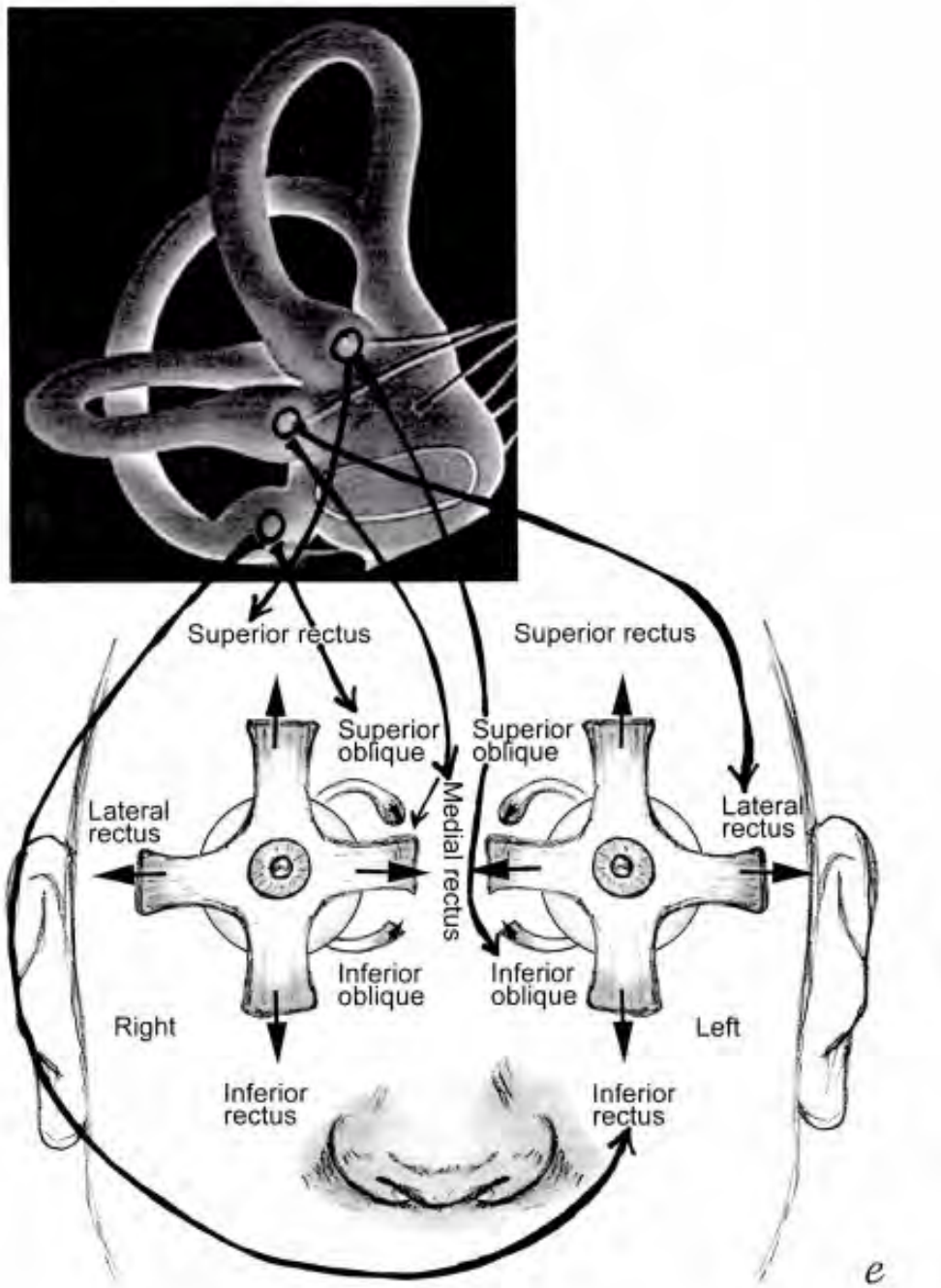
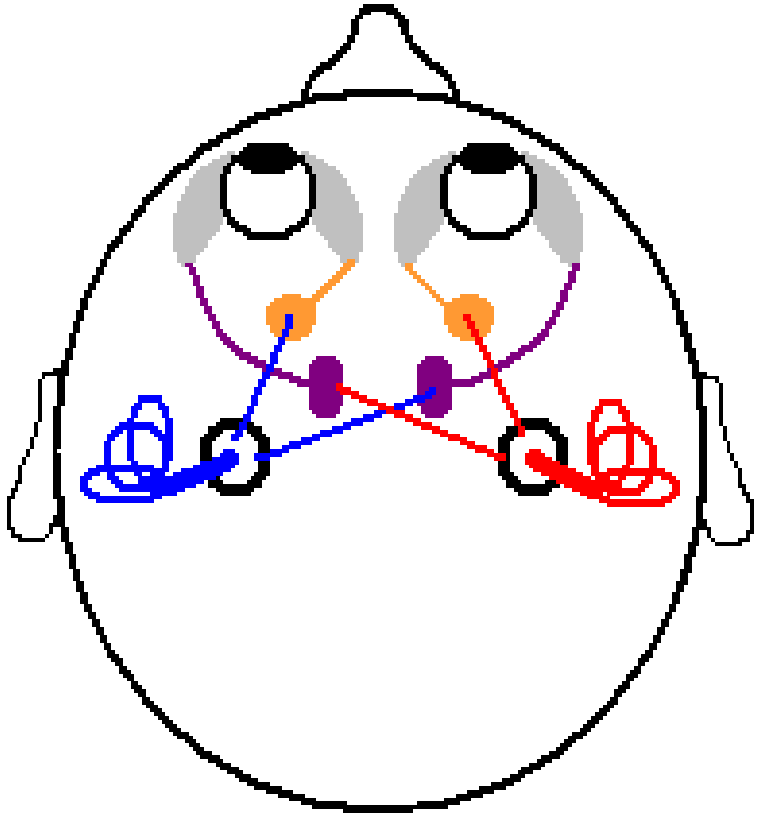
Eye Muscle Anatomy



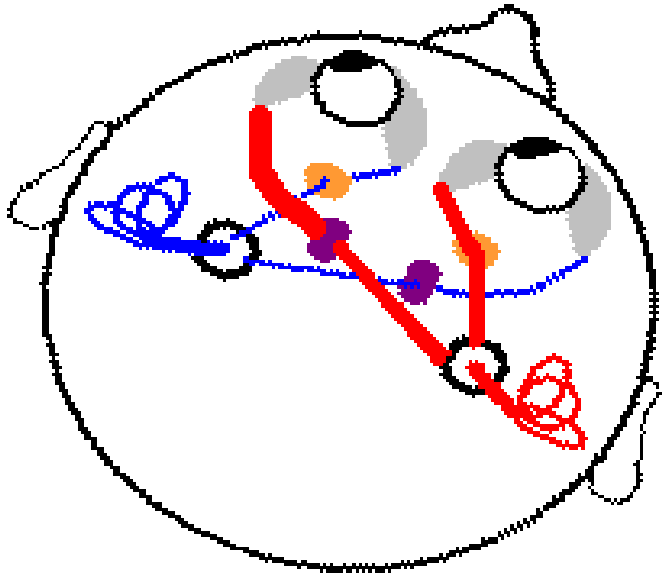




contra VI nucleus→lateral rectus

HC→vestibular nucleus

ipsi III nucleus→medial rectus



- Head turns right = endolymph moves left (Ewald's 1st law)
- This is utriculopetal for the right ear = excitatory
  - Left lateral rectus contracts
  - Right medial rectus contracts
- This is utriculofugal for the left ear = inhibitory
  - Right lateral rectus relaxes
  - Left medial rectus relaxes

Vestibulo-Ocular Reflex (VOR)

- Limited range of eye deflection
  - Eyes can only move so far
  - If body continues to rotate, eyes will eventually reach limit
  - CNS will cause eyes to rapidly move back to center to establish new focal point = saccade

Vestibulo-Ocular Reflex (VOR)

- If body continues to rotate after eyes have moved to center:
  - Repeat of slow movement in direction opposite of head/body movement
  - Eyes will again reach their limit, and have saccadic rapid movement back to center
  - This alternating slow and rapid eye movement is called nystagmus
  - Note: slow movement of eyes occurs at same speed as head/body movement (equal and opposite)

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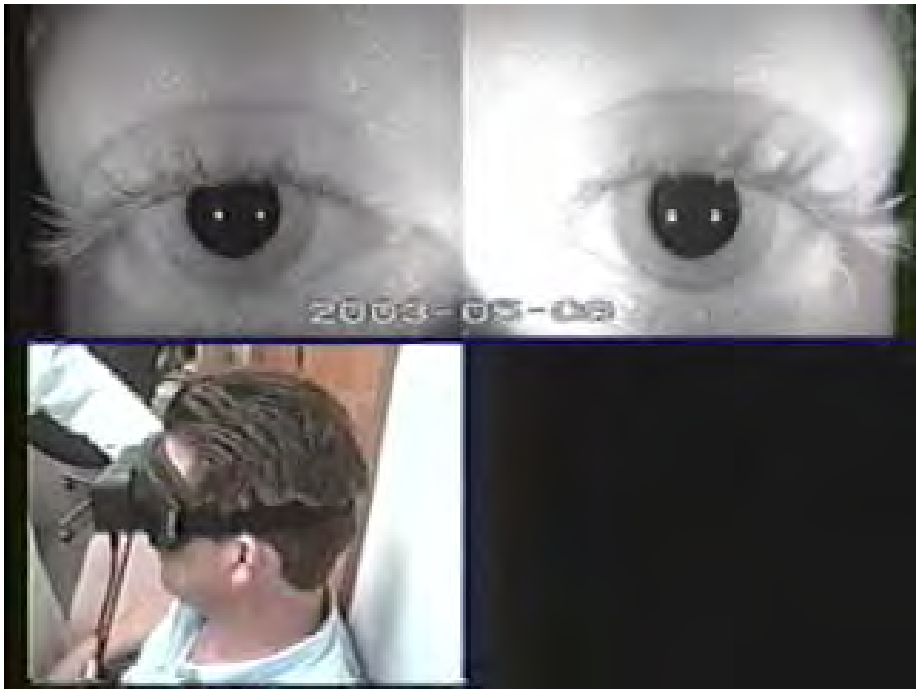
Nystagmus

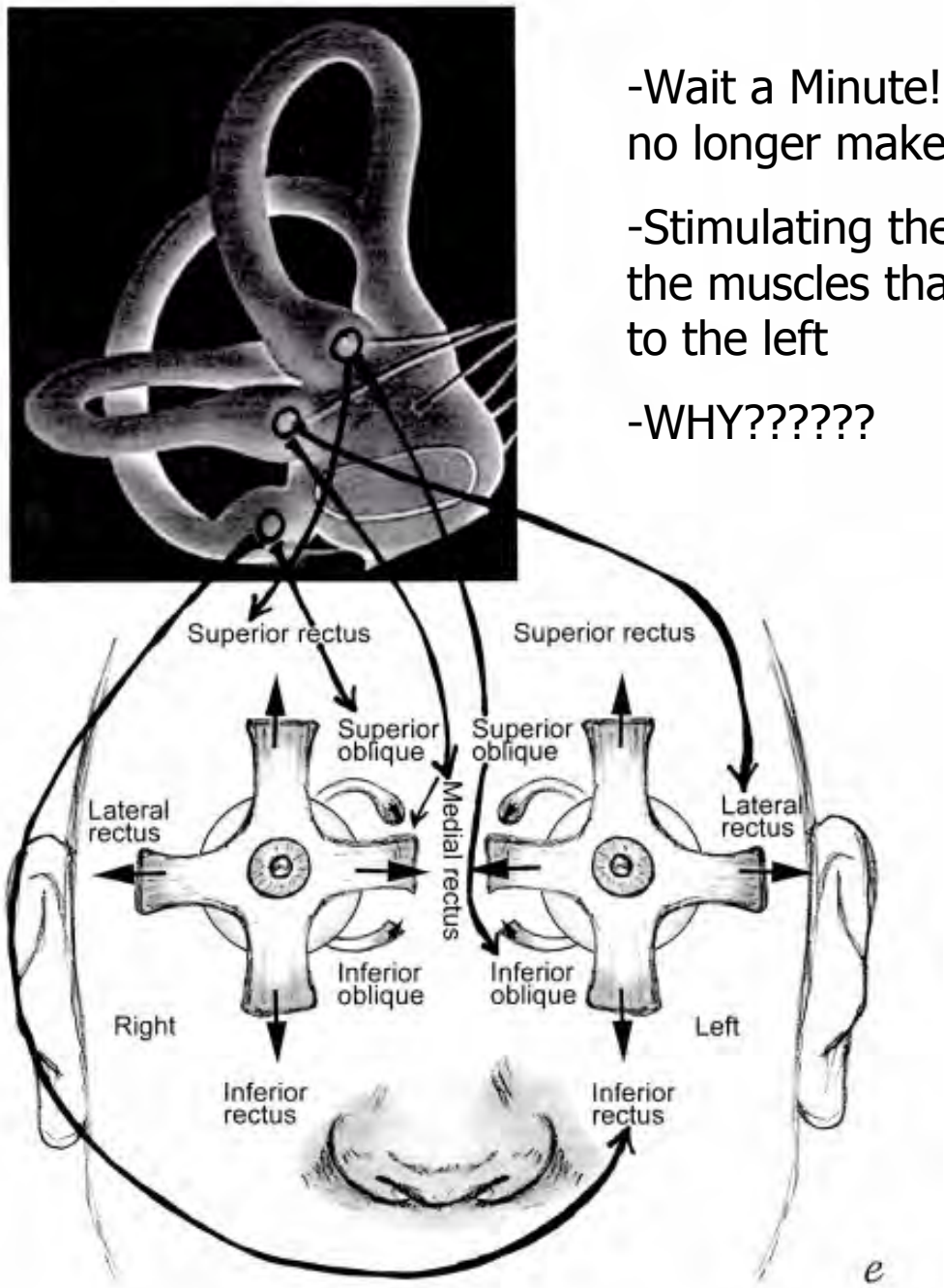


Note: this patient is not moving…

Why might he have nystagmus?

Nystagmus

- We describe nystagmus in reference to the fast phase (direction "beating")
- Remember:
  - Slow phase is driven by the ears
  - Fast phase is driven by the CNS
  - Nystagmus beats towards a stimulated ear
  - Nystagmus beats away from an inhibited ear



-Wait a Minute!!!!! This diagram no longer makes sense.

-Stimulating the right HC excites the muscles that move the eyes to the left

-WHY??????

Back to Ewald's 2nd Law

- Excitation is stronger than inhibition
- Let's measure a patient's nystagmus
  - Patient has a deficit in the right vestibular system
  - Spin the patient right
  - Spin the patient left
  - How would the nystagmus compare between directions? Why?

Nystagmus w/ Peripheral Vestibular Lesions

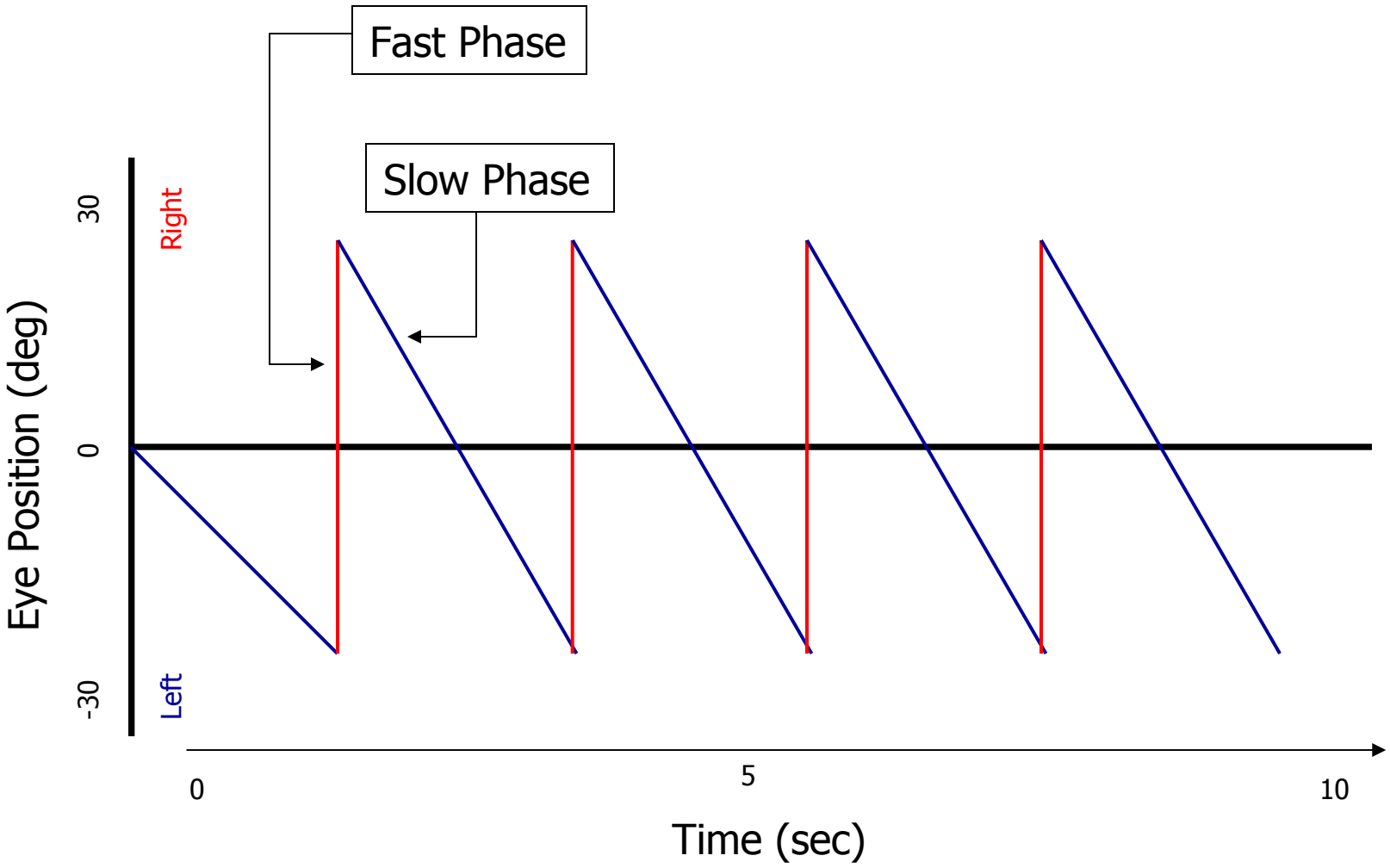
- How does nystagmus look with a deficit on one side?
- The vestibular nucleus does not understand that one side is lesioned when integrating sides
- Nystagmus beats away from the side with a deficit-causing lesion
- Nystagmus beats towards the side with an irritative lesion. Example????

Clinical Correlate

- If we know our ear anatomy and the specifics of the VOR, we can identify the side of lesion by watching a patient's eyes for nystagmus

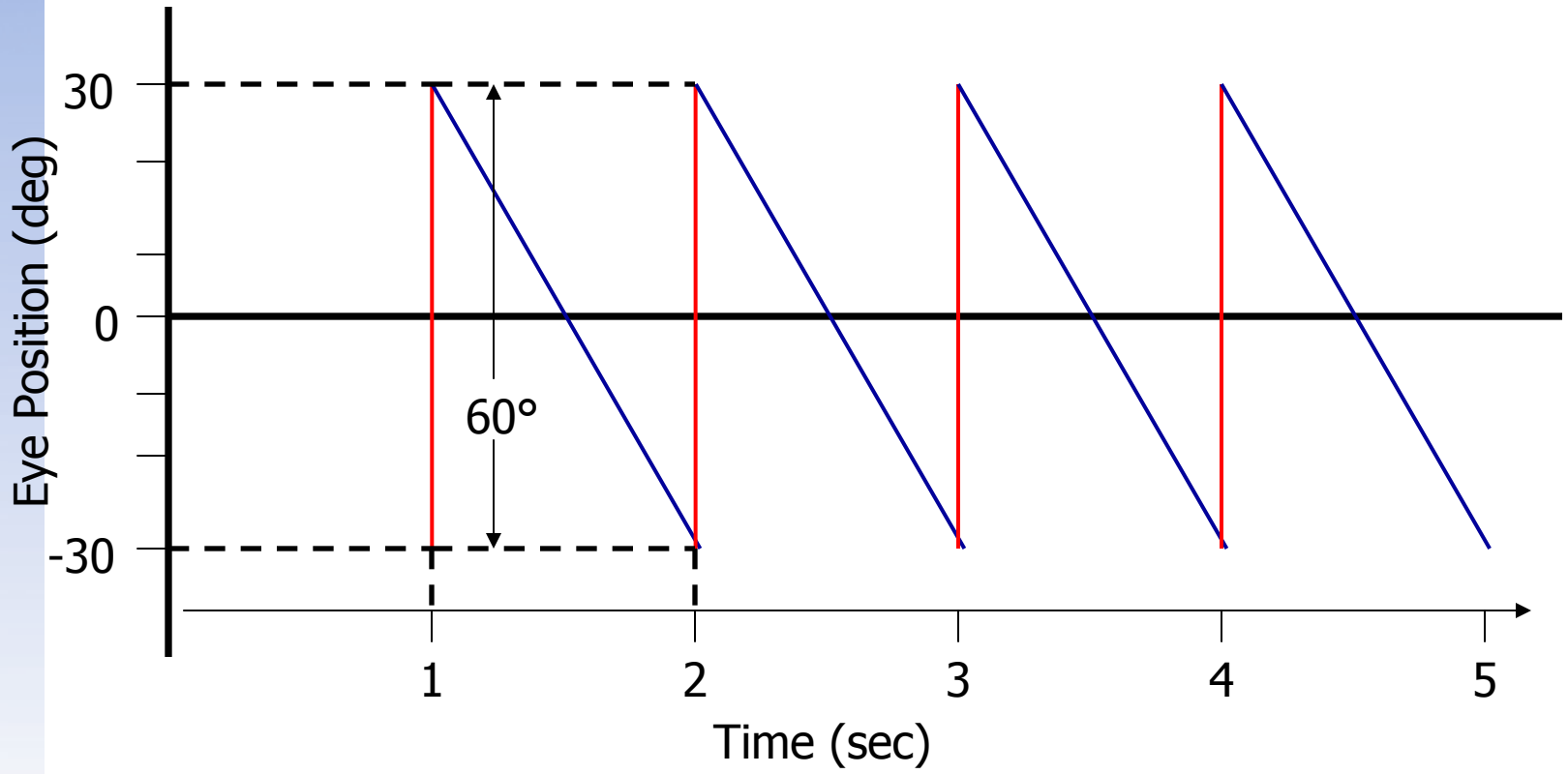
- We have clinical tests (calorics) that can stimulate/inhibit an ear individually, and we can measure nystagmus

Nystagmus



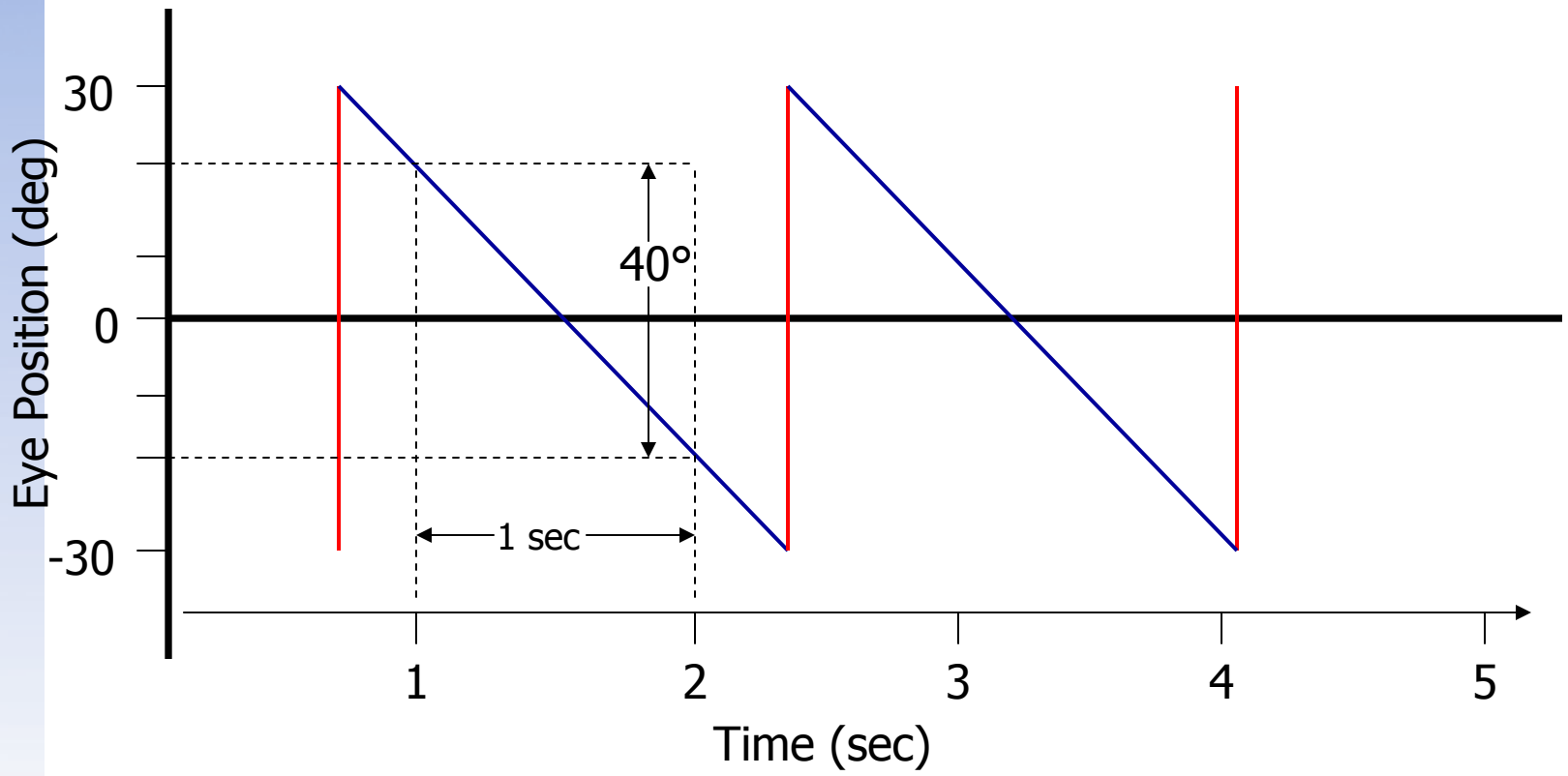
Nystagmus

- Defined as degrees/second
- How many degrees does slow phase move over a one second interval?



Nystagmus

- What if the beats do not line up with the second marks?



Nystagmus

- A few final thoughts/questions:
  - What would nystagmus look like with a bilateral and equal vestibular deficit?
  - Does nystagmus persist indefinitely after a permanent lesion?
  - Can spontaneous nystagmus be caused by problems in places other than the ear?

Nystagmus

- Let's consider a unilateral left vestibular lesion (decreasing output)
  - What is the patient's subjective experience?
  - Why would a patient have these symptoms?
  - Describe the patient's nystagmus.

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Vestibulospinal Reflex

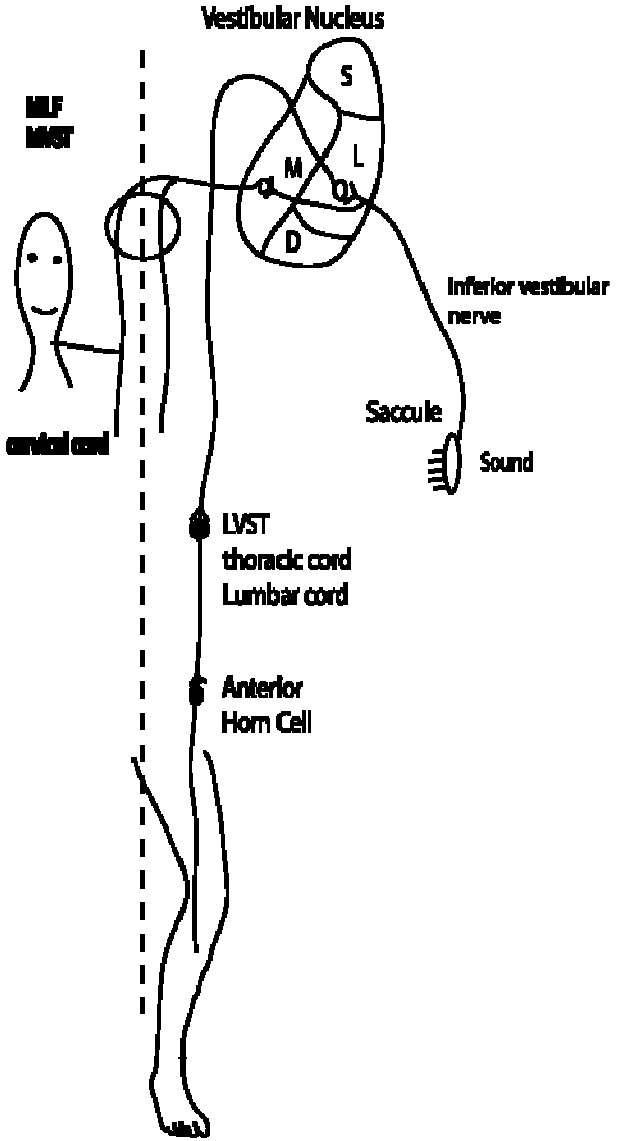
Vestibulospinal Reflex (VSR)

- Connection between vestibular system and stability muscles of the torso and lower extremities (below the neck)
- Vestibular system detects movement and postural sway and corrective signal sent to muscles to maintain balance and coordinate movement
- Can be volitional or reflexive

Vestibulospinal Reflex (VSR)

- Volitional
  - Conscious shifting of weight from center of gravity to move
  - Modified throughout time through learning
- Reflexive
  - Short-latency response to perturbation
  - Ankle strategy = small/slow perturbation
  - Hip strategy = weight shifts
  - Suspensory strategy = lowering COG
  - Stepping strategy = new COG

Vestibulospinal Reflex (VSR)



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Vestibulospinal Reflex (VSR)

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Vestibulocollic Reflex

Vestibulocollic Reflex (VCR)

- Vestibular system's connection to stabilization muscles of cervical spine
- Righting reflex
- Helps maintain upright head position
- Independent of trunk movement
- Mediated through otolithic organs and medial vestibulospinal tract

Vestibulocollic Reflex (VCR)

- Tested through cVEMP (will discuss more later)
  - Auditory stimulus saccule and creates neural impulse
  - Impulse sent through inferior vestibular nerve to vestibular nucleus
  - Vestibular nucleus sends signal to SCM through descending medial vestibulospinal tract

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Pathologies

Vestibular/Equilibrium Problems

- Many Symptoms:
  - Dizziness/Vertigo
  - Falls/Imbalance
  - Problems walking in dark or on uneven surfaces
  - Blurred vision with head movement
  - Dizziness or sense of motion with a change in position
  - Discomfort looking at moving objects
  - Veering when walking

Dizziness Facts at a Glance

- Vertigo/dizziness:
  - Will affect 76,000,000 Americans at some point
  - Yearly, 5 million people consult with their doctors for dizziness
  - Number 1 malady for those over 70
  - Average dizzy patient sees 4.5 physicians before getting a diagnosis, let alone Tx.

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BPPV

BPPV

- **Benign**: does not threaten life; responds well to treatment
- **Paroxysmal**: sudden, recurrent, and frequent attacks
- **Positional**: provoked by changes in position
- **Vertigo**: sensation of spinning or movement

BPPV

- Most common cause of true vertigo
- 50% of those 70+ will get at least once
- Although age related, can also be related to:
  - Head trauma
  - Illness (vestibular neuritis, migraine, etc.)
  - Change in medication
  - Surgery
  - Often idiopathic

BPPV Causes

- Head trauma:
  - Traumatic loosening of particles
- Vestibular neuritis:
  - Superior vestibular nerve innervates utricle, so damage = biomechanical changes
- Surgery:
  - Extended head position
  - Vibration, trauma, etc.
- Sudden SNHL:
  - 12.7% develop

Consequences of BPPV

EVALUATION FACTORS	GROUPS	
	BPPV	Without BPPV
Activities of Daily Living (ADL)	Greater Impairment (ADL Score = 2.89)	Less Impairment (ADL Score = 4.17)
Diagnosis of Depression	78%	36%
Falls in Prior 3 Months	78%	35%

Oghalai, J., Manolidis, S., Barth, J., Stewart, M., & Jenkins, H. (2000). Otolaryngology – Head and Neck Surgery, 122, 630-634.

BPPV

- Caused by otolithic debris in any of the 6 semicircular canals
- Most commonly located in PC:
  - PC: 80-96%
  - HC: 2-16%
  - AC: 1.2-12%
- In my clinical experience, AC-BPPV rarely (if ever) occurs in the wild

BPPV

- Typically unilateral: 85-96%
- Bilateral involvements are often the product of head trauma
- Most often affects the right side: 1.41 times more common
  - Sleeping side/position
  - Cardiovascular connections???

BPPV

- Hallmarks
  - Short-duration episodes (less than 1 min.)
  - Positional provocation
  - True vertigo
  - Fatigable
  - Slight delay in Sx onset
- Hallmarks easily explained by pathophysiology

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Otoliths in Posterior SCC

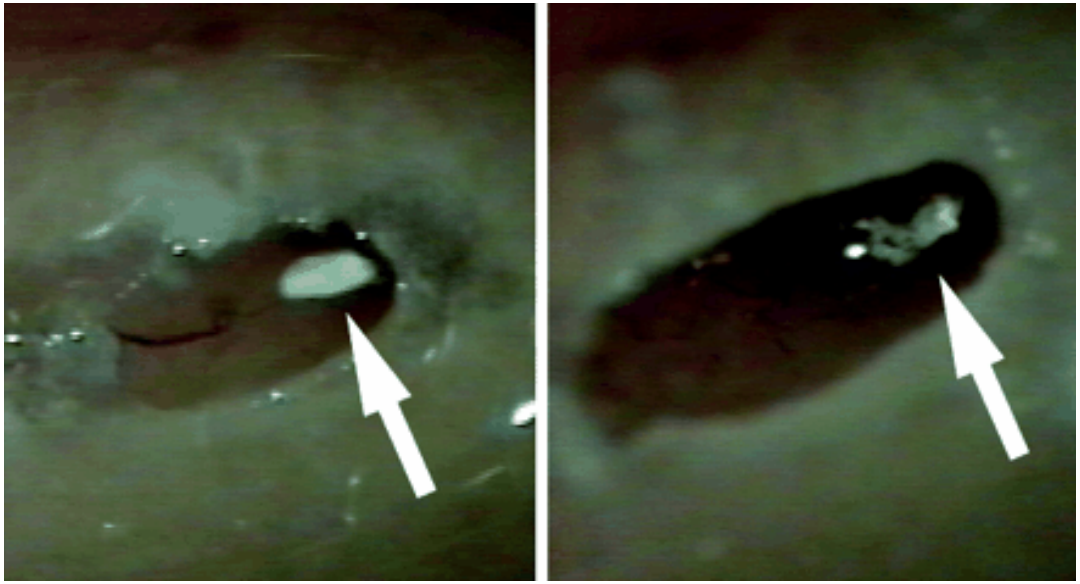
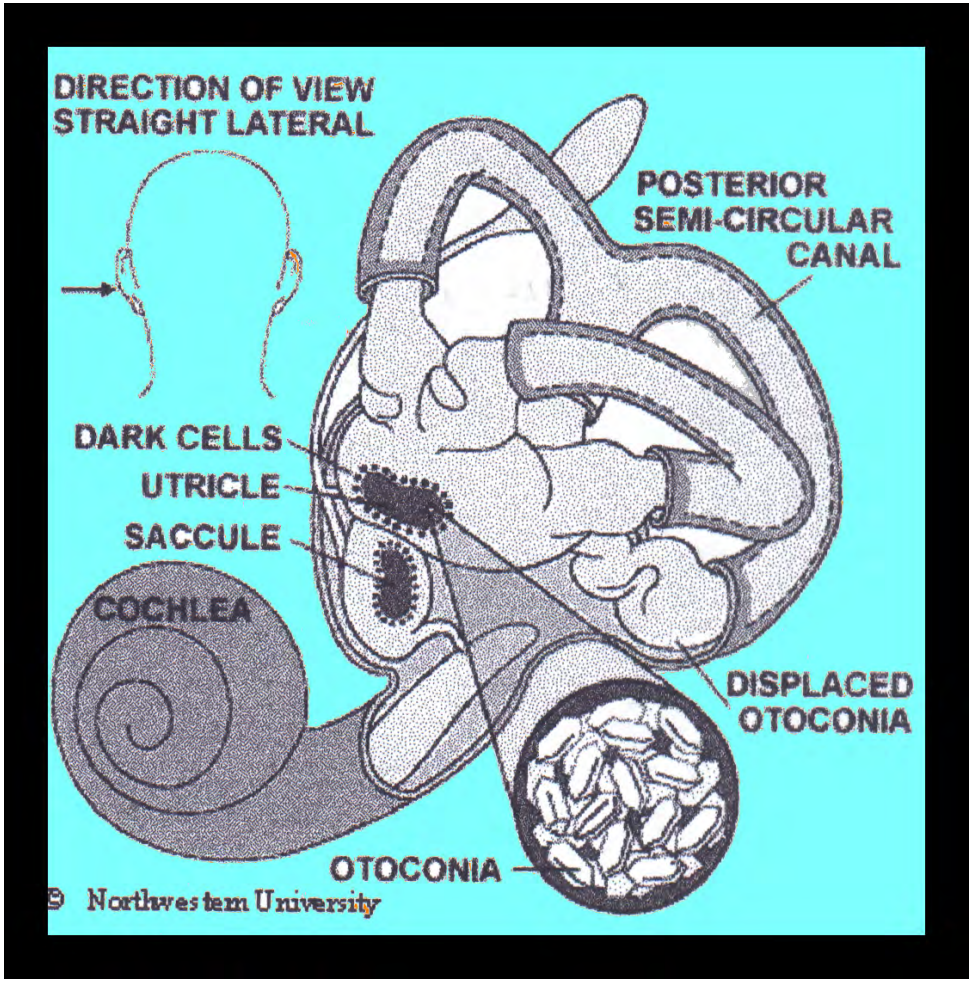


Photo courtesy of Lorne S. Parnes, M.D.



-BPPV is a mechanical phenomenon: hallmarks can easily be explained

-Tx involves moving otoconia from SCC to utricle

-BPPV easily treated by trained person; if not trained, can be issues

-Otolith jam

-Crisis of Tamarkin

Pathophysiology

- Canalithiasis
  - Otoconia free moving within SCC
  - More modern view of pathophysiology
  - Most closely explains traditional symptoms of BPPV

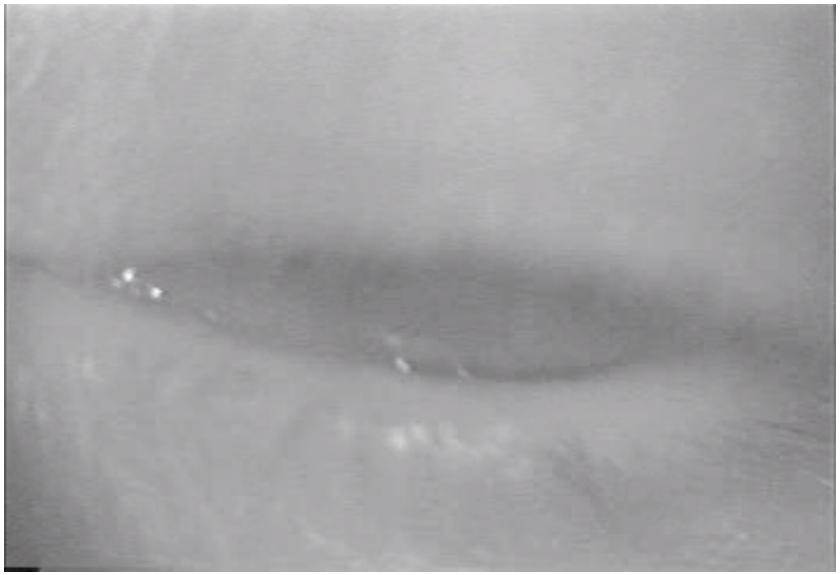
Cupulolithiasis vs. canalithiasis

- With PC and AC BPPV, many clinicians do not believe cupulolithaisis exists (including me)
- Cupulolithaisis does occur with HC BPPV
  - Longer duration attacks
  - No symptom onset delay

Localization

- Posterior canal
  - Rotary geotropic nystagmus
  - Affected ear down
- Anterior canal
  - Rotary ageotropic nystagmus
  - Affected ear up
- Horizontal canal
  - Horizontal geotropic/ageotropic nystagmus
  - Affected ear up or down

Rotary Nystagmus



- This must be BPPV of the left posterior SCC
- Based on left posterior SCC's connection to extra-ocular muscles
  - Excitatory- left inferior oblique, right superior rectus
  - Inhibitory- left superior oblique, right inferior rectus

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Left PC-BPPV

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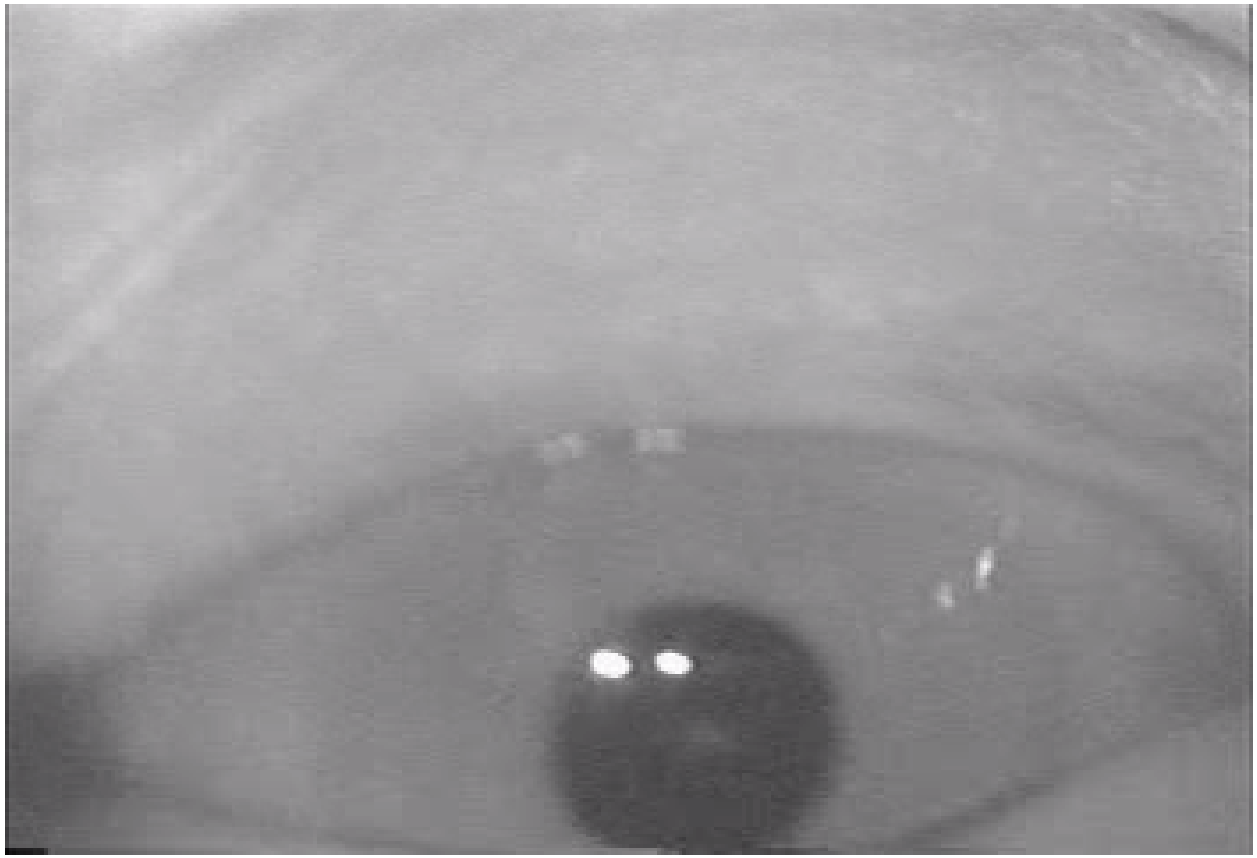
HC-BPPV



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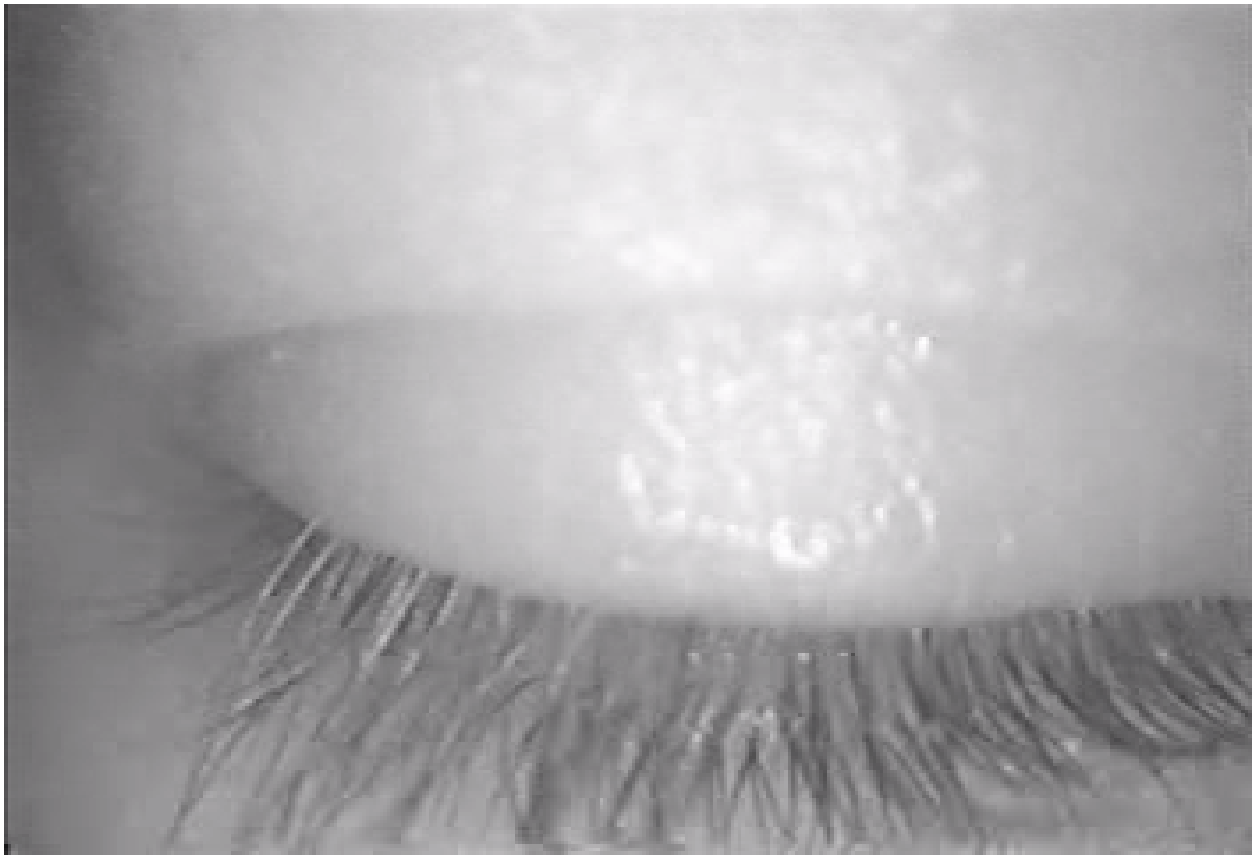
AC-BPPV

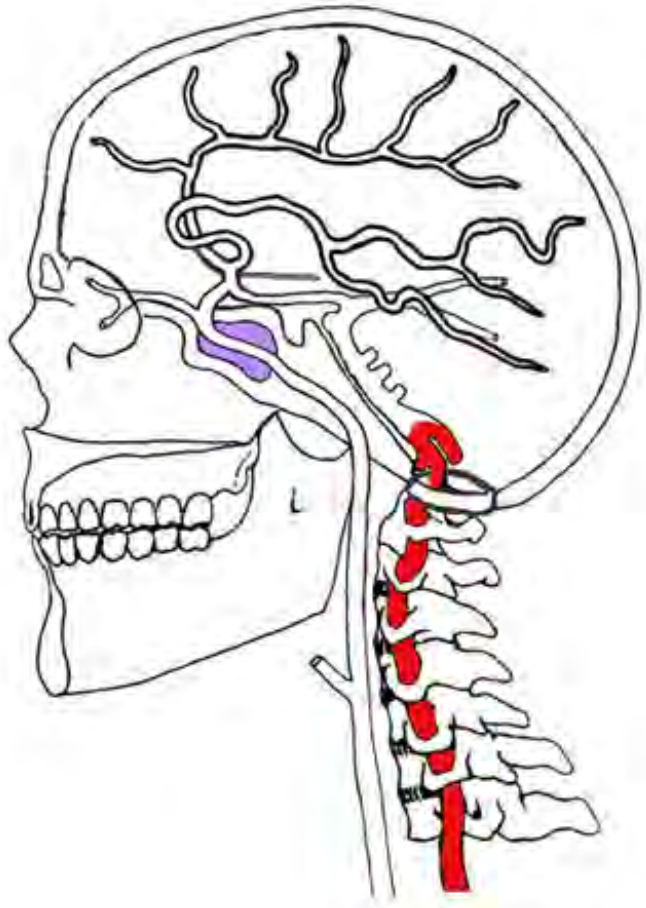


Localizing HC

- Geotropic nystagmus
  - Canalithiasis
  - Side w/ more intense nystagms = involved
- Ageotropic nystagmus
  - Cupulolithiasis
  - Side w/ weaker nystagmus = involved

Testing for BPPV

- Prior to testing, you must complete a vertebral artery screening test
- Vertebral artery dissection is a very real concern



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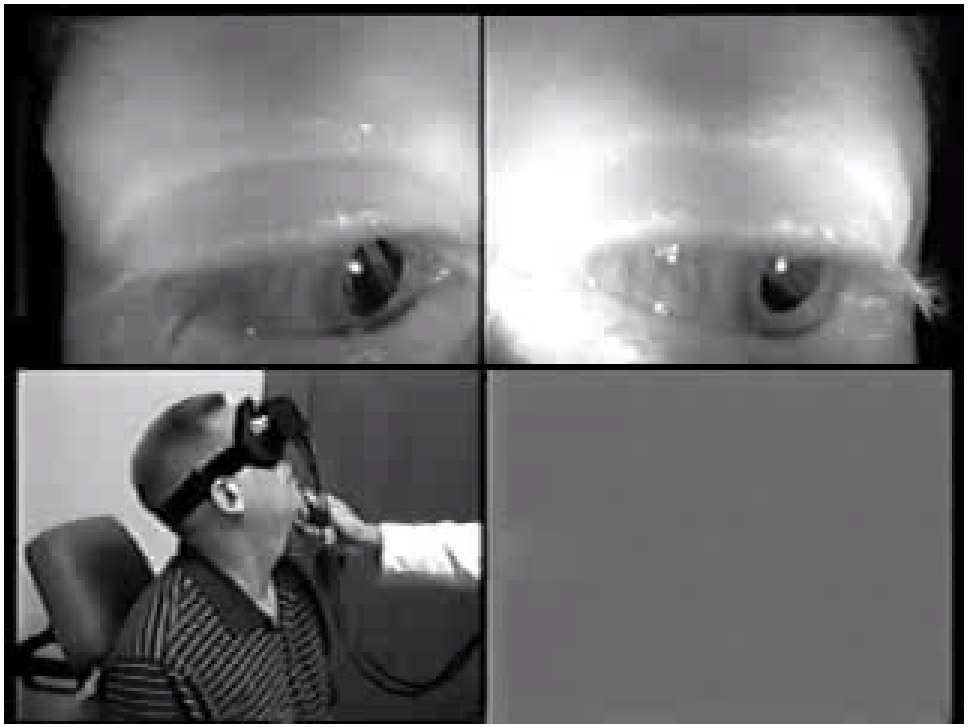
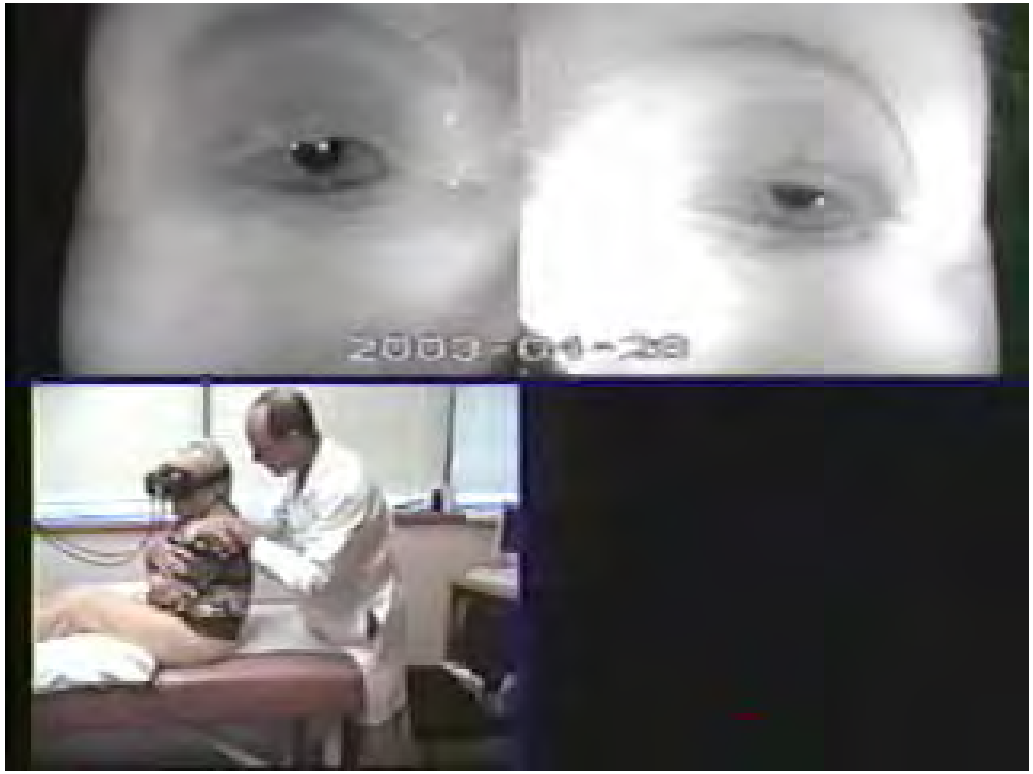
VAST



This screening should be done immediately after case history and before any test is performed

Testing for BPPV – PC & AC



Dix-Hallpike Maneuver

Testing for BPPV

- Horizontal BPPV
  - Side-lying test
- Positive VAST
  - Fully supported Hallpike
- Orthopedic issues
  - Side-lying Hallpike

BPPV Tx. Options

- Posterior Canal:
  - CRM
  - GRM
  - Semont-Liberatory
  - "True" Epley rarely used (not even by Dr. Epley-poor patient/clinician biomechanics)
- Horizontal Canal:
  - Appiani
  - Cassani
  - BBQ-Roll
- Anterior Canal:
  - Same as posterior

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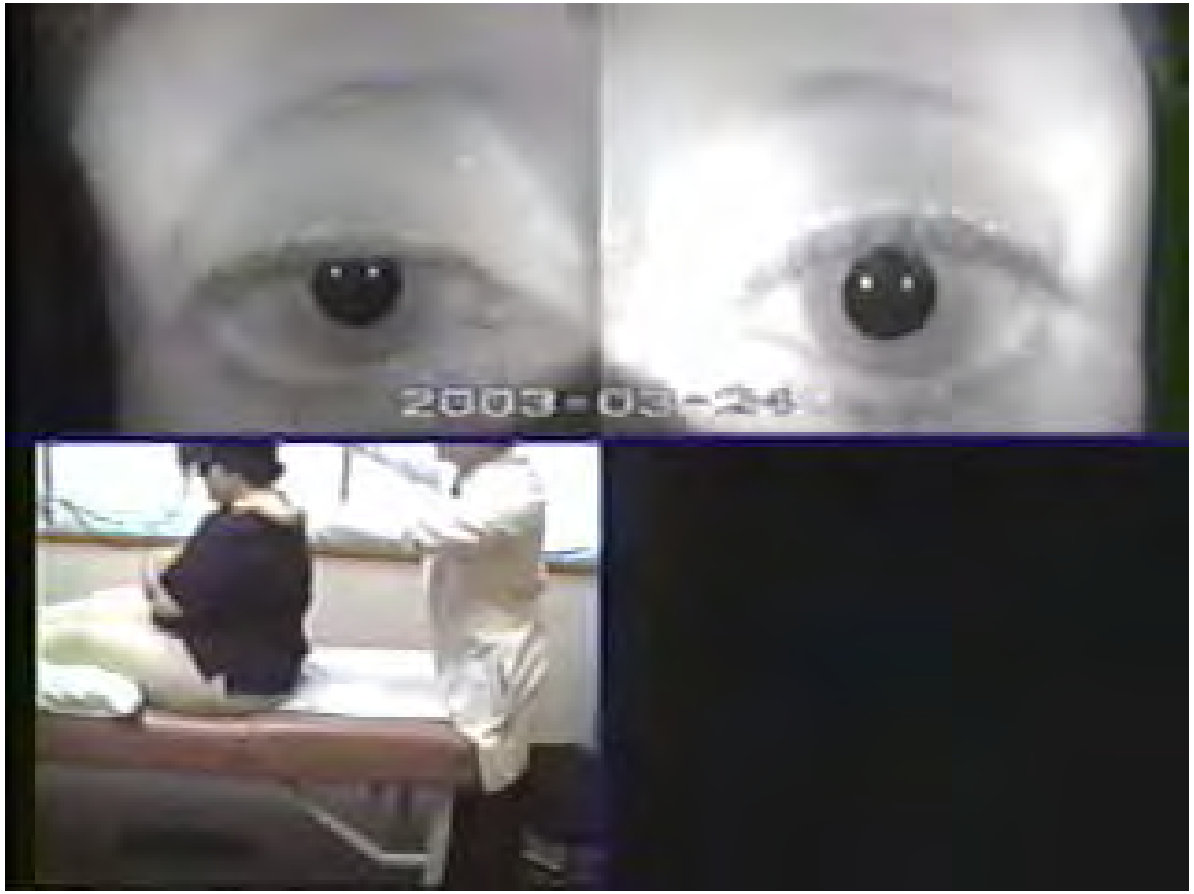
Canalith Repositioning (left ear)



Patient kept in each position for 1-3 minutes

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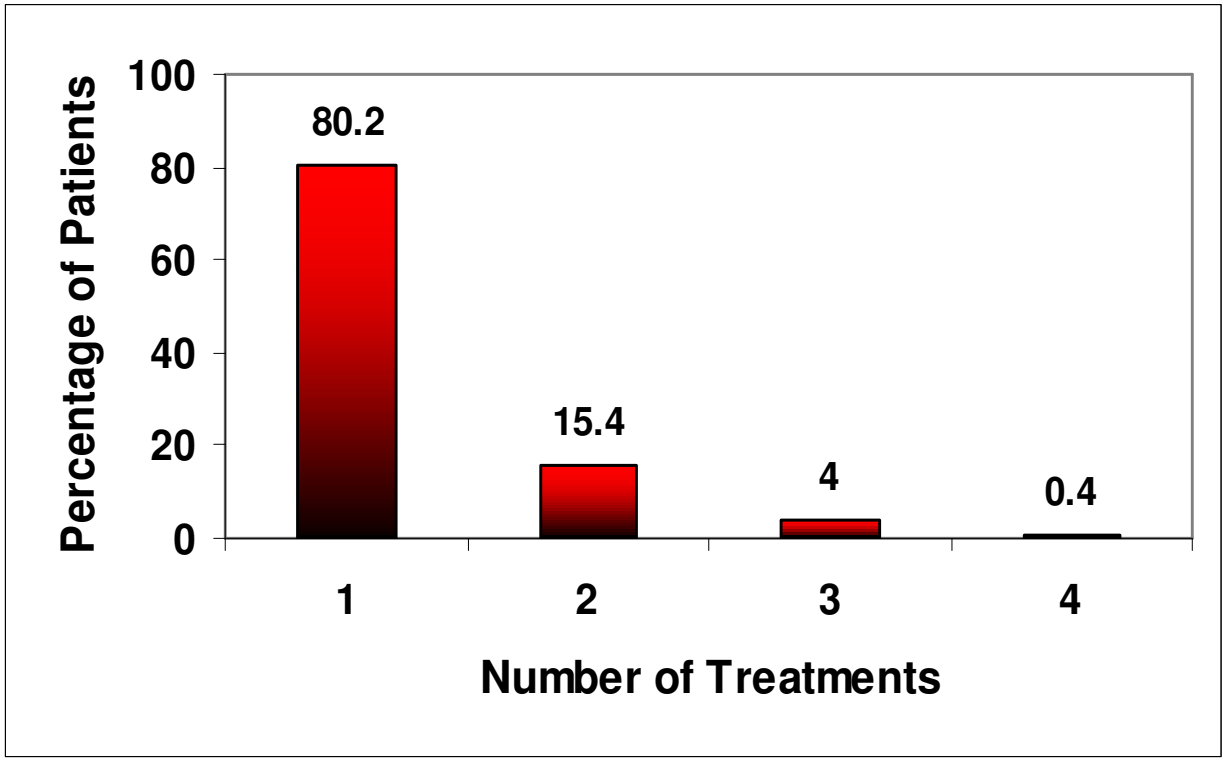
Gans Repositioning Maneuver

One of many treatment for PC-BPPV

BPPV Tx. Options

- Horizontal Canal:
  - Appiani
    - Away from affected ear
  - Cassani
    - Towards affected ear
  - BBQ-Roll
    - 90-degree steps away from affected ear

Results



- **95.6%** cleared of Positional Vertigo after two GRM treatments

Roberts, R., Gans, R., and Montaudo, R. (2006). "Efficacy of a new treatment for posterior canal benign paroxysmal positional vertigo", *Journal of the American Academy of Audiology, 17,* 598-604.

Is it always this easy?

- Horizontal canal migration
  - HC-BPPV is often the product of canalith repositioning for the posterior canal
- Crisis of Tamarkin
  - Caused by otoconial debris "bumping" the macula
- Otolith jamb
  - Otoconia become stuck at crus of PC and AC

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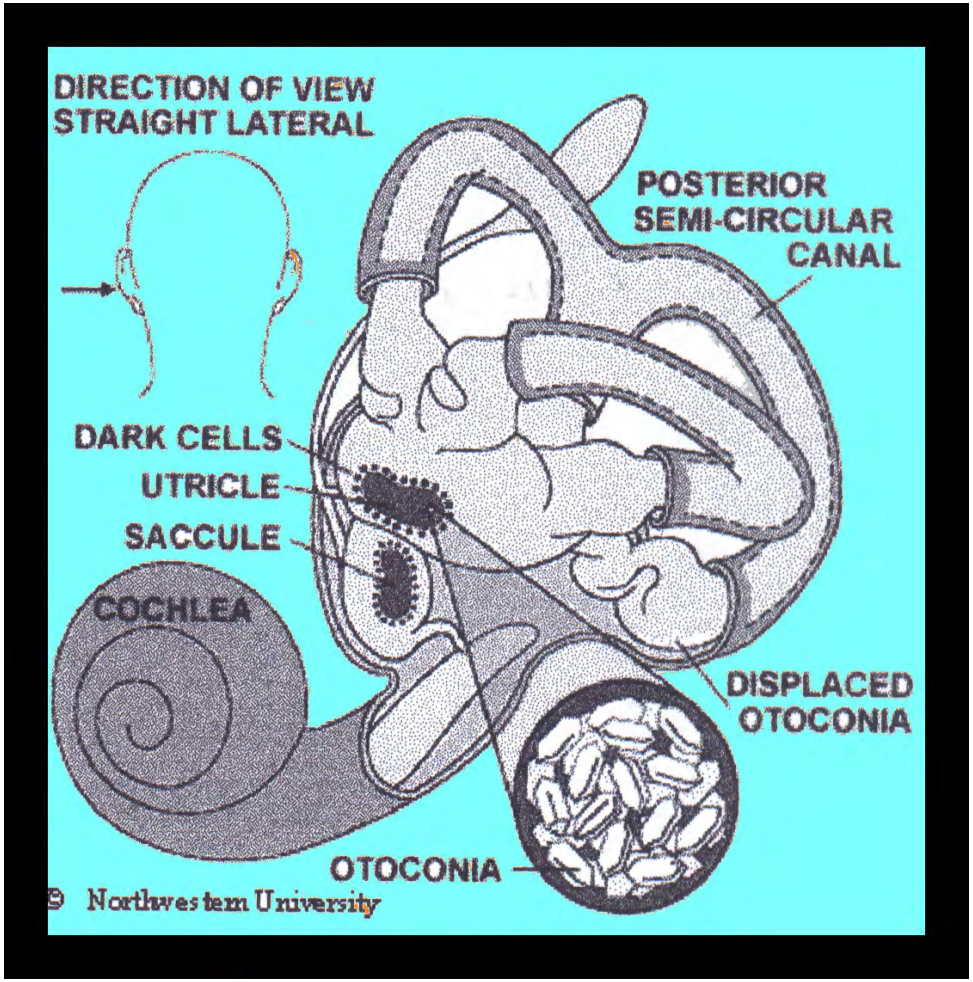
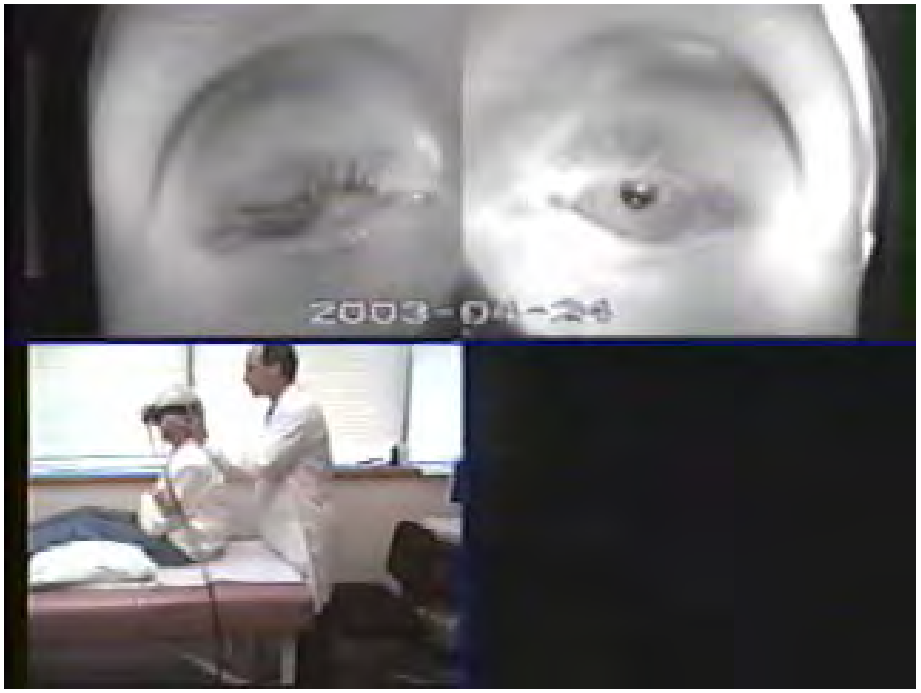
Is it always this easy?



Crisis of Tamarkin



→This is why not everyone should be treating BPPV

→This is also why we NEVER give patients home-based BPPV Tx

Additional Thoughts

- BPPV is 3x more prevalent in individuals with migraine
- After appropriate Tx, the recurrence rate of BPPV is ~10%
- BPPV is absolutely within our scope of practice, and should be conducted by audiologists
- BPPV can cause abnormal spontaneous & positional nystagmus, and disequilibrium

Additional Thoughts

- Path. that mimics symptoms/findings:
  - Migrainous positional vertigo
  - Inner ear 3rd window
  - Cerebellar mass lesion
  - Central vestibular involvement

- BPPV may self resolve over days to months (years in some rarer cases)

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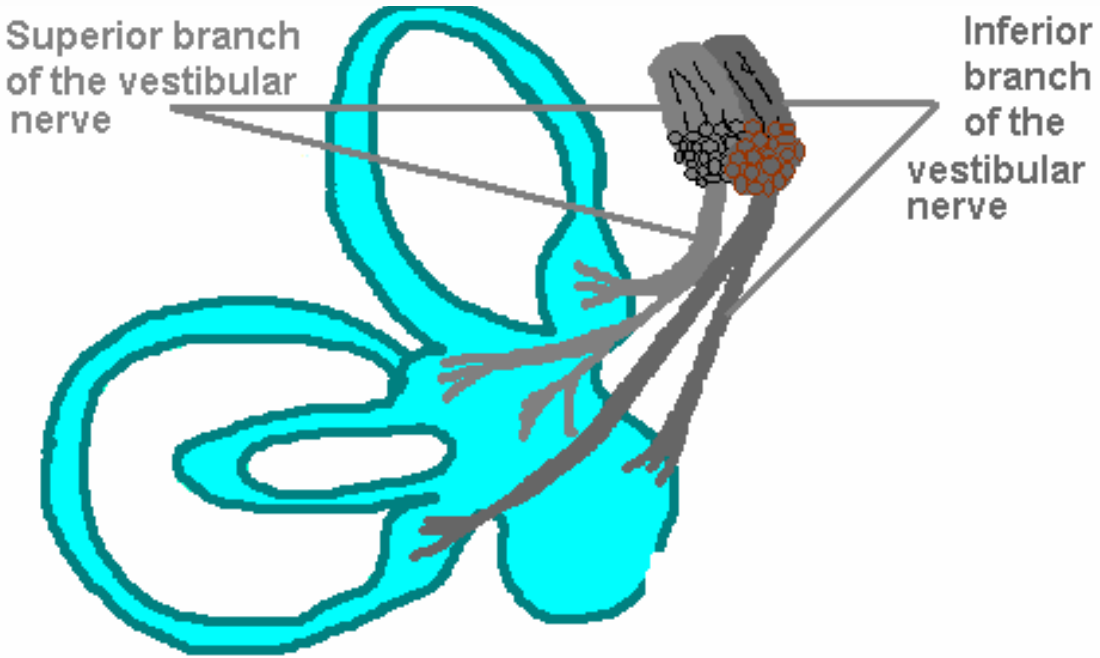
Vestibular Neuronitis

Vestibular Neuronitis

- 2nd most common cause of vertigo
- Often related to the herpes simplex virus (chicken pox)
- Hallmarks
  - Several day attack of true vertigo
  - Can be very traumatic
  - Often accompanied by nausea/emesis

Vestibular Neuronitis

- Typically only affects superior branch of the vestibular VIIIth nerve
- Can be diagnosed by tests that separate superior from inferior nerve function
  - VEMP vs. calorics, VAT, CD-VAT
- Can be diagnosed by specific spontaneous/positional nystagmus patterns and post-HFHS nystagmus



- With vestibular neuronitis, superior branch affected
- At rest, the central vestibular system achieves homeostasis by receiving equal input from each ear
- When one side is damaged, the input is no longer equal and homeostasis is lost
  - Patients experience vertigo (stimulation/inhibition = mismatch)
  - Brain sends signal to eyes via reflex arc (VOR)

Vestibular Neuronitis

- Nystagmus and acute vertigo occur because of this neural mismatch at level of brainstem
- Vertigo typically only occurs during acute stage- inflammation in bony channel
- Spontaneous and provokable nystagmus only occur when lesion remains uncompensated

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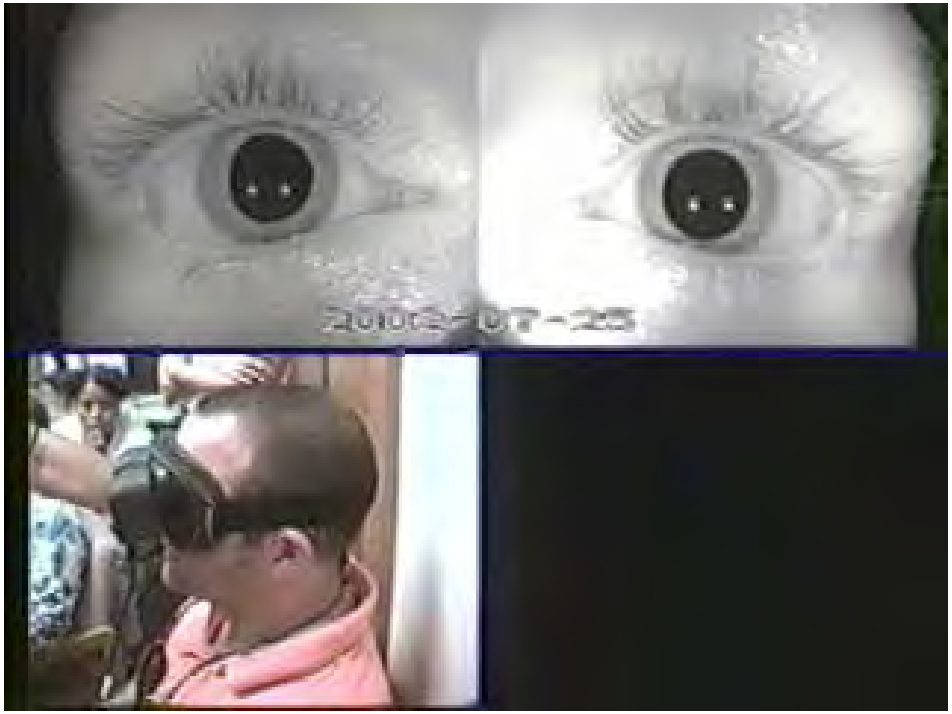
Vestibular Labyrinthitis

Vestibular Labyrinthitis

- Can look a lot like vestibular neuronitis
- Its defining characteristic is that it also affects hearing on the same side
- Almost always unilateral (as is v.n.)
  - If bilateral, patient will NOT experience an acute attack of vertigo
    - Remember, equal input = homeostasis
    - Vertigo/nystagmus caused by mismatch

Post HFHS Nystagmus



Uncompensated vestibulopathy

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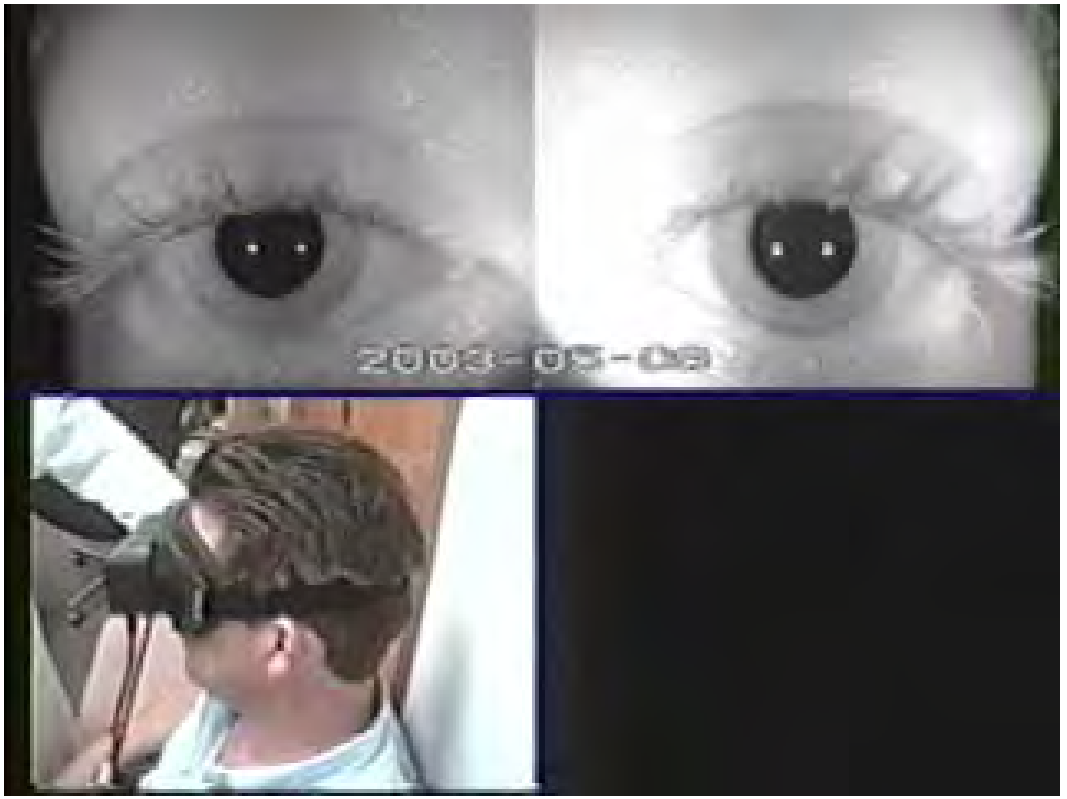
Spontaneous Nystagmus



Acute Vestibulopathy, CNS, or Congenital

Central Compensation

- Vestibular mismatch does not have to be only related to V.N.
- We can speed up and encourage compensation
- Vestibular rehabilitation therapy (VRT)
- "The brain can't fix what the brain can't see."
- We will address VRT later…

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Migraine

Migraine

- Migraine can occur in the absence of headache
  - Headache-free migraine (acephalgic)
  - More common in those w/ hx of aura
- New research in migraine and vestibular-like symptoms
- Aura can be vertigo/dizziness

Migraine

- Pathophysiology: 2 theories
  - Vasoconstriction: basilar migraine affecting internal auditory artery
  - Spreading wave of depression:
    - Can be separate from headache
    - Release of neuropeptides* causes excitation of base spontaneous firing rate
    - Asymmetrical release results in sensation of vertigo

*neuropeptide substance P, neurokinin A, calcitonin gene–related peptide

Migraine

- Vestibulopathy in 44% of migraineurs
- Meniere's disease is 2x more common
- BPPV is 3x more prevalent
- Can look like Meniere's disease
  - Fluctuating HL (permanent)- less common
  - Episodic vertigo
  - Episodic tinnitus
  - Often unilateral

Migraine

- Migraine: 11,350 / 100,000 people
- Meniere's: 210 / 100,000 people

- In other words, migraine is 54 times more common than Meniere's disease

- Further reading:
  - Cutrer FM, Baloh RW. Migraine-associated dizziness. Headache. Jun 1992;32(6):300-4.

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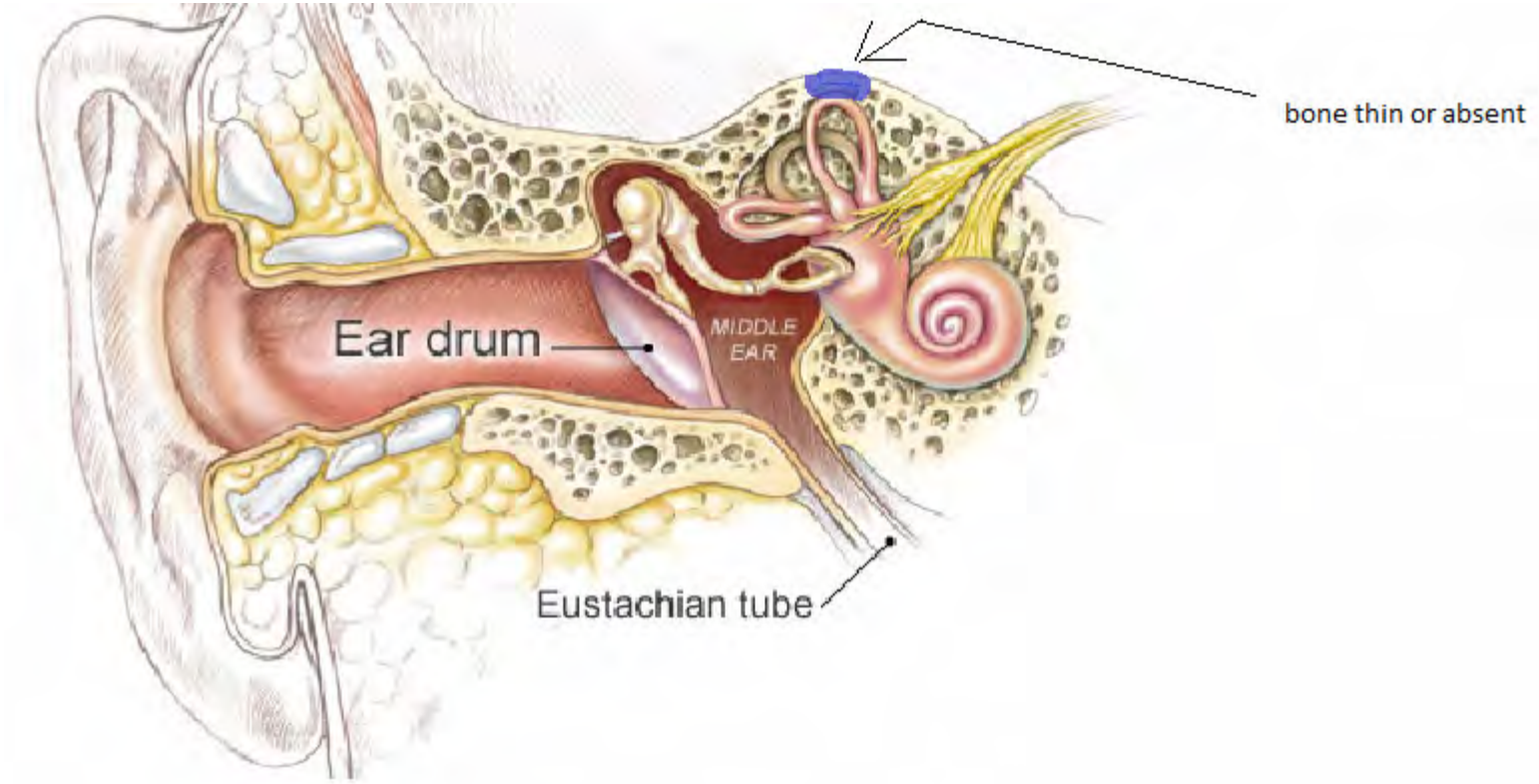
Superior Canal Dehiscence



SCDS

- Possible symptoms:
  - Tullio's phenomenon
  - Hennebert's sign
  - Autophony
  - Hyperacusis
  - Dizziness/disequilibrium
  - Aural distortion
  - Pulsatile tinnitus

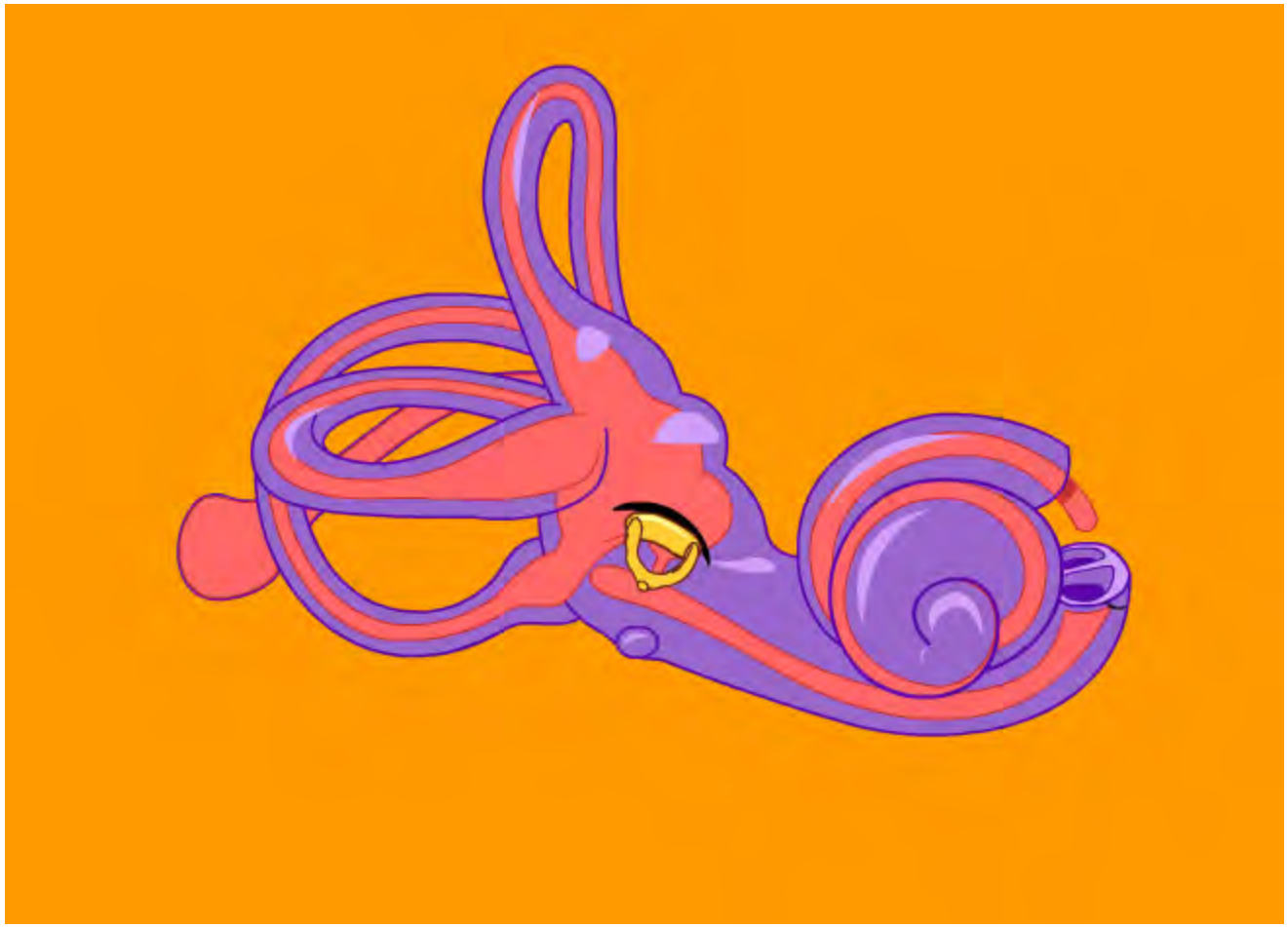
SCDS

- Clinical findings:
  - Multifactorial disequilibrium
  - Nystagmus with vocalization
  - Positive perilymphatic fistula test
  - Pseudo conductive hearing loss
    - BC at 250 Hz too good
    - Conductive HL w/ present reflexes
  - Abnormally low VEMP threshold
  - Air-bone gap w/ present VEMP
  - Abnormally large VEMP response

SCDS Pearls

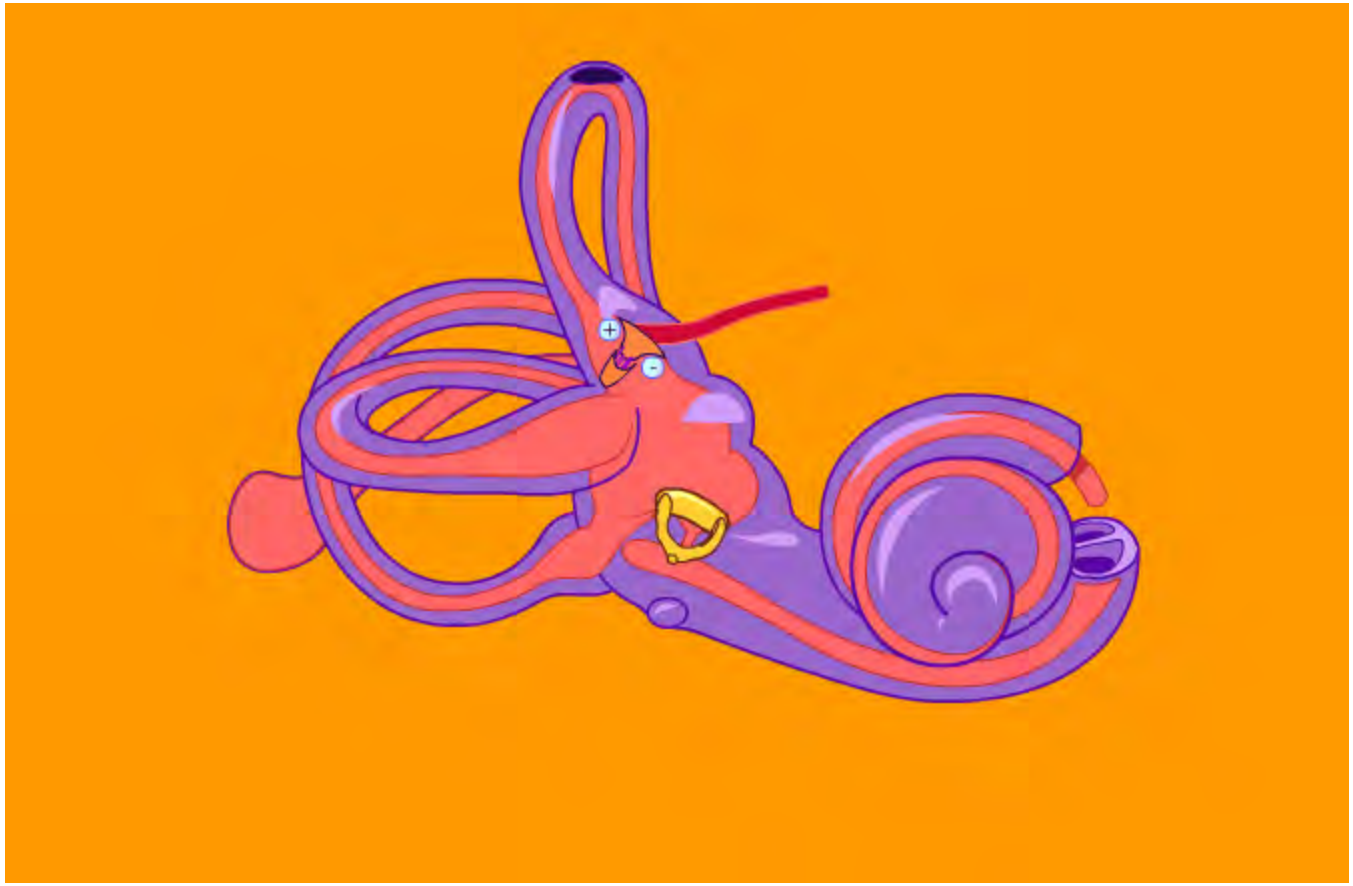
- Should always consider SCDS if:
  - Air-bone gaps with present stapedial reflexes
  - Air-bone gaps with present VEMP
- Can occur after head trauma

SCDS – Normal Cochlea



From: www.dbi.udel.edu/MichaelTeixidoMD/DownloadableTeachingMovies.html

SCDS – Tullio's Phenomenon



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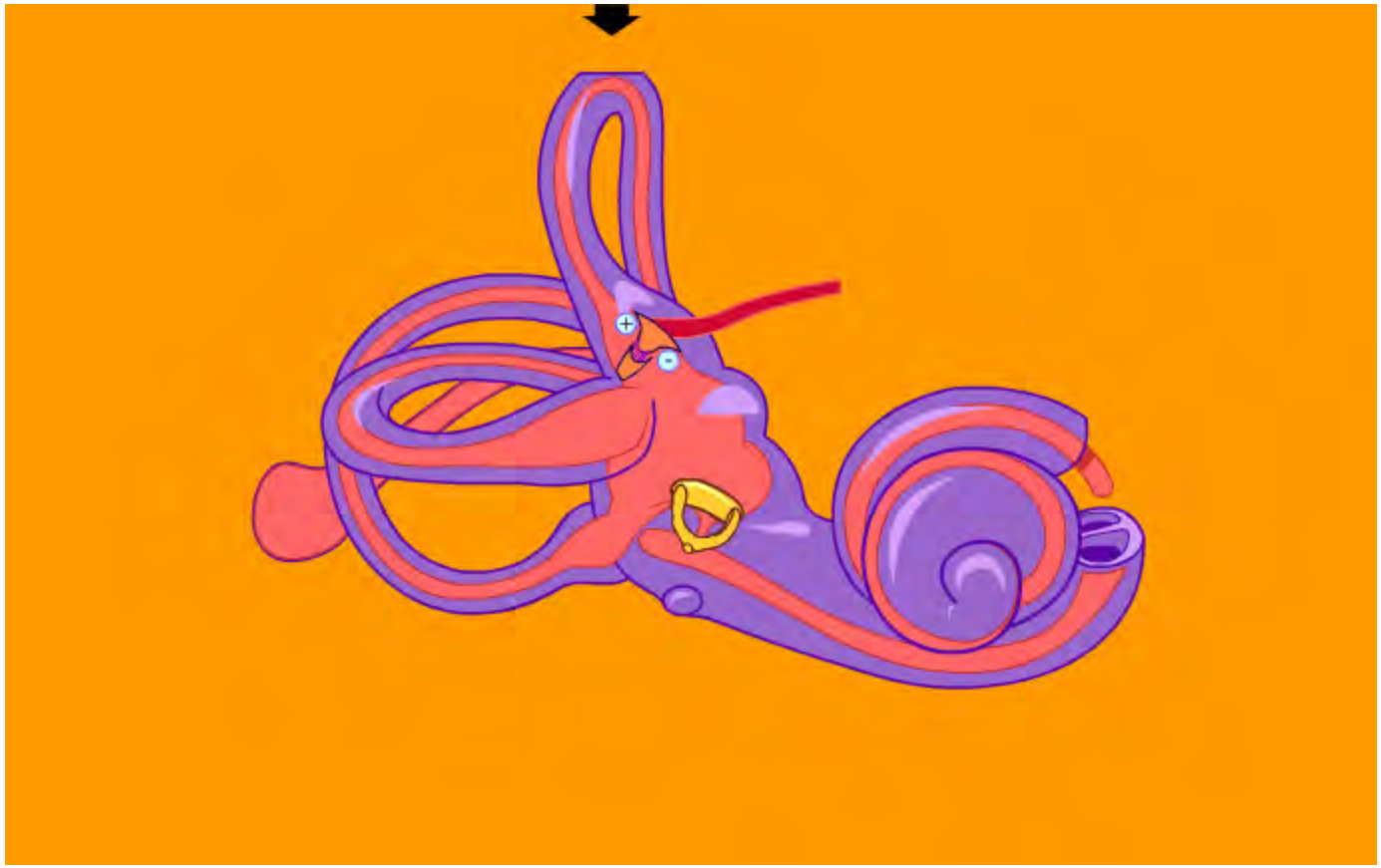
SCDS – Hennebert's Sign



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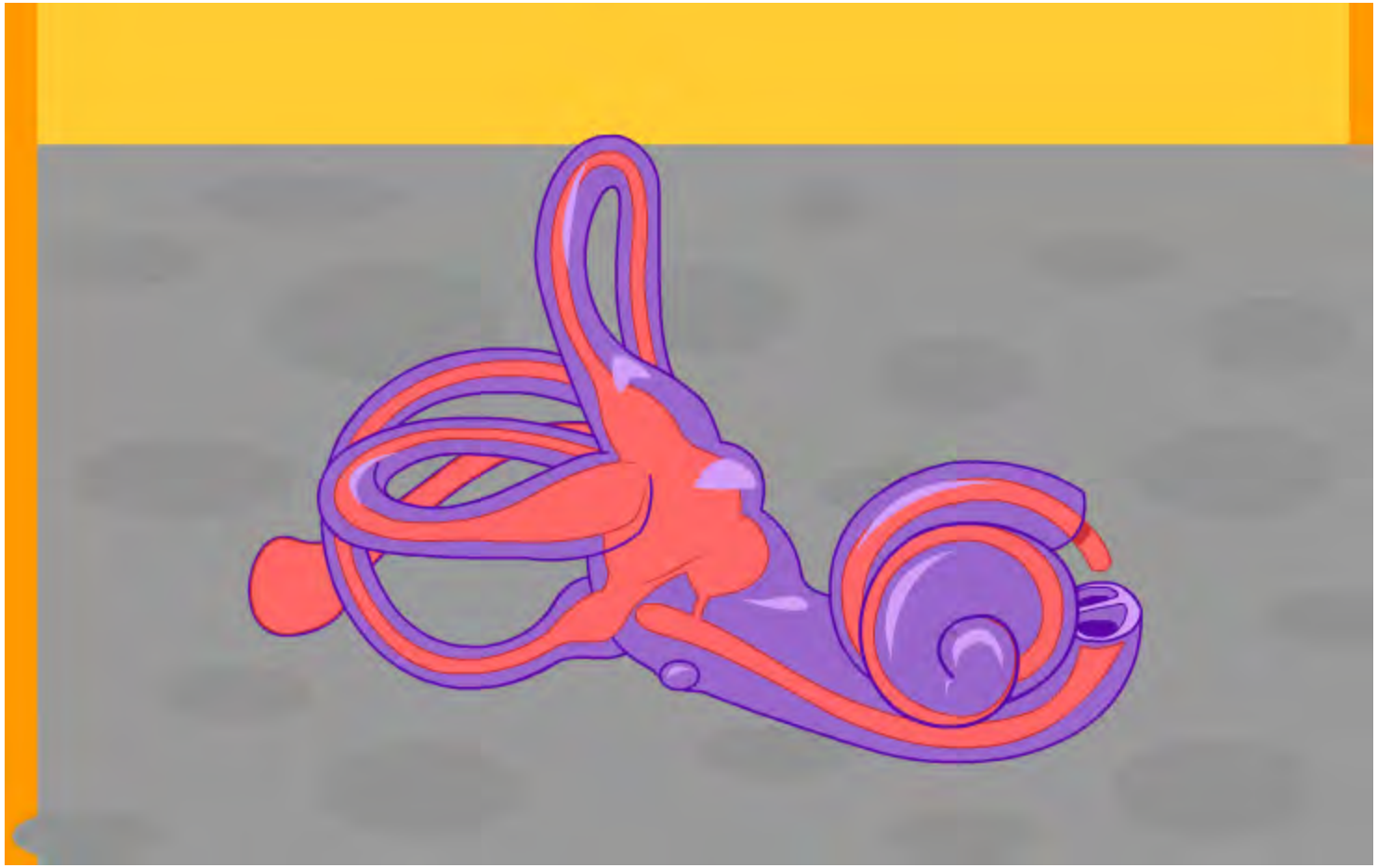
SCDS – Autophony



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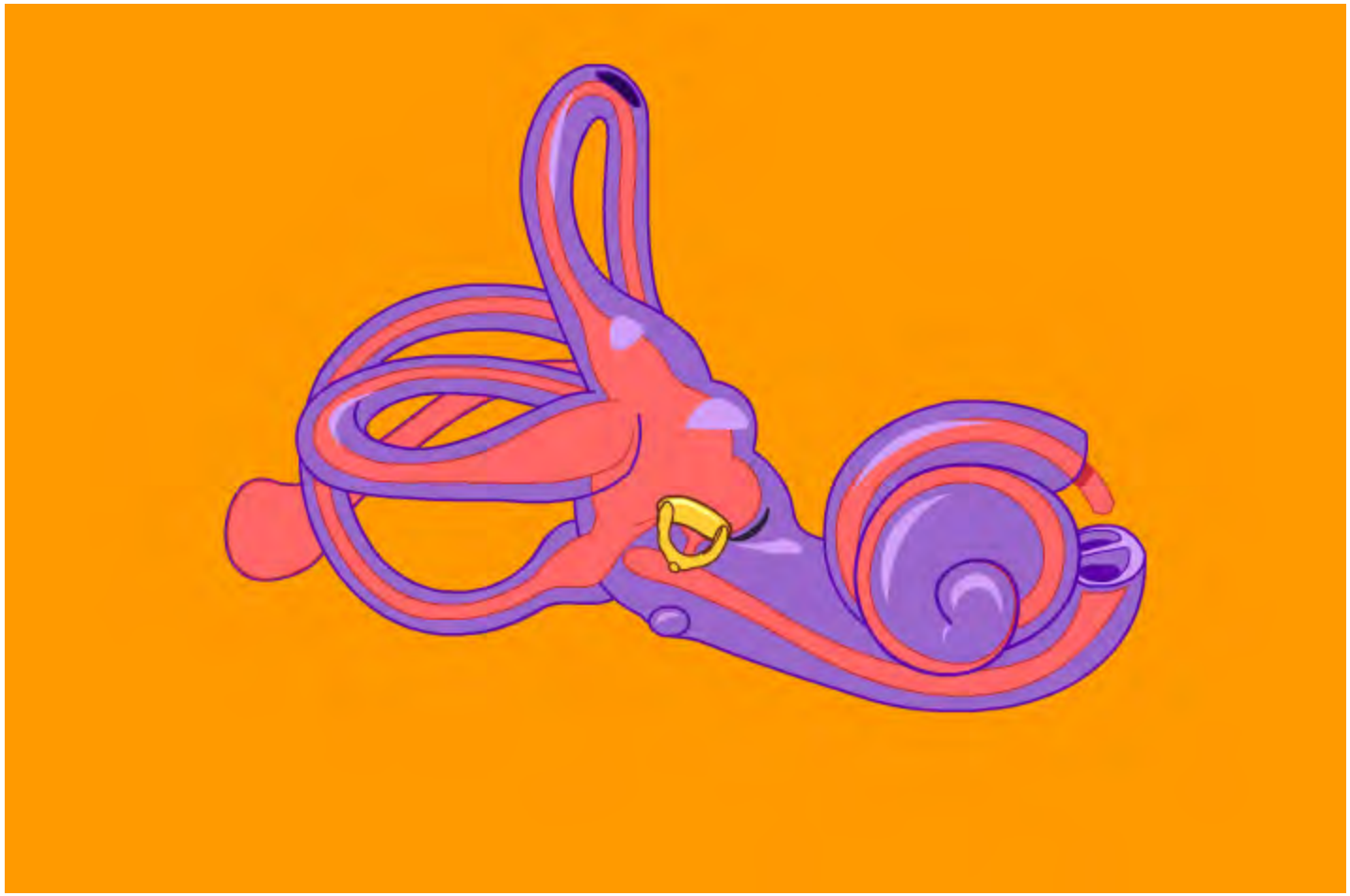
SCDS – Conductive HL



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SCDS Repair

- Typically surgical
- Symptoms have to warrant intervention
- Most of my patients live w/ it

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Perilymphatic Fistula

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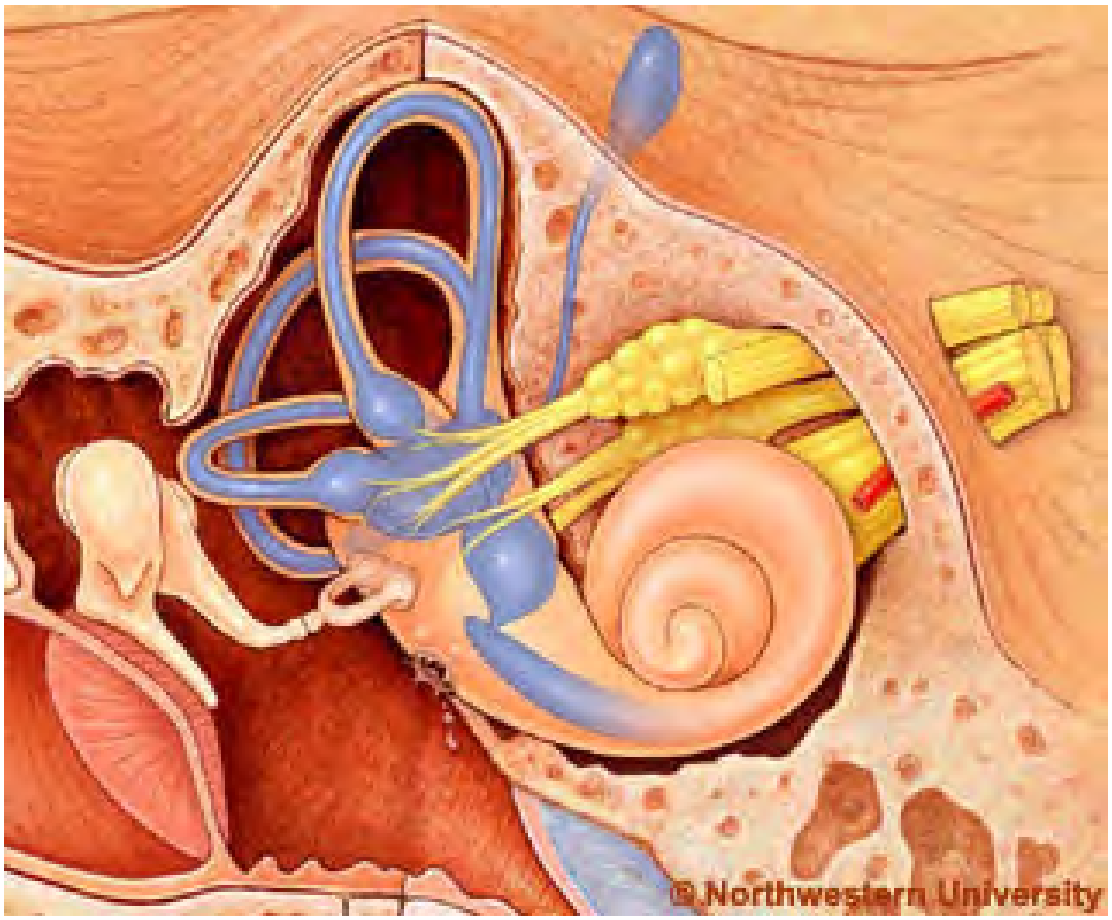

Perilymphatic Fistula

- Possible symptoms:
  - Tullio's phenomenon
  - Hennebert's sign
  - Dizziness/disequilibrium
  - Aural distortion
  - Hearing loss (SNHL)
  - Dizziness when straining or with physical exertion

Perilymphatic Fistula

- Causes:
  - Traumatic
    - Implosive: airplane descent, barotrauma
    - Explosive: coughing, straining, increased intracranial pressure
  - Following middle ear surgery
    - Particularly stapedectomy
  - Congenital, idiopathic, secondary

Perilymphatic Fistula

- Diagnosis:
  - Perilymphatic fistula test
    - Immittance bridge and goggles
    - Pneumatic otoscope
  - Surgical exploration
    - When conservative measures fail
    - Search for leaks

Perilymphatic Fistula

- Treatment:
  - Traumatic acute cases
    - 2-3 days of bed rest
    - Spontaneously heal
  - Chronic
    - Surgical repair
    - Patching with fascia

Perilymphatic Fistula vs. SCDS

- Similar symptoms
- Differentiating not difficult
  - SCDS = CHL; Fistula = SNHL
  - SCDS = nystagmus w/ vocalization
  - SCDS = abnormal VEMP

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Meniere's Disease

Meniere's Disease

- This is something you all know about
- However, this condition is often considered a "waist-basket" diagnosis
- It is often over diagnosed and misdiagnosed
- Let's review incidence, symptoms, diagnosis, etc.

Meniere's Disease Diagnosis

- Diagnosis is based on symptoms
- AAO released position statement due to over Dx
- MUST have triad of symptoms
  - Episodic vertigo lasting 30 min to 1 day
  - Tinnitus in effected ear
  - Fluctuating hearing in effected ear
- May also have aural fullness in effected ear

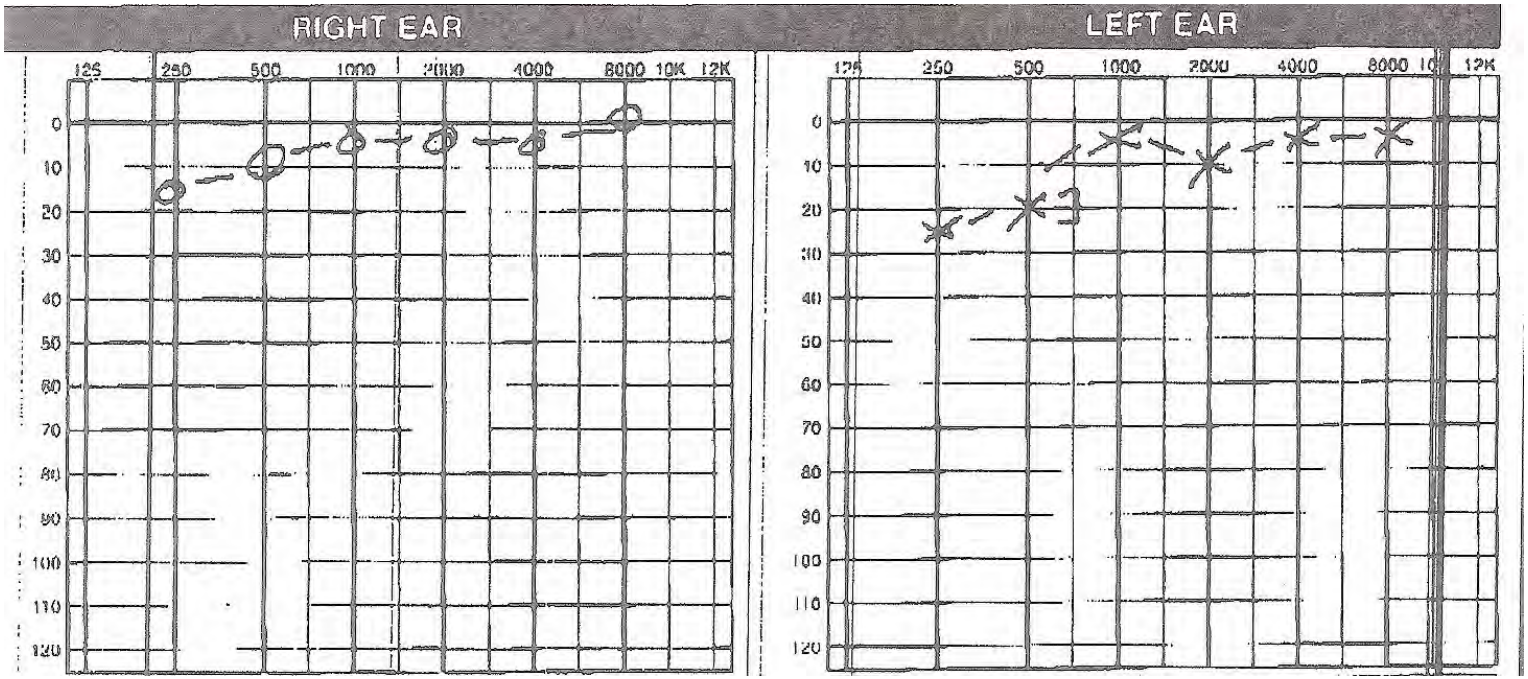
Meniere's Disease Diagnosis

- Although clinical testing may aid in Dx, its clinical definition requires a diagnosis to be made on Sxs and not testing
- Hearing loss is sensorineural (bone scores should be considered) and typically low frequency
- If you have any asymmetry at 250 Hz and you have not completed masked bone, you will misdiagnose your patients!!!

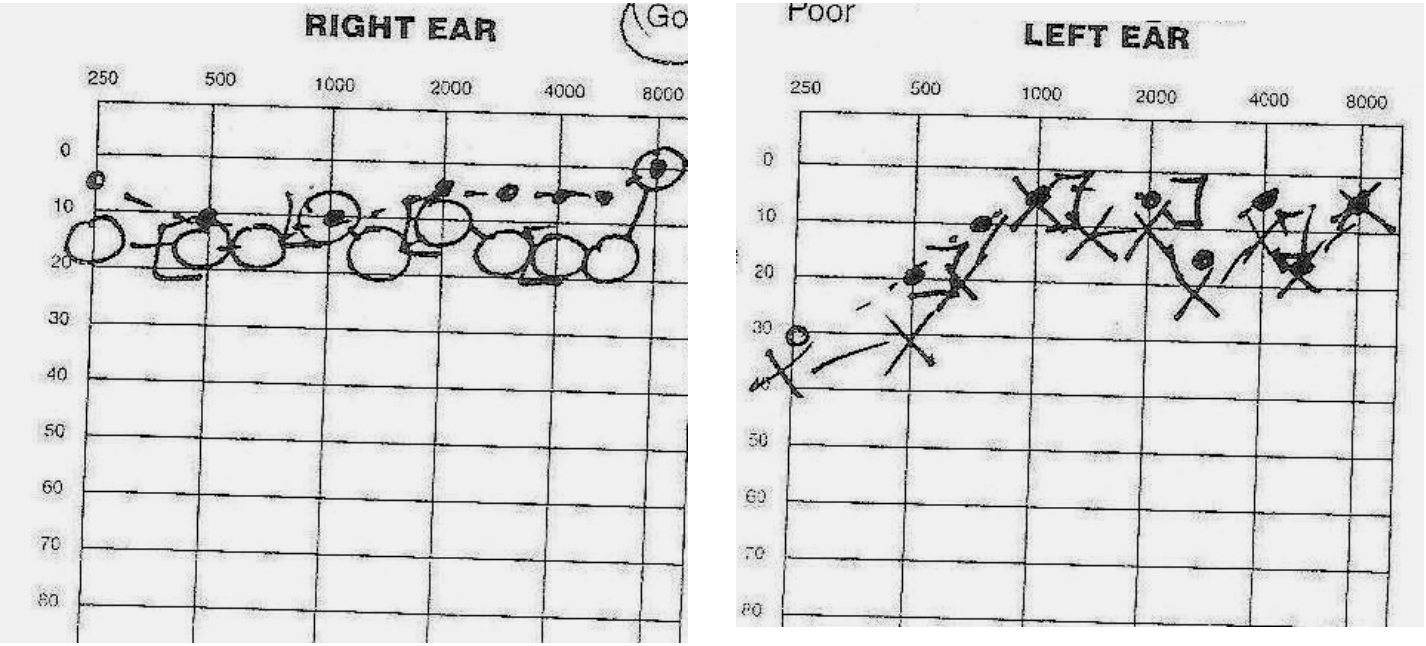
Meniere's Disease Diagnosis

- ...and now an example of a mis-diagnosis of Meniere's disease based on poor audiology
- This patient was evaluated in TWO different neurotology clinics
- Diagnosis was delayed for years because audiologists doing the testing do not complete their due diligence and did not understand complaints and vestibular disorders

Audiogram 6/06



Audiogram 9/07



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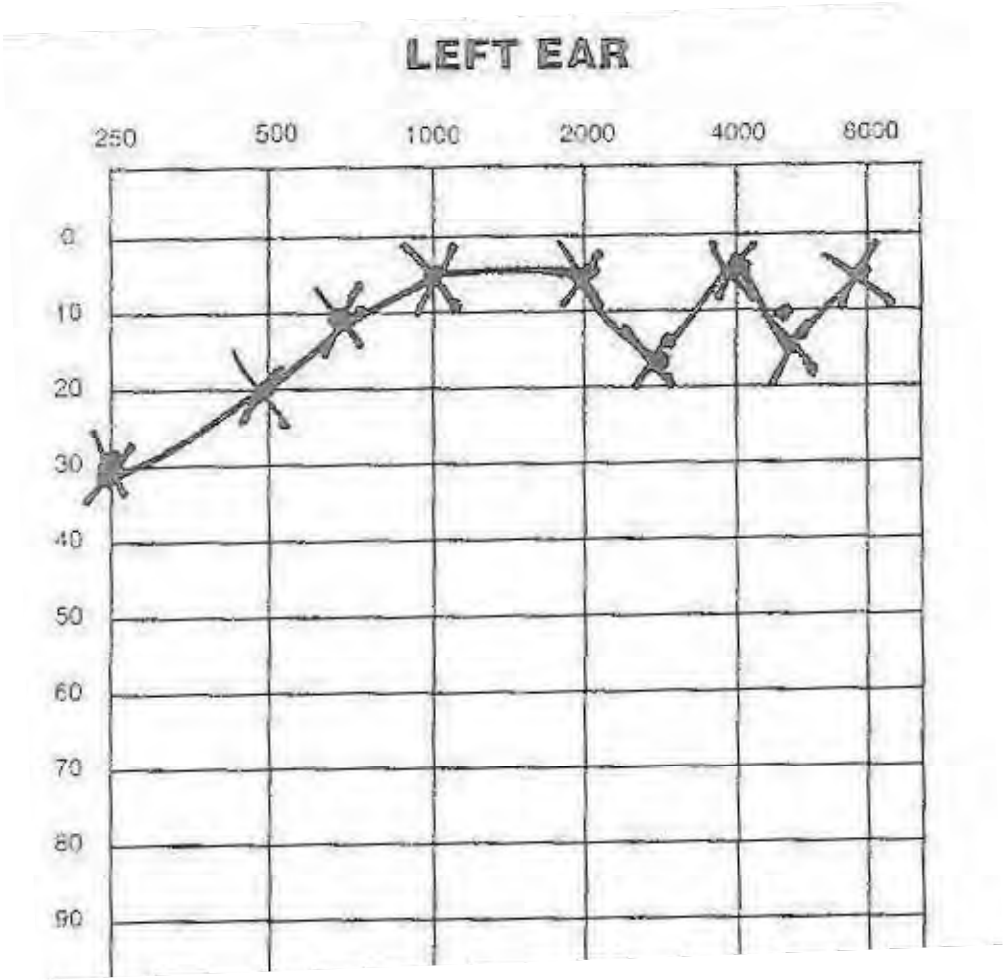
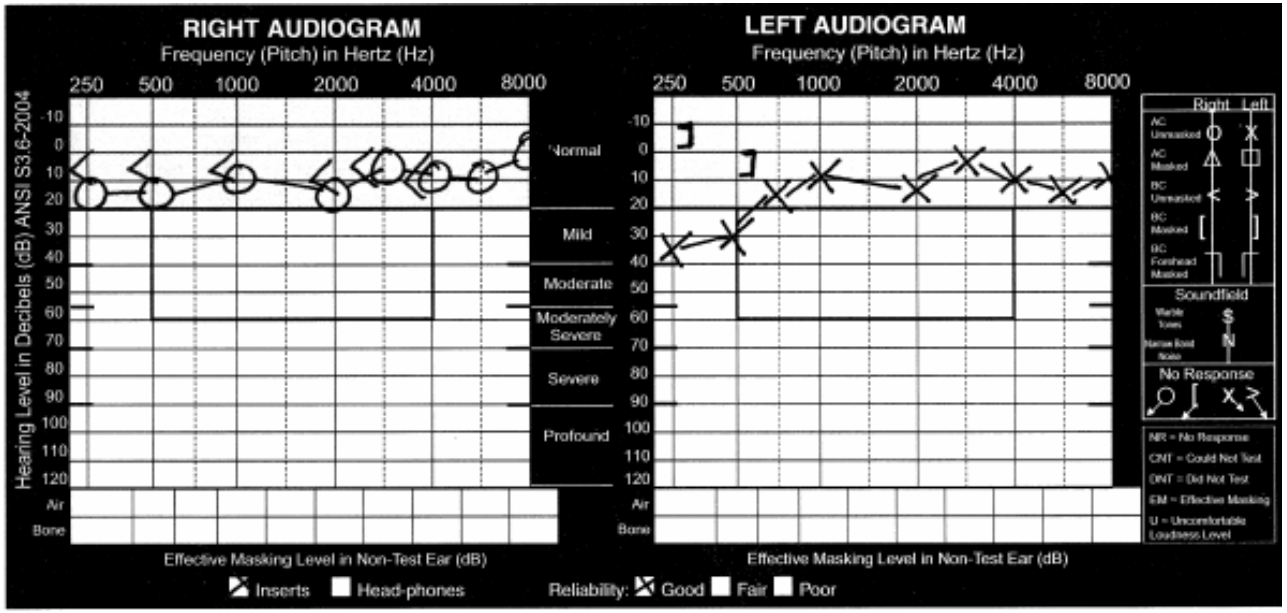
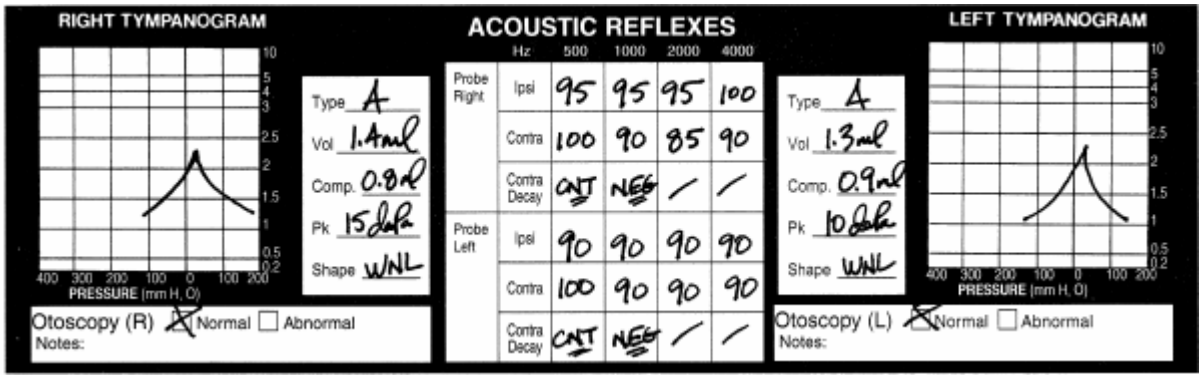
Audiogram 9/09



Current Evaluation





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Vestibular Schwannoma

Vestibular Schwannoma

- AKA acoustic neuroma
- Typically arises from inferior vestibular nerve
- Benign and slow growing
- Patients typically will not experience vertigo: slow growth allows central compensation

Vestibular Schwannoma

- Symptoms:
  - Unilateral hearing loss
  - Unilateral tinnitus
  - Poor word recognition
  - Aural distortion
  - Aural fullness
  - Disequilibrium
  - Dizziness

Vestibular Schwannoma

- Findings
  - Unilateral SNHL
  - Sudden HL
  - Fluctuating HL
  - Characteristic reflex pattern
  - Poorer word recognition than expected
  - Abnormal ABR = what???
  - Unilateral vestibular test findings (peripheral)
  - Gold standard = MRI of IACs with/without

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Patient D.R.

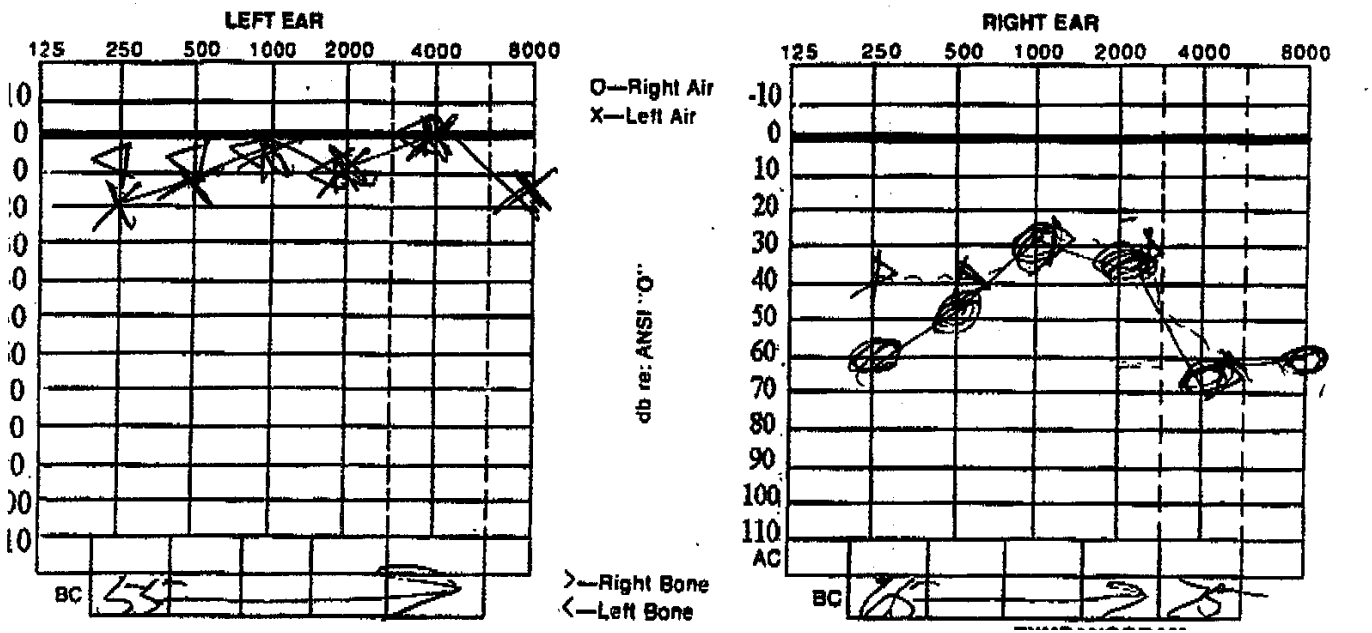
Patient D.R.

- 49 y/o female
- 6-month hx of significant reduction in right hearing, right tinnitus, episodic vertigo
- Patient believes Sxs began after URI
- Episodes last 10 min – several hours
- Cannot provoke
- No fluctuations in tinnitus and/or hearing during episodes

Patient D.R.

- Evaluated by 2 ENTs
- Diagnosed with Meniere's disease
- No formal vestibular testing or imaging since the onset of these Sxs
  - MRI ~1 year ago for sudden vision loss
  - MRI unremarkable at that time
- Two audiograms
- Placed on low sodium and caffeine diet, prescribed Valium

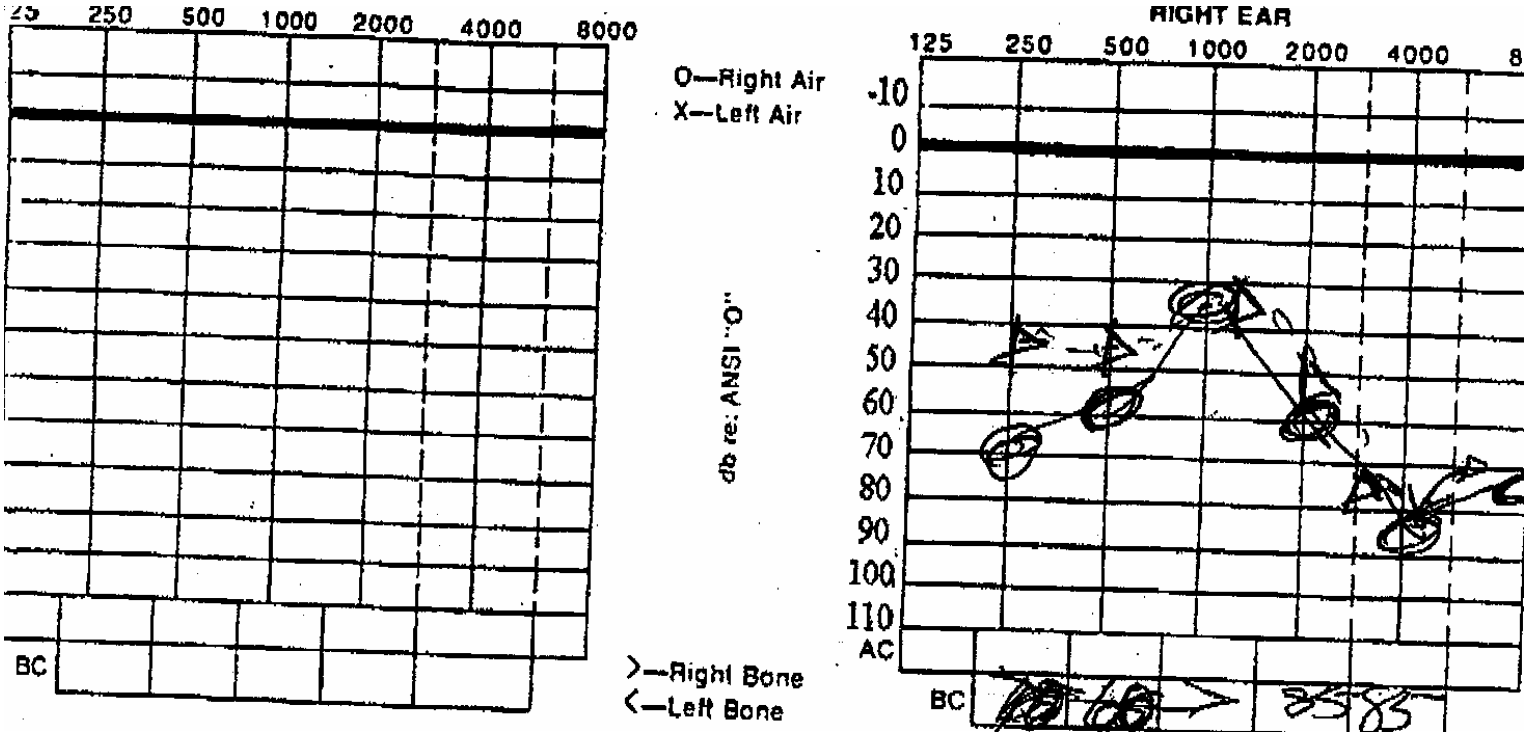
Patient D.R. Outside Audio #1



-Audiogram performed 03/04/2011

-Transducer??? → masking

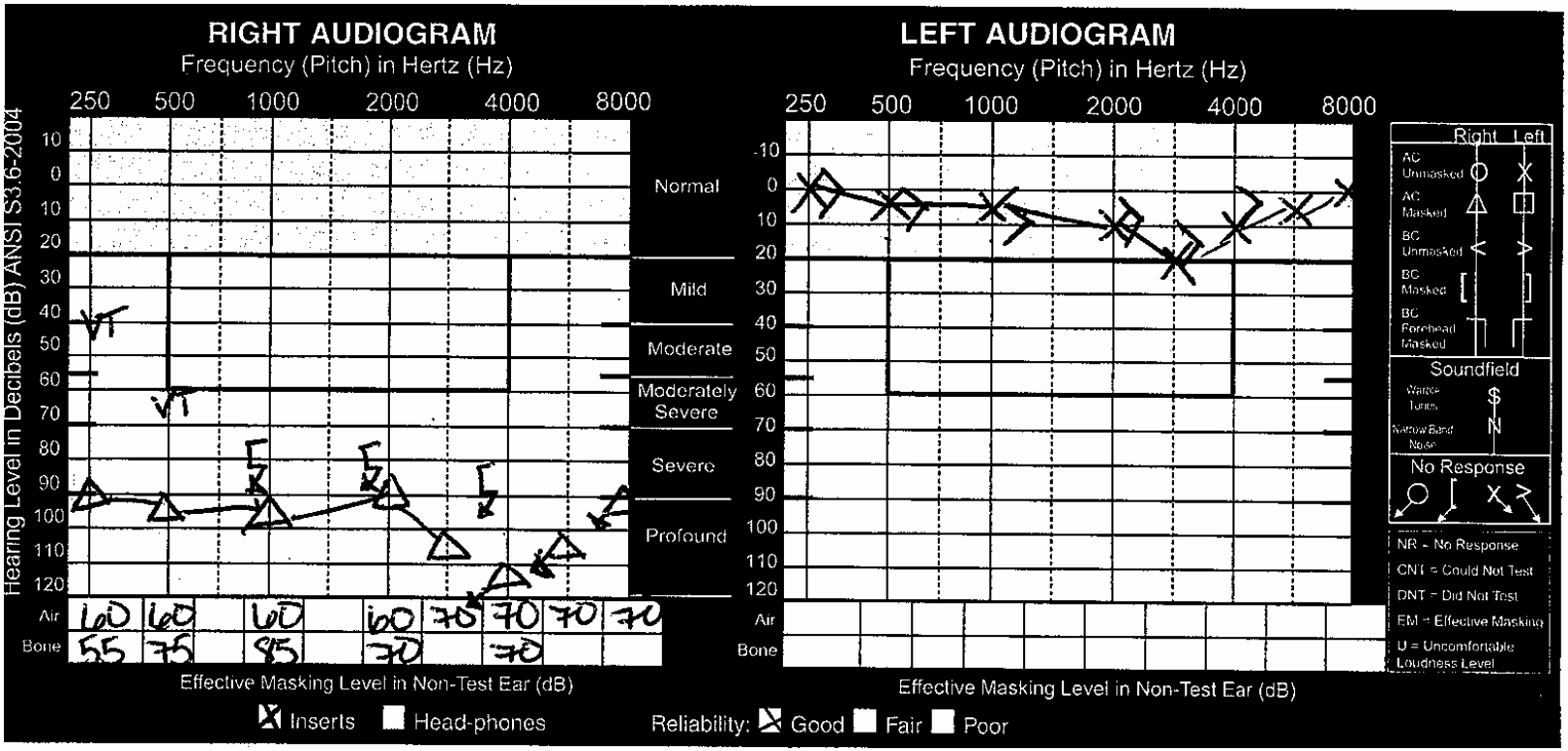
Patient D.R. Outside Audio #2



-Audiogram performed 05/02/2011

-Transducer??? → masking

Patient D.R. Our Audio



-Audiogram performed 06/23/2011

Patient D.R.

- Dx Immittance
  - No reflexes with stimulation right
- SOP
  - Vestibular pattern (fall on #6)
- VAST – negative
- VEMP
  - reduced amplitude right
- VNG
  - Left beating gaze and positional nystagmus- suppressed with vision and enhanced dynamically
  - No caloric response right, left WNL

Patient D.R.

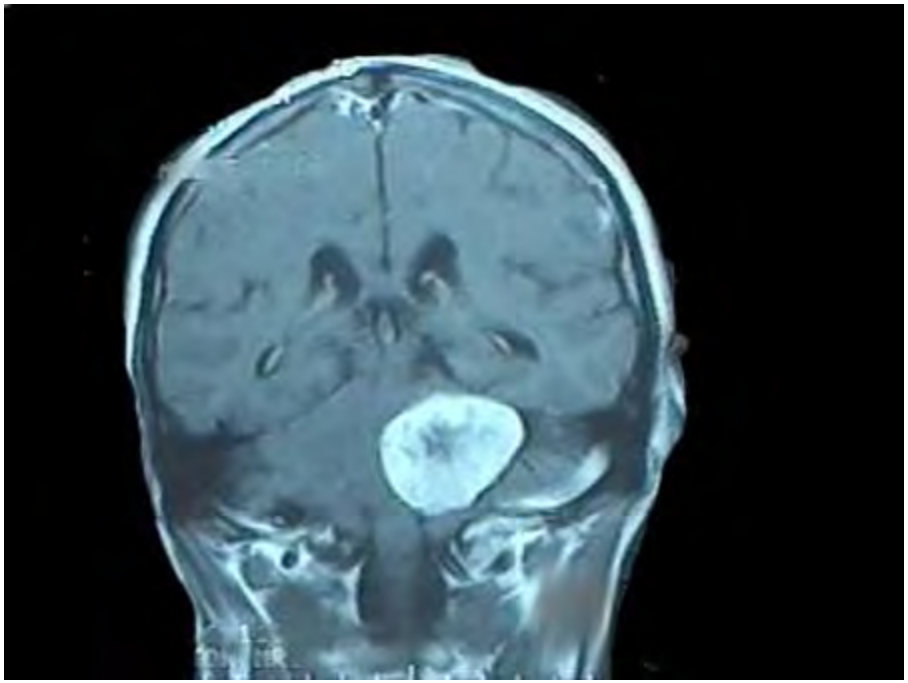
- Here is what we have:
  - Asymmetrical HL with right poorer
  - HL in excess of Meniere's disease
  - No reflexes with stim right
  - Reduced right VEMP
  - No caloric response on the right
  - Vestibular pattern on SOP
  - Negative MRI prior to symptom onset
  - Dx of Meniere's disease by ENTx2
- So, what are your recommendations?

Patient D.R.

- My preliminary diagnosis:
  - Either a reduction in function of several right-sided end organs or multiple branches of cranial nerve VIII
  - Retrocochlear lesion appears likely
- Patient referred for MRI of IACs with and without contrast
- Patient referred to neurotology

Patient D.R.

- Imaging revealed a right vestibular schwannoma
- Patient elected to defer surgery, so being monitored by neurotology…



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Disequilibrium

Equilibrium Facts at a Glance

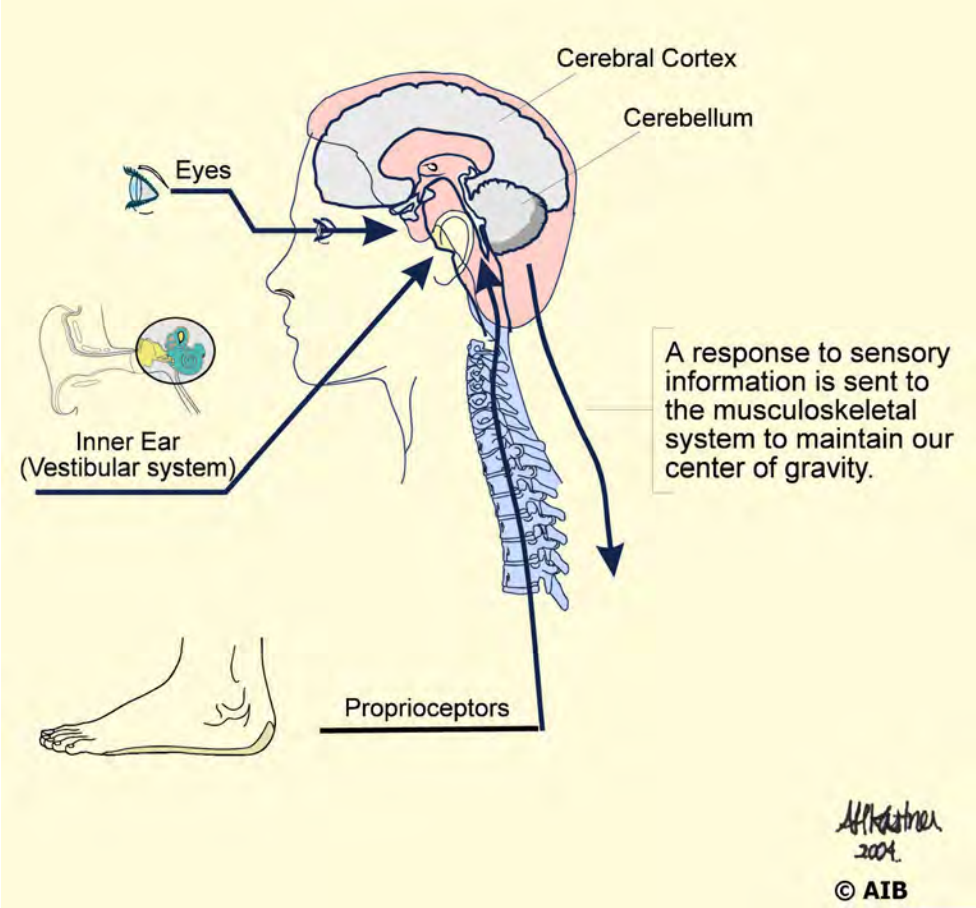
- Falls:
  - Leading cause of injury deaths in those 65+
  - Leading cause of nonfatal injuries
  - 1/3 of those 65+ suffer a fall every year
  - 30% cause injuries, which require medical Tx
  - Cause 300,000 hip fractures annually
  - Result in $3,000,000,000 in medical expense

Equilibrium

- Humans use 3 modalities to maintain their equilibrium
  - Vestibular
  - Vision
  - Somatosensory
- These systems can easily be isolated and differentiated clinically

Common Balance Symptoms Associated with Vestibular Problems

-Unsteadiness
-History of Falls
-Surface Dependence
-Visual Preference



Equilibrium

- A certain degree of disequilibrium is expected w/ age (presbystasis)
  - Lower extremity deconditioning
  - Comorbid conditions
  - Medications
  - Vestibular changes

Equilibrium

- Treatment:
  - Balance retraining therapy (more later)
  - Medication consultation
    - Benzodiazepines
    - Tricyclic Antidepressant
    - Hypnotics
    - Anti-psychotics
    - Alcohol
  - Environmental Modifications
  - Assistive device
  - Counseling

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Questions/Comments???

Let's take a 10 minute break.

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Hearing Your Best for Life

Case History

Samuel N. Bittel, Au.D.

Case Hx

- Focus areas for Hx:
  - When did symptoms start
  - Symptom specifics
  - Provoking factors
  - Concomitant symptoms

When did symptoms start?

- Date, time, etc.
- Event that predates complaints
  - Change in medication
  - Head trauma
  - Illness
  - Psychological stressor
  - Hospitalization
  - Injury
  - Surgery, medical procedure, etc.

Symptom Specifics

- Areas that need to be addressed:
  - Subjective experience
  - Duration of attacks
  - Frequency of episodes

Symptom Specifics

- Subjective quality of Sxs
  - Important to not lead patient
  - Dizziness is broad term, so must ask patient to be more specific
  - Let patients tell their story
  - Direct conversation, but leave the description of subjective quality open ended

Symptom Specifics

- Subjective quality of Sxs
  - True vertigo
  - Syncope, pre-syncope
  - Lightheadedness
  - Disequilibrium
  - Discomfort looking at moving objects
  - Issues in crowds or open spaces

Subjective Experience

- Vertigo
  - Illusion of spinning
  - Internal vs. room
  - Hallmark of vestibular dysfunction
  - Not specific central vs. peripheral
  - Product of nystagmus

Subjective Experience

- Syncope and pre-syncope
  - Losing consciousness or feeling faint
  - Typically cardiovascular
    - Orthostasis
    - Decreased cardiac output
    - Dehydration
  - Also vasovagal
    - Parasympathetic activity in limbic system
    - Decreased b.p. and pulse
    - Fight or flight response
  - Not vestibular!!!

Subjective Experience

- Lightheadedness
  - Fairly non-specific
  - Usually means disorientation
  - Can be reported w/ uncompensated vestibulopathy
  - If no vertigo at any point, probably not vestibular

Subjective Experience

- Disequilibrium
  - Environments that are challenging:
    - Ambulating in dark
    - Ambulating on uneven surfaces
  - Peripheral neuropathy and visual status
  - Falls: when, how, injuries, etc.
  - Leg strength, exercise (stand up w/o help)
  - Previous PT, assistive device, etc.
  - Fear of falling

Subjective Experience

- Disequilibrium
  - Important to remember that disequilibrium is often multifactorial
  - Often combination of decreased sensory input and deconditioning
  - Our job is to reduce falls and increase quality of life

Symptom Specifics

- Duration of attack (vertigo)
  - Very useful in narrowing down differential
  - Important to differentiate between actual attacks and residual, baseline, or concomitant symptoms
  - Duration alone cannot determine central vs. peripheral

Duration of Attack (Vertigo)

- Brief
  - Instantaneous (less than 1 second)
    - Indicative of unequal vestibular input
  - Seconds to minutes
    - BPPV
    - SCDS
    - Perilymphatic fistula
    - Arnold-Chiari malformation
    - MS
    - Cerebellar lesion

Duration of Attack (Vertigo)

- Intermediate
  - 20 minutes to 2 hours
    - Meniere's disease
    - Migraine
    - TIA
    - Acute intoxication
    - Panic attacks
    - MS

Duration of Attack (Vertigo)

- Long
  - Days
    - Vestibular neuritis
    - Labyrinthitis
    - MS
    - Infarct- brainstem, cerebellum
    - Labyrinthine concussion
    - Labyrinthine ischemia

Symptom Specifics

- Frequency of episodes (vertigo)
  - Single vs. recurrent
  - Typically inversely proportional to duration
    - Single = longer duration
    - Recurrent = shorter duration
  - If recurrent, how often???

Frequency of Episodes (Vertigo)

- Single
  - Vestibular neuritis (unless recurrent)
  - Labyrinthitis
  - Stroke/TIA
  - Labyrinthine concussion/ischemia

Frequency of Episodes (Vertigo)

- Recurrent
  - BPPV
  - Meniere's disease
  - Migraine
  - Panic Attack
  - SCDS
  - MS

Duration and Frequency

Duration	Central	Peripheral
Acute Long (days)	Cerebellar Infarct Cerebellar Hemorrhage Brainstem Infarct Multiple Sclerosis	Vestibular Neuritis Labyrinthitis Autoimmune Inner Ear Disease Labyrinthine Ischemia Labyrinthine Concussion
Recurrent Long (minutes – hours)	Vertebrobasilar Ischemia Multiple Sclerosis Migraine	Autoimmune Inner Ear Disease Meniere's Disease Recurrent Vestibular Neuritis
Recurrent Brief (seconds)	Cerebellar Tumor Cerebellar Atrophy Multiple Sclerosis	BPPV Superior Canal Dehiscence

Adapted from: Baloh, 1998; Baloh & Honrubia, 1990; Roberts, Richard

Provoking Factors

- Can your symptoms be provoked?
  - Positioning
  - Complex visual stimuli
  - Motion
  - Open spaces, crowds, situational
  - Loud sound, increased intracranial pressure
  - Environmental
  - Foods, smells, sleep deprivation, hormonal

Provoking Factors

- Positioning
  - BPPV = lying supine, rolling, head pitch, rising from supine, bending at waist
  - Orthostatic hypotension = rising from supine or seated; NOT lying supine
  - Head pitch = neck hyperextension?
    - Vertebrobasilar insufficiency
    - Cervicogenic

Provoking Factors

- Complex visual stimuli – Optic flow
  - Driving down road w/ trees
  - Walking down grocery isle
  - Video games

  - Can be seen w/ vestibular lesions
  - Psychogenic?
  - Migraine?

Provoking Factors

- Motion
  - Self vs. environment
    - Self = vestibular system activated
    - Environment = optic flow, motion intolerance
- Open spaces, crowds, situational
  - Psychogenic
  - Agoraphobia

Provoking Factors

- Environmental
  - Ambulating in dark environments or uneven surfaces
  - Dark environments = visual preference
  - Uneven surfaces = surface preference
  - Falls = inside vs. outside
  - Weather change = migraine

Provoking Factors

- Migraine
  - Smells = perfume, cleaning solutions, smoke (do not wear perfume in clinic)
  - Sleep deprivation
  - Foods = tannins, chocolate, aspartame, MSG, hard cured meet & cheese
  - Hormonal (females) = menses cycle
- Meniere's disease
  - Alcohol, caffeine, salt

Concomitant Symptoms

- Concomitant symptoms
  - Nausea and emesis common w/ vertigo
  - Otologic symptoms = ear
  - Neurological symptoms = central
  - Psychogenic symptoms = positive review of systems

Otologic Symptoms

- Aural fullness
  - Meniere's disease
  - Vestibular schwannoma
- Otalgia or Otorrhea
  - Middle ear disease
  - Temporal bone disease
- Tinnitus
  - Meniere's disease
  - Labyrinthitis
  - Vestibular schwannoma

Otologic Symptoms

- Hearing loss
  - Meniere's disease
  - Vestibular schwannoma
  - Labyrinthitis
  - Perilymphatic fistula
  - Vestibular ischemia

Otologic Symptoms

- Aural distortion
  - Vestibular schwannoma
  - SCDS
- Autophony, misophonia, hyperacusis
  - SCDS
  - Perilymphatic fistula
  - Psychogenic

Neurological Symptoms

- Facial symptoms
  - Weakness = tumor, MS
  - Asymmetry = vestibular schwannoma, stroke, Bell's palsy, MS, migraine
  - Numbness/tingling = MS, stroke, migraine
- Ataxia
  - Poor control over lower extremities
  - Never vestibular
- Headache

Symptoms Central vs. Peripheral

Symptom	*Central*	*Peripheral*
Imbalance	Severe	Mild-Moderate
Nausea/Emesis	Variable	Severe
Auditory Sxs	Rare	Common
Neuro Sxs	Common	Rare
Central Comp	Slow	Rapid

Adapted from: Baloh, 1998; Baloh & Honrubia, 1990; Roberts, Richard

Psychological Symptoms

- Positive review of systems
  - Every symptom
  - Precipitating stressful event
  - Phobic or anxious temperament
  - Avoidance
  - Overly emotional during Hx

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Hearing Your Best for Life

VEMP

VNG- Oculomotor Testing

Samuel N. Bittel, Au.D.

Vestibular Evoked Myogenic Potential (VEMP)

- Background:
  - Otolithic organs may be sound (or vibration) sensitive
  - Saccule serves as hearing organ in some animals
  - In humans, there might be basic low-frequency hearing (primitive response)
  - Otolthic organs can be stimulated by both AC and BC signals

VEMP

- Two VEMP responses being utilized clinically
  - Cervical VEMP (cVEMP)
  - Ocular VEMP (oVEMP)
- These tests differ in several important ways
  - Administered differently
  - Originate in different otolithic organs
  - Represent reflex in differing nerve branches
  - Differ in ipsil vs. contra response

cVEMP

- Stimulating the saccule with sound generates a reflex arc:
  - Ipsilateral afferent pathway
  - Creates inhibition in the contraction of the SCM
  - This arc is independent of hearing
  - Must have sufficient sound/vibratory energy to stimulate the saccule
  - This reflex arc is responsible of sending information for equilibrium down to postural control muscles of neck, torso, and legs

cVEMP

- The cVEMP is the only clinical test we have to measure the vestibulocolic reflex
- It can also help give us additional information about the descending motor tract
- Deficit = issues with postural stability

Vestibulocollic Reflex (VCR)

- Vestibular system's connection to stabilization muscles of cervical spine
- Righting reflex
- Helps maintain upright head position
- Independent of trunk movement
- Mediated through otolithic organs and medial vestibulospinal tract

Vestibulo-Spinal and Collic Reflexes

Sound stimulates saccule;

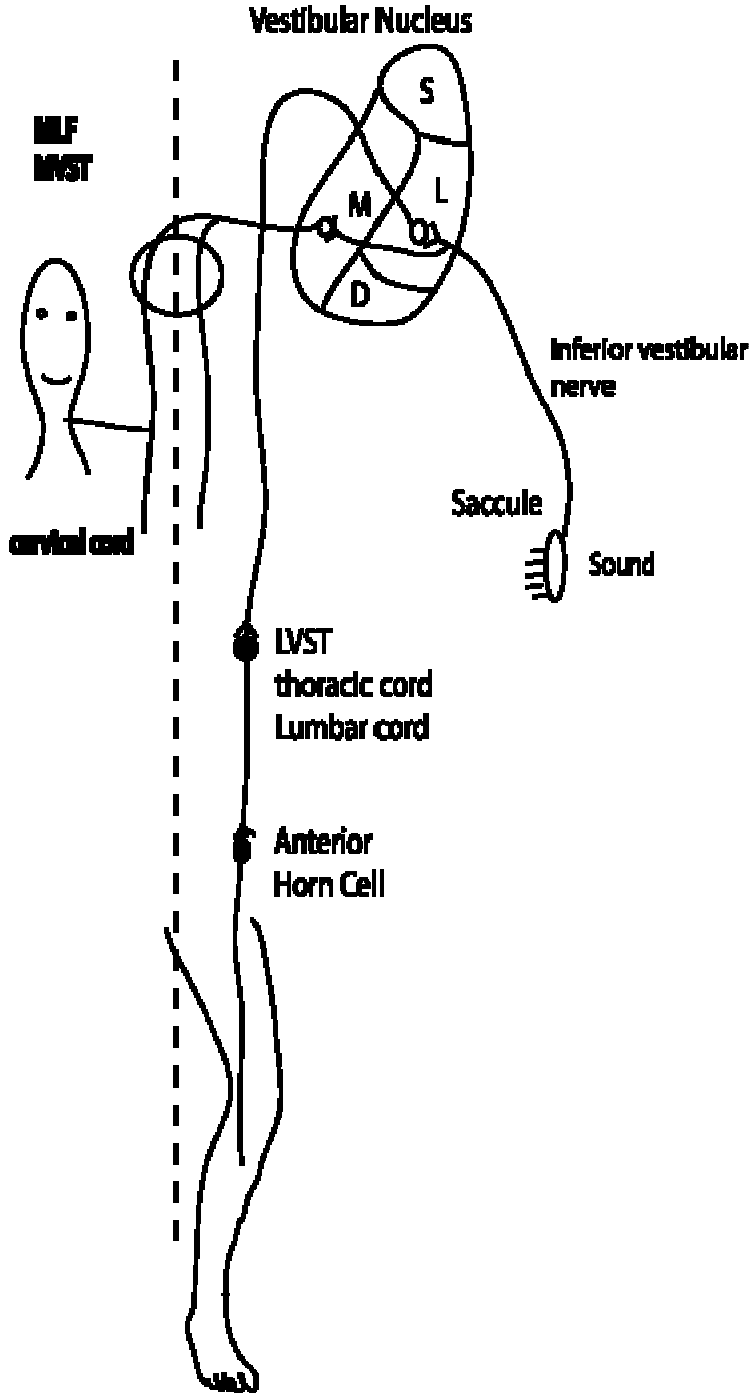
Activates inferior vestibular nerve;

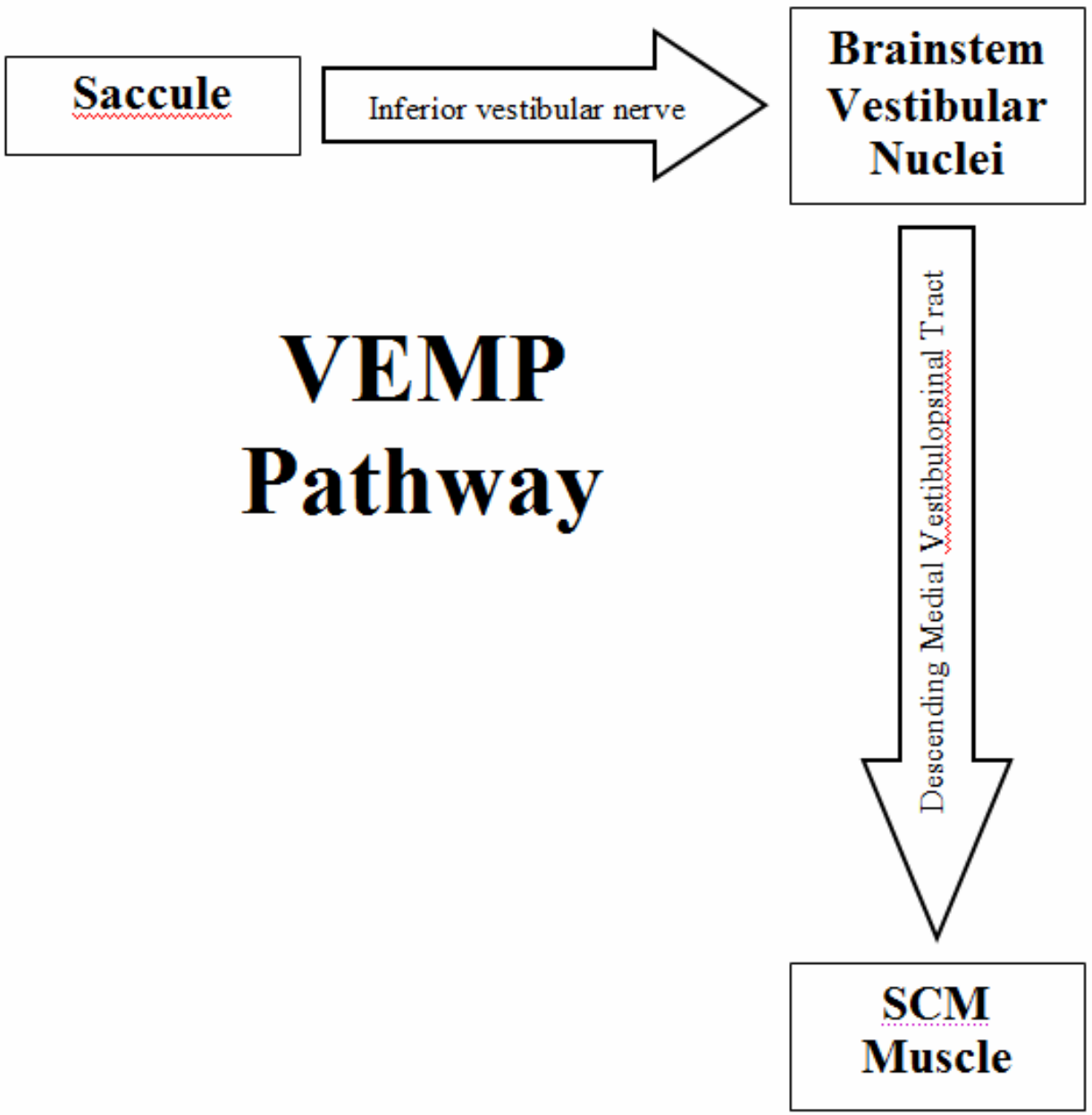
Lateral vestibular nucleus;

Medial vestibulospinal tract (ipsi);

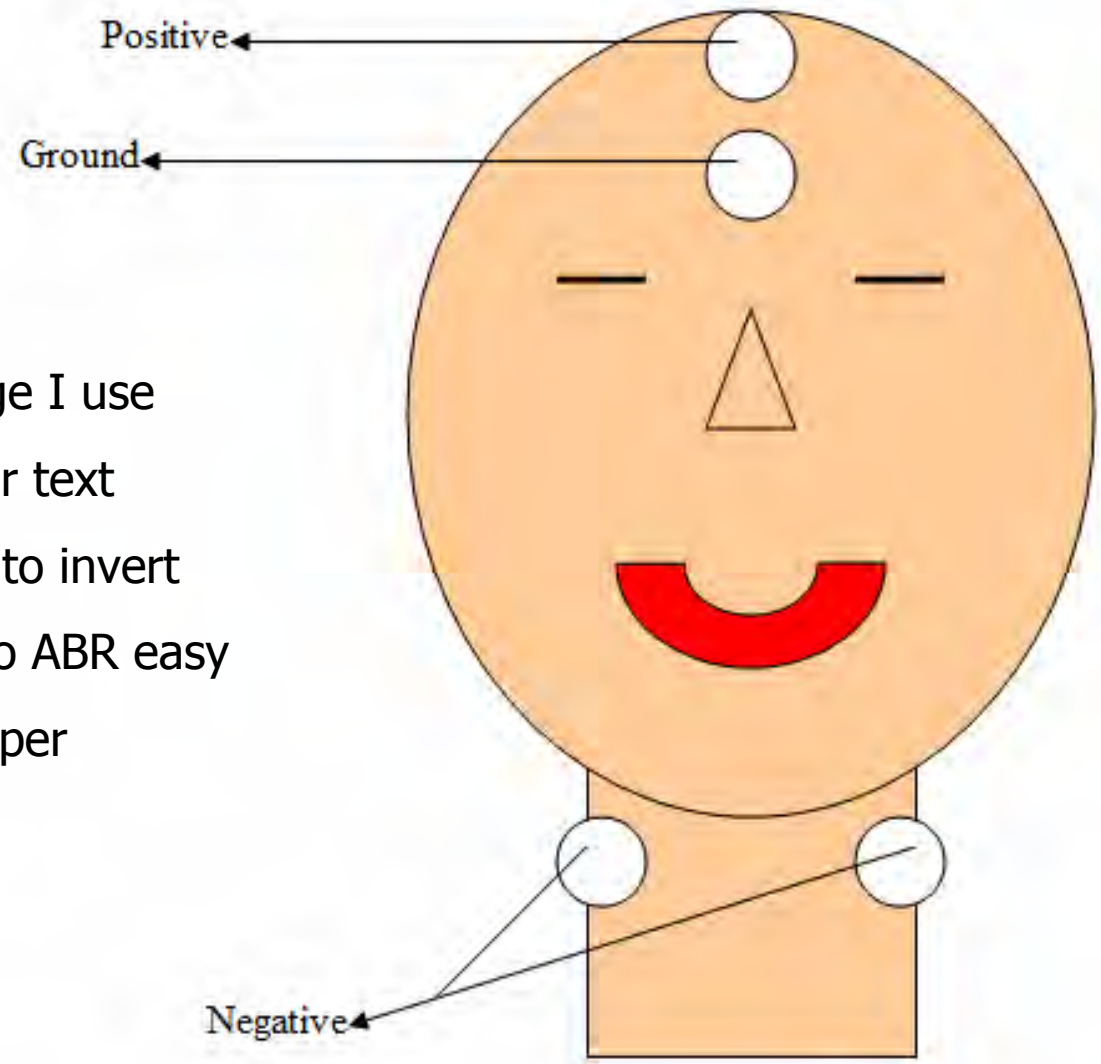
SCM;

Lateral vestibular spinal tract





VEMP Pathway



-This is the montage I use

-Different than your text

-Causes waveform to invert

-Makes switching to ABR easy

-Common + = jumper

cVEMP

- Connecting patient:
  - This is a large response = impedance much less important
  - Scrubbing with alcohol appropriate
- Electrode placement
  - Negatives should be just above midpoint of SCM (belly of muscle)
  - Placement should be mirrored between sides (location important for latency)

cVEMP

- Test procedure:
  - Patient placed supine w/ head slightly elevated
  - Insert earphone placed in test ear
  - Patient asked to lift head 2-3”
  - Patient should turn head in direction contralateral to test ear
  - Stimulus started when head in ideal position

cVEMP

- Test procedure:
  - Patient should hold head as steady as possible
  - ~60 runs
  - Ask pt. to return to neutral and rest
  - Replicate
  - Complete same procedure for other side
    - Switch earphone, etc.

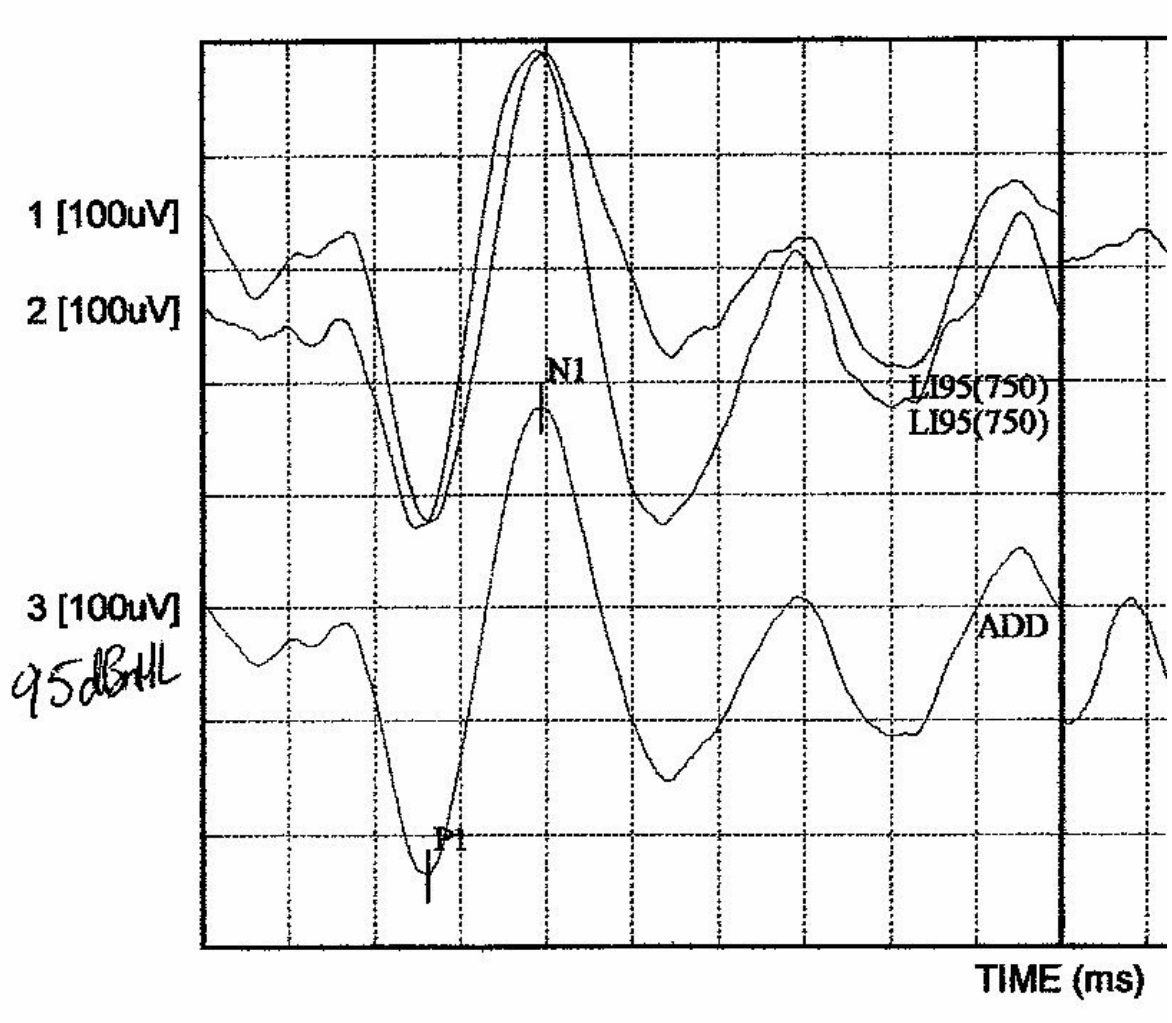
cVEMP

- Test procedure:
  - Watch for equal SCM contraction
  - I start with a stimulus at 100 dBnHL
  - I will go as loud as 105 dBnHL
  - Refer to text for equipment settings
  - Summate responses and mark…

cVEMP

- Normal response:
  - With **my** montage, you would expect a downward wave at ~13 msec. and a positive wave at ~23msec.
  - First part of wave called "p" for positive (downward/negative w/ my montage)
  - Second part of wave called "n" for negative
  - AKA- P13-N23 waveform

cVEMP



cVEMP

- What we measure:
  - Latency of P13
  - Latency of N23
  - Amplitude of wave (P13 to N23)
  - Threshold (if applicable)
  - Asymmetry in any of the above between sides

cVEMP

- My clinical norms:
  - Latency: within 3 msec. of 13 and 23 msec.
  - Amplitude: under 500 µvolts
  - Threshold: threshold above 90 dBnHL
  - Asymmetry:
    - P13 and N23 should be within 3 msec.
    - Amplitude should be no less than 1/2

cVEMP

- Technical way to calculate amplitude asymmetry (aka asymmetry ratio):

AR=

(AS amp – AD amp)/(AS amp + AD amp) x 100%

cVEMP

- Abnormal findings:
  - Latency = central (typically)
  - Amplitude = peripheral (typically)
  - Absent = peripheral (typically)

cVEMP

- Absence means saccular or inferior vestibular nerve involvement
  - Vestibular schwannoma
  - Neuritis w/ inferior nerve involvement
  - Damage to saccule
  - Meniere's disease
  - Etc…

cVEMP

- Atypically large response:
  - SCDS
  - Vestibular schwannoma
  - Meniere's disease
  - WHY????
- Atypically low threshold:
  - SCDS

cVEMP

Pathology	cVEMP Responses			
	Absent	*Reduced*	*Enhanced*	*Delayed*
Otologic				
Meniere's Disease	X	X	X	
Superior Canal Dehiscence			X	
Labyrinthitis	X	X		
Vestibular Neuritis	X	X		
Neurologic				
Migraine	X	X		X
Spinocerebellar Degeneration	X			X
Multiple Sclerosis	X			X
Brainstem Stroke	X			X

Roberts & Gans (2005

cVEMP

- Considerations:
  - Technique matters!!!!
  - Must have fairly equal SCM contraction
  - SNHL does not influence this response, as it is not an auditory potential
  - Sound = vibration
  - Conductive HL can obliterate VEMP, as impedes vibratory energy

cVEMP

- Considerations:
  - I do not typically search for threshold
  - Low thresholds w/ SCDS
  - I do a search if pt. symptoms and other test findings suggest SCDS
    - Air-bone gaps
    - Tullio's phenomenon, Hennebert's sign, etc.

cVEMP

- Considerations:
  - Latency does not typically change with a decrease in intensity
  - Position of electrode can influence latency (distance between electrodes)
  - Response is result of attenuation of tonic SCM contraction
  - Need proper contraction to see attenuation

cVEMP

- Considerations:
  - Stimulation type matters
  - VEMPs can be elicited a number of ways
    - Skull taps
    - Bone conduction
    - Air conduction
  - Best to use 100 µsec tone burst
  - 500-750 Hz
  - At least 60 sweeps (if possible)

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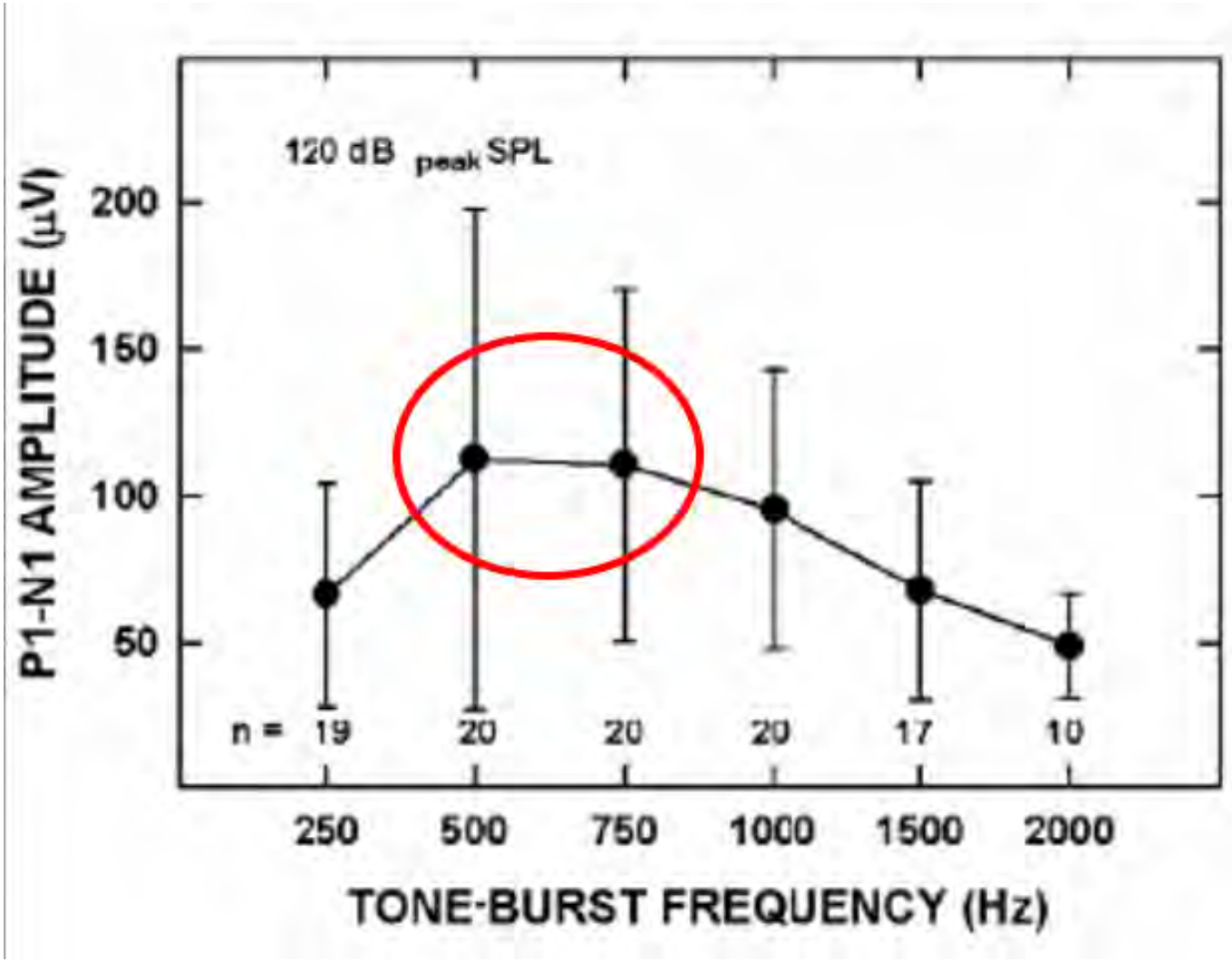
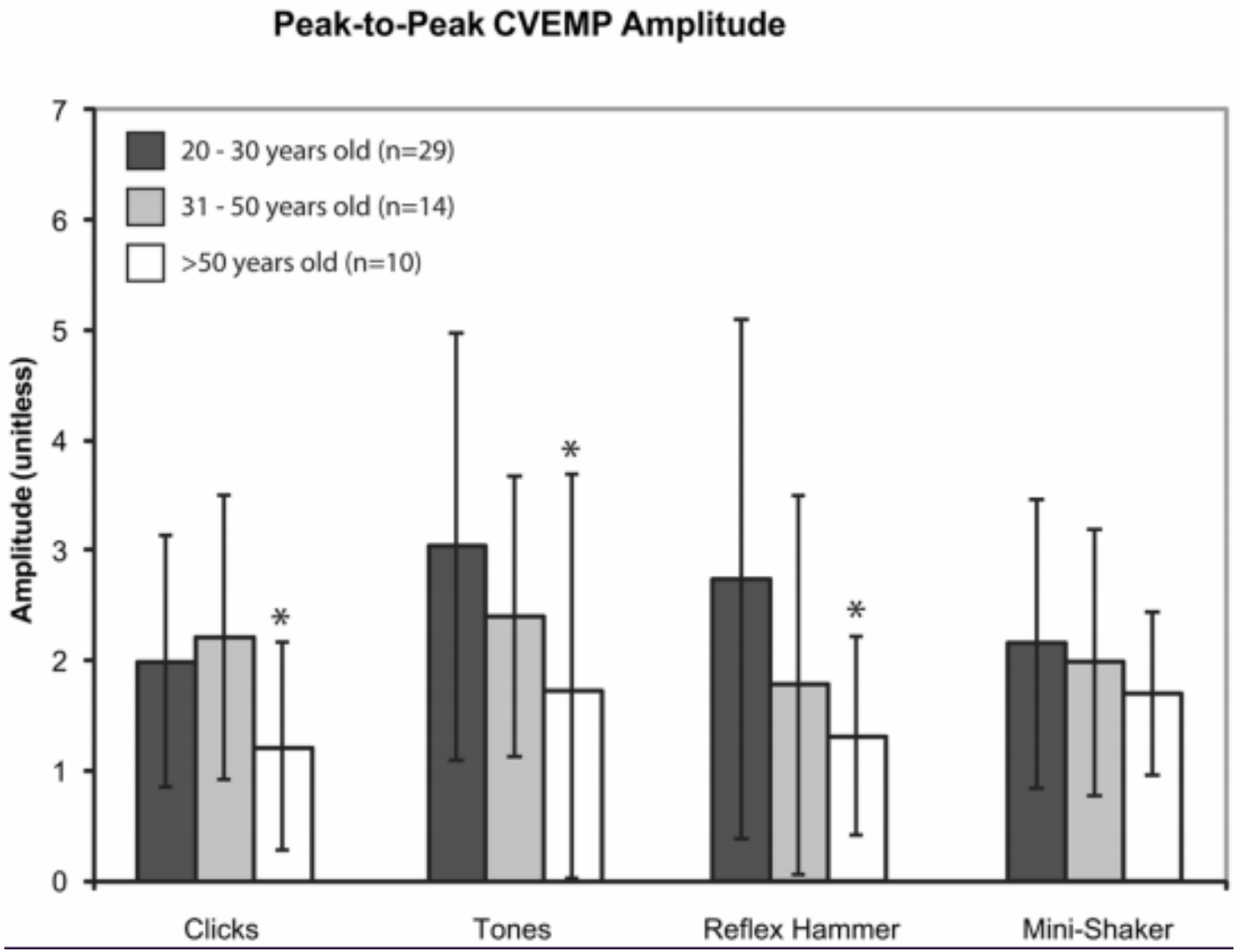
cVEMP



cVEMP



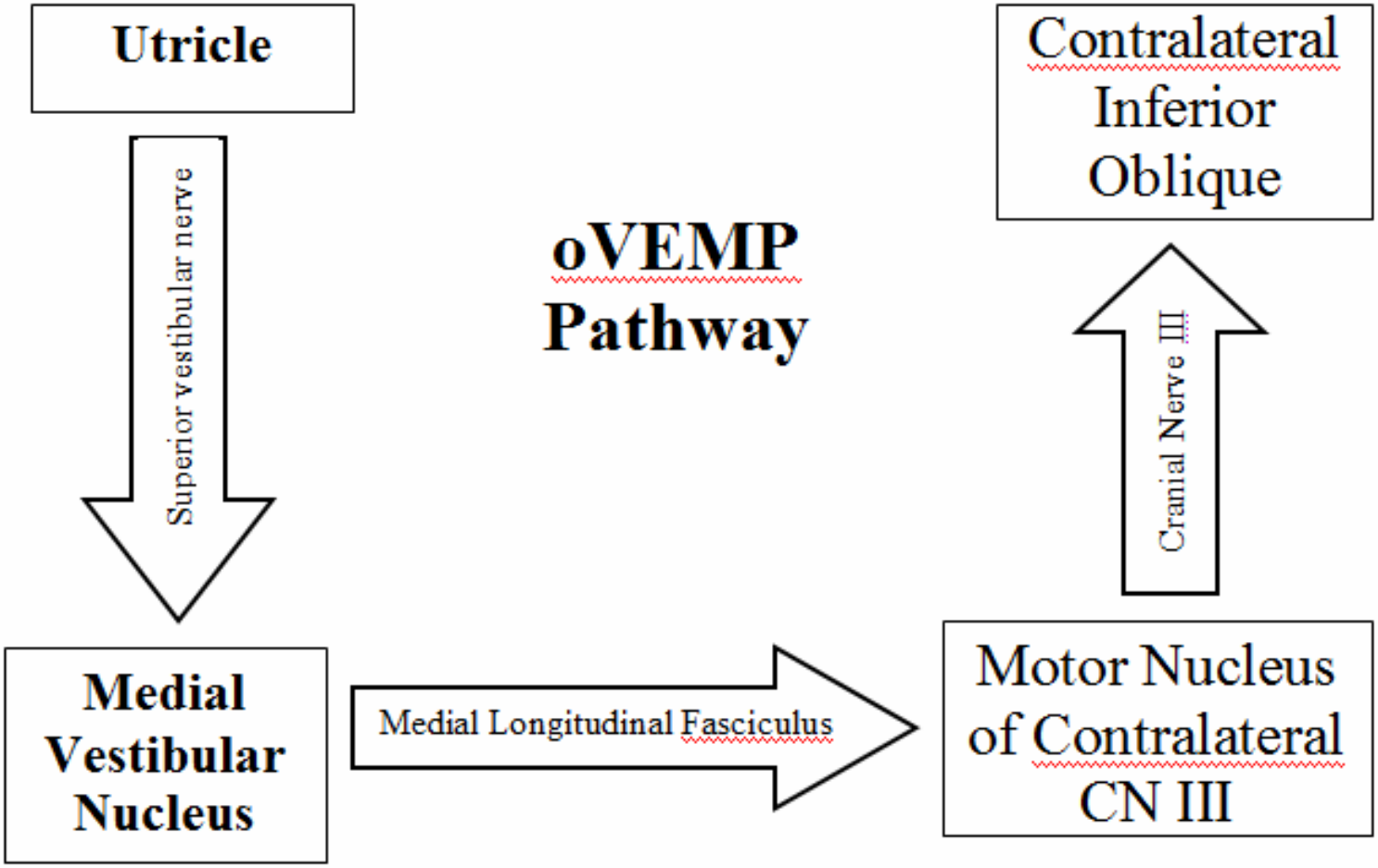
cVEMP

- Clinical utility:
  - Only test looking at VCR
  - Helps identify saccular damage
  - Looks at inferior vestibular nerve
  - Gives information about descending motor tract, etc.
  - Correlation w/ vestibular-related disequilibrium

oVEMP

- Stimulating the otolithic organs (specifically the utricle) with sound can also create an ocular response
- Seems to reflect a VOR pathway
- Mechanical vibration (like cVEMP) creates a reflex arc that activates contralateral inferior oblique

oVEMP



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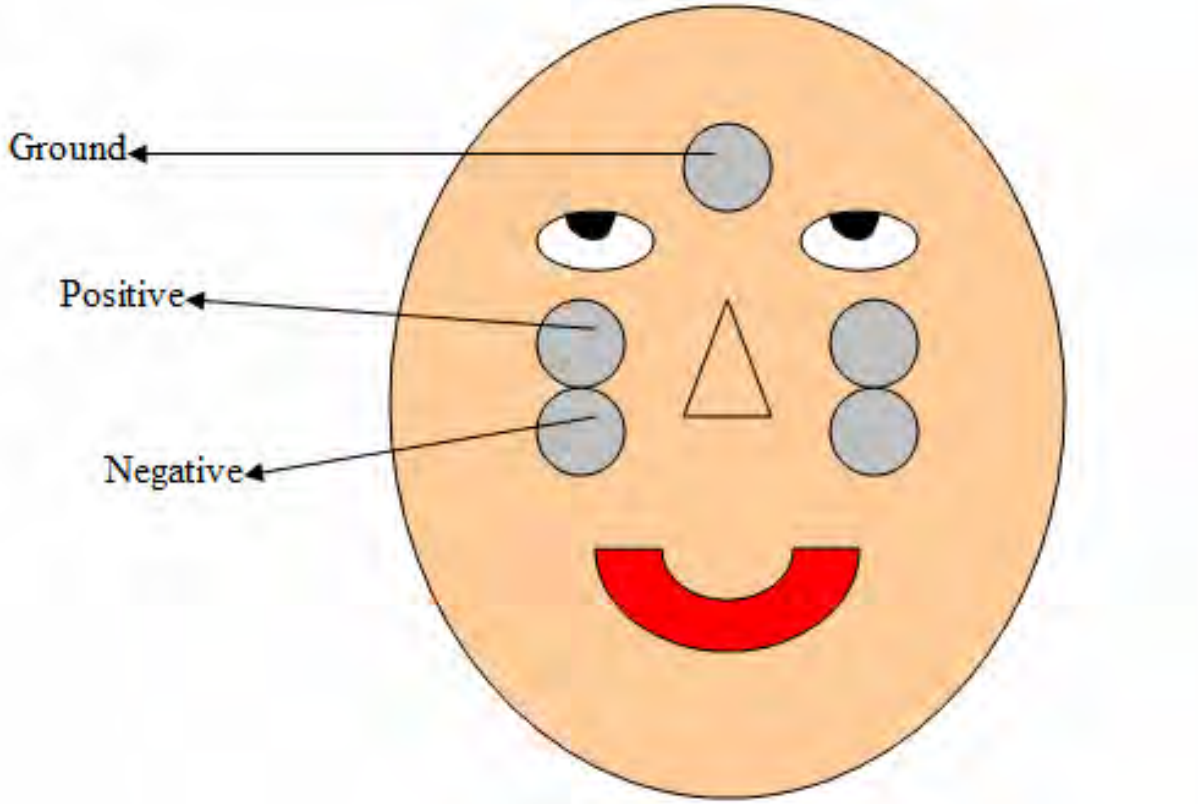
oVEMP



oVEMP

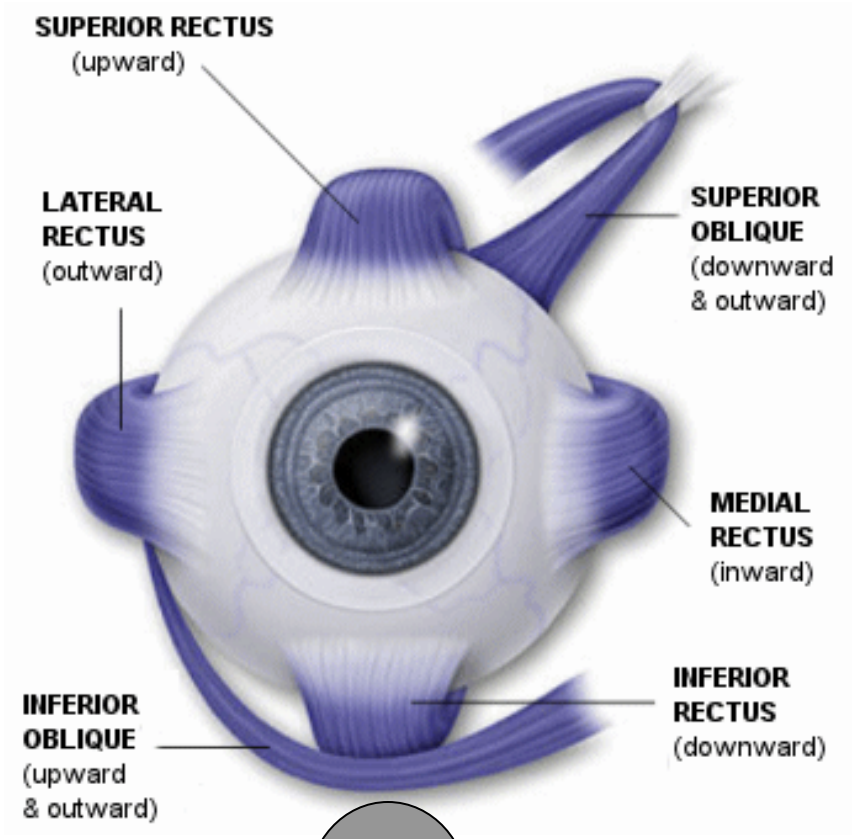
- Recording parameters
  - Gain = 100,000 (cVEMP = 5,000)
  - Sampling rate = ~3,000 Hz
  - Epoch = 100 msec.
  - Artifact rejection =on at 40 µvolt
  - Filtering = 10 – 2000 Hz
  - Stimulus similar to cVEMP

oVEMP

- Waveform negative (N1) and positive (P2)
- Negative wave 10-12 msec.
- Positive wave 15-20 msec.
- Norms not as well established as cVEMP – I look for presence vs. absence

oVEMP

- Patient needs to gaze upwards…



oVEMP

- Considerations
  - Response fairly small
  - Large amplification = more noise
  - Can be influenced by conductive HL
  - 25% of "normal" patients over 60 may not have
  - Looks at entirely different pathway than cVEMP
  - Norms not as established as other tests

oVEMP

- Considerations
  - May be easier to perform for patients
  - Not dependant on tonic muscle contraction
  - Multiple runs might be easier
  - Verify superior vestibular nerve involvement
  - Sensitive to SCDS – easier to establish threshold

Clinical Utility

- cVEMP = saccule and inferior vestibular nerve
- oVEMP = utricle and superior vestibular nerve
- Calorics = horizontal SCC and superior nerve

Clinical Utility

- Present oVEMP and calorics w/ absent cVEMP = saccular or inferior nerve lesion
- Present cVEMP with absent oVEMP and calorics = superior nerve lesion
- Present cVEMP and calorics with absent oVEMP = utricular lesion
- Present cVEMP and oVEMP with absent caloric = horizontal SCC lesion
- Note- keep in mind that the superior and inferior vestibular nerves have separate branches for separate structures

	oVEMP	cVEMP
Origination	utricle	saccule
Pathway	contralateral	ipsilateral
Vestibular Nerve	superior branch	inferior branch
Response	excitation of inferior oblique	relaxation of SCM

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VNG:

Oculomotor and Gaze testing

Oculomotor Testing

- Purpose:
  - Recognize central impairments
  - Differentiate between central vs. peripheral
  - Identify spontaneous/revocable nystagmus
  - Identify factors that may confound other tests

Oculomotor Testing

- Can be further divided into subtests:
  - Saccades
  - Smooth pursuit tracking
  - Optokinetics (OPKs)
  - Gaze testing (and HFHS)

Saccades

- Ability of eyes to rapidly move to an object of interest to focus on fovea
- Target moves randomly and patient moves eyes quickly to target
- Pt. asked to keep head stationary
- Target should move randomly
  - 5° - 40°
  - Fixed interstimulus interval

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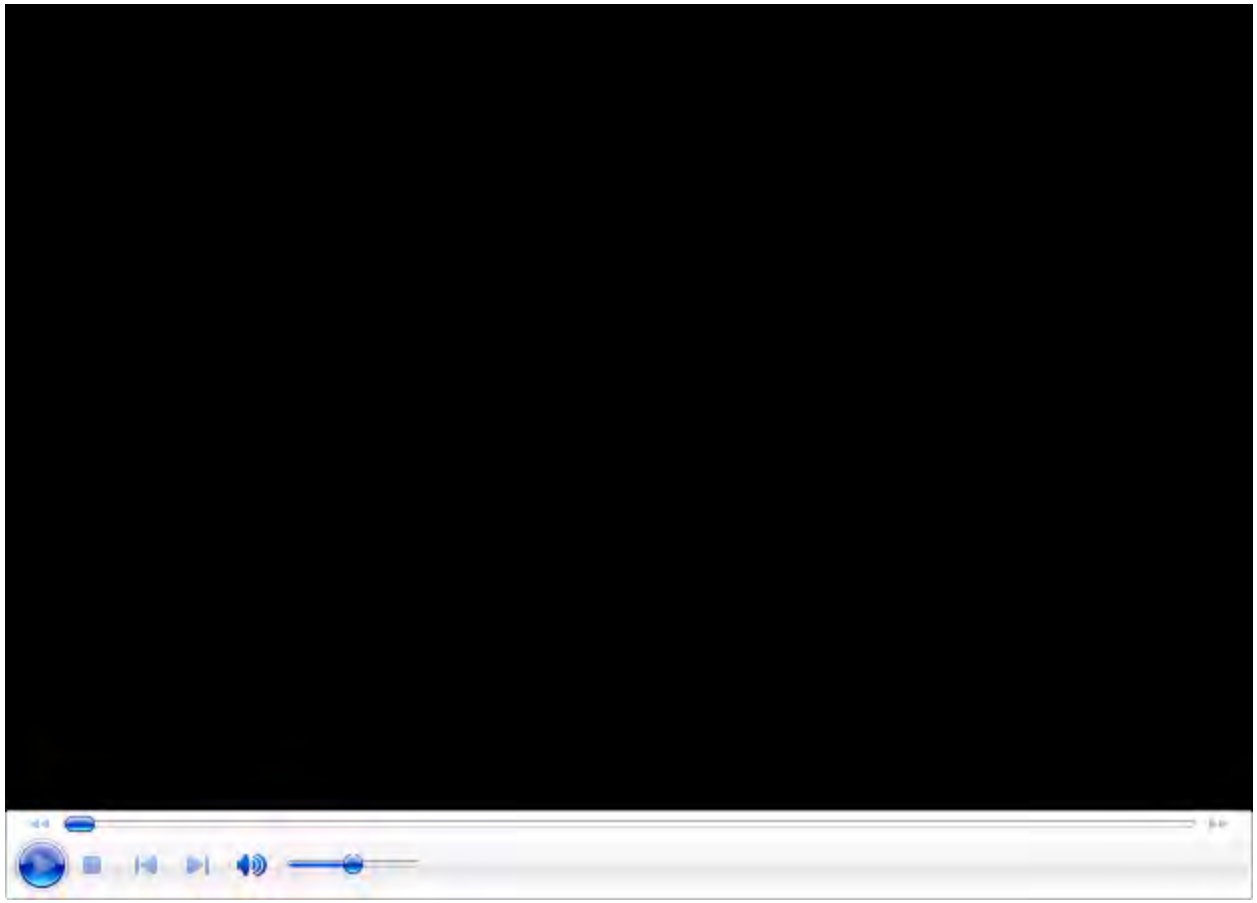
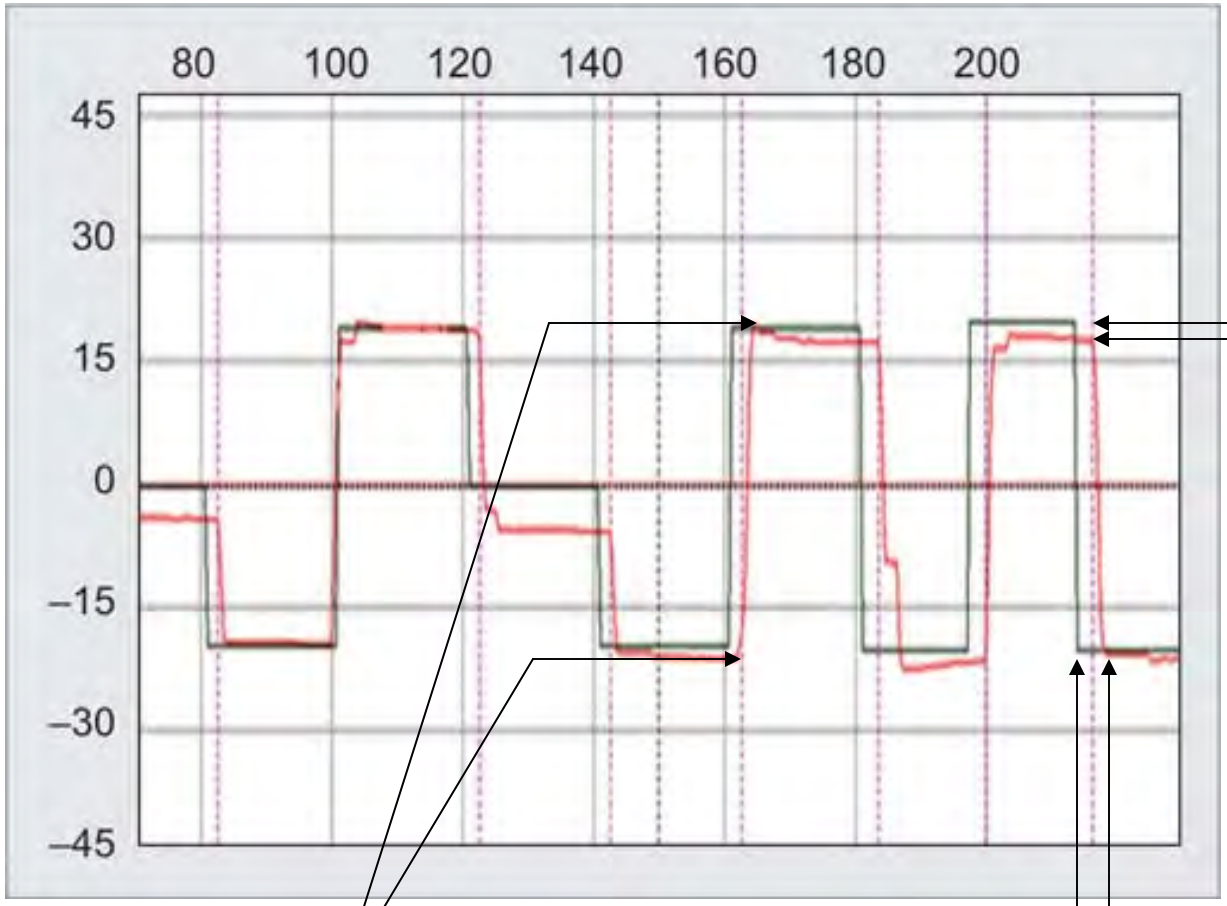
Saccades



Saccades

- Analysis
  - Velocity: how fast eye moves to target after the saccade has been initiated (how fast eyes move once start moving)
  - Accuracy: how far the eyes moved over/under the target during their excursion
  - Latency: difference in time in milliseconds from the movement of the target to the initiation of eye movement (how long it takes for eyes to start moving)

Saccades



Accuracy

Velocity

Latency

Saccades - Velocity

- Abnormal slowing caused by:
  - Impairment of the brainstem neural network that generates saccadic pulse
  - Might be (fairly diffuse)-
    - Basal ganglia
    - Brainstem
    - Cerebellum
    - Oculomotor nerves/muscles
  - Can also be caused by fatigue/drowsiness, CNS depressant meds.

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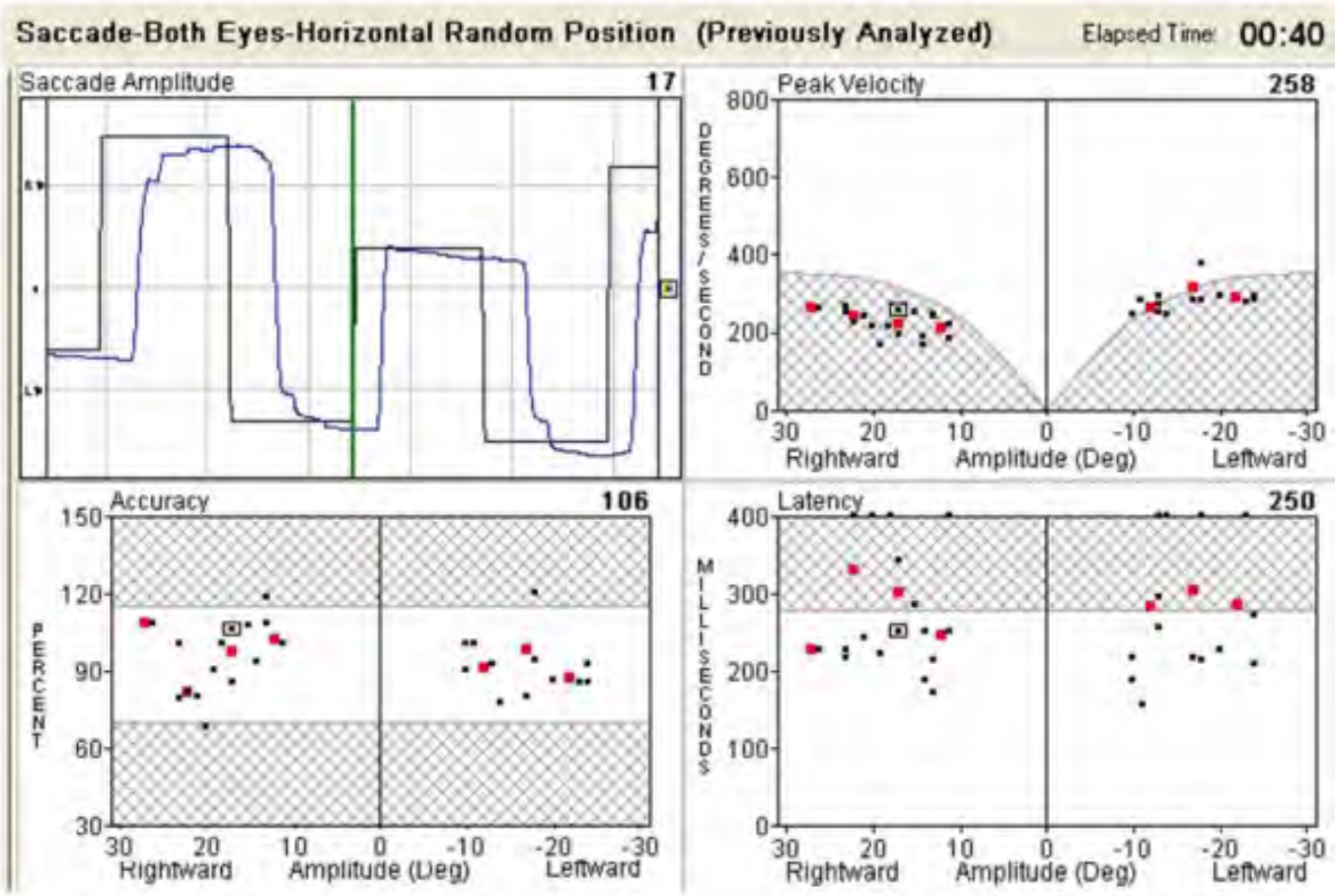
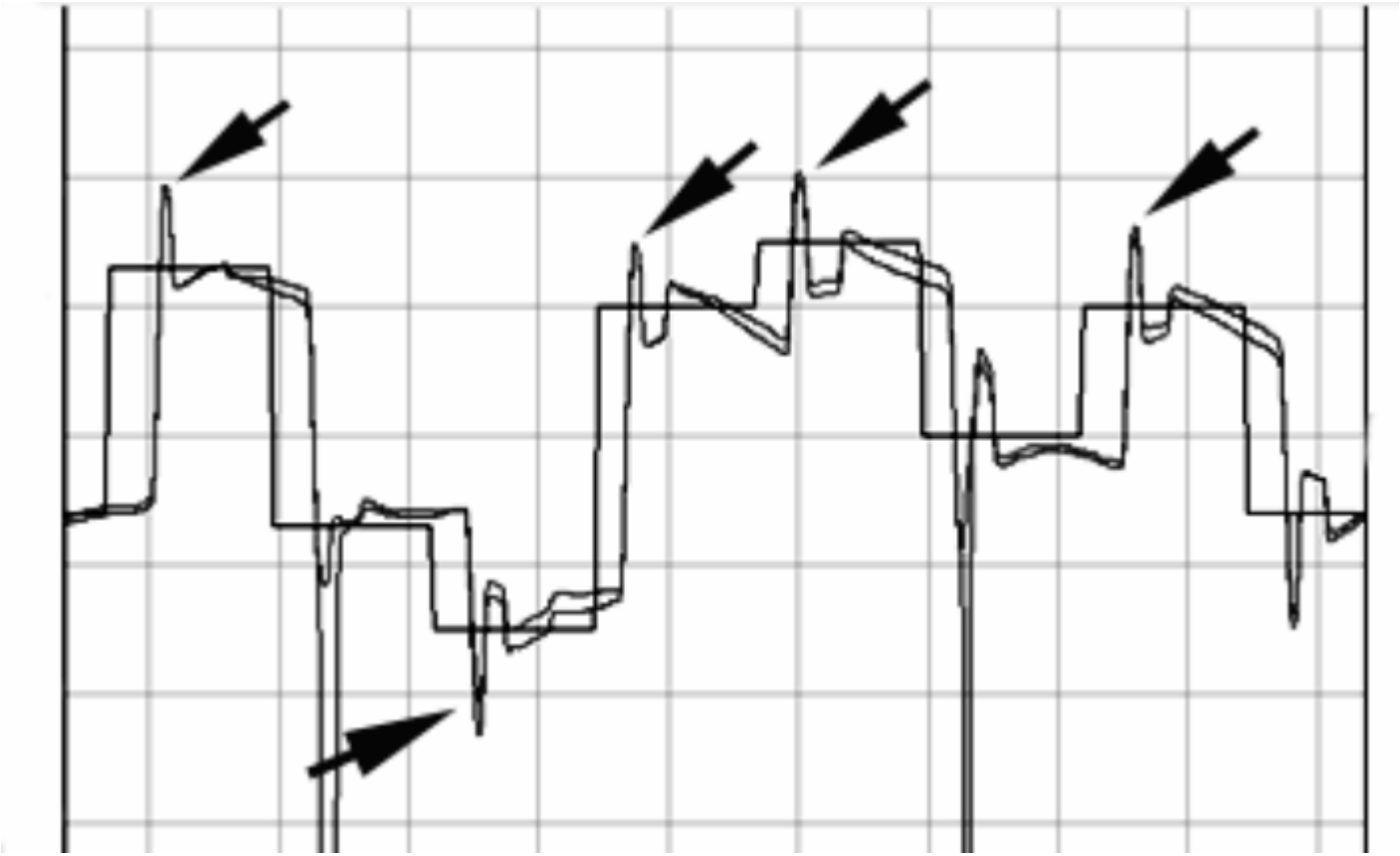
Saccades - Velocity



Saccades - Accuracy

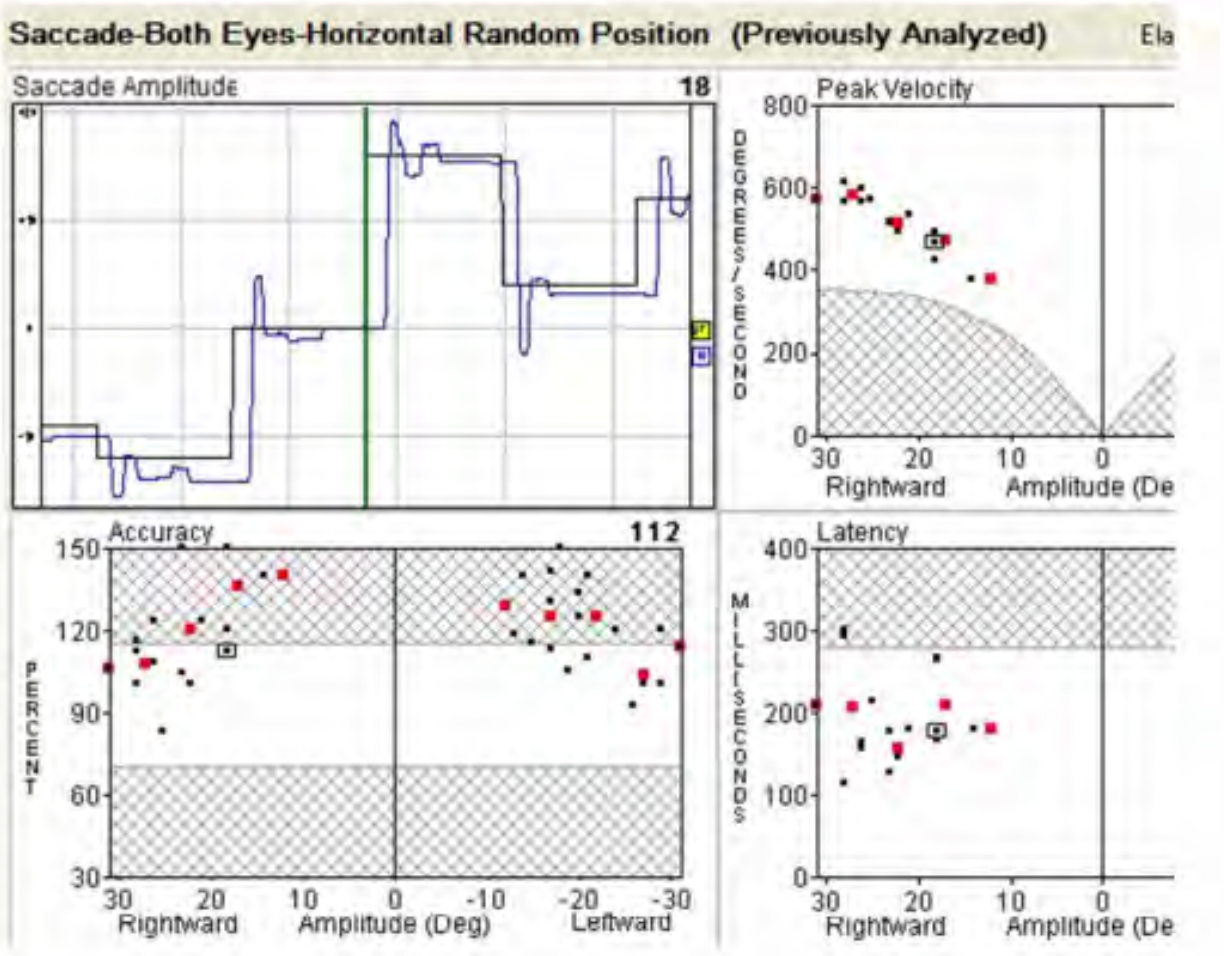
- Abnormal accuracy caused by:
  - Hypometria (undershoots)
    - Cerebellar dorsal vermis (bilateral)
    - Ipsilateral cerebellar/brainstem (unilateral)
    - MG
  - Hypermetria (overshoots)
    - Cerebellar
    - Cerebellar fastigial nucleus (bilateral)
  - Medications, fatigue, attention

Saccades - Accuracy



Overshoots

Saccades - Accuracy



Saccades - Latency

- Abnormal latency caused by:
  - Incorrect calculation by pulse integrator
  - Basal ganglia in disorders of motor initiation
  - Visual deficits
  - Medication, fatigue, state of arousal
  - More significant if unilateral

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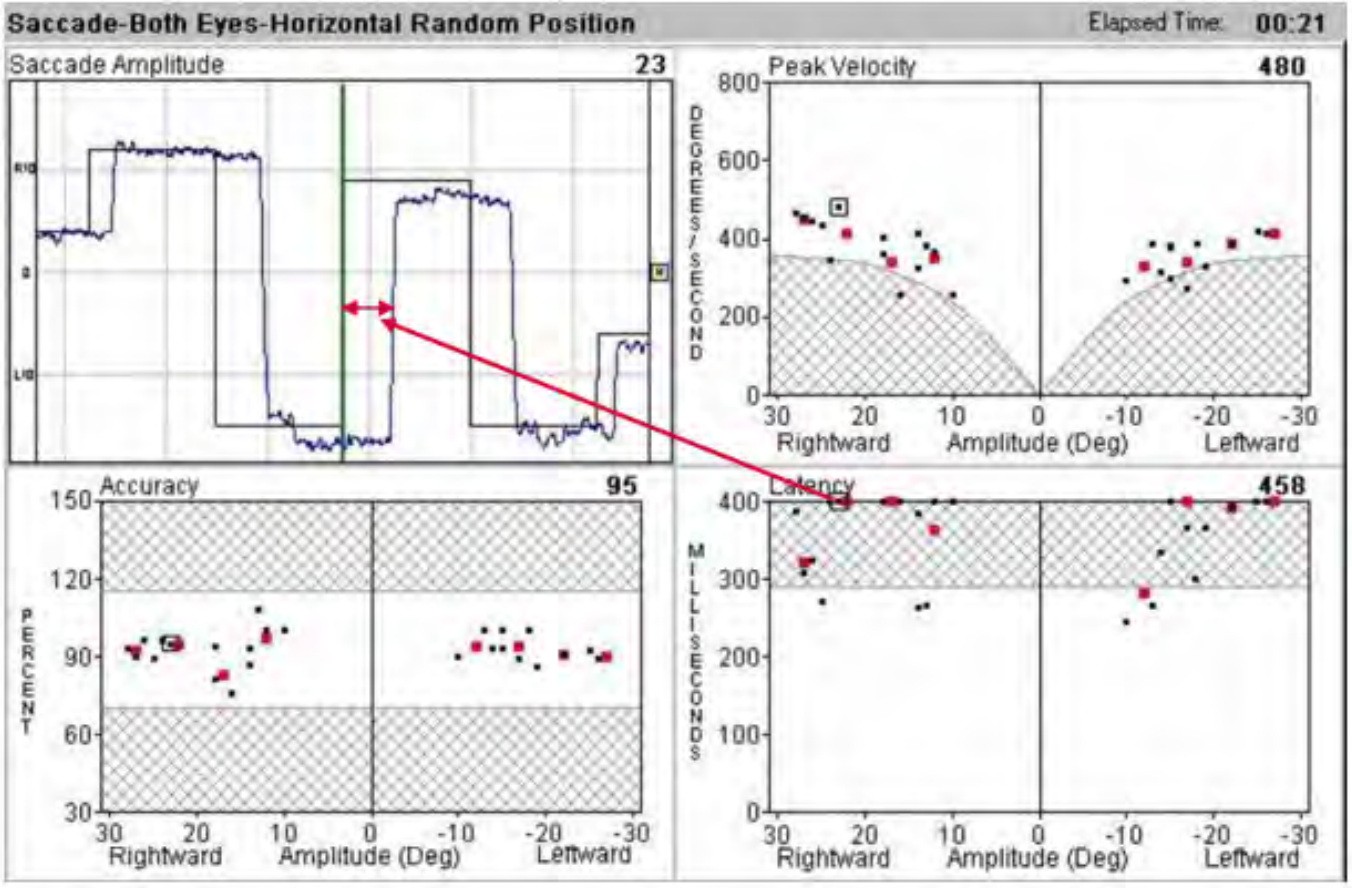
Saccades - Latency



Smooth Pursuit

- Smooth moving stimulus to the right and left up to about 30°
- Target moves from .2 - .8 Hz
- Age corrective normative data should be used
- Patient participation is important for this and saccades; if abnormal, you should repeat with re-instruction

Smooth Pursuit

- Generation and sustaining smooth pursuit:
  - Multiple areas of cortex
  - Projections through pontine area of brainstem and cerebellum, onto nuclei of extraocular muscles
  - Multitude of pathways
  - Probably reflects issue with cerebral cortex with VNG
  - Cannot easily identify site of lesion

Smooth Pursuit

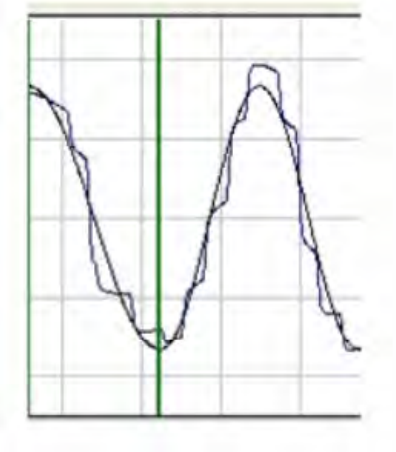
- Interpretation:
  - Velocity gain- how sinusoidal eye movement was compared to target
  - Asymmetry- velocity gain right vs. left; performance w/ right vs. left moving target
  - Phase- how much eye is leading or lagging behind target

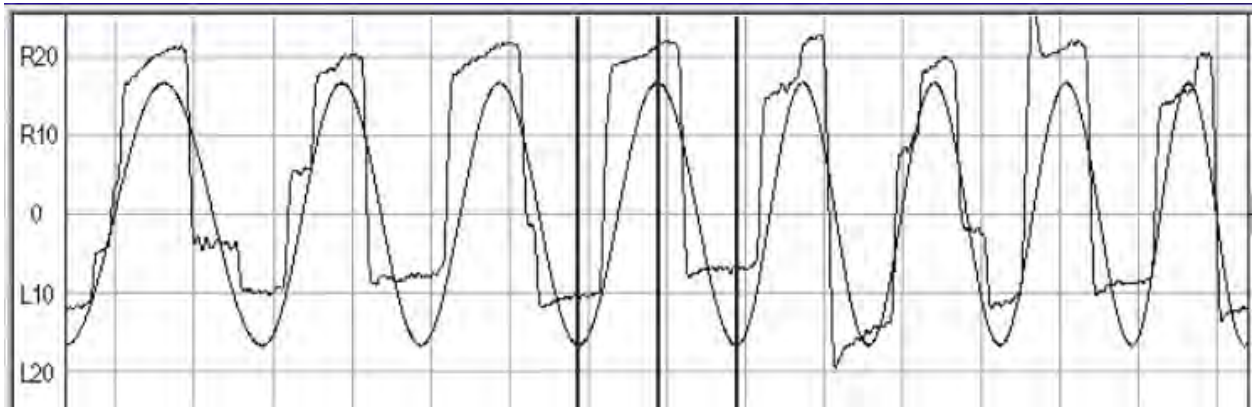
Smooth Pursuit

- Interpretation:
  - Highly age dependant; age-related changes can be seen as early as 3rd decade
  - Leading target is not pathologic and is test related (anticipatory)
  - Performing poorer with lower frequencies is not expected, and should be considered task related
  - Normal high frequencies = normal study

Smooth Pursuit

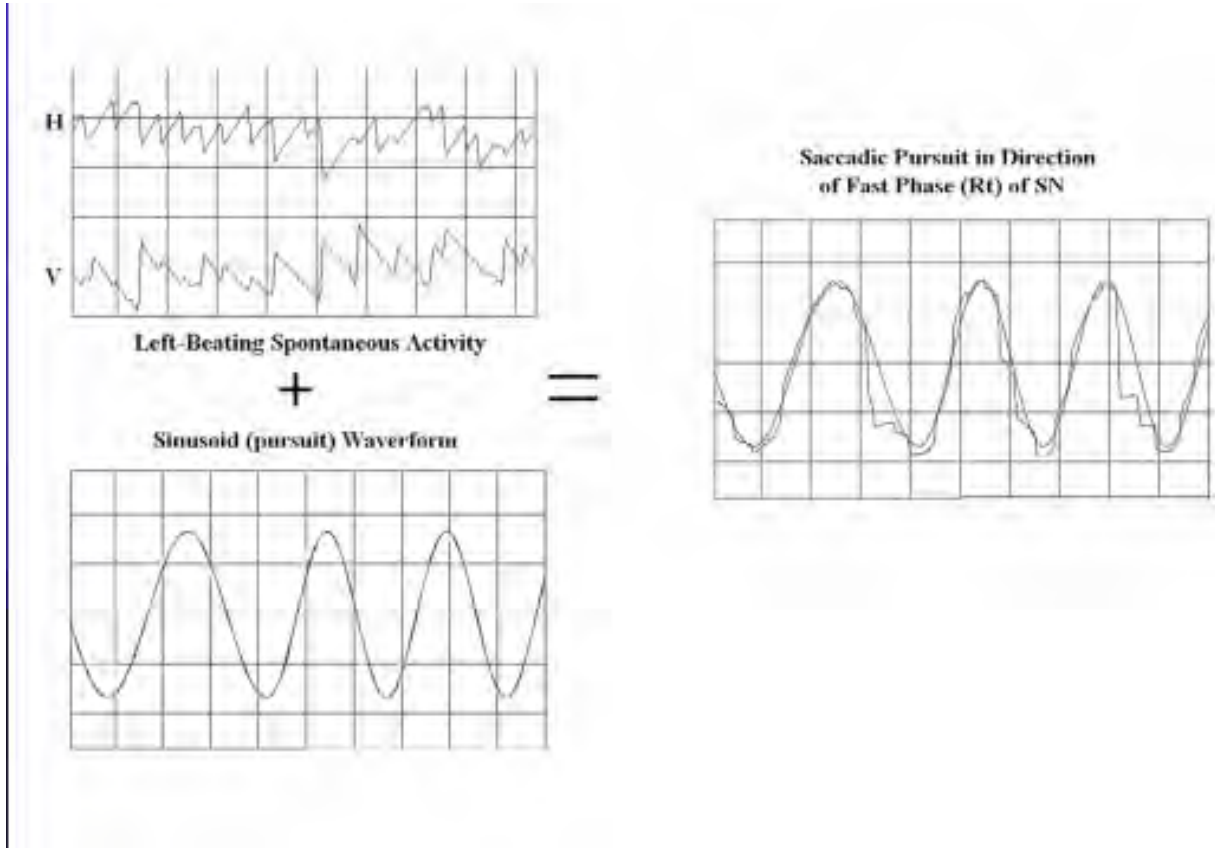
- Abnormalities:
  - Cog-wheeling/stair-stepping





Smooth Pursuit

- Can be influenced by spontaneous nystagmus:



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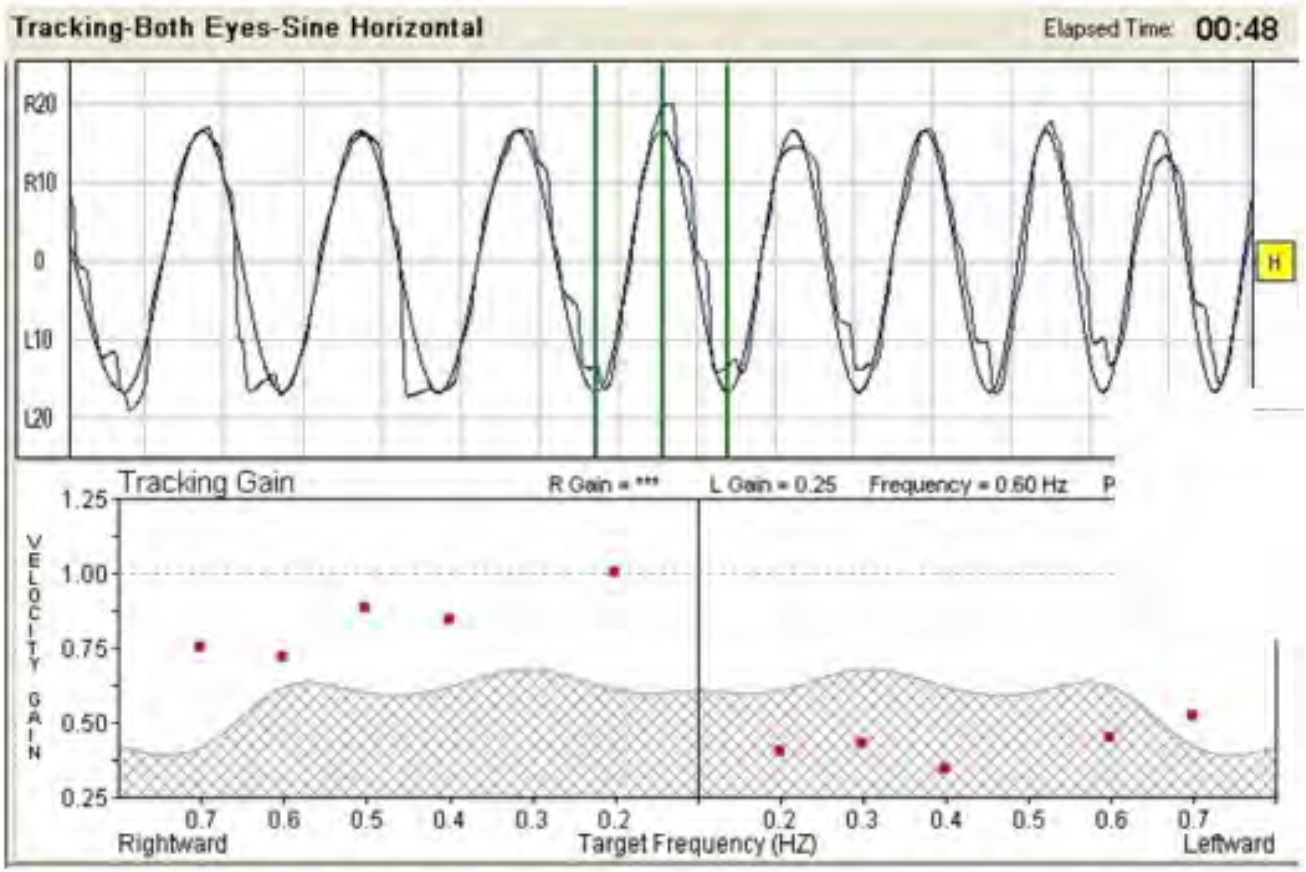
Smooth Pursuit



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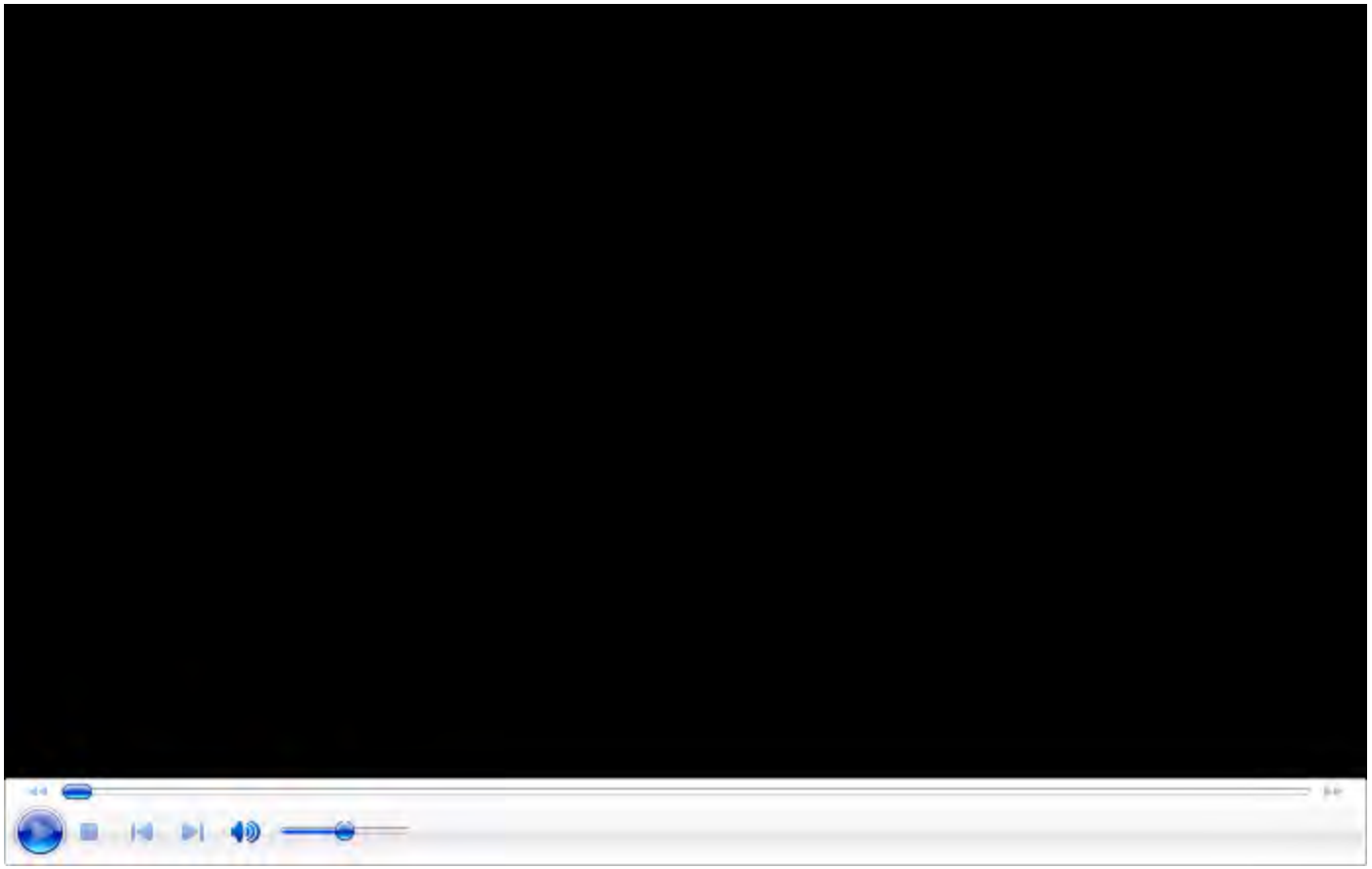
Smooth Pursuit



Optokinetic Testing (OPK)

- Creates nystagmus when following target
- Involves combination of neural systems responsible for smooth pursuit and following moving objects (OPK areas)
- Initiation of nystagmus from smooth pursuit system
- As target continues to move, central OPK system adds movement

Optokinetic Testing (OPK)

- Stimulus MUST fill 90% of visual field
- VNG/ENG systems that utilize a light bar are not really testing OPK
  - Smooth pursuit
  - Saccadic pursuit

Optokinetic Testing (OPK)

- Testing can be highly influenced by instruction
- "Keep your eyes in the middle and watch the dots as they pass by. Do not focus on any one dot."
- Patients can "focus through" stimulus
- Following one dot will just measure smooth pursuit

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Optokinetic Testing (OPK)

Optokinetic Testing (OPK)

- Analysis involves by calculating velocity gain of eye movements
- Peak eye velocity divided by target velocity

$$\text{OKN Velocity Gain} = \frac{\text{Peak eye velocity (degrees/sec)}}{\text{Peak target velocity (degrees/sec)}}$$

Optokinetic Testing (OPK)

- Interpretation:
  - This is the least sensitive of our oculomotor tests
  - If abnormal, but smooth pursuit is normal
    - Remember smooth pursuit is part of true OPK
    - Most likely result of uncompensated peripheral hypofunction
    - May be product of visual pathways (vision loss)
  - If OPK truly abnormal, should be seen with smooth pursuit and saccades (more sensitive measures)

Optokinetic Testing (OPK)

- Interpretation:
  - If OPK truly abnormal, should be seen with smooth pursuit and saccades (more sensitive measures)
  - OPK is actually dependant on overlapping neurological systems = smooth pursuit and saccadic pursuit
  - OPK gain decreases w/ age (like smooth pursuit)

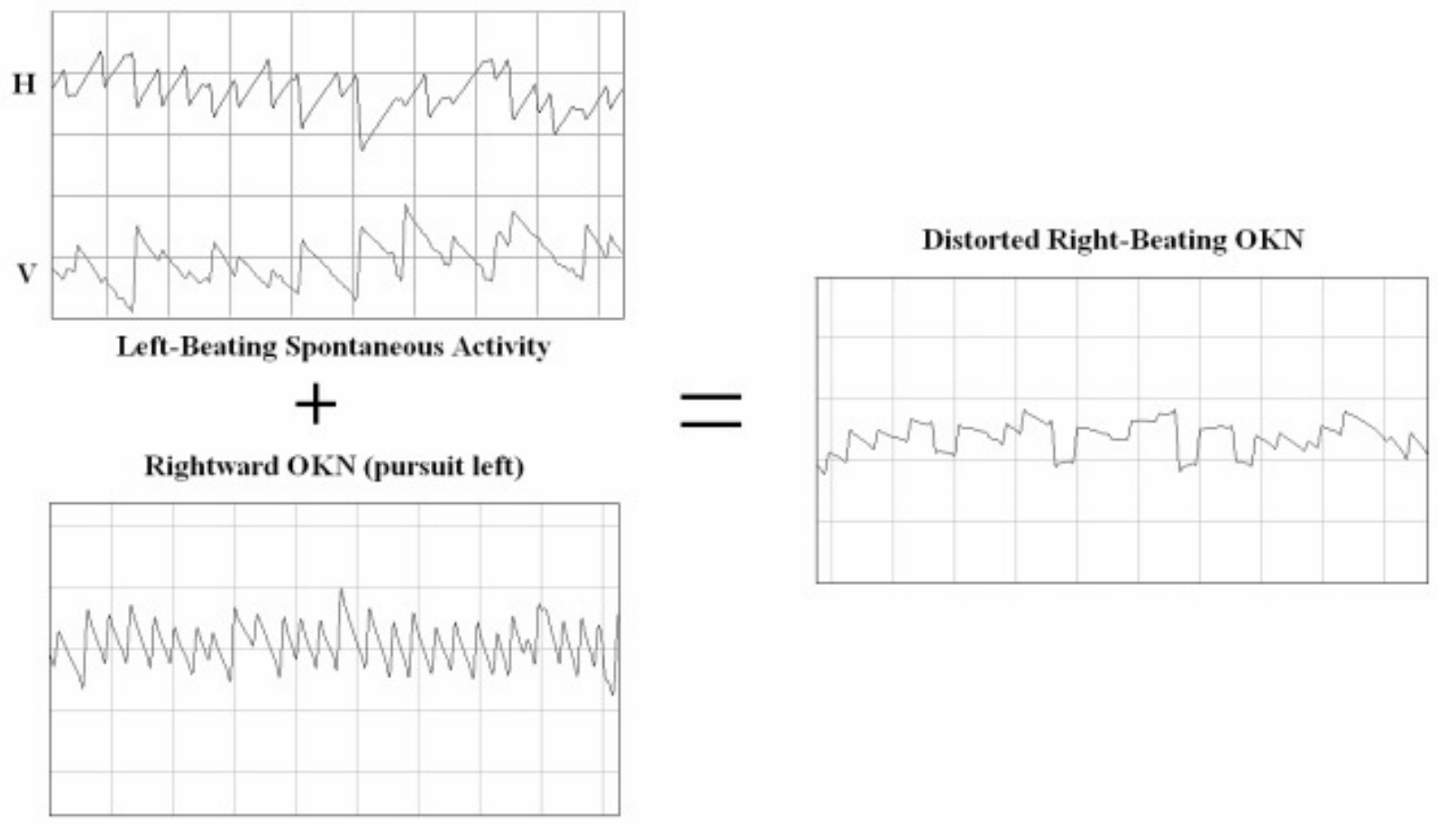
Optokinetic Testing (OPK)

- Interpretation:
  - If OPK normal, but smooth pursuit and saccadic pursuit abnormal = you have a problem (pt. effort???)
  - If OPK abnormal, but smooth pursuit and saccadic pursuit normal = do you care about OPK results???

Optokinetic Testing (OPK)

- Clinical utility:
  - Cross-checking better studies
  - Evaluating smooth pursuit in children, as it is hard to get infants to track a smooth pursuit stimulus
  - Evaluating central compensation???
  - Abnormal with stimulus movement in direction of lesioned ear???

Optokinetic Testing (OPK)



Gaze Testing

- Patient placed in dark (goggles closed) and asked to gaze:
  - Straight ahead
  - To right (30°)
  - To left (30°)
  - Up (30°)
- If nystagmus observed, open goggles and see if nystagmus persists

Gaze Testing

- If nystagmus seen, need to note the following:
  - Direction of nystagmus
    - Horizontal- right or left
    - Vertical- up down
  - Which gaze direction has nystagmus
  - Quality/magnitude of nystagmus

Gaze Testing - Peripheral

- Typically only observed w/ vision denied if acute
- Direction fixed
- Alexander's law
- Suppresses with vision
- Enhances dynamically
- Linear slow component
- Horizontal nystagmus (right/left beating)

Gaze Testing - Central

- With vision enabled (fixation) nystagmus persists
- Does not lessen over time (no central compensation)
- Direction fixed or changing
- Rarely in primary
- Vertical/rotary
- Enhanced with vision enabled
- Vertical nystagmus post HFHS
- Does not enhance dynamically

Gaze Testing - Central

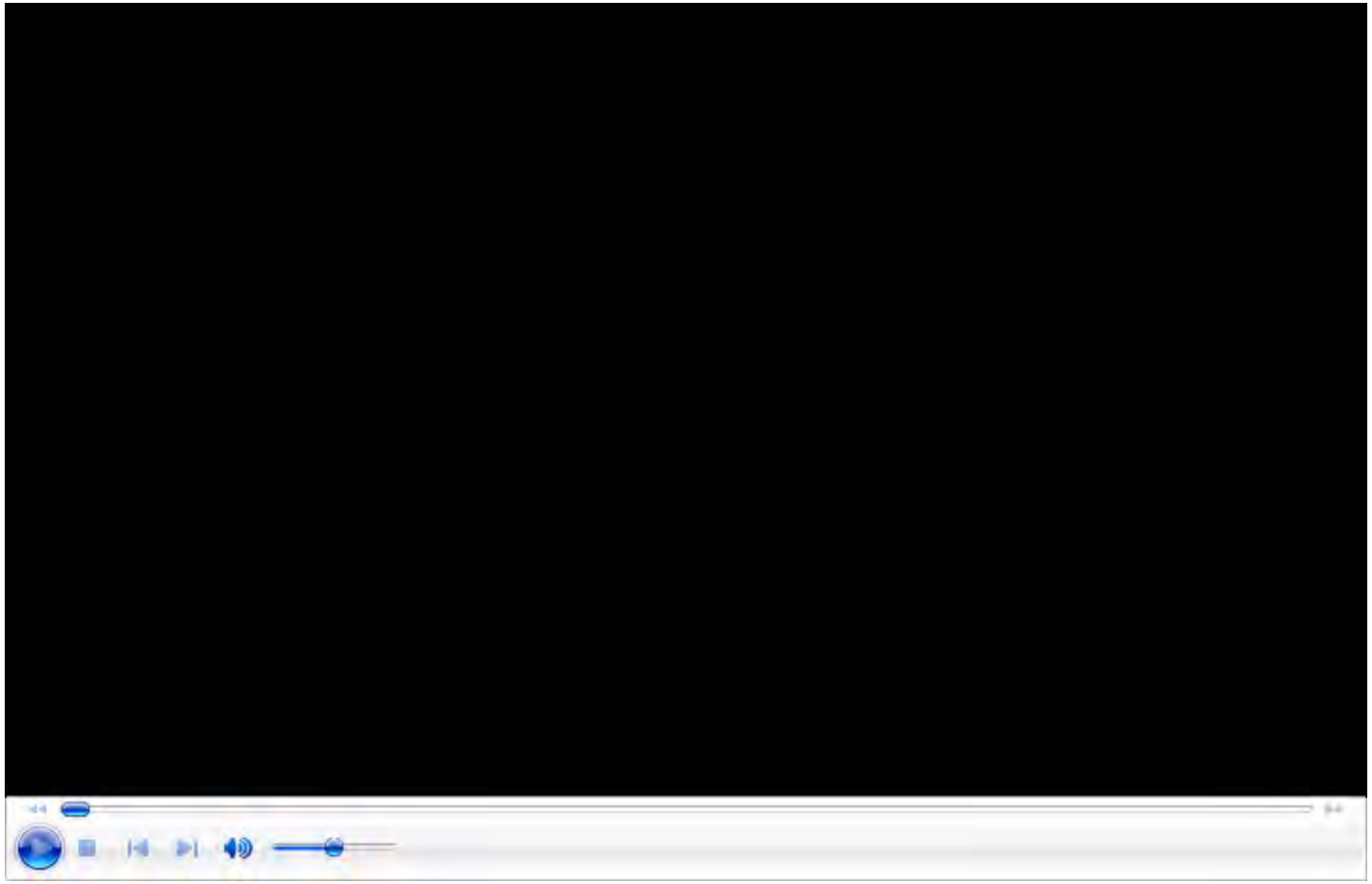
- If vertical and/or rotary during gaze testing, lesion should be considered central until proven otherwise
- Only known peripheral condition to cause pure vertical down-beating nystagmus = bilateral SCDS

Gaze Testing - Central

- Rebound nystagmus:
  - Nystagmus beats in direction of most recent eye movement after eyes returned to center
  - For example, after completing right gaze and eyes returned to center (moving left), a left-beating nystagmus is observed
  - If eyes held eccentrically for an extended period (~ 20+ sec.), normals may have a few (2-3) beats

Gaze Testing - Central

- Rebound nystagmus:



Gaze Testing - Central

- Vertical:
  - Down-beating nystagmus:
    - Craniocervical junction pathology
    - Arnold-Chiari malformation
    - Low posterior fossa
    - Vestibulocerebellum
  - Up-beating nystagmus:
    - Lower brainstem
    - Medullary regions

Gaze Testing - Central

- Vertical:
  - Important points-
    - Lesions in posterior fossa that produce purely down-beat nystagmus may not cause abnormal oculomotor tests (saccades and smooth pursuit)
    - If only persistent vertical nystagmus is noted, posterior fossa lesion should be ruled out

Gaze Testing - Central

- Rotary:
  - In primary gaze, lesion lateralized to the right or left in the pontomedullary/medullary area ipsilesional or above the pons contralesional
- Saccadic intrusions and oscillations
  - Square-wave jerks:
    - Slow saccadic movement during gaze
    - If occurs with vision enabled = cerebellar
    - If occurs with vision denied = normal

Nystagmus

Characteristic	Central	Peripheral
Fixation	**Does not suppress with vision**	**Suppresses with vision**
Direction	**-Vertical Nystagmus (upbeating or downbeating)** **-Rotary Nystagmus**	**-Horizontal Nystagmus** **-Fast phase towards intact ear (except Irritative or Recovery Nystagmus)** **-Rotary (BPPV or HC involvement)**
Positionals	**Direction Changing: Geotropic or Ageotropic**	**Direction Fixed (except with BPPV)**
Gaze	**Direction Changing in Neutral Position**	**Alexander's Law**
Dynamic	**Rare Enhancement**	**Frequent Enhancement**

Adapted from: Baloh, 1998; Baloh & Honrubia, 1990; Roberts & Gans, 2008

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Questions/Comments???

Let's take lunch
45 minutes

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Hearing Your Best for Life

VNG/ENG-Positional/Positioning Calorics

Samuel N. Bittel, Au.D.

Positional/Positioning

- Positional-
  - Recordings taken in a static position
  - Does patient have nystagmus in a specific position without movement
- Positioning-
  - Patient moved into a specific position
  - Does changing position create dizziness/vertigo and/or nystagmus

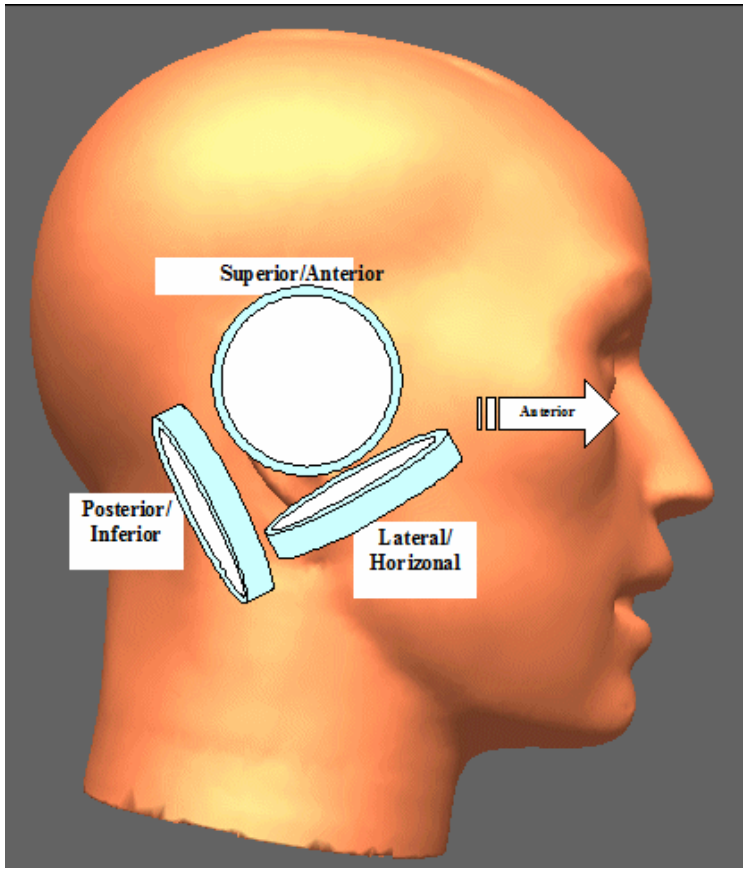
Positional Testing

- Patient placed in several positions:
  - Supine
  - Head/body right
  - Head/body left
- Nystagmus recorded via VNG goggles
- Direction, intensity, and quality of nystagmus recorded
- Compare to neutral position (seated)

Positional Testing

- Should start with vision denied
- Supine position:
  - Patient's head should be raised to 30°
  - Horizontal canal should be perpendicular to gravity
  - Support patient's head and cervical spine
  - If nystagmus observed, open goggles

Positional Testing



Positional Testing

- Head right/left
  - Head should be turned parallel to gravity
  - If patient can not turn head 90° elevate should slightly
  - Start w/ vision denied, if nystagmus observed you should open goggles. Does nystagmus suppress?

Positional Testing

- Body right/left
  - If nystagmus seen in head position, you should place patient in lateral position
  - Again, start vision denied and open goggles if nystagmus observed
  - Head vs. body position = different based on neck hyperextension, better head placement

Positional Testing

- Recording should be completed for long enough to get good understanding of nystagmus
- Need to note:
  - What nystagmus looks like (type/quality)
  - What direction does nystagmus beat
  - How big is nystagmus
  - Does nystagmus suppress with vision

Positional Testing

- Interpretation:
  - Geotropic = beats towards ground
  - Ageotropic = beats towards the sky
  - Vertical up-beating = beats towards eyebrows
  - Vertical down-beating = beats towards nose

Positional Testing

- Interpretation:
  - Peripheral hallmarks
    - Direction fixed
    - Suppressed with vision
    - Enhanced dynamically
    - Horizontal (in relation to eyes)
- Interpretation:
  - Peripheral hallmarks
    - Usually beats away from affected ear
    - Unless irritative lesion or recovery nystagmus
      - Recovery nystagmus = after recovery for a temporary lesion, CNS over compensates

Positional Testing

- Interpretation:
  - With peripheral, nystagmus will often be larger w/ affected ear undermost
  - Lateral HFHS
    - More sensitive than seated headshake
    - W/ peripheral, nystagmus may enhance (or appear)
    - Again, larger with affected ear undermost

Positional Testing

- Interpretation:
  - Central hallmarks
    - Direction changing
    - Remains or enhances with vision
    - Does not enhance dynamically
    - Vertical (in relation to eyes)

Positional Testing

- Interpretation:
  - If you have geotropic/ageotropic nystagmus in each lateral position, is this direction fixed or direction changing?
  - Remember, direction fixed is in reference to the ear the nystagmus is beating towards…

Positional Testing

- So, if you see nystagmus:
  - Open goggles and look for suppression
  - If in head right/left, check lateral positions
  - Perform HFHS to see if it enhances
  - How does it compare to gaze testing
    - Direction changing can be gaze vs. positionals

Positional Testing

- Central nystagmus:
  - Age-related
  - Alcohol/drug related
  - VBI
  - Migraine
  - Other central vestibular lesions

Positional Testing

- Central nystagmus – Age related
  - Not atypical to see geotropic nystagmus in the absence of pathology
  - Age-related changes to cerebellum
  - Do not over-interpret in patients over 70
  - If suppresses w/ vision in elderly patient = may not be clinically significant

Positional Testing

- Central nystagmus – Alcohol/Drug
  - Alcohol can cause abnormal positional nystagmus; patients should refrain for 48 hrs. = positional alcohol nystagmus (PAN)
  - Central-acting medications can also cause abnormal nystagmus
  - Nicotine can cause abnormal nystagmus

Positional Testing

- Migraine
  - A certain amount of central-type nystagmus has been reported in individuals with migraine
  - This nystagmus is variable, but will typically follow a central pattern
  - Can be horizontal, vertical, rotary
  - Can be transient, persistent, etc.

Positional Testing

- Vertical nystagmus:
  - Indicates damage to cerebellum, Arnold-Chiari malformation, MS, VBI, and pharm
  - If patient with BPPV gazed away from involved ear (when provoked), the nystagmus may appear more vertical due to enhancement of contra rectus muscle
  - Was this seen during gaze testing?

Positional Testing

- Other central/pharm indicators:
  - Nystagmus does not fatigue (BPPV should fatigue in less than 1 minute)
  - No associated vertigo
  - Remember, you can also see horizontal nystagmus w/ CNS issues- make sure to correlate with Hx/Sx and other findings

Positional Testing

- Can a certain degree of positional nystagmus be considered normal?
- Wide variation in the literature
  - Some of suggested that less than 5 deg/sec is not clinically significant
- Research by Roberts, Bittel, and Gans shows lower rates than the general literature (explained by path)

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Positioning

Positioning Testing

- Dix-Hallpike
  - Moving patient's ear into a position that would provoke BPPV
  - Multiple variations:
    - Modified Hallpike
    - Fully-supported Hallpike
    - Side-lying Hallpike
- Complete VAST prior to Hallpike!!!

Positioning Testing

- Modified Hallpike
  - Patient sits with feet up table w/ back to clinician
  - Patient positioned back far enough that head will be off end of table
  - Clinician stand behind patient and supports head
  - Patients head turned 45° towards test ear
  - Patient placed in supine position

Positioning Testing

- Modified Hallpike
  - Patient's head hyper-extended over edge of table
  - Position held for approximately 1 minute
  - Watch up-most eye for nystagmus
  - Watch capillaries in sclera for movement
  - Return head to neutral before sitting up
  - Guide patient up (not by neck/head)

Positioning Testing

- Modified Hallpike
  - If nystagmus observed, note direction, duration, and type/quality
  - If positive for BPPV, patient may have vertigo/nystagmus when sitting back up

Positioning Testing

- Hallpike pearls
  - Pinna oriented in similar plane to posterior canal = can visualize what you are attempting to do
  - KEEP FINGERS TOGETHER WHEN SUPPORTING HEAD

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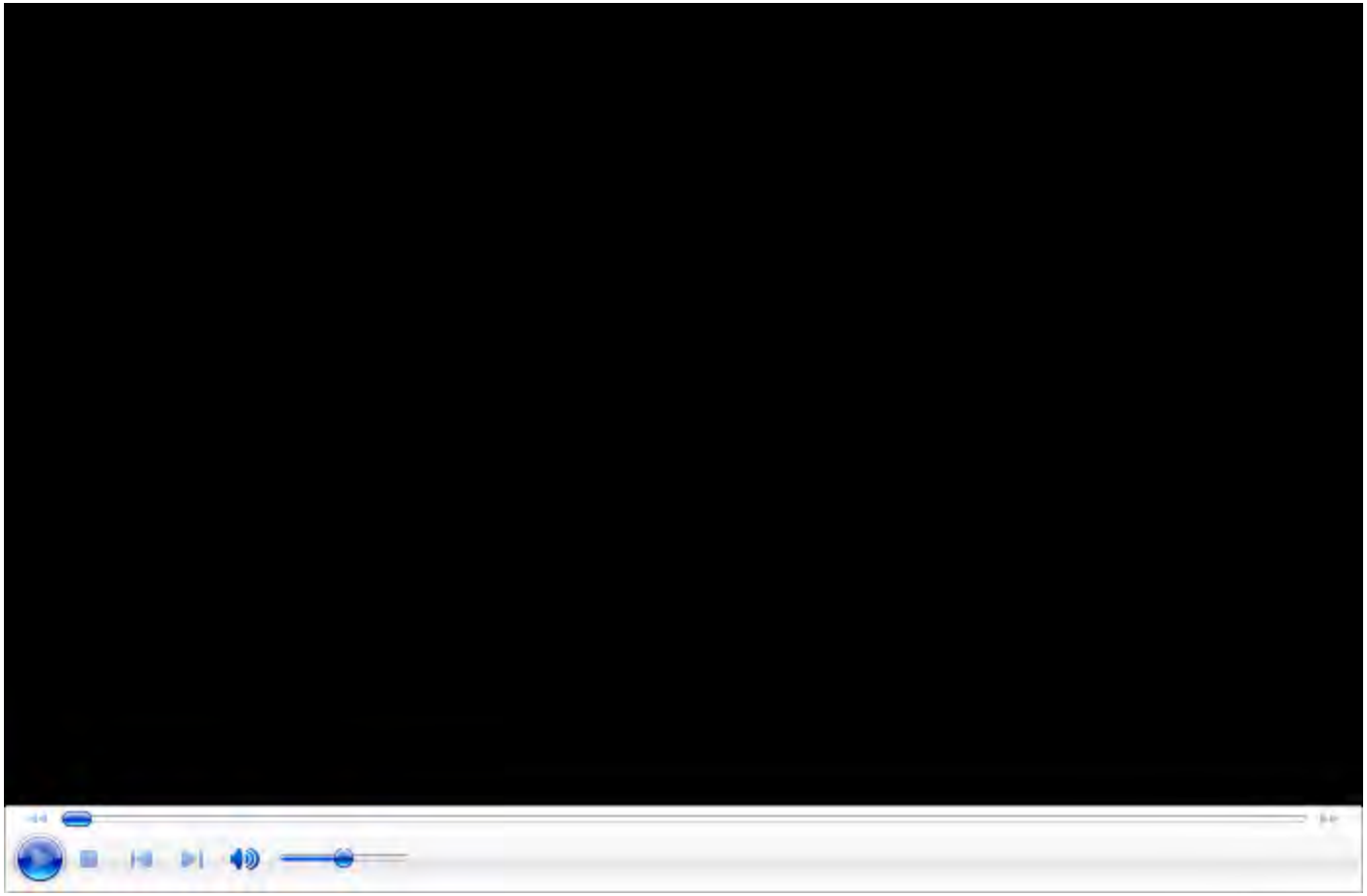
Positioning Testing Modified Hallpike



Positioning Testing

- Fully-supported Hallpike
  - Useful w/ cervical spine or vertebral artery issues
  - Patient's head not hyperextended off of edge of table
  - Head still turned 45° towards test ear
  - Will this provoked BPPV???

Positioning Testing

- Side-lying Hallpike
  - Useful for back pain, neck issues, and compromised vertebral artery
  - Patient placed on side of table
  - Head turned away from test ear
  - Place patient on side w/ test ear undermost
  - Legs guided up onto table

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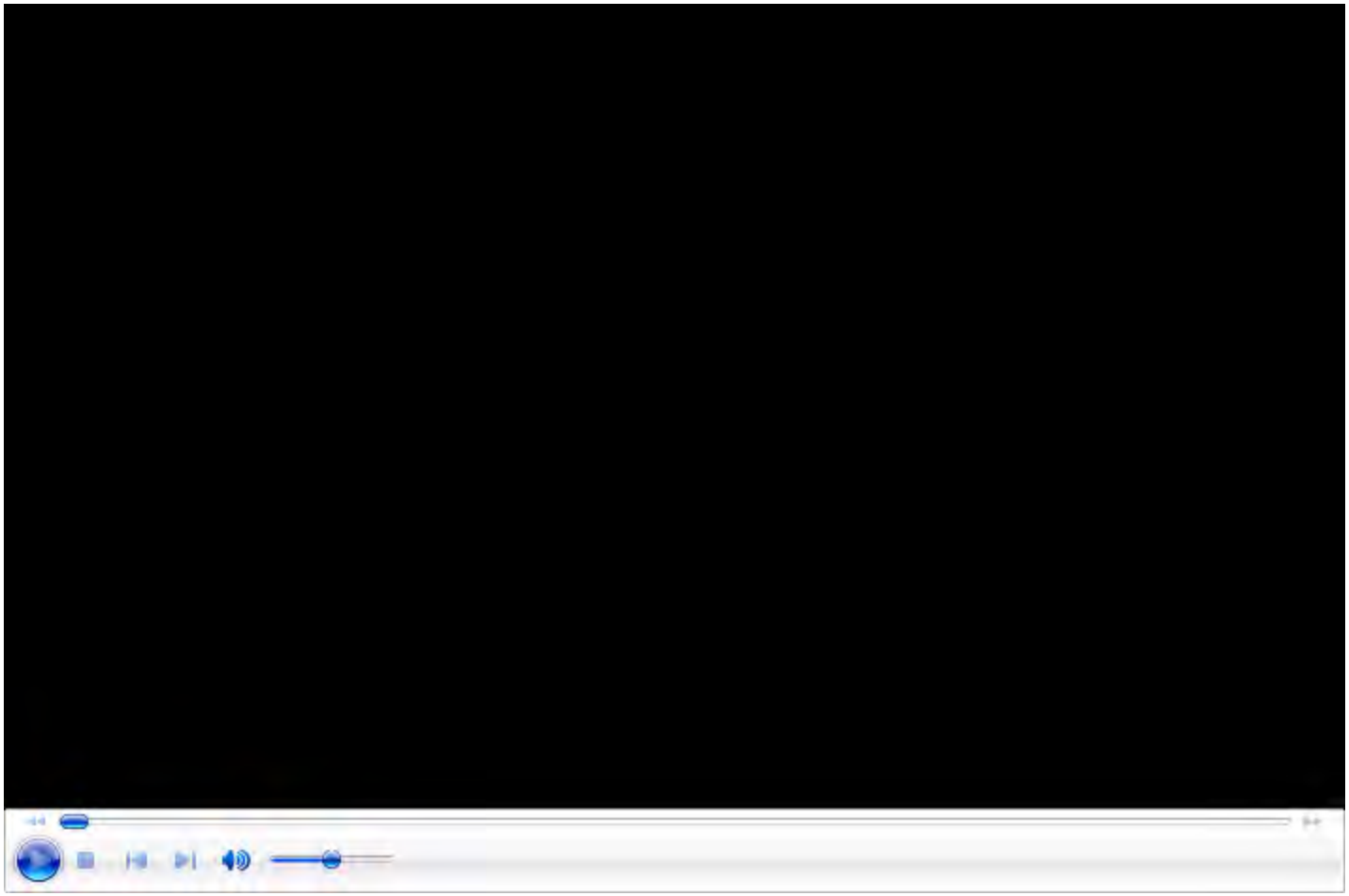
Positioning Testing
Side-lying Hallpike



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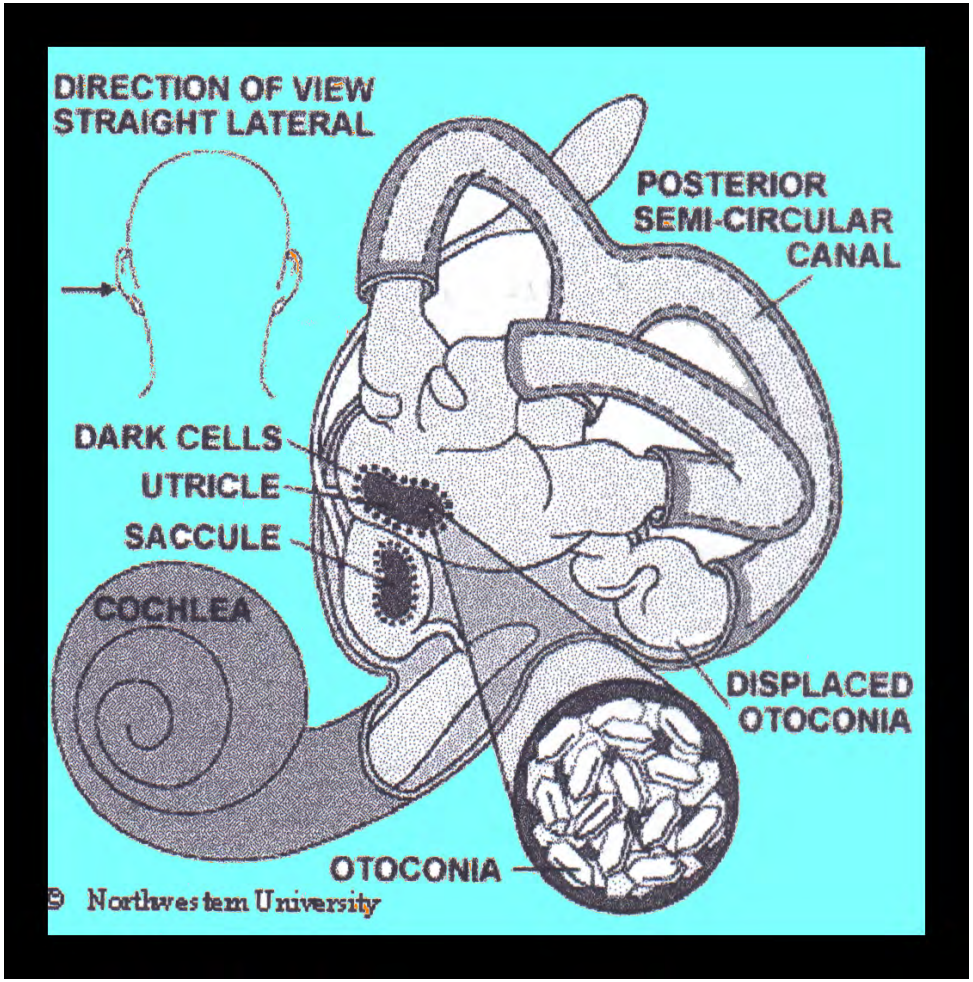
Positioning Testing Interpretation BPPV

Localization

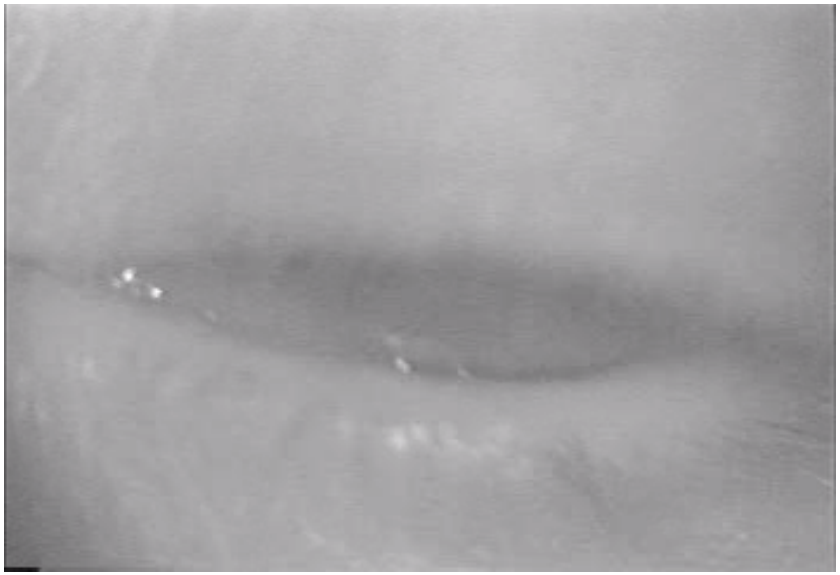
- Posterior canal
  - Rotary (upward) geotropic nystagmus
  - Affected ear down
- Anterior canal
  - Rotary (upward) ageotropic nystagmus
  - Affected ear up
- Horizontal canal
  - Horizontal geotropic/ageotropic nystagmus
  - Affected ear up or down



-BPPV is a mechanical phenomenon: hallmarks can easily be explained

-Tx involves moving otoconia from SCC to utricle

Rotary Nystagmus



- This must be BPPV of the left posterior SCC
- Based on left posterior SCC's connection to extra-ocular muscles

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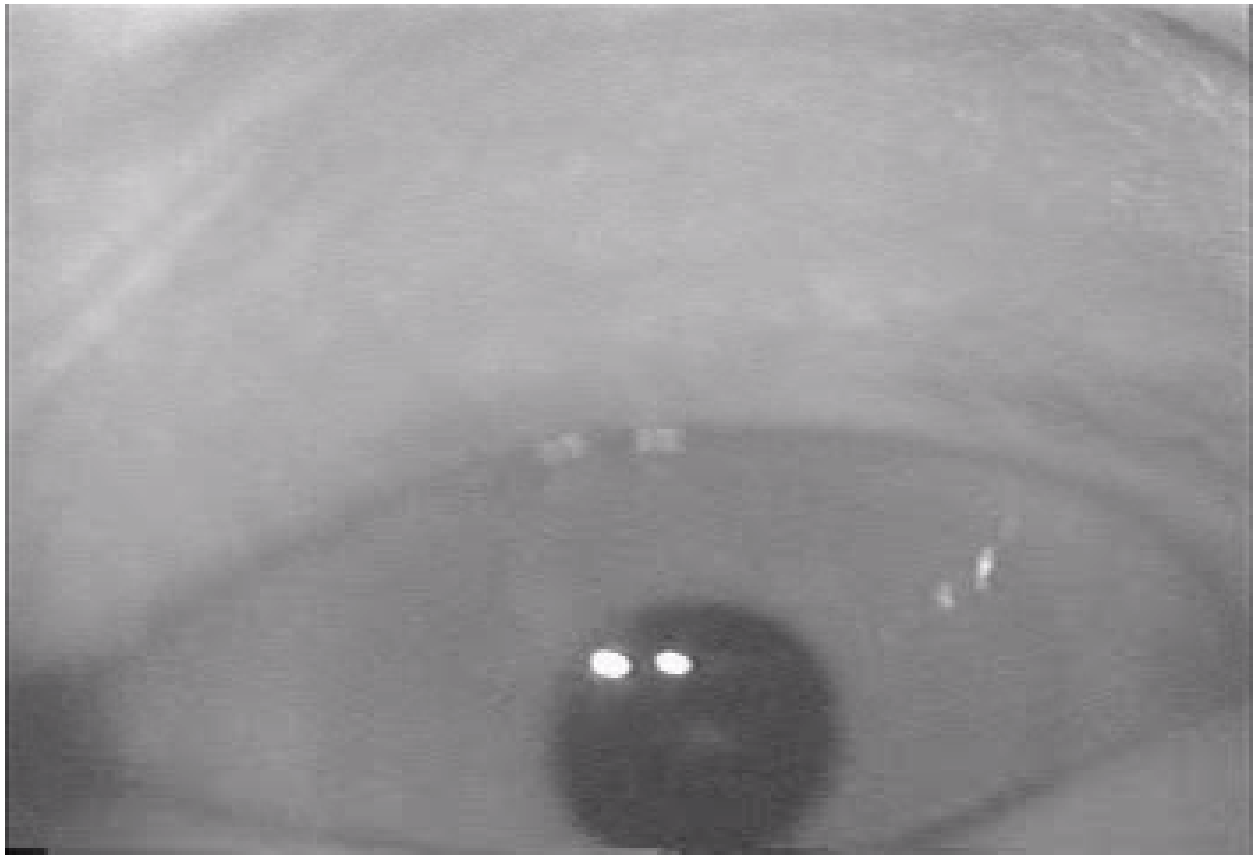
HC-BPPV



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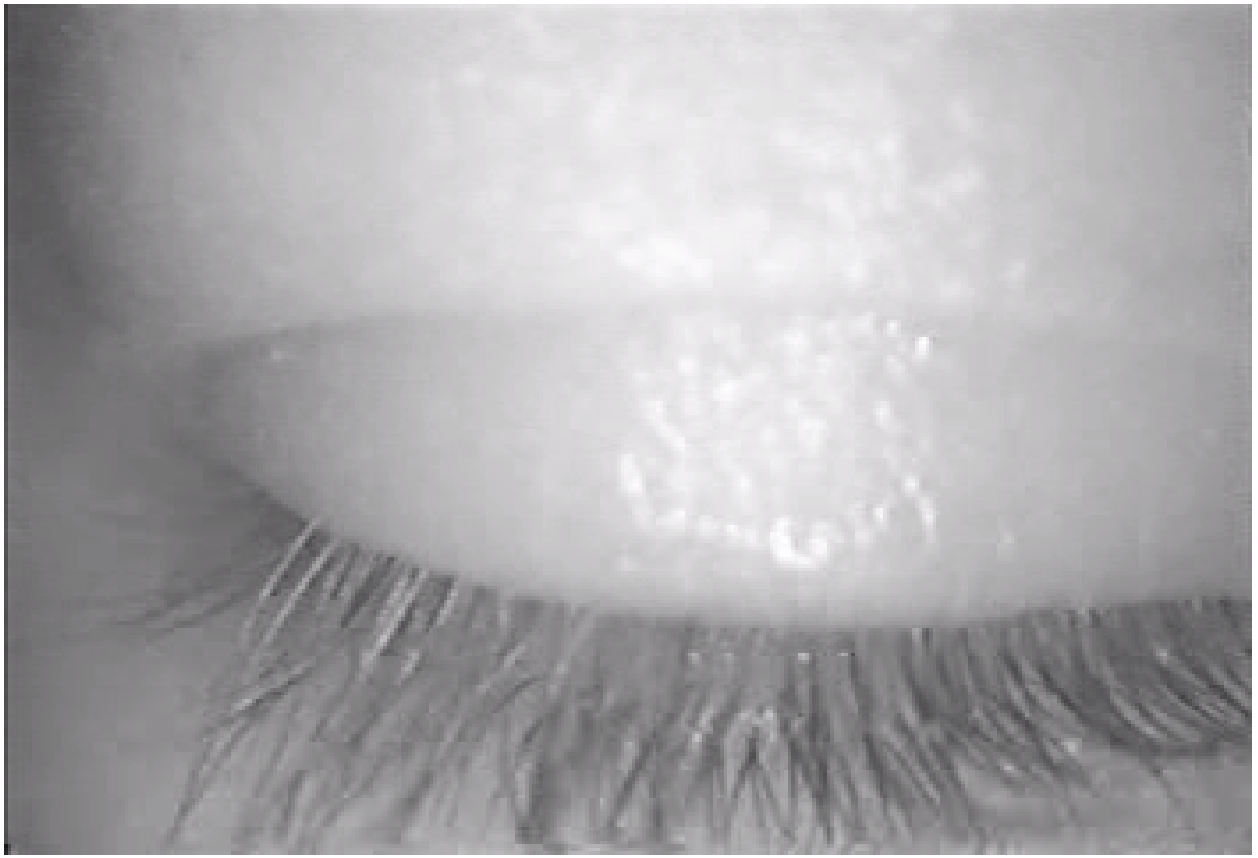
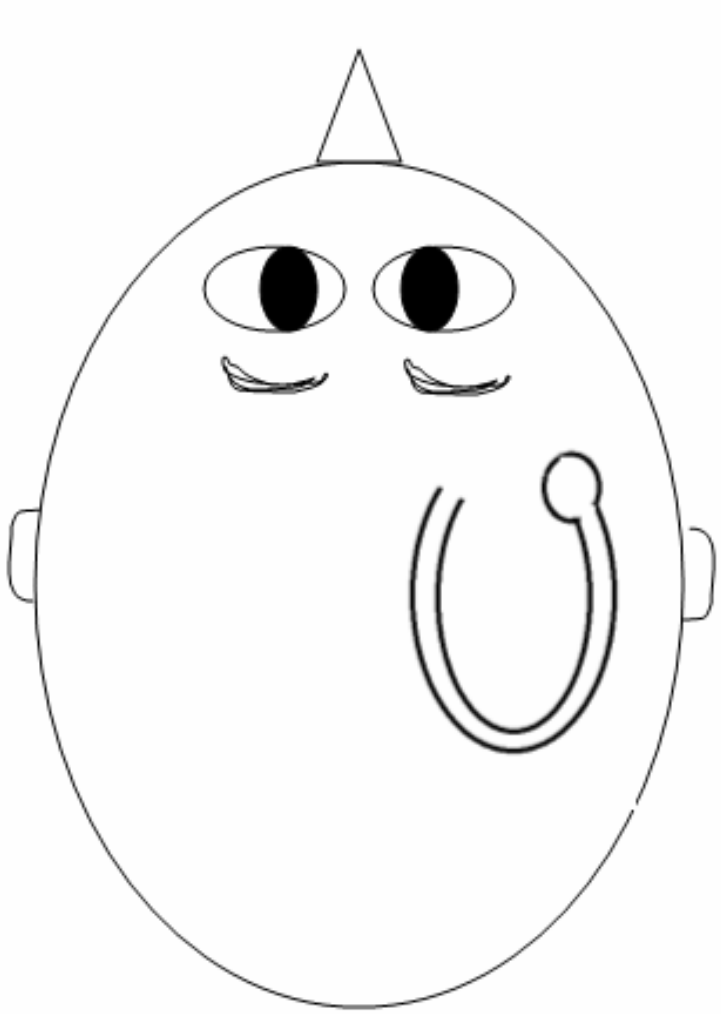
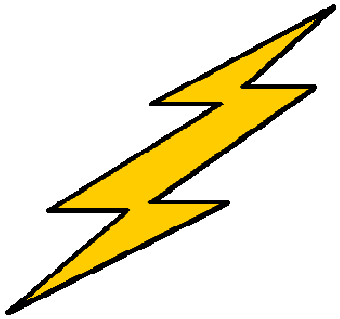
AC-BPPV



Localizing HC

- Geotropic nystagmus
  - Canalithiasis
  - Side w/ more intense nystagmus = involved
- Ageotropic nystagmus
  - Cupulolithiasis
  - Side w/ weaker nystagmus = involved

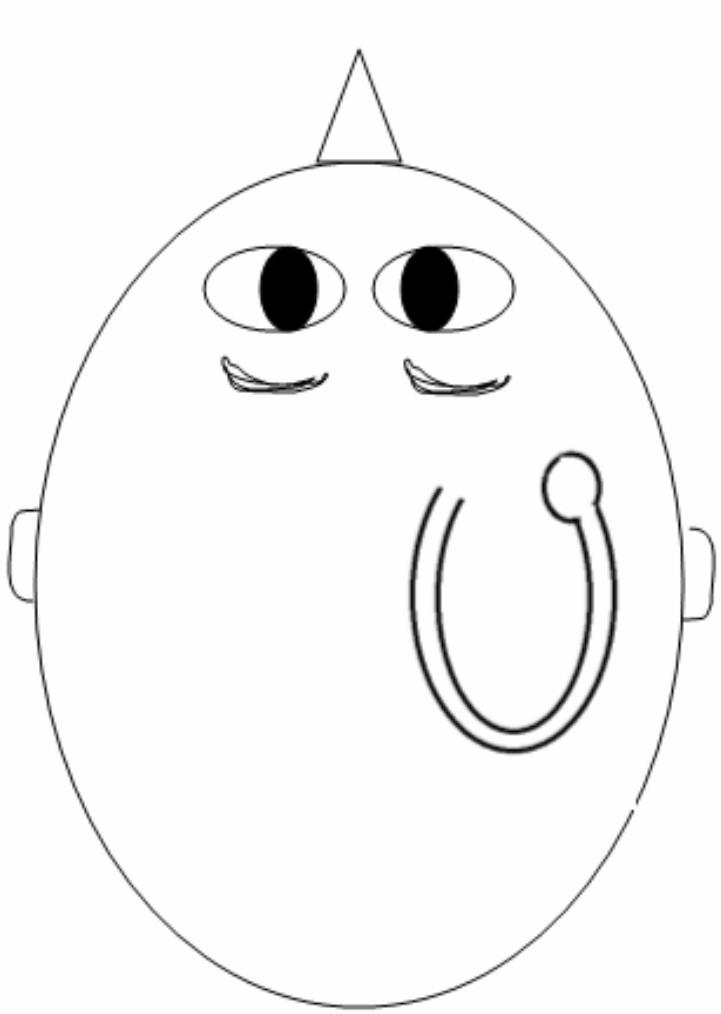


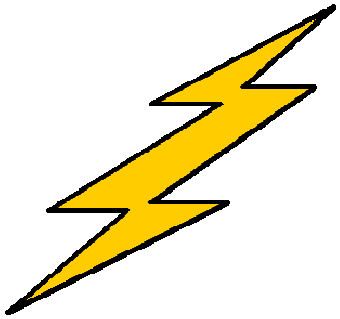


→Utriculopetal = stimulatory for HC

→Utriculofugal = inhibitory for HC

→Ewald's 2nd law = excitation stronger than inhibition





→Utriculopetal = stimulatory for HC

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→Ewald's 2nd law = excitation stronger than inhibition

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Caloric Testing

Caloric Testing

- The last test in the VNG
- This test is considered the gold standard for identifying unilateral vestibular dysfunction
- Allows independent and individual measurement of each side

Caloric Testing

- Water/air introduced into EAM
- By changing temperature of EAM, a temperature change is created in the middle ear space and across the ossicles
- After long enough, the endolymph in the HC will begin to change temp

Caloric Testing

- Warmer endolymph rises towards cupula = utriculopetal (stimulates)
- Cooler endolymph sinks away from cupula = utriculofugal (inhibits)
- Measure VOR response and calculate nystagmus

Caloric Testing

- COWS:
  - Warm is stimulatory
  - Cool is inhibitory
  - Clinical correlate:
    - Vestibular lesion
    - Head/body rotation
  - Warm caloric often larger than cool
    - Why???

Caloric Testing

- Reflects horizontal canal and superior vestibular nerve
- Remember promontory (from HC) protrudes into middle ear
- Should be last test in your battery:
  - Can influence other subtests…

Caloric Testing

- Technique:
  - Patient placed supine with head elevated 30°
  - HC perpendicular to gravity
  - Eyes recorded with vision denied
  - Three methods:
    - Open-loop water
    - Closed-loop water
    - Air

Caloric Testing

- Prior to completing caloric:
  - Otoscopy to evaluate shape/size of EAM, status of TM, cerumen, etc.
  - Remove cerumen
  - Explain test procedure to patient
    - Be careful with verbiage
    - Explain possible dizziness, but do not say anything that can lead to nausea/emesis
    - Explain response is controlled and reversible
    - Let patient feel air on hand

Caloric Testing

- Interpretation:
  - Factors that can by analyzed
    - Unilateral weakness- right ear vs. left ear
    - Directional preponderance- right beating nystagmus vs. left beating nystagmus
    - Visual fixation
    - Frequency of nystagmus

Caloric Testing

- Interpretation:
  - Largest beat of nystagmus is identified for each irrigation
  - Compare warm/cool calorics between ears (unilateral weakness)
    - AS cool and warm vs. AD cool and warm
  - Compare right beating vs. left beating nystagmus (directional preponderance)
    - AS cool and AD warm vs. AS warm and AD cool

Caloric Testing

- Unilateral weakness:
  - Looking for vestibular hypofunction
  - Reflects unilateral reduction in function
  - Permanent damage
  - Has nothing to do with central compensation

Caloric Testing

- Unilateral weakness calculation:

$$\frac{(\text{AS warm} + \text{AS cool}) - (\text{AD warm} + \text{AD cool})}{(\text{AS warm} + \text{AS cool} + \text{AD warm} + \text{AD cool})} =$$

Unilateral Weakness

Caloric Testing

- Unilateral weakness:
  - For clinical significance, the literature reports a range
  - Typically varies from 20% to 25%
  - My clinic uses greater than 23% as significant
  - Significant for a reduction in function on one side…

Caloric Testing

- Directional preponderance:
  - Comparing how well the eyes beat right vs. left
  - The literature is all over the place with the clinical significance of this test
  - Often times the product of user error (outlier)
  - If good technique, no error, and spontaneous nystagmus factored in, this should be considered a central finding

Caloric Testing

- Directional Preponderance Calculation:

$$\frac{(\text{AS warm} + \text{AD cool}) - (\text{AD warm} + \text{AS cool})}{(\text{AS warm} + \text{AS cool} + \text{AD warm} + \text{AD cool})} = \text{Directional Preponderance}$$

Caloric Testing

- Directional Preponderance:
  - For clinical significance, the literature reports a range
  - My clinic uses greater than 30% as significant
  - MUST BE CAREFUL THAT THIS IS A TRUE FINDING (more later)

Caloric Testing

- Interpretation:
  - Visual fixation-
    - Remember, calorics are completed with vision denied
    - If vision is not denied, there will be very reduced nystagmus
    - After a caloric, we enable vision and calculate the reduction in nystagmus
    - Nystagmus should reduce by 50% in approximately 10 seconds

Caloric Testing

- Interpretation:
  - Factoring for spontaneous pre-existing nystagmus:
    - You must adjust calculations for pre-existing nystagmus
    - For example, a pre-existing right-beating nystagmus will make all right-beating calorics look bigger and all left-beating calorics look smaller
    - Will create an erroneous DP, but not UW

Caloric Testing

- Does the order of your calorics matter?
  - In one word, NO
  - The order of your calorics does not matter
  - We often start with warm (larger response)
  - We will start in weaker ear (presumed)
  - If patient becomes emetic, we want to have as much information as possible
    - Weaker ear = smaller response
    - Warm = ear acting at "its best)

Caloric Testing

- How long should you wait between calorics?
  - I was taught at least 5 minutes
  - However, you need to wait until stimulation from last caloric is gone prior to moving on to next
  - Put the patient back in the dark and watch for nystagmus
  - If nystagmus gone = precede

Caloric Testing

- Do I need to complete all 4 calorics?
  - You need enough data to compare sides
  - You can complete monothermal calorics
    - If all other testing appears normal
    - If you do not expect a weakness
    - If both calorics are robust and symmetrical
    - In my clinic, I will only complete monothermal if response over 11 deg/sec and within 1-2 degrees (between ears)

Caloric Testing

- Do I need to task my patient?
  - In one word, YES
  - The nystagmus from calorics is enhanced if the patient is mentally tasked
  - Should use tasks that require the patient to actively use memory
  - Have conversation or ask to list names, places, etc.

Caloric Testing

- Can a caloric response be too big?
  - You can have hyperactive responses
  - Total right ear responses greater than 140 deg/sec AND total left ear responses greater than 140 deg/sec
  - May reflect cerebellar lesion
  - Cerebellums inability to "clamp down" on response

Caloric Testing

- Can all caloric responses be too small?
  - You can have a bilateral vestibulopathy
  - Caused by ototoxic meds, mid-line cerebellar stroke, poor health (diabetes, cardiovascular disease), trauma, and sometimes age
  - Total right ear responses less than 12 deg/sec AND total left ear response less than 12 deg/sec

Caloric Testing

- Bilateral vestibulopathy
  - Should be correlated with other clinical tests:
    - Would expect fall on SOP condition #6
    - Oscillopsia on VAT, CD-VAT
    - Should correlate with mechanical torsion chair
    - MAKE SURE NOT TECHNIQUE OR PHARM
  - If a patient has a bilateral weakness, you cannot define directional preponderance accurately (leave it out of report, etc)

Caloric Testing

- What if your calorics beat the wrong way?
  - First, check to see if EAM is damp; if EAM wet, warm air can actually cool; do not irrigate ear during cerumenectomy
  - Look at status of TM; you can occasionally see reversal with perf.
  - Factor in/out pre-existing nystagmus

Caloric Testing

- What if your calorics beat the wrong way?
  - Called caloric inversion if true response
  - Can reflect lesion to posterior fossa
  - Should rule out other factors before considering this a true response
  - Out of the thousands of calorics I have completed (~15,000), I have only truly seen this one time

Caloric Testing

- What if your calorics beat in the vertical plane?
  - First check patient's head position = should be straight or might stimulate other canals
  - If vertical nystagmus greater than horizontal, you might have vestibular nuclei lesion
  - A certain amount of vertical nystagmus is fairly typical, so an abnormality should either be only vertical nystagmus or vertical > horizontal nystagmus

Caloric Testing

- What if you have no caloric response?
  - If you truly expect no function (as per history and other test findings), you need to verify
    - Complete rotary chair
    - Complete ice calorics

Caloric Testing

- Other technical considerations
  - Watch for outliers
  - Although warm calorics are often larger than cool calorics, you would expect consistency between ears
  - The first caloric can be erroneously large
    - Remember, state of patient arousal influences response. If patient surprised by hallucination of vertigo, they may have higher state of arousal

Caloric Testing

- Watch for outliers
  - Having a single really large/small caloric can throw of your calculation
  - Directional preponderance and unilateral weakness = technical error
  - If you have an outlier, that caloric must be repeated
  - If you do not have consistency between runs, repeat!!!

Caloric Testing

- Other technical considerations
  - Vertigo can cause nausea/emesis
  - You must have a plan in place if a patient becomes emetic (this is true for all vestibular patients)
  - Remember proper infection control
  - The brain does not do cool and emetic well together = ice down your nauseated patients
  - Stay calm and supportive

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Hearing Your Best for Life

Integrating Test Results

Samuel N. Bittel, Au.D.

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Case Studies

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Patient R.H.

Patient R.H.

- 62 y/o female self referred (no PCP)
- Unremarkable medical history
- No current medications
- 2-month hx of general dysequilibrium, veering when walking, general lightheadedness, bilateral aural fullness
- Visual aura = ophthalmic migraine dx by optometrist
- Left eye ptosis = began several yrs ago (maybe)

Patient R.H.

- Was evaluated at walk-in clinic by internist several weeks ago for sinus infection = told to schedule formal appt. for ptosis (possible stroke)
- Optometrist and patient think ptosis related to contact lenses
- No current vertigo and/or positional provocation
- Issues w/ depth perception & ambulating in dark environments
- No recent MRI
- No auditory symptoms

Patient R.H.

Ptosis (párpado caído)

A.D.A.M.

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What Questions Would You Ask????

Patient R.H.

- If the ptosis is from your hard contact lenses, was there a recent change?
  - No, same contacts for 20+ years
- Have you had any eye irritation?
  - No
- Was there a preceding event?
  - No
- Have you EVER had vertigo?
  - No
- Have you had a recent medical evaluation?
  - No

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What Tests Would You Perform???

Patient R.H.

- SOP
  - Normal = no dysequilibrium
- ABR
  - Prolonged conduction times at 21.1 clicks/sec AU
  - Prolonged conduction times and poor morphology at 77.7 clicks/second AU
- Audio
  - High-frequency SNHL AU
- Immittance
  - Normal tymps
  - Contralateral reflexes raised – absent AU

Patient R.H.

- VAST
  - Negative
- CD-VAT
  - No movement = 100%
  - Horizontal movement = 48%
  - Vertical movement = 100%
- VEMP
  - WNL

Patient R.H.

- VNG
  - Occulmotors:
    - Saccades, OPKs, and Smooth Pursuit abnormal
  - Gaze, Positionals, HFHS, Hallpikes = WNL
  - Calorics:
    - Right total = 8 deg/sec
    - Left total = 7 deg/sec
- Now What????

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What Is Your Preliminary Diagnosis and Recommendations?

Patient R.H.

- My Diagnosis
  - Based on abnormal video-oculography, raised to absent contralateral stapedial reflexes, CNS-type issue can be ruled out
  - Bilaterally reduced calorics and oscillopsia on CD-VAT most likely related to neuro-conductive or brainstem issue (remember, calorics and oscillopsia can occur with central issues)
  - When clinical findings are correlated with left-eye ptosis, general dizziness w/o true vertigo, and general dysequilibrium, a neurology consult and/or MRI are HIGHLY suggested

Patient R.H.

- What happened…
  - R.H. thought I was over-reacting and did not want to take my recommendations seriously
  - I was very forthright with my concerns
  - I spoke with her 5 times over the course of one week and finally convinced her to get a primary care doctor
  - She spoke with her optometrist who disagreed with me about ptosis being a red flag
  - Patient schedule appt. w/ new PCP 7 days after seeing me = I called PCP prior to her appt. and informed him of my concerns and need for immediate MRI and R.H.'s reluctance

Patient R.H.

- Final Outcomes:

- Did not hear back from R.H., but received a records request from a local ENT group

- 5 weeks after appointment w/ me, R.H.'s obituary came through our fax machine (we track obituaries for our patients)

- R.H. had metastatic stage IV breast cancer, w/ extensive tumor growth within brain, brainstem

Patient R.H.

- Take home messages:
  - You need to take your CNS tests very seriously
  - When completing vestibular work, you need to consider other pathologies
  - You must pay attention to subtle details
  - As a vestibular audiologist, you must take the time to call patients and make specific recommendations to patient's physician (by telephone)
  - Dizziness alone may be a red flag!!!

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Patient I.F.

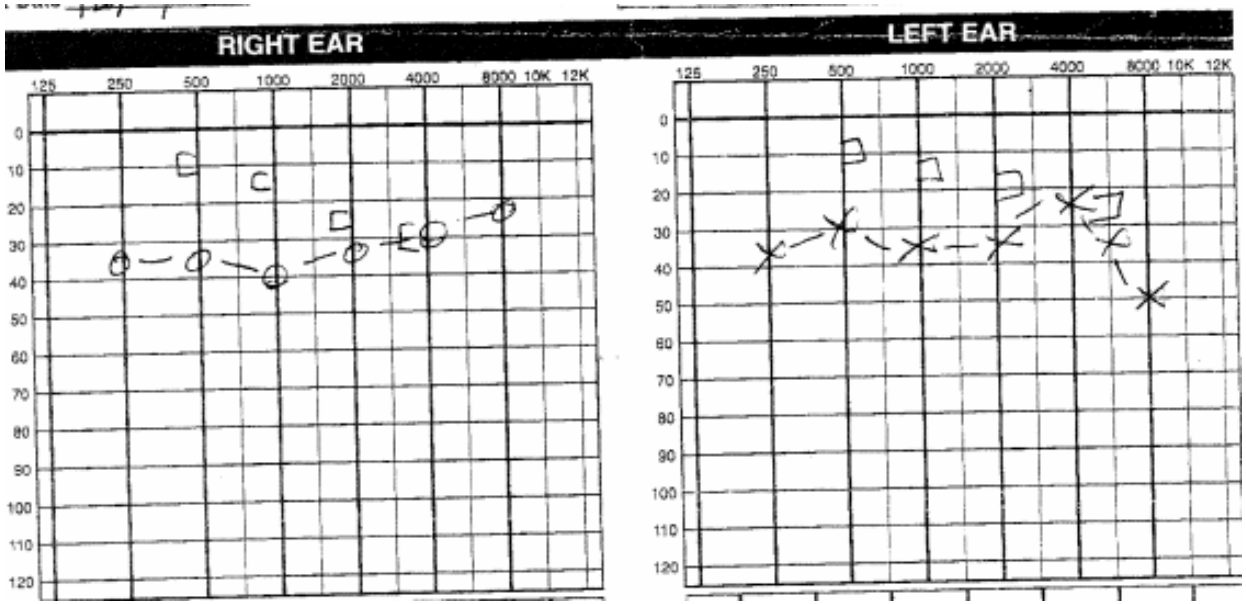
Patient I.F.

- 56 y/o male physician
- 6-year hx of internal dizziness (not vertigo), lightheadedness, disequilibrium
- Symptoms provoked w/ quick head movements, rapid changes in position, loud sounds, and w/ pressure changes
- Symptoms started after head trauma
- Issues seeing patients, as very off balance

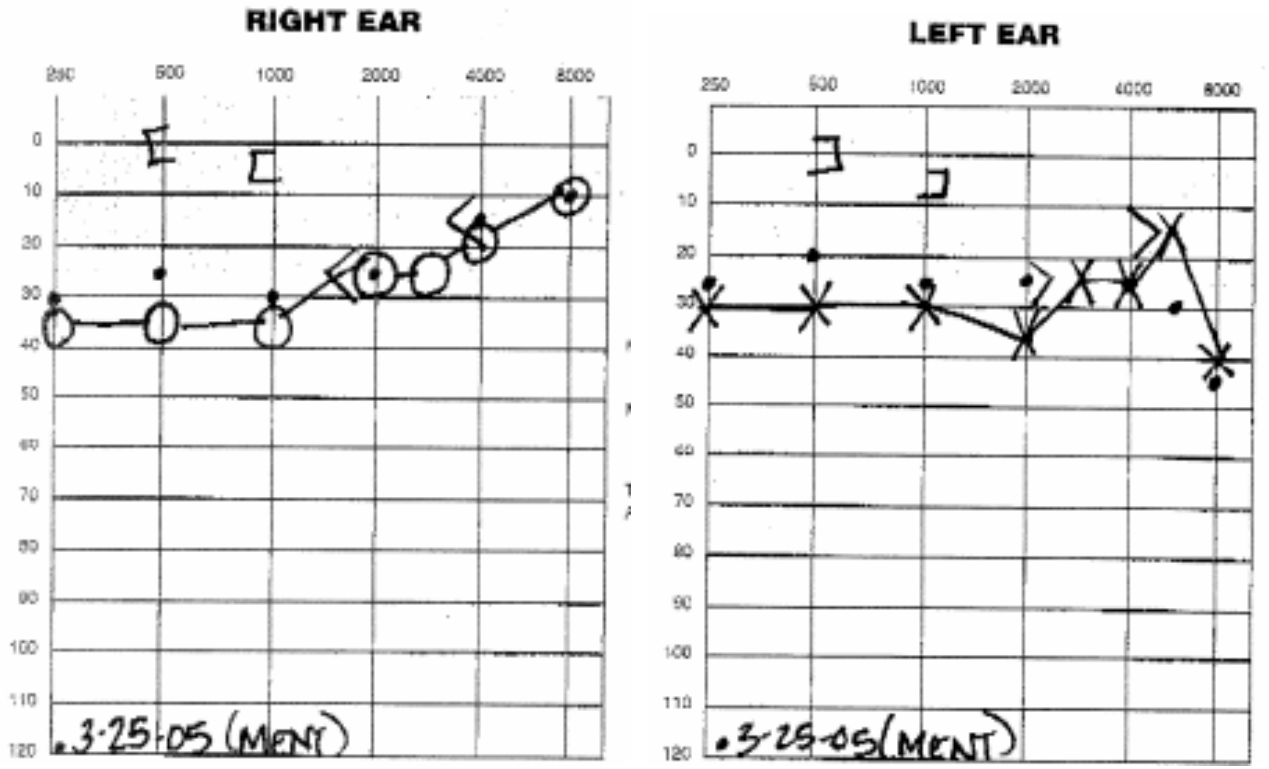
Patient I.F.

- Has been extensively evaluated for these symptoms
  - Two otologists – not vestibular
  - ENT – possible Meniere's, so low salt diet
- Recent MRI of brain WNL
- Bilateral tinnitus, which does not fluctuate
- Moderately controlled hypertension
- Now, let's take a look at his audios.

Patient I.F. Audio #1



Patient I.F. Audio #2



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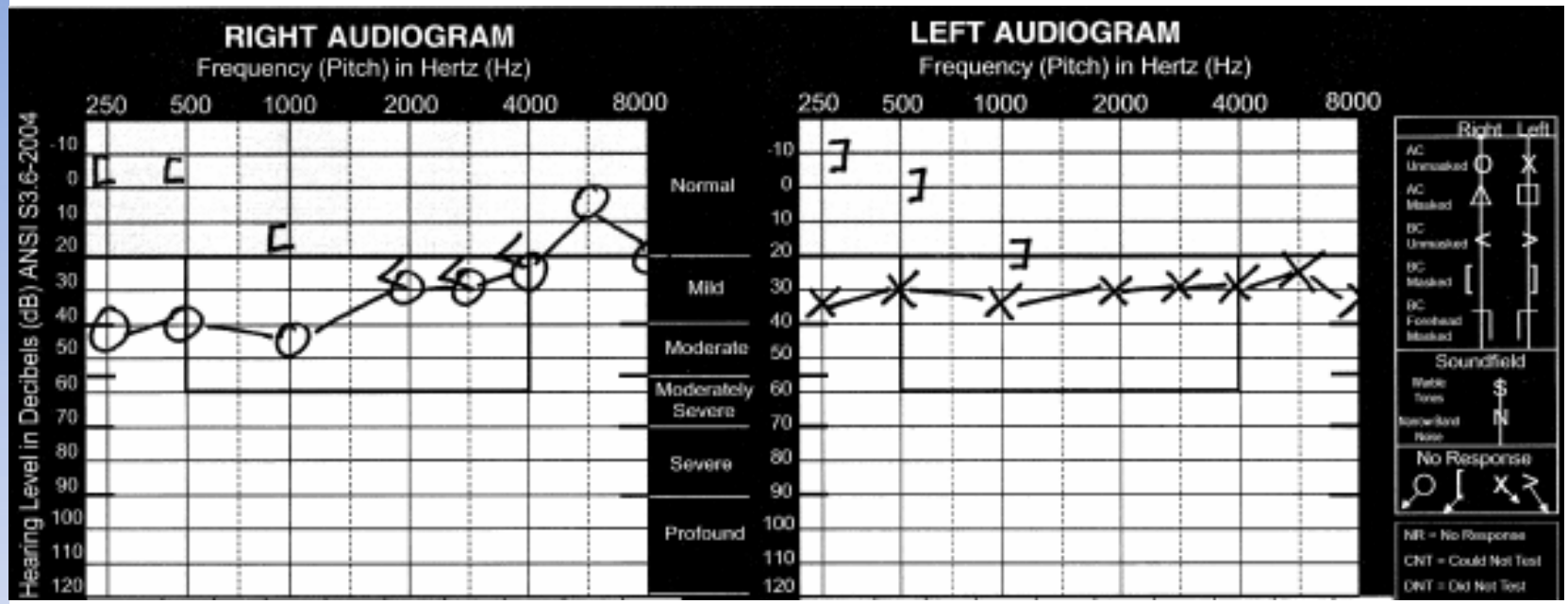
Patient I.F. Audio #3



Patient I.F.

- Patient was previously diagnosed with perilymphatic fistula and had bilateral middle ear surgery
  - What do you think?
  - Why or why not?
- Let's start thinking about our differential diagnosis – any ideas?
- What tests????

Patient I.F.

- Audio/Immittance
- VAST – negative
- VEMP – low thresholds (80 dBnHL)
- Perilymphatic fistula test
  - Low-intensity right-beating nystagmus w/ dizziness on right
  - Left negative
- VNG = all findings unremarkable
- Any other important tests…

Patient I.F.

- Gaze w/ vocalization
  - Oblique right-up nystagmus
  - I.F. experienced intense dizziness and nausea
- What does the above finding mean?
- What 2 conditions was I trying to differentiate between?

Patient I.F.

- Has anyone observed any potential diagnosis indicators?
- Does this patient really have a conductive hearing loss?
  - Why or why not?
  - Do any findings disagree?
- Does this patient have a perilymphatic fistula?
- How about Meniere's disease?

Patient I.F.

- Here is what we have:
  - Hennebert's sign
  - Tullio's phenomenon
  - Low VEMP thresholds
  - Positive right perilymphatic fistula test
  - Really good BC w/ low frequencies
  - Present VEMPs w/ air-bone gaps
  - Dizziness and nystagmus w/ vocalization
- So, what are your recommendations?

Patient I.F.

- I was concerned with an abnormal inner-ear third window
  - May not be SCDS
  - What about fistula?
  - What other pathologies can cause third window? (mention patients w/ Staecker)
- I suggested high-contrast CT-Scan of temporal bone:
  - Bilateral large SCDS

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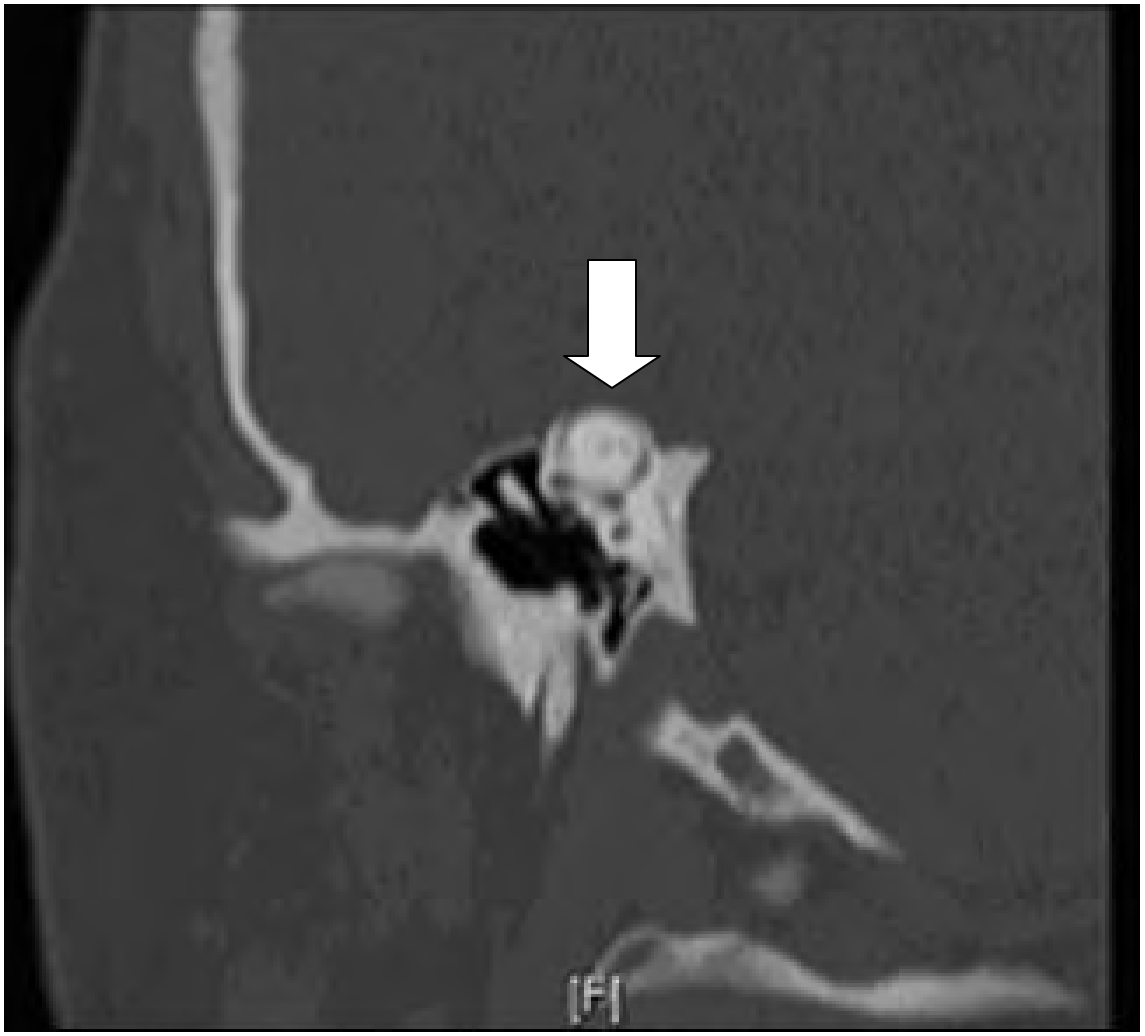
Left CT-Scan



Patient I.F.

- I.F. updates:
  - Spoke w/ him by phone 2 weeks ago
  - Has spoken to all local otologists, but does not want surgery here
  - Had surgery appt. at Johns Hopkins in 2/2011 to have right-sided SCDS fixed
  - Do you recall what surgery entails?
  - SCDS was discovered by Lloyd Minor at John Hopkins = cutting edge for surgery

Patient I.F.

- Take home messages:
  - Should not always take things at face value
  - Other audiologists were not thinking about vestibular when completing audiograms
  - Audio for a vestibular patient might be for a different purpose
  - You need to have differential prior to beginning your testing
  - You have a responsibility to your patients in making recommendations and completing testing (you are not a tech)

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Patient J.U.

Patient J.U.

- 73 year old male
- History of hypertension & diabetes
- 2-week history of nausea w/ emesis (intense, basin at appt), general dysequilibrium, and general lightheadedness
- Initial preceding attack of mild vertigo, w/ very heavy emesis
- Seen at E.R. = told vestibular problem, but no testing was completed

Patient J.U.

- No auditory changes
- No preceding event
- Placed on meclizine and oral antiemetic, which did not help
- Still acutely symptomatic during appt. w/ me
- Very ataxic when ambulating

Patient J.U.

- Let's start with a differential dx – any ideas?
- What testing???

Patient J.U.

- SOP
  - Romberg eyes open – sway
  - Romberg eye closed – sway
  - Sharpened Romberg e/o – sway
  - Sharpened Romberg e/c – fall
  - Dynamic surface e/o – fall
  - Dynamic surface e/c – fall
  - Stepping Fukuda – sway (no rotation)
- Audiogram – presbycusis w/ normal immittance

Patient J.U.

- VAST – negative
- VNG
  - Oculomotor - all abnormal
    - Smooth pursuit – stair stepping
    - Saccades – superimposed left beating nystagmus
  - Gaze – low intensity direction-changing nystagmus in static position (vision denied)
  - HFHS – low intensity left beating
  - Positionals – Ageotropic nystagmus in head and body right positions (left beating)

Patient J.U.

- Calorics:
  - I did not test, any ideas why???
- What are we thinking?
- Recommendations?

Patient J.U.

- My concerns:
  - Symptoms had not improved over initial 2 weeks
  - Nausea/emesis disproportionate to vertigo, as vertigo not primary complaint
  - Direction changing nystagmus is a BIG red flag
  - Patient's history of diabetes and hypertension can be concerning
  - No testing done in E.R.
  - History sounded atypical to me

Patient J.U.

- My recommendations:
  - I called PCP and suggested immediate MRI
  - Would consider calorics if (and only if) negative MRI
- What was found:
  - Bilateral cerebellar stroke…
    - Vertigo
    - Ataxia
    - Gait issues
    - Nausea w/ emesis
    - Prognosis fair

Patient J.U.

- Take home messages:
  - You can have a direction-fixed nystagmus w/ central lesions
  - You need to listen carefully to case history
  - It is your responsibility to make appropriate recommendations – again, I called PCP and suggested MRI
  - With the dizzy patient, there is often more going on than just the ear
  - If you are a doctoral-level practitioner and a "dizzy expert," you have a higher level of responsibility

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Patient J.S.

Patient J.S.

- 41 y/o male
- Referred by PCP
- Initially seen by one of my colleagues who specializes in tinnitus and misophonia
- Had audio, DPOAEs, and dx immittance prior to appt. with me

Initial Chief Complaints

- Life-Long Hx of decreased sound tolerance
  - Left > Right
  - Sensitivity to "S" sound
- Unilateral left tinnitus
- Dizziness (not vertigo)
  - Physical exertion
  - Straining
  - Valsalva
- Autophony

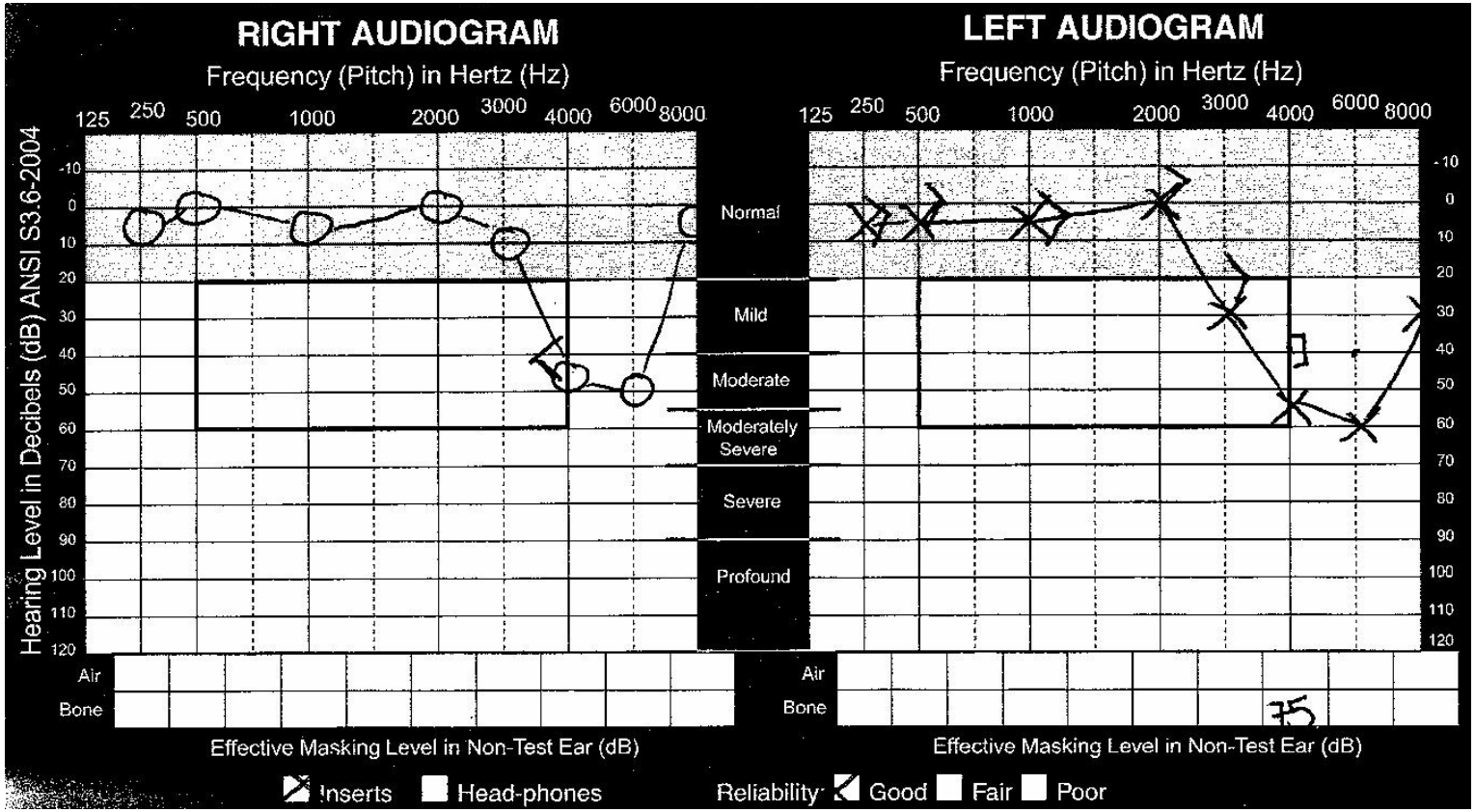
Medical Hx

- Medical Diagnoses
  - Migraine- no photophobia, phonophobia, and or nausea w/ headaches
  - Anxiety- treated with Zoloft
  - Hypertension- treated with Furosemide
- Other Medications
  - Allegra & Flonase for allergies
  - Valium as needed for dizziness

Prior Test Findings

- MRI of brain
  - Negative
  - No contrast and/or concentration on IACs
- Audiometric testing:
  - Mild sloping to moderate SNHL, w/ possible noise notch
  - Slight asymmetry with left poorer
  - Tymps and immittance WNL
- DPOAEs:
  - Reduced with high-freq stimulation

Audiogram



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Vestibular Evaluation

Vestibular Hx

- J.S. denied:
  - Any history of vertigo
  - Disequilibrium
  - Otalgia, otorrhea, aural fullness
  - Preceding event
    - Barotrauma
    - Head trauma
    - Middle ear lesion/surgery

Vestibular Hx

- J.S. denied Tullio's phenomenon
- Hennebert's sign
  - No issues with external pressure changes
  - No problems/dizziness when flying
  - OK when introducing pressure to EAM
  - However, does have dizziness when straining and during Valsalva

My Differential Dx Prior to Eval

- Perilymphatic fistula
  - Dizziness with pressure changes
- Abnormal inner ear third window (SCDS)
  - Sound sensitivity, autophony, Hennebert's
- Retrocochlear VIIIth nerve mass lesion
  - Unilateral Sxs and hearing asymmetry
- Arnold-Chiari malformation
  - Dizziness when straining (increased intracranial pressure)

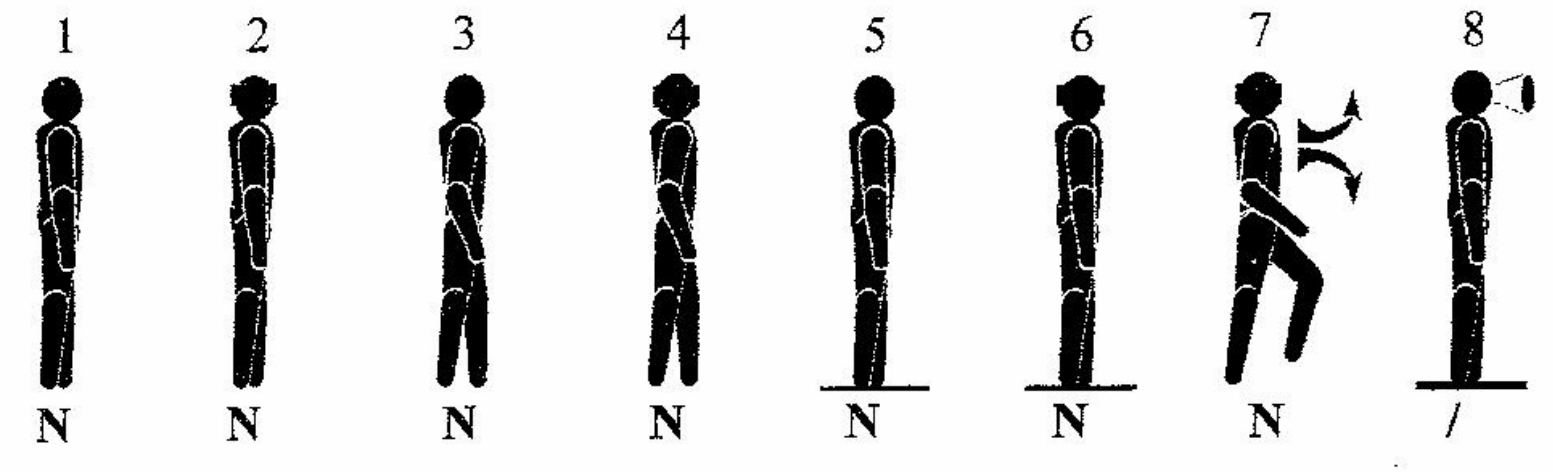
Differential Dx- Caveats

- Perilymphatic fistula-
  - No preceding event
  - Negative middle ear hx
- Abnormal inner ear third window (SCDS)
  - No low-frequency air-bone gaps
  - No Tullio's phenomenon
  - No reported disequilibrium

Differential Dx- Caveats

- Retrocochlear VIIIth nerve mass lesion
  - Previous negative MRI (although did not look closely for small tumor)
- Arnold-Chiari Malformation
  - Negative MRI
  - Cannot explain auditory symptoms

Current Evaluation - VSR



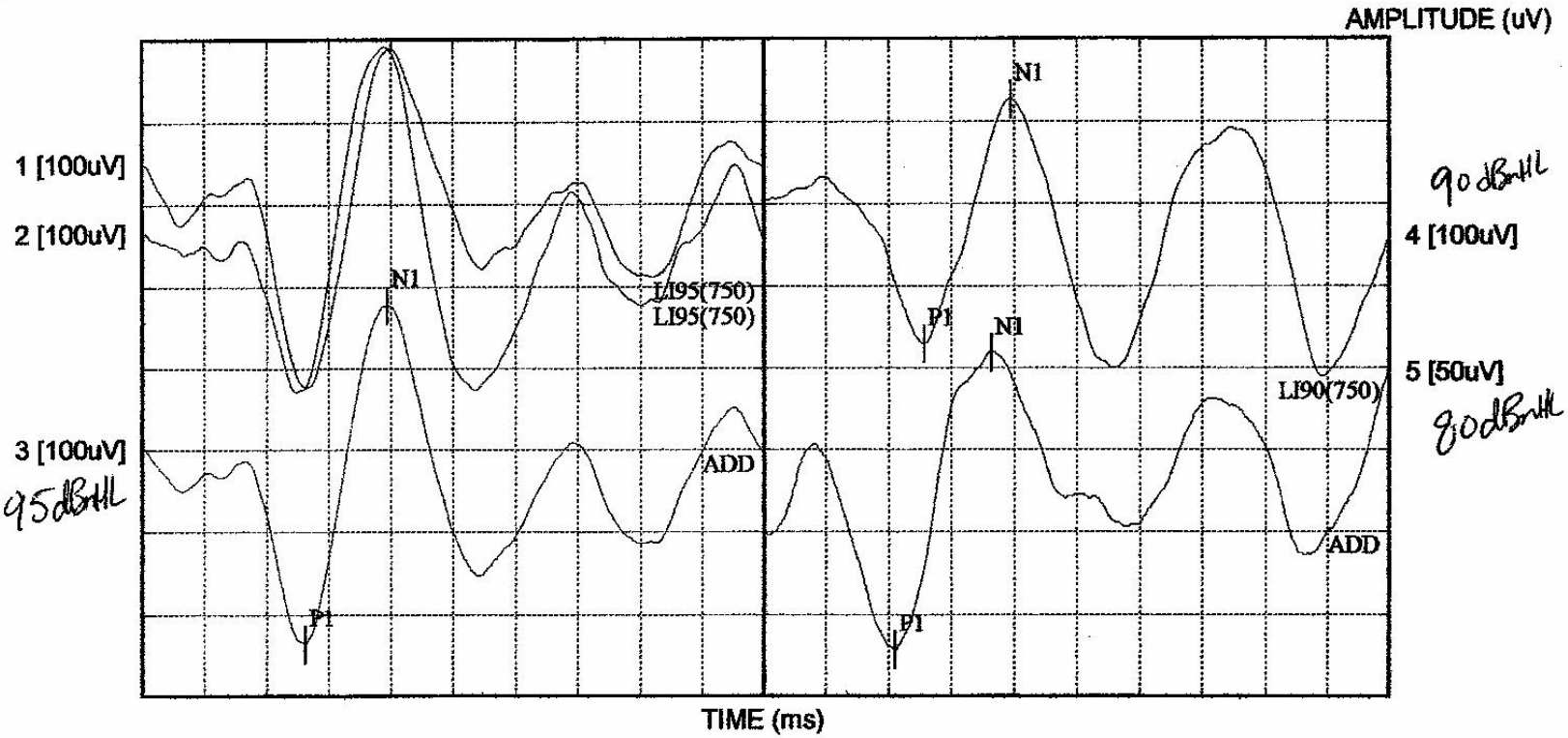
Current Evaluation - VOR

- VAT:
  - Intact VOR fx over the frequency range with both horizontal and vertical movements
- CD-VAT:
  - No significant degradation in visual acuity with volitional head movements in vertical and/or horizontal planes

Current Evaluation - VCR

- VEMP:
  - Left-
    - Amp at 100 dBnHL: 414 µV (abnormal)
    - Threshold: 80 dBnHL (abnormal)
  - Right-
    - Amp at 100 dBnHL: 134 µV
    - Threshold: 95 dBnHL

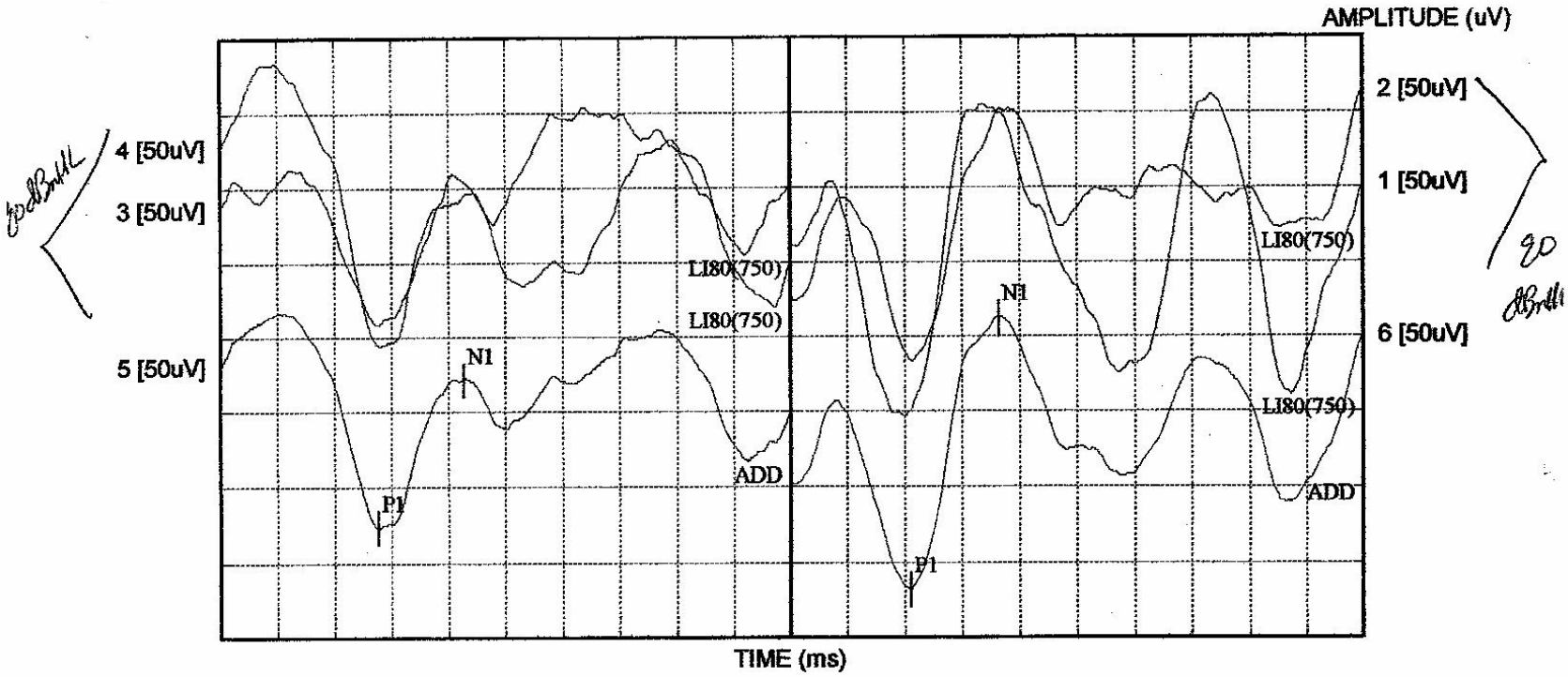
Current Evaluation



LATENCIES (ms)						
ALR						
	P1	N1	P2	N2	P3	N3
3	13.82	20.86	***	***	***	***
4	13.56	20.86	***	***	***	***
5	11.09	19.18	***	***	***	***

Left VEMP

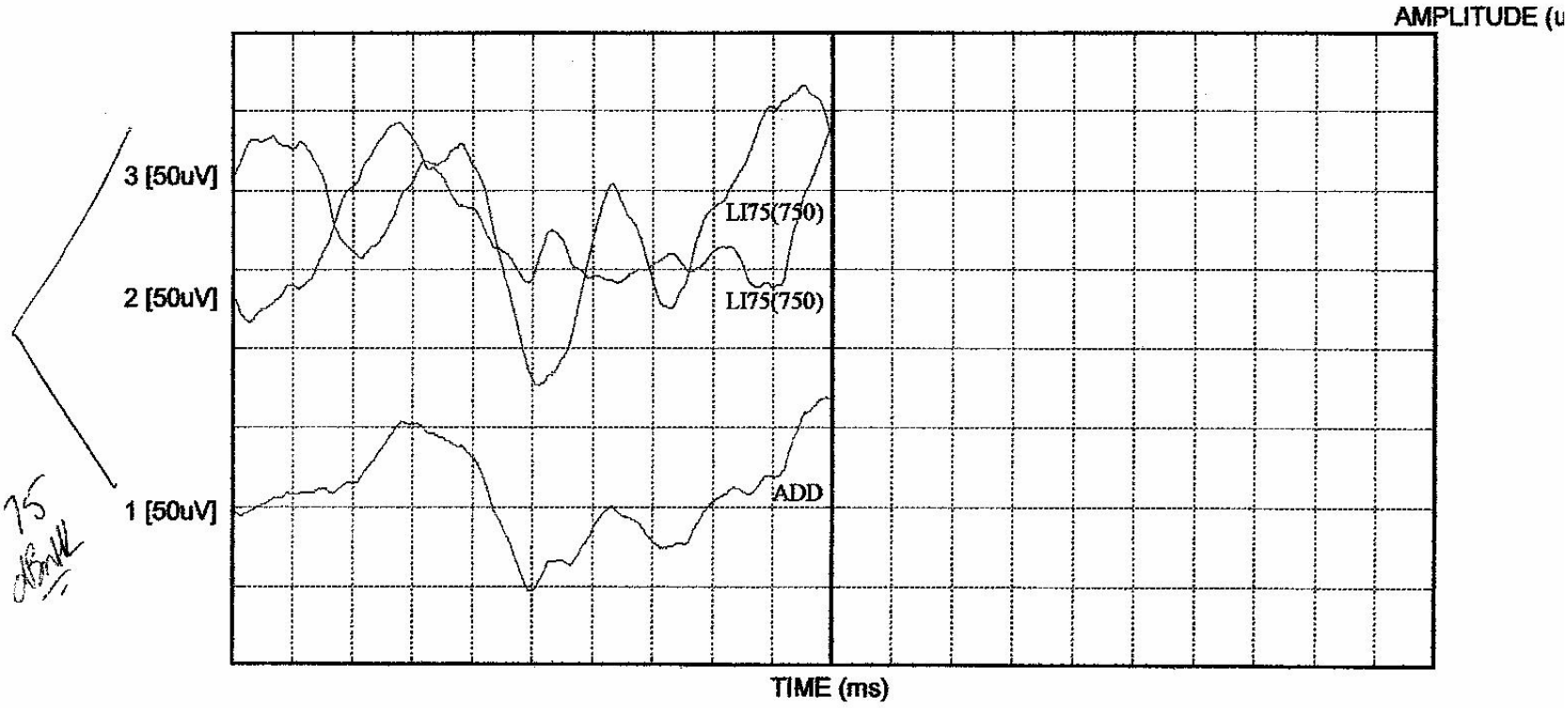
Current Evaluation



LATENCIES (ms) ALR						
	P1	N1	P2	N2	P3	N3
5	14.61	22.53	***	***	***	***
6	11.09	19.18	***	***	***	***

Left VEMP

Current Evaluation



Left VEMP

Current Evaluation – VOR, etc.

- Binocular Video-oculography
  - Gaze: no clinically significant nystagmus
  - HFHS: no provokable nystagmus
  - Hallpike: negative
  - Positionals: no clinically sign. nystagmus
  - Calorics: robust and symmetrical

Current Evaluation

- Perilymphatic Fistula Test:
  - Negative w/ rarefaction and condensation of pressure in EAM
- Gaze With Vocalization:
  - No nystagmus and/or dizziness
- ABR:
  - Symmetrical waveforms
  - Rate study WNL

Clinical Thoughts

- Possible inner ear third window
  - Abnormal left VEMP
  - Left autophony
  - Left sound sensitivity
  - Hennebert's sign
- What about unilateral tinnitus AS?
- Is this SCDS?

Recommendations

- High-contrast temporal bone CT-Scan to r/o abnormal inner ear third window
- Consultation with neurotologist for possible dehiscence
- Continued management w/ my colleague for misophonia & tinnitus

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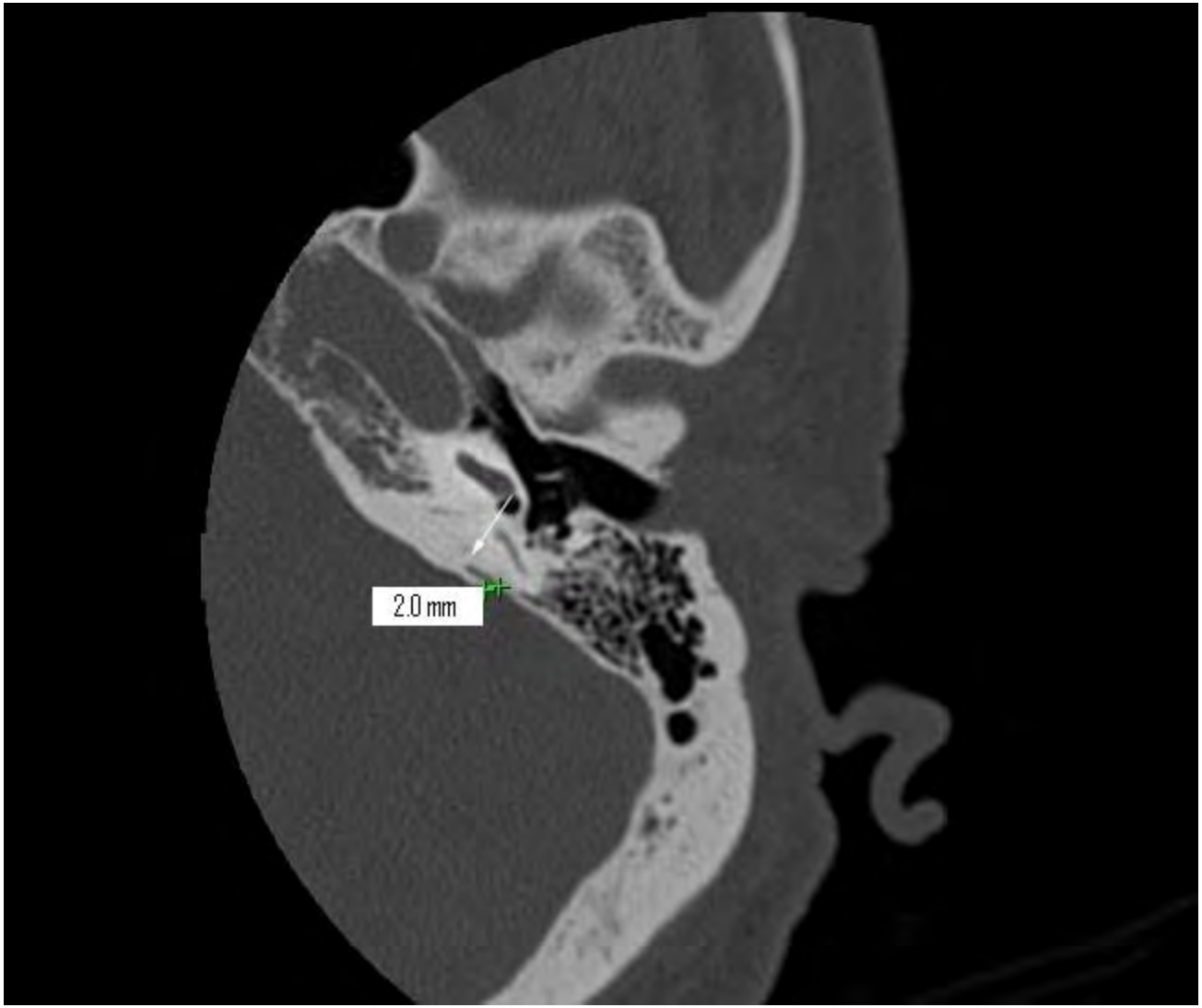
Left CT-Scan



Diagnosis and Management

- Diagnosis
  - Mildly enlarged left vestibular aqueduct
  - Semicircular canals WNL
  - Otologist re-reviewed MRI= negative for vestibular schwannoma
  - No other structural abnormalities noted
- Management
  - Treat symptomatically
  - No surgical intervention

Mildly Enlarged VAS

- Enlarged Vestibular Aqueduct Syndrome
  - Abnormal above 1.5mm at midpoint (our patient 2.0mm)
  - Hearing loss pattern can be variable
    - Degree- mild to profound
    - Type- mixed, sensorineural, low-freq conductive

EVAS and 3rd Window Effects

- Low freq air-bone gaps w/o ME path
  - Present in 66% - 97% of pediatric pts.

Zhou & Gopen (2011); Lan et al. (2007); Mimura (2005); Merchant & Rosowski (2008)

- Atypical VEMPS:
  - Abnormally low thresholds
  - Abnormally large amplitude
  - Present VEMP with air-bone gaps

Zhou & Gopen (2011); Sheykholeslami et al. (2004); Li el al. (2011); Wu el al. (2010)

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Questions/Comments???